PEMPHIGUS VEGETANS (NEUMANN).

BY

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THE case I am about to relate is the first recognised instance in England. There are, however, at least two other English cases on record, but their nature was not fully appreciated, owing to Neumann's observations not having been promulgated in this country.

As most of the known cases are from German sources, I propose, in addition to my own case, to give an abstract of all the cases related by Neumann and others which I have been able to find, and a general outline of the subject, so far as our present knowledge enables us to go.

Neumann was not the first to relate cases of this disease; Hebra and Kaposi had already described cases as vegetating syphilides, and Auspitz had met with two cases, and recorded them as herpes vegetans. Neumann, however, was the first to recognise the relation of the affection to pemphigus, and drew up such a clear and distinct account of it, that no one can have any further difficulty in avoiding the mistake of treating it for syphilis, which has generally happened.

Emma H—, æt. 43, was admitted into University College Hospital on June 18th, 1887. There was nothing of interest in her family history. Until attacked by the present disease she had never suffered from any illness which laid her up. There was some reason to think that she was not strictly temperate. On April 9th, 1887, she went to Dr. Phillips, of the Southgate Road, who kindly informed me that she then complained of great weakness and stomatitis. Nine days later, the first bulla appeared on the abdomen; others soon followed, and from the first, each bulla, instead of healing, left an ulcer at its site. From the abdomen, the eruption spread to the thighs, back, and, indeed, all over the body, the groins and axillæ being especially involved. Large doses of quinine, with and without arsenic, were given, but with no good effect, and opium, which was also tried, seemed to act prejudicially on her appetite and general condition.

On June 19th, the day after admission, the following notes were taken: The patient is a fairly well-nourished woman, with grey eyes and dark complexion. The upper lip has a single scabbed patch; the lower is thickly scabbed and dry; the rest of the face is clear. Inside the mouth the tongue is white almost all over from loosening of the epidermis; there are excoriated patches on the soft palate. The inside of the cheeks and lips are nearly denuded of epithelium, but in spite of this condition of mouth she can eat meat without much discomfort, but nothing dry or hard. The scalp is nearly covered with yellowish fatty crusts, and has an offensive smell. On the occiput is an irregular excoriated patch with unhealthy-looking edges, but there are no bullæ, and she says they began as scabs. On the neck are several excoriated patches, and where the folds of skin are in contact there is a thick white secretion. Round the excoriations, which are the site of previous bullæ, are numerous small vesicles and bullæ which are not very tense, and contain a cloudy fluid. Just above the left breast is a brown, slightly scabbed patch, two and a half by one and a half inches in area, which is a superficial ulcer in process of healing. On the right breast is a more recent patch, in

which the epidermis has been evidently raised up, but the covering is still there. The rest of the chest is quite clear. The left axilla is entirely occupied by a papillomatous, irregularly projecting mass, raised about a third of an inch above the surface. The lower half is dry, and covered with a brown crust, broken up into irregular masses by deep furrows; the upper half, where the surfaces are in contact, is covered with a white secretion with a penetrating nauseous odour like that of the scalp. In the right axilla is a similar condition, but the mass is not quite so prominent. In the arms, the left upper third is nearly clear, but near the axillæ two bullæ have just appeared, half an inch in diameter, with perfectly clear contents. Near the elbow, both front and back, the whole circumference is covered with more or less perfect bullæ and scaly crusts, and over the internal condyle is a superficial ulcer three quarters of an inch in diameter, with raised edges and red smooth surface which is now dry. The right arm is more affected, and has several fresh bullæ; over the biceps is a patch two inches square of bullæ both old and new, together with scaly crusts. Underneath the arm, continuous with the papillomatous part in the axillæ, is a large excoriated patch partly covered with epidermis. At the back of the arm near the elbow, is a raw surface about four inches by two and a half, and at the flexure is another; both are bright red, moist, and secreting serum only. Just above the flexure is a large crusted patch adjoining the raw surface at the flexure, the crust being mainly epidermic. On the left forearm, is a nearly free triangular surface over the ulna, extending about an inch and a half each side nearly to the wrist, where it ends in a point. All the rest is covered with not quite tense bullæ with opaque contents, except at the wrist, where there is an excoriated but dry surface. The right forearm is affected similarly to the left both in distribution and character, but is nowhere denuded of skin. On the extensor aspect of both wrists the bullæ are so crowded as almost to touch. The backs of both hands as far as the first interphalangeal joint are uni-

formly covered with thick epidermic crusts, and here and there, even beyond these limits, are signs of past bullæ, but the nails are unaffected. In the centre of both palms the epidermis is irregularly raised and dry, from the formation of imperfect bullæ, but the hands are not so bad as they have been. Over the arms and hands the disease as a whole is very symmetrical in its distribution. On the abdomen, large nearly continuous patches extend almost across it and upwards to just above and including the umbilicus. At the sides they are confluent, and at the centre, one patch is three inches across, with some brown stains round it, and an area of clear skin of only one inch across separates the centre and side patches. These patches are covered with brown, irregular, warty-like, crusted masses of epidermis. The crust has become detached on the left side over part of the area, leaving a smooth excoriated surface. Below the umbilicus, the skin is free as far down as a line drawn two inches above the pubes. At this point commences a large brown discolouration extending across the abdomen, the site of former lesions. In the left groin, extending back over the hip, is a large excoriated surface. On the right side, a previously diseased area has healed, except a small patch on the right flank. Towards the inner side of both inguinal regions, and extending for about three inches down the inner side of both thighs, is a papillary hypertrophy similar to that of the axillæ, but covered with dry brown crusts. On the front and outer side of the left thigh is a large, crusted, and excoriated patch, but there is no corresponding lesion on the right side. The front of the legs is quite clear to the ankles, but there have been two bullæ on the left leg. The ankles and the whole upper surfaces of the feet, as far as, but not including the toes, are covered with thickly-set bullæ in all stages, very much resembling the hands, but evidently more recently affected.

The disease extends round on the inner side to the soles, but there are only small bullæ in the centre.

The whole back is more or less covered with excoriated

patches, worst about the shoulders and lower part, and the skin of the whole back is sodden. The patches are quite irregular, and surrounded by whitish sodden epidermis. The excoriations are very abundant over the sacrum, but the whole of the back of the lower extremities is free. She says the back is not painful, even to lie on, but itches very much. Her arms are painful and burning, as well as itching, the latter being especially troublesome just before the bullæ appear.

The urine is of a very dark brown colour, very acid, sp. gr. 1040 ; no deposit, no albumen, and no sugar ; no pain on micturition.

The odour from the excoriated and sodden skin was so strong and offensive that the patient was obliged to have a ward to herself.

The first care was to endeavour to remove this foetor. The patient was placed on a water bed, and for the back and abdomen lint dipped in carbolic acid oil, 1 in 20, was applied. The papillomatous axillæ and groins were freely dusted with iodoform and the mouth washed out frequently with chlorinated soda lotion, and internally, in accordance with Mr. Hutchinson's suggestion in somewhat similar cases, Liq. Opii Sedativus $\mathfrak{m}\mathfrak{v}$ was given three times a day. This was subsequently increased to $\mathfrak{m}\mathfrak{x}$.

The antiseptic measures were quite successful, the foetor ceased almost entirely, and the fungations in the axillæ and groins became much less prominent ; altogether the patient was much more comfortable, but without any real improvement in her general condition.

By June 22nd she was unable to manage solid food, and as she missed her stimulants, six ounces of port and six ounces of brandy were allowed her.

On the 29th the eyes were painful and discharging, and she was ordered boracic acid lotion.

On July 2nd her general condition was decidedly worse, but her back showed some signs of healing. The axillæ and groins were still in a raw condition, but the fungation was less prominent.

On July 4th her temperature could no longer be taken in the axillæ or mouth.

By July 6th on the inside of the arms, on the axillæ, and on the back of the hands and feet several fresh bullæ had appeared, ruptured, and left excoriated patches. The back was better at the upper part, but quite raw over the buttocks, as was nearly the whole of the lower part of the trunk near the groins.

The patient was emaciating, manifestly weaker, wandering at times, and sleeping but little; but this was not due to pain, of which she complained very little, except when her back was being dressed, or when she was moved in any way. As she could not well use a mouth-wash for herself, her mouth was sprayed with permanganate of potash solution.

On July 7th she had a rigor at 3 a.m., the temperature in the vagina was 102° and the whole condition was worse.

On the day before, a large bulla appeared on the back of the left hand, which burst and left a raw surface. The next day another bulla had developed on the back of the right hand, and another came out on the right heel.

On July 8th the patient was evidently sinking, the forehead was covered with a clammy perspiration, and the ulceration on the right side was deeper. She died at 1 p.m. on July 9th, her general condition having gone from bad to worse, quite uninfluenced by the opium. The disease had lasted only three months.

Post-mortem (twenty-six hours after death).—Slight rigor mortis present. The body was not very much emaciated, the abdominal walls containing a large quantity of fat. The eruption was now represented by brown crusting on the abdomen, pubes, and sides; and by excoriated patches in the iliac regions and over the flanks and hips, round the neck down to the manubrium. The lips, lower portion of the orbits, and the edge of the upper eyelids were also excoriated.

On section.—Pleuræ: The left contained a considerable

quantity of thin, dirty-looking sero-purulent fluid, and there were adhesions at the lower part of the lung. The right contained a small quantity of similar fluid.

The lungs : On the left side, the posterior surface of the lower lobe was covered with greenish-white lymph, and the lung substance was collapsed, extremely softened, and diffuent at this part, but the upper lobe was normal. The right lung was normal in the upper lobe, but the lower lobe was cedematous from hypostatic congestion.

The liver weighed fifty ounces, the surface was pale, the substance soft, the lobules of a yellowish tint at the periphery, with central bluish-red points.

The spleen weighed two ounces, and was soft and diffuent.

The kidneys : The left weighed six ounces, the right seven ounces ; the capsule stripped easily, the cortex was slightly thickened, but the kidney substance was apparently normal.

The heart : The valves were all healthy, but the walls of the left ventricle were thin and softened.

There were no signs of peritonitis or disease of the intestines.

The back : On turning the body over there were seen numerous excoriations about the shoulders, and a few smaller ones in the centre of the back, below the angle of the scapula, and several not larger than peas in the inter-scapular region.

There were large sloughy-looking excoriations just above the ischial tuberosity on both sides, most marked on the right.

There were similar lesions on the buttocks as far as the margin of the anal cleft, but none between the buttocks, and no condylomatous condition about the buttocks and perinæum.

On the inner side of the thighs below the buttocks the papillary hypertrophy of the skin was very marked. There was a large excoriation over the occipital tuberosity, and a few smaller near it and near the ear. Portions of skin were removed from the groin, where the papillary hyper-

trophy was most marked, for microscopical examination, and the spinal cord was also removed and placed in bichromate of ammonium for further investigation. It showed no macroscopical changes.

The spinal cord was subsequently very carefully examined in every region by my friend Dr. Frederick Mott; the appearances were, however, quite normal.

Microscopical examination of the skin removed from the groin and abdomen showed enormous filiform papillomatous processes, many with a bulbous extremity springing from the upper layer of the corium. Each process consisted of delicate fibrous tissue containing a vascular loop and covered with epidermis; some of the processes were simple, others compound, and here and there in these compound structures two processes, separated below but united above, enclosed oval layers of horny cells, due doubtless to the continued growth of what had been external epidermis, but was now enclosed by the growing processes.

In the corium, below the hypertrophied papillæ, the vessels were dilated, and there was moderate cell-effusion round them. This was also visible round some of the sebaceous glands and sweat-coils, and round the vessels of the papilliform processes which were wide enough to show it.

The following cases, given in abstract, are all that I have been able to find of the affection under consideration :

CASE 1.—The first known case is recorded by Moritz Kohn (Kaposi), in an article on "Frambœsia" in the 'Archiv für Derm. und Syph.," vol. i, 1869, p. 406, as a case of "Syphilis cutanea papillomaformis (vegetans; frambœsia syphilitica)." It is also represented in figs. 63 and 64 of his 'Atlas of Syphilis of the Skin.'

The patient, æt. 43, was admitted into Hebra's Clinique on September 2nd, 1860, his disease dating one year before his admission, at which time he was cachectic and œdematous. The disease occupied the legs from the back of the

foot to the patella in front, and to the upper third of the leg behind, the pubic region and lower part of abdomen, coccygeal region, and from the trochanters over the anterior surface of the thigh, the axillæ, scrotum, &c. The skin of the fingers from the tip to beyond the middle, over the metacarpus, and the nails of both fingers and toes were deformed, flaky, and bent, and there were proliferations on the nail fold. The skin lesions on other parts presented the characters described in my own case. No mention is made of the condition of the oral or other mucous membranes. The disease extended considerably, and in some places there were deep ulcers. The patient was treated energetically for syphilis, but without benefit; nevertheless after suffering from albuminuria, dysentery, and pneumonia, he recovered perfectly, and he was discharged on April 8th, 1862. The disease had lasted three years and he had been twenty months in hospital.

Six years later (May 8th, 1868) he was readmitted; he had remained well up to nineteen weeks previously.

His second attack was exactly like the first; he did not improve, and a year later, May 8th, 1869, he was worse than ever; all varieties of the lesions described were abundant and had extended more widely. He had various treatment, chiefly of an antisyphilitic character, and when Kohn's report was made he had improved slightly; but eventually he died, as Neumann relates in his paper on "Pemphigus Vegetans."

CASE 2.—Reported by Auspitz as "Herpes Vegetans," in the 1869 vol. of the 'Archiv für Dermatologie,' p. 246. A married lady, æt. 28, consulted Auspitz in May, 1868, for onychogryphosis of all the fingers; there was also marked fissured psoriasis palmaris, but no other syphilitic symptoms. She was treated for syphilis, and Auspitz did not see her again, but in January, 1869, her family doctor told him that the psoriasis had got well, the nails had improved and that she became pregnant, and was going on well until November, when grouped bullæ appeared on the abdomen in successive

crops, and subsequently became pustular, crowded, and crusted, and beneath this the surface appeared as fleshy, wart-like growths or fungations. Meanwhile the patient became feverish, then the foetus ceased to show signs of life, oedema of the lungs set in, and she died collapsed.

CASE 3 was also reported by Auspitz in the same place. The patient was a married woman, æt. 26, admitted into Hebra's clinique, in the sixth month of pregnancy. She was dyspeptic and had had hæmoptysis, but no cause could be assigned for the eruption, which came out fourteen days previously, on the abdomen and lower limbs. She had no albuminuria, but had had occasional clonic spasms for the last thirteen months, and these recurred ten days after admission in the upper and lower extremities, lasting for some seconds, but without loss of consciousness. The spasms got worse in the next few days, and on the seventeenth day lasted half an hour, with loss of consciousness. Meanwhile raspberry-like proliferations had developed in the right groin, where Hebra had cauterised the crusted and excoriated patches; but they extended beyond the cauterizations, and a few days later appeared in the lower part of the abdomen, on the inner surface of the thighs, and on a palm-sized patch on the left leg, and also between the labia. Beyond the border of the growth was a dark, excoriated, weeping base.

On the eighteenth day the amnion burst, and foetal life ceased; and on the nineteenth a dead foetus, sixteen inches long, was expelled, without any eruptions on it. On the twenty-fourth day there was restlessness, delirium, and diarrhoea, without tenderness, and on the twenty-seventh day she died, after six weeks' illness.

Post-mortem.—Purulent peritonitis, inflammation of the whole of the whole of the organs of generation and some bronchitis.

Auspitz thought that two similar cases had been recorded by Köbner, a man and a woman, under the name of "Berry-like Multiple Papillary Growths of the Skin;" a male

case, of Alibert, quoted by Köbner; and two male cases by Bazin, but these were all probably cases of *Mycosis fungoides*.

CASE 4 is the text case of Neumann's paper on pemphigus vegetans in vol. xiii of the 'Viertelj. für Derm. u. Syph.,' p. 157,—a strongly-built, healthy woman, æt. 31, who only complained at first of pain on swallowing. When first seen by Neumann there was a thaler-sized patch of the usual characters in the axilla; soon after a bulla came on the lower lip, followed by numerous bullæ on the mucous membrane of the lips, mouth, and pharynx, so that nutrition became difficult. The condylomatous character of the axillary regions led to the diagnosis of syphilis. Soon after, bullæ appeared, at first of the ordinary character, but running the course of other cases of pemphigus vegetans. The oral mucous membrane became more and more affected *pari passu* with the skin, and feeding had to be effected through a tube. The conjunctiva was next involved, hiccough supervened, very scanty urine was passed, and the air of the room became very strongly impregnated with ammoniacal odour. On March 22nd, as the greater part of the skin was involved and decomposition could not be arrested, she was put into a bath at 29° R. for three hours a day; at first with some perchloride of mercury and afterwards without any, but for eight hours at a time. In the intervals salicylic wool was found to be the best to remove fœtor. Food and drugs had to be administered through the rectum, and she died on March 30th of progressive emaciation after four months' illness.

CASE 5 was an engineer, æt. 50. He was at first treated for syphilis, but later on a correct diagnosis was made. He died, but Neumann gives no further particulars.

CASE 6 was a case in Kaposi's clinique seen once by Neumann, but I have not been able to find out further particulars. The next four were more closely observed by Neumann.

CASE 7, related in Neumann's paper, was that of a lawyer, æt. 30, who had been previously treated for syphilis with iodide of potassium. He came under Neumann in April, 1884, the disease having begun in October, 1883, with pain in the throat; the fingers and toes were affected, there were the usual papillary growths, the bullæ came out continually in crops, and he died emaciated and exhausted on August 2nd, 1884, having been ill ten months.

CASE 8 was a woman, æt. 56. She first came under observation February 23rd, 1885, having been ill four months. The disease began with throat symptoms, and she was treated for syphilis by inunctions. The whole oral and pharyngeal mucous membranes were affected, and also the vulva. The eruption presented the usual characters, but one bulla on the foot became gangrenous. About a month before death, which took place early in August, painful contractions appeared in the right knee and left elbow-joints, lasting only a few moments, but recurring frequently. The reflexes were heightened, both on the patella and biceps. There was cedema of the whole left leg to the hip, and considerable emaciation. The duration of the case was between nine and ten months.

CASE 9 was a woman, æt. 41. Her illness began in September, 1884, with pain in the palate; she was admitted into the hospital on September 30th, 1885. The under lip was enlarged; both it and the tongue were denuded of epithelium. There was continual salivation, flattened bullæ at the angles of the mouth, and brown crusts round the nostrils. The skin lesions presented the usual characters in the genito-crural, anal, axillary folds and elsewhere.

The eruption continued to appear in crops at intervals, and she could only take fluids. On November 13th the patella and biceps reflexes were increased, there was delirium followed by collapse, and she died on November 16th, with a total duration of the disease of fourteen months.

CASE 10 was a woman, æt. 61, admitted July 15th, 1885,

after nine months' illness. She had a similar eruption in the same positions, and running a course like the rest, until September 15th, when she had pains in the neck, dysphagia, hoarseness, and dyspnoea. The hoarseness continued, and on the 21st bronchitis developed. On October 13th it was noted that the nails of the big toes were fissured, uneven, and furrowed, both longitudinally and transversely; the nail was discoloured, and the nail-fold surrounded by crescentic papillomatous, exuding, and crusted fungations.

On November 11th there were pains in the head, complete dysphagia for solid food, partial even for liquids, and there was right ptosis. On November 21st she refused food, and was delirious; and she died the next evening, the duration of the disease having been thirteen months.

In the '*Medicinische Jahrbücher*' for 1885, p. 539, G. Riehl publishes two cases observed in Kaposi's clinique. His paper, "*Zur Kenntniss des Pemphigus*," is translated in a full abstract in the '*Annales de Dermatologie*' for 1886, p. 163. It is possible that one of them is Case 5 of my series, of which Neumann gave no particulars.

CASE 11.—A previously healthy man, æt. 24, was admitted into the Vienna Hospital on August 15th, 1883. He had suffered for four weeks from a bullous eruption which began on the face and anterior border of the axillæ and then spread by successive crops over the abdomen and limbs. The bullæ were for the most part of the usual characters, but in the lower part of the abdomen, the axillæ and wrists, they underwent the papillomatous change and extended at their borders. The mucous membrane of the palate, cheeks, and lips was involved, continuous crops appeared on the eyelids, neck, hands, and at the periphery of the older plaques. The patient emaciated in spite of all treatment and died on September 21st, the disease having lasted nine weeks. At the post-mortem, the oral mucous membrane and tongue are described as ulcerated superficially, but the visceral changes, with the exception of bronchitis, were insignificant.

CASE 12 was a man, æt. 45. He had never been ill until eight weeks previously to his admission to the hospital on January 6th, 1884. The first symptom was intense dysphagia which lasted two days and was followed by the appearance of bullæ on the palate and tongue. They soon extended to the circumference of the mouth, the lips, the eyelids, and then gradually to the trunk and limbs. The patient was a strongly built man, but on admission was emaciated and weak. The skin round the orbits and mouth and nearly the whole surface of the body was covered with excoriated crusted patches, but the palms and soles were free. There were not only extensive excoriations in the mouth, tongue, palate and cheeks, but in the epiglottis and in the larynx, so as to produce aphonia. There was severe conjunctivitis and the cornea was dull. The bullæ were of the ordinary characters with opalescent contents, whilst others in the flexures had the papillomatous base already described. There was a similar papillomatous condition of the anterior portion of the skin of the penis and of the prepuce. There was no albumen or sugar in the urine, and no visceral disease, but the man became weaker and weaker, and died on January 13th, the disease having lasted only nine weeks. The only post-mortem visceral change of importance was left broncho-pneumonia.

The next two cases are reported by Mr. Jonathan Hutchinson in the seventieth volume of the 'Medico-Chirurgical Transactions,' p. 421, under the title of "A Form of Chronic Inflammation of the Lips and Mouth." When the paper was read I suggested that they were really the disease already described by Neumann.

CASE 13 was a master currier, æt. 58, who began to be ill at Christmas, 1879, and died in the beginning of May, 1880. His illness began with a sore mouth followed by swollen hands and feet like "chilblains." Then a vesicular eruption appeared on his hand with onychitis and accompanied by spontaneous ptyalism, sore-throat and sore lips. Next a bullous eruption extended over the whole body, his nails

fell off, large granulation-masses developed on the sores left by the bullæ, and some of these became pedunculated. Death occurred from exhaustion in less than five months from the commencement.

Post-mortem.—Broncho-pneumonia and left pleurisy were found; and a large lympho-sarcomatous tumour in front of the spine embedding the pancreas and large vessels, and weighing three and a half pounds. Of this last he presented no symptoms during life, and it did not appear to have contributed materially to the fatal result.

CASE 14 was a gentleman whose illness began in the winter of 1882. On February 18th he had "a severe form of inflammation of the mucous membrane of the mouth," but he had been under previous treatment and was taking iodide of potassium; a single mercurial pill was given and produced profuse ptyalism. Soon after this, papillary growths appeared in the groins; inflammation, first of the toe-nail and then of the finger-nails, ensued. Then small bullæ appeared in succession, on the feet, legs, trunk and arms, and when they broke, in the flexures, papillary excrescences sprouted up from their base; the mouth continued very sore, there was constant ptyalism, he became greatly emaciated, and very weak, and sank from exhaustion.

At the autopsy there was only recent hepatisation of the base of the right lung.

Mr. Hutchinson also relates three cases, which although probably, do not certainly, belong to the form of disease under discussion; and the important feature is, that they, unlike all the rest, recovered under the administration of opium in the form of *Liq. Opii Sedativus* ℥viii to ℥x three times a day. Moreover, the skin lesions were slight, not characteristic, or absent.

CASE 1 was a master tanner, æt. 50, of previously good health. His illness began with a sore mouth. When seen by Mr. Hutchinson on March 25th he had sores on his lips, inside the cheeks, and on the palate, consisting of large

ulcerated patches with red margins, and covered by a pellicle ; the tongue was not much affected. After the sore mouth had lasted about three months "some patches of a sort of a papillary psoriasis appeared on the backs of his hands, his toe-nails inflamed, and several of them were lost." He became very weak and lost much flesh, but after other treatment had failed he recovered under 10 minims of Liq. Opii Sedativus three times a day, and ceased attendance on June 30th. Two years later he had had no relapse.

CASE 2 was a clergyman, æt. 46, previously healthy, whose illness began with a sore mouth, the tongue was first affected and afterwards the gums, cheeks and lips. He lost flesh and strength, and when seen by Mr. Hutchinson four months after his illness began there was much ulceration of the mouth, but no skin eruption. Subsequently he came under the care of Mr. Pollock, and got well as a result of treatment by opium.¹

CASE 3 was a farmer, æt. 66, who had a condition of mouth similar to that of the last case, but had no disease of the skin or nails. He came under treatment six weeks from the onset, and small doses of Battley's solution were given at first, but without improving his condition. After six weeks' treatment, he became so thin and weak that his life was in danger ; then the opium was increased to 8 minims, and in a fortnight he was very much better. The dose was further increased, and in a few months he recovered completely, and a year later there had been no recurrence.

CASE 4.—The particulars were insufficient for a diagnosis. A gentleman under the care of Mr. Charles Macnamara was seen once by Mr. Hutchinson. He had a very sore mouth, an eruption on the skin, of which the character was not stated, and he died a few weeks after Mr. Hutchinson saw him. Previous to the last illness his health had been bad, in consequence of chronic diarrhoea.

¹ "On the Use of Opium as an Aid to Surgery," 'Brit. Med. Journ.,' 1884, vol. i, p. 801.

An analysis of these fourteen undoubted cases, in addition to my own, enables the following account of this rare and formidable disease to be drawn up.

Without any preceding illness or other apparent cause, the disease commences in the majority of cases by pain on swallowing, or at all events soreness of the mouth; examination shows that this is due to the formation of bullæ on the tongue, pharynx, or other part of the oral mucous membrane. After a few days or weeks, skin manifestations develop either in the form of tense bullæ with clear contents of the ordinary pemphigus type, or the contents may be sero-purulent and imperfectly fill the bulla. These bullæ frequently appear first upon the abdomen, but there is no rule upon this point; the outside of the lips, the chest, especially at the borders of the axillæ, are also places where they have commenced in more than one case. It is, however, the course that the bullæ run which constitutes one of the most striking features of the disease.

Instead of drying up in the ordinary way, leaving only the roof of the bulla as a thin scab, which soon falls off, the base of the lesion remains, in the great majority of bullæ, as an excoriated surface, which may continue raw for an indefinite period, or if it does heal eventually, leaves deep brown pigmentation. These raw surfaces are, as might be expected, largest and deepest and most obstinate on points of pressure, such as the occiput, shoulders, sacrum, and hips.

In the axillæ, groins, gluteal fold, the lower part of the abdomen, and in women about the vulva, in short, in warm moist situations where folds of skin are in contact, another even more striking phenomenon occurs on the site of the ruptured bullæ, viz. papillary proliferations of the derma projecting from a quarter to half an inch above the surface, and secreting a muco-purulent offensive fluid.

These circumstances, and their occurrence about the genito-anal region, naturally suggest condylomata, and coupled with the ulceration of the tongue and mouth, have

frequently led to the affection being diagnosed as syphilis, and treated accordingly, very much to the detriment of the already serious condition of the patient.

The course of the disease, in all the undoubted cases but one, has been steadily downwards; in one, recovery occurred, but some years later another attack carried off the patient. The rule is, that fresh crops of bullæ continue to come out at intervals; and as the condition of the mouth continues and becomes aggravated, the administration of food becomes more and more difficult. Nutrition, therefore, suffers from this cause as well as from the disease itself, which, as in all forms of dermatitis involving a large surface of the skin, produces great exhaustion and emaciation. The fœtor from the raw surfaces and papillary growths is very penetrating and almost insupportably nauseous, and after a course of weeks or months the patient dies exhausted from pain, difficulty of ingestion of food, and the large extent of the skin eruption; delirium, collapse, or some intercurrent inflammation closing the scene.

Variations.—In some cases, the skin eruption precedes the affection of the mouth, but it is difficult to ascertain how often this occurs, as the early history is defective in some of the cases, and the ulceration of the mouth is not always mentioned. In five instances at least, there has been a suppurative onychitis of some or all of the fingers and toes, and consequent shedding of the nails. Clonic convulsions and heightened reflexes of the patellar and other tendons have been observed by Neumann in more than one instance, and then death occurred by coma. Ptyalism has been noted in both of Mr. Hutchinson's and in one of Neumann's cases, of course aggravated if mercury had been given. Elevation of temperature has seldom been a conspicuous feature. Besides the oral mucous membrane, those of the conjunctiva and vulva are generally affected.

Diagnosis.—The diagnosis ought now to present no difficulty. It is true that the presence of ulceration of the mouth and lesions like condyloma about the genito-anal region is strongly suggestive of syphilis; and this dia-

gnosis has been made by experienced observers before the disease was understood. The following form a distinctive group of symptoms to anyone cognisant of the existence of the disease: The onset with dysphagia or ulceration of the mouth, followed by a pemphigoid eruption on the skin, in which the bullæ leave superficial ulcerations on their site; while in warm and moist situations like the groins and axillæ, exuberant fungating masses spring up after the bullæ have ruptured.

The post-mortem records do not throw any light upon the origin of the disease. In a puerperal woman there was peritonitis and inflammation of all the organs of generation, and in one case there was a large lympho-sarcoma in front of the spine, unsuspected during life. Bronchitis, pleuropneumonia, and croupous pneumonia by itself have been described, but these are all consequences rather than causes of the disease. The spinal cord was examined in my case with a negative result, but in future cases it would be worth while to examine the spinal ganglia and peripheral ends of the nerves. In the majority of the cases there has been little to account for death, except the extensive skin lesions.

To assume, what is probable enough, that the disease is a neurosis, is practically no real advance in our knowledge as to its nature, and for the present we must confess ourselves utterly ignorant both of its pathology and etiology.

Treatment.—With one possible exception, all treatment has proved utterly futile to arrest the fatal course of the disease. Much, however, may be done to alleviate the suffering of the patient, and to remove the fœtor due to the decaying epithelium. Such measures, as already detailed, were quite successful in my own case, and Neumann obtained a similar result by using the continuous bath for several hours a day, and salicylic acid wool. Antiseptic washes for the mouth such as chlorate of potash lotions, or, when the patient is too weak to use them, permanganate of potash solution sprayed in, are also useful. The one possible exception to the futility of internal treatment is that

suggested by Mr. Hutchinson, namely, opium. It is for this reason that I have included an abstract of his three patients, who recovered under its use ; but although probably of the same nature, it is not certain that they were so, as the characteristic skin symptoms had not developed. In my own case opium was tried in vain, both in the early stage, before I saw her, and after she came to me ; it was then given in full doses, but probably the disease had gone too far, while at the beginning the opium certainly failed to relieve, but possibly it was not given in adequate doses, ten minims of the *Liq. Opii Sedativus*, three times a day, being the least dose likely to be beneficial.

Table of all recorded cases of Pemphigus Vegetans.

No.	Sex.	Age.	First symptoms.	Mucous membrane affected.	Nails involved.	Previous health.	Duration.	Post-mortem.	Reporter.	Name given to disease.	Where published.	Remarks.
1	M.	43	?	No mention	Yes	Good	1st attack 3 years. 2nd over 1 year	?	M. Kohn	Syphilis vegetans	Archiv für Derm. u. Syph., vol. i, 1869, p. 406, and plates lxiii and lxiv in Kaposi's Atlas of Syphilis der Haut	The patient was in Hebra's Clinique; second attack 6 years later than first; was alive when first reported, but Neumann says he died subsequently.
2	F.	28	Grouped bullæ on abdomen	?	?	Pregnant when attacked; probably had syphilis	A few weeks	None	Auspitz	Herpes vegetans	Loc. cit., vol. i, p. 246	—
3	F.	26	Bullæ on abdomen and lower limbs	Vulva	?	Pregnant	6 weeks	Purulent peritonitis and inflammation of all organs of generation	"	"	Loc. cit., vol. i, p. 248	Clonic convulsions and heightened reflexes.
4	F.	31	Dysphagia	Lips, mouth, pharynx, and conjunctiva	—	Very good	4 months	?	Neumann	Pemphigus vegetans	Vierteljahrsschrift für Derm. u. Syph., vol. 1886, p. 159	—

No.	Age & Sex	First symptoms.	Mucous membrane affected.	Nails.	Previous health.	Duration.	Post-mortem.	Reporter.	Name given to disease.	Where published.	Remarks.
5	M. 50	?	?	?	?	?	?	Neumann	Pemphigus vegetans	Loc. cit.	No particulars, except that at first syphilis was diagnosed, and that he died.
6	?	?	?	?	?	?	?	"	"	Loc. cit.	Seen once by Neumann in Kaposi's Clinique.
7	M. 30	Pain in the throat	—	Yes	?	10 months	?	"	"	Loc. cit.	Was treated for syphilis at first.
8	F. 50	"	Mouth, pharynx, vulva	?	—	9—10 months	?	"	"	Loc. cit.	Had clonic and tonic contractions and exaggerated reflexes.
9	F. 41	Pain in palate	Mouth, tongue, nostrils	?	—	14 months	?	"	"	Loc. cit.	Pyralism, exaggerated reflexes.
10	F. 61	Bullæ on skin	Mouth, pharynx	Yes	—	13 months	?	"	"	Loc. cit.	Dysphagia, ptosis, delirium.
11	M. 24	Bullæ on face and axillæ	Mouth, conjunctivæ	No	Good	9 weeks	Bronchitis	Riehl	Pemphigus	Med. Jahrbücher, 1885, p. 539	In Kaposi's Clinique.
12	M. 45	Intense dysphagia	Mouth, tongue, conjunctivæ, epiglottis, larynx	No	"	9 weeks	Left broncho-pneumonia	"	"	Loc. cit.	"

13	M.	58	Sore mouth	Mouth	Yes	"	5 months	Broncho-pneumonia, pleurisy, and large sarcoma	J. Hutchin-son	Chronic in-flammation of lips and mouth	Med.-Chir. Trans., vol. lxx, 1887, p. 421	Ptyalism.
14	M.	?	"	"	Yes	"	—	Right pneumonia	"	"	Loc. cit.	"
15	F.	43	"	Mouth, nose, conjunctivæ, vulva	No	"	3 months	Left pleuro-pneumonia	Radcliffe Crocker	Pemphigus vegetans	Loc. cit. present vol.	—
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Analysis of the 14 available cases shows 7 males and 7 females, their ages varying from 24¹ to 58. In 12 cases in which the mode of onset is mentioned, it began with dysphagia or sore mouth in 8, and with bullæ on the skin in 4; but in at least 10 the oral mucous membrane was affected at some time. There was suppurative onychitis in 5, ptyalism in 3, exaggerated reflexes in 3, clonic convulsions in 2. The general health before the onset of the disease was nearly always good, but 2 were pregnant when attacked. Excluding Case 1, in whom the first attack lasted three years, and six years later his second fatal illness lasted thirteen months, all the cases were fatal in from six weeks to fourteen months, and the post-mortem changes were for the most part insignificant and clearly secondary.

¹ In Dr. Mapother's case mentioned in the discussion, the patient was a woman, æt. 21, who was treated for syphilis, and died after nine weeks' illness.

(For report of the discussion on this paper, see 'Proceedings of the Royal Medical and Chirurgical Society,' Third Series, vol. i, p. 95.)

DESCRIPTION OF PLATE V.

Pemphigus Vegetans (Neumann) (H. RADCLIFFE CROCKER, M.D.).

Section of skin taken from groin, showing enormously hypertrophied papillæ.

- a.* Papillary outgrowth.
- b.* Cell infiltration in papillary layer.
- c.* Dilated vessels.
- d.* Sweat coil, with cell infiltration between the coils.