TWO CASES
IN WHICH
THE SKIN, HAIR AND TEETH
WERE VERY IMPERFECTLY DEVELOPED.

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Received Dec. 20th, 1847—Read Feb. 8th, 1848.

IN the same work in which the founder of Inductive Science noted the “deficiencies of medicine,” he likewise complained that he could find “no sufficient or competent collection of the heteroclites or irregulars of nature well examined and described.”* Since the days of Lord Bacon, and particularly of later years, much has been done for the advancement of that department of physiology which refers to original defects of structure and malformations; but, notwithstanding the labours of the St. Hilaires and Vroliks of our times, it must still be admitted that the modern-named science of Teratology requires large additions of correctly-observed and well-described facts for its complete development. I shall probably not overrate the importance of this class of facts, when I express the opinion that practical medicine itself, no less than the natural history of our species, may hope to receive additions from the study of these exceptional cases of original structure and function. Hence I think that the cases which I propose now to describe are not merely

* Advancement of Learning.

interesting as medical curiosities, but are calculated to throw some light on the structure and functions of the skin and its appendages.

Case 1.—The gentleman who was the subject of this case, died during the year 1847, aged about 58. He was distinguished, throughout life, by the almost complete absence of hair; by the teeth being not more than four in number; by the delicate structure of the skin; and by the absence of sensible perspiration and tears.

During the last ten years of his life, he was more particularly known to myself. At that period he may be described as a person of middle stature, delicate form, and, generally, of a somewhat valetudinarian appearance. His countenance had a rather foreign aspect, difficult to describe; the features being rather small and flat, the nostrils small and compressed, the complexion yellow and freckled, and the face much marked, as if by the small-pox, from which, however, it does not appear that he had ever suffered. Possessed of average intellectual powers, his character was marked by a certain amount of sensitiveness and timidity, and by occasional irritability of temper. These peculiarities of mental constitution may have been in part induced by the physical defects under which he laboured: but, in connection with this subject, it should perhaps be stated that a sister, an uncle and a cousin on the father's side died insane. His habits of life were regular and temperate. He was never married.

On his head, there was not so much hair as on that of an infant three months old. What hair there was, was confined to the crown and back of the head, was very fine, short and soft, and of a light brown colour. The hair of the eyebrows was absent, and the eye-lashes were defective. The hair of the face and chin was relatively more developed than that of the head; but though he practised shaving daily, the beard and whiskers, particularly the latter, were very scanty. In the armpits, there was a small amount

of soft, downy hair; and on the pubes were a few scattered hairs, which were longer and stronger than any others on the body. Elsewhere, indeed, scarcely a hair, however small, was to be observed.

He had only four teeth, which were in the upper jaw, and consisted of the second and third molars of each side. So far as I can learn from the dentist who supplied his deficiency in this respect, there was no peculiarity of any kind in these teeth. I have not been able to ascertain the age at which these teeth appeared, nor yet whether any of the deciduous teeth were ever developed: it is thought they were not. The nasal passages were remarkably narrow, and it was only by extraordinary attention to cleanliness, in drawing water through the nostrils, that he was able to prevent their being completely occluded with mucus. Another peculiarity, of which I was not informed until some time after his death, was, that he had never been known to shed tears, and that the painful emotions, generally relieved in this way, had in him an unusually distressing appearance.

The skin generally was very fine and thin, and very delicate, both as regards colour and texture; as much so, indeed, as in the most delicate female. The surface of the body was often dry and harsh, and exhibited no traces of sensible perspiration: it was remarkably free from any trace of unctuousity. During hot weather, and after taking unusual exercise, the skin often became very hot and red, without being in the least degree bedewed with moisture. Under these circumstances, it was frequently observed that his breathing was oppressed; and, whilst thus heated, he often resorted to the free ablution of the entire surface with cold water, from which he appeared to derive great relief. The only occasion on which his nearest connections or medical attendant ever observed any trace of moisture on the surface of the body, was during the last week or ten days of his life, when in a very exhausted state, and particularly during the hour immediately preceding death.

Sudorific remedies had no sensible effect on the skin, as

regards moisture ; but, under their use, changes from a dry and harsh, to a comparatively relaxed and soft, state were observed.

In connection with this part of the case, I am indebted to his medical attendant for the information that he had been long subject to transient attacks of what may be described as feverish heat and excitement. During these attacks, the skin became harsh, dry and intensely hot ; the tongue, dry and red, but not coated ; the pulse, full and quick ; the respiration, difficult and laboured ; and his feelings, those of great restlessness and discomfort. These paroxysms were most effectually relieved by small and repeated doses of tartarised antimony, which, in the course of a few hours, produced a state of comparative comfort and tranquillity, the skin regaining its natural temperature and becoming soft and relaxed. Both in health and during indisposition, the urinary secretion was very abundant and usually clear. He had a considerable bronchocele, though this was not so large as to attract much attention during life when covered with the cravat.

He had never enjoyed very robust health, and, about his 35th year, suffered from a severe attack of pulmonary inflammation, which does not appear to have been treated with sufficient activity. After this, he was more or less subject to difficult breathing, which was liable to be aggravated by cold and damp weather. During the last ten years also of his life, he suffered several times from symptoms of severe affection of the stomach, which were chiefly characterised by severe pain, without vomiting, and were relieved by medical treatment.

In the autumn of 1845, he had an unusually severe pulmonary attack, attended with much dyspnœa, cough and debility, and with repeated sanguineous expectoration. He rallied for a while during the warm summer of 1846, but the disease made steady progress during the winter, and he died March 1847.

Inspection twenty-four hours post mortem.—Many of the

peculiarities of the tegumentary system already described were now particularly noted. The body generally was much emaciated.

Head.—The calvaria was thick, and of almost ivory-like density. The dura mater adhered firmly to the cranium. The internal Pacchionian glands were rather large and numerous, and around them there was a considerable patch of opacity, with trifling adhesion of the arachnoid. The consistence of the brain was perhaps hardly so firm as normal. The blood-vessels were not more loaded than in health. In the lateral ventricles there was about an ounce of serum. The weight of the brain was $49\frac{1}{2}$ ounces; of the cerebrum, 44 ounces; of the cerebellum and medulla oblongata, $5\frac{1}{2}$ ounces.

Thorax, &c.—In the left lobe of the enlarged thyroid gland was much calcareous deposit, of a spicular kind. The upper three fourths of the trachea were compressed between the lobes of the enlarged gland, and its area was reduced by about one-half. The mucous membrane of the larynx and trachea was pale and thin, covered by muco-purulent secretion, and studded with minute miliary tubercles. There were old, firm adhesions of the pleura. Two or three vomices, surrounded by scattered tubercles, existed in the upper portions of both lungs. In the apex of the left lung was a cavity, the size of a large orange, containing a grumous fluid, resembling the lees of red wine. There were œdema and congestion of the lungs generally. The pericardium contained nearly 4 ounces of turbid serum. The heart was soft and flabby, and weighed $9\frac{3}{4}$ ounces.

Abdomen.—Two ulcers were found in the stomach; the smaller and more recent, in the lesser curvature; the other, the size of a shilling, about an inch and a half lower, in the anterior surface. This ulcer had round, smooth edges, and had completely perforated the mucous membrane. At the point corresponding to it externally, the peritoneum was adherent. Around the cæcum were old, general adhesions of the peritoneum. The vermiform appendage was at least

double the usual length, and was turned up between the cæcum and spine. The liver and spleen were of normal size. The pancreas was large. The kidneys were larger than usual, and healthy. The ensiform cartilage was bifurcated.

It appeared desirable that the microscopic character of the skin should be correctly ascertained and described. I, therefore, transmitted small portions from the axilla, epigastrium and pubes to Mr. Erasmus Wilson, who has favoured me with the following observations, as the results of his repeated careful examination :—

“ The peculiarities of the portions of skin sent to me are their whiteness, thinness and softness, and the tenuity of, and absence of pigment in, the epiderma. The number of pores upon the surface and the grooves of motion are natural, though probably smaller than usual. The chief peculiarity, however, is that which is discerned by the microscope,—namely, a state of extreme laxity and apparent atrophy of the derma, with a total absence of fat and of the usual filling-up material. The corium has the appearance of a delicate sponge, every areolar space being transparent and filled with a colourless fluid, and the parietes of the spaces formed of distinct and separate filaments of fibrous tissue. The object which thin sections of the derma most nearly resemble is bone affected with interstitial absorption, all the cell-walls being excessively and beautifully delicate and fine. The sudoriparous glands are not wanting, but their ducts are unusually delicate and deficient in substance, and their epithelial and consequently secreting lining very thin. The portion of skin from the axilla is richly supplied with sebiparous glands, and there is the accustomed cluster of these glands in connection with each hair. The subcutaneous adipose tissue, moreover, has entirely lost its oily matter, that which remains being a mass of shrivelled cells and nuclei, retaining the yellow colouring pigment of fat.”

From these observations we may, I think, infer with Mr. Wilson, that the pathological state of the skin in this case

was one of atrophy and atony, and that the absence of sensible perspiration depended, in part, on the defective state of the sudoriparous apparatus; and, in part, on the want of proper action of the skin itself.

I may further state, that Mr. Bowman, who was also good enough to examine portions of the skin with the microscope, did not obtain satisfactory evidence of the presence of sweat-glands or ducts. The result of Mr. Bowman's examination would thus be that of giving a still more defective character to the skin. Mr. Wilson's examination was, however, perhaps more extended, and was repeated on a second set of specimens.

Many curious and not unimportant physiological and pathological questions arise in connection with the case which has now been narrated. Upon these, however, I do not intend to enter at length. I will briefly express the opinion, that the defective structure and function of the skin not only exerted a generally unfavourable influence on the system, but that it, probably, likewise contributed materially to the fatal result. It no doubt rendered the patient less able to resist the chronic disease of the lungs under which he laboured, and which, with a general surface of ordinary transpiring power, might have been more amenable to treatment, or, at least, might have longer remained in a quiescent form. The bronchocele doubtless also acted as an additional cause of pulmonary disturbance.

What appears to constitute a general principle in the laws governing malformations, is likewise illustrated by this case. It will, I think, be found that considerable malformations are seldom single, but are usually associated with one or more minor defects. Thus acephalous fœtuses very commonly present examples of club-foot, supernumerary fingers and toes, and other malformations of the limbs. I have also observed similar malformations to be associated with those defects in cerebral organisation which constitute idiocy. In the case before us, we may observe that the general teleological defect in the structure of the skin and its appendages

was associated with bifurcation of the ensiform cartilage, and with unusual size and position of the vermiform appendage of the cæcum.

Not being, at the time, informed of the absence of tears in this case, the opportunity was lost for examining the condition of the lachrymal glands and apparatus, supposing them to be present.* It is hardly probable that the gland was present, unless in a merely rudimentary form. And it may be also observed, that the narrow and often occluded condition of the nostrils renders it not improbable that the lachrymal duct was itself absent. In his recent "Report on the Homologies of the Vertebrate Skeleton," and in his "Lectures on Comparative Anatomy," Professor Owen concludes that the lachrymal bone really belongs to a different category from the rest of the bones of the head and face; that it is developed in connection with the skin, and constitutes the solitary bone of the exo-skeleton in man.† Had the absence or defective development of this "muco-dermal" bone been established in the instance before us, the conclusion of Professor Owen as to its homologies would have received valuable and curious confirmation.

Case 2.—The circumstance of perhaps most interest in connection with the case which has been narrated, remains to be adverted to. A cousin-german, on the mother's side, who was born only a year or two before the subject of the preceding case, presented almost precisely the same peculiarities. His head was equally bald; the skin had a similarly delicate texture; there was the same absence of sensible perspiration, and of tears, and the teeth were similarly deficient. Which of the teeth were present cannot now be ascertained. His nostrils were more contracted than his cousin's, and were

* I am not aware whether any cases, in which the lachrymal secretion was wanting, have been recorded.

† "Report of British Association for 1846," pp. 210, 300, 308; and "Lectures on Comparative Anatomy of Vertebrate Animals," Part I. 1846. See also "British and For. Med. Review," vol. xxiii. p. 490.

at one time the seat of some ulceration. He had very delicate health, suffered much from inflammatory affections, and died of croup at the age of 13 years.

In early life, the two cousins were almost precise counterparts of each other; and the friends of the family of the youth last described were not a little perplexed when, a year or two after his death, the other came on a visit to his uncle's family. In mental endowments and characteristics, however, there was a great distinction; and whilst the one was timid and sensitive, the other was distinguished for boldness and fearlessness of character, and by his addiction, as a boy, to reading and other intellectual pursuits.

It was only natural to inquire whether similar peculiarities in the cutaneous system had been observed in any former generation of the family. The only approach to such peculiarity, which is known, was in the maternal grandmother, who was distinguished by the extreme delicacy of her skin, and by the very limited amount of sensible perspiration which she presented, under any circumstances. There was, however, no defect or peculiarity in the hair, nor, so far as is known, any in the teeth of this lady; and with the exception of the two grandsons, whose cases have been described, and who were the children of her only two daughters, no such peculiarities have been observed in any of her descendants, who have long reached the fourth generation, and are upwards of thirty in number. For the most part, indeed, an abundance of hair characterizes the family.

The physical defects described in the two cases now related, even when separately considered, must be regarded as rare. As combined, however, in the same individual, and still more as occurring in two persons closely connected by consanguinity, they constitute an anomaly of a still more curious and remarkable kind.

Congenital defect of the hair is, perhaps, less rare than is any considerable deficiency in the teeth. Several of the cases described as congenital have, however, really been examples of loss of hair from disease. Thus, of four cases cited by

Otto, two at least, those of Heister and Wells, are cases of general alopecia.* In his translation of Otto's work, Mr. South briefly alludes to the case (I presume, congenital) of "a man, aged 20 years, employed at the Courier newspaper office (London), in 1841, who had not any hair upon the head, eyebrows, eyelids, or chin, and was said to have had none on the pubes."

Rayer speaks of congenital deficiency of the hair as a malformation very rarely met with. He states that "it very seldom continues longer than the few first years of life, and ought rather to be regarded in the light of a late development of this appendage of the skin." He describes one remarkable case observed by himself in 1827, at the Hospital la Charité, in which this congenital defect was very complete:—"The skin of this man's cranium appeared completely naked; although on examining it narrowly, it was found to be set with a quantity of very fine white and silky hair, similar to the down that covers the scalp of infants; here and there upon the temples there were a few black specks, occasioned by the stumps of several hairs, which the patient had shaved off. The eyebrows were merely indicated by a few fine and very short hairs; the free edges of the eyelids were without cilia, but the bulb of each of these was indicated by a small point. The beard was so thin and weak that Beauvais (this was his name) only clipped it off every three weeks. A few straggling hairs only were observed on the breast and pubic region, as in young people on the approach of puberty. There was scarcely any under the axillæ. It was rather more abundant on the inner part of the legs. The voice had the pitch and intonation of that of a full-grown and well-constituted man. He had had syphilis twice. He stated that his mother and both his sisters had fine heads of hair, whilst his father presented the same defect in regard to the hair which he did himself."†

* Pathological Anatomy, by South, p. 120. The case by Augustin, in Wolfart's Asklepieion, [quoted by Otto,] I have had no opportunity of verifying.

† Diseases of the Skin, by Willis. 1835. p. 1049.

The permanent absence of one, two, or even a greater number of the teeth, is a defect respecting which the dentist will be more frequently consulted than the physician or surgeon, and which is probably not of very rare occurrence. The incisors (of the lower jaw particularly) and the bicuspid, appear to be more especially liable not to be developed.* Mr. Fox mentions an instance in which this defect attached to several members of the same family, none of whom had ever cut the incisors of the lower jaw.

Three cases are recorded in which all the teeth remained wanting through life. The first is that of a woman, sixty years of age, and is recorded by Borelli, who merely observes that the case proves "it is not always true, as has hitherto been believed, that paucity of teeth portends a short or feeble life."†

The next instance which I find recorded is more particularly interesting in connection with the cases which are the subject of this paper, and presents an example of the entire deficiency of both teeth and hair in the persons of two brothers. One cannot but regret the absence of further particulars of these cases, which, though promised by the author who narrates them, do not appear to have been ever given. The brief notice which exists is headed, "Concerning men without hair or teeth;" it is as follows:—"In our neighbourhood lives a Jewish family, of which two adult sons neither have, nor ever have had, hair or teeth. The more particular details are yet unknown to me."‡

The absence in the same cases of both teeth and hair, an associated defect, as we thus find, not now for the first time observed, is confirmatory of the opinion, at the present day entertained by many physiologists, that the teeth, hair, sebaceous follicles and mucous follicles constitute a common class of organs, subsidiary to the integumentary system; and that

* Otto, op. cit., pp. 185, 188. Good's Study of Medicine, vol. i. p. 61.

† Hist. et Obs. Med. Phys. Cent. II. Obs. xli. p. 144. 1676.

‡ Danz, in Stark's Archiv. f. d. Geburtshülfe. Vol. iv. p. 684. Jena, 1792.

they are closely connected in their development, growth and decay.

Cases have been previously recorded in which the sensible perspiration was stated to be entirely absent; but it may be doubted whether they rest on accurate observation.* In the cases which are described in this paper we may regret that no observations on the character and amount of the *insensible* perspiration were made during life.

Postscript, July 14, 1848.—Though such cases as those described in the foregoing paper are no doubt rare, they are probably less so than would generally be supposed. Thus, on the occasion of the paper being read to the Society, Dr. C. J. B. Williams mentioned the case of a young female to which I am happy to refer in this postscript. The young lady was described to Dr. Williams as being without hair and without teeth. When seen by him, however, he found that she was not entirely so. She was fifteen years of age; her hair was fine, scanty and white, and she had scarcely any hair on her eyebrows, or any lashes. She had three or four projections resembling teeth, which were decayed. The nose was imperfectly developed, and she breathed with difficulty through the nostrils. She suffered from chronic ulceration of the lining membrane of the nares, attended by a discharge, having an unpleasant odour. She had great delicacy of constitution and appearance; and her manner was so timid, that she was thought by some to be imbecile. This, however, was the result of a neglected education. Menstruation was not yet established, although there were indications of its approach. She had never perspired. A tonic plan of treatment was resorted to, and under proper care she improved.†

* Good's Study of Medicine, vol. iv. p. 531.

† London Medical Gazette, vol. xli. p. 336.