GLIOMATOUS ENLARGEMENT OF THE PONS VAROLII IN CHILDREN.

BY

ANGEL MONEY, M.D.,

FELLOW OF UNIVERSITY COLLEGE, LONDON; REGISTRAR TO THE HOSPITAL
FOR SICK CHILDREN.

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I WISH to place on record two more cases of “gelatini-form enlargement of the pons Varolii.” Readers of the thirteenth and seventeenth volumes of the ‘St. Bartholomew’s Hospital Reports’ will find a case of this disease described in each volume—one by Dr. Percy Kidd and the other by Dr. Gee, though both were under the care of the latter physician during life. I had the good fortune to be charged with the microscopical examination of the specimen, of which the clinical account may be found in the seventeenth volume of the ‘Reports.’ Concerning the affection Dr. Gee wrote thus: “So far as I know, the disease has been hitherto undescribed.” This statement is the chief cause of the appearance of the present paper.

My special thanks are due to Dr. Gee for primarily directing my attention to the disease, which I need hardly say was entirely new to me.

CASE 1.—Alfred James B—, æt. 11, was admitted into the Hospital for Sick Children in November, 1873, under the care of Dr. Dickinson. His illness began three months before admission, when it was noticed that the head was held to the left side, that there was loss of control over the left foot, that the left hand dropped; he had vomited especially in the morning, had grown dull, and walked badly. The bowels were said to have been regular. There was some complaint of headache at the postero-dextral region. There had been otorrhœa on the right side since the child was six months old. The patient had had a cut on the head four years ago. The father's grandmother died insane. No alcoholism. He was one of ten children, of whom two had epileptic fits, one died of "brain fever," and one died of "fever," and had otorrhœa. There seemed to be no evidence of struma, syphilis, or tubercle. The patient was said to have always been excitable.

State on admission.—Well nourished. Holds head to left shoulder. Deaf of right ear from which a discharge of matter issues. Can stand and walk, but drags left leg. Grasp of left hand weaker than right. Sensation diminished in left arm and leg. The left calf measures 9 inches, the right $9\frac{1}{2}$ inches. The bowels and bladder act naturally. Bowels are confined. Vomiting. Left pupil dilated and sight is worse in the left eye. Ocular movements perfect and equal. Sensation and motion of face perfect. Tongue protruded in mid line.

On November 14th the patient was more drowsy and the intellect was duller.

21st.—Complains of feet feeling cold, especially of the left.

27th.—Pain in back of head and giddiness were noted.

28th.—Giddy but no headache.

December 7th.—Left optic disc too pink, outline distinct, vessels not very tortuous nor dilated; right optic disc natural.

26th.—Further loss of power in left arm and leg, cannot draw left leg up to abdomen without the help of his hand. Grasp of left hand much weaker than right. Other symptoms continue.

January 15th.—Has lost all power in left hand ; can only move left arm a little ; thumb is turned into palm, and fingers are flexed thereon. Only slight voluntary power over left leg. Food lodges between gums and left cheek ; there is constant dribbling.

February 4th.—Face often flushes. Cannot swallow so well. Speaks indistinctly. Abdomen retracted.

6th.—*So weak and faint* that death was hourly expected. Mucus on both eyeballs ; takes but little notice when shouted at. Left arm and leg are rigid. Left ear discharging. Sordes on teeth. Dirties and wets bed.

10th.—Bedsore on left ischial tuberosity. Unconsciousness. Less otorrhœa.

12th.—More conscious. More otorrhœa.

24th.—Conscious. Bowels not open for eighteen days.

March 11.—Boy continues to be sensible. No action of bowels for thirty-five days.

24th.—Left arm rigid and helpless. Right arm less rigid (no previous note of rigidity here), some power in it ; can move right but not left leg.

29th.—Death.

Post-mortem (thirty-five hours).—Weight of brain 45½ ounces. No sign of inflammation anywhere in cranium. There was enormous expansion of the pons Varolii and crura cerebri. The “bridge” measured 2 inches from side to side, 1½ inches from before back, or about half as much again as natural for that age. Dr. Dickinson said that in shape it was what mineralogists call “botryoidal,” and resembled in colour, &c., “chalcedony ;” further, it looked like a “soft package tightly corded.” The depressions corresponding to blood-vessels. The basilar artery lay in a deep furrow. There was a great prominence at the anterior part of the left side of the pons, behind the root of the third nerve ; being greatest between the third

and the fifth nerve. The cerebral peduncles were swollen, the right the more; also both cerebellar peduncles. Both third nerves were much flattened. The medulla oblongata was natural. The tuba cinereum expanded. The arachnoid on the convexity of both hemispheres was thickened and opaque. There was about four ounces of fluid in the lateral ventricles. Some pneumonia of the right lung. The spinal cord was natural.

For the use of the above notes my thanks are due to Dr. Dickinson. As the microscopical examination is precisely like that found in the other cases, a single description will serve for all, and this will be given later. A drawing made by Dr. Westmacott gives a fair representation of the pons and neighbourhood of this case (see Plate IX).

CASE 2.—Emily H—, æt. 6½, admitted into the Hospital for Sick Children on December 18th, 1882, under the care of Dr. Cheadle, who has kindly given me permission to make use of the case. Her illness began about four weeks before entry, with occasional vomiting, with headache, and much giddiness, and some degree of languor; the gait had been noticed to be staggering for the space of three weeks, and a squint had been observed for two weeks. She is said to have been "convulsed." She was a studious child. No cause could be given for the illness. The patient had had measles at the age of three years, this was followed by whooping cough, and at the age of four years and a half scarlet fever was said to have affected the patient. She had never had teething convulsions. The family history showed that there had been eight children with two miscarriages of no special relation to one another, and not in any way suggestive of syphilis. There were seven children living, the seventh out of eight had died from "effusion into brain" at the age of eleven months. Emily H— was the sixth child. Some of the children had thrush for two weeks from birth, but no snuffles. There was no history of rickets or consumption; some of the children had had an abscess in the neck.

The father's brother was supposed to have died of consumption; he was a soldier. The father had never been in the army or navy and denied all forms of venereal disease. The mother has been and is still suffering from "rheumatic iritis" for four years.

On December 21st the present state was taken, the patient having been watched since entry. The girl was rather drowsy, did not play, and remained quietly lying down all day. She had vomited once and thrice on the 19th and 20th respectively. The temperature was 99.2° in the evenings (about 6 p.m.) and about 98.4° in the mornings (about 10 a.m.). Pulse 132, regular (several observations). Breathing 24, regular. Urine acid, no albuminuria, no glycosuria, no increased frequency of micturition, the amount, however, was not measured. Bowels constipated, not open for two days. The belly was not full, the lower edge of the spleen was palpable.

Sensation and special senses were apparently natural, but this examination was difficult to carry out owing to the patient's impaired mental state. The speech was decidedly altered, being of a spasmodic character. Walking was just possible with support, but the gait was very jogging and unsteady; the patient could not stand alone. There was a convergent squint; the eyeballs moved about in every direction except outwards. There was probably paralysis of both external recti. The pupils were of medium size, equal, and acted, but whether to accommodation as well as to light could not be ascertained. Mr. Marcus Gunn detected some abnormality in the shape of a spot situate to the inside and rather above the right optic disc; there was a little hypermetropia, no optic neuritis. The muscles of mastication seemed to act naturally. There was no paresis of either half of the face. The tongue was protruded fairly well and symmetrically, had many large papillæ, and was rather dry. The patient had a stupid look, the mouth was half open, there was no dribbling of saliva. When the patient sat up in bed, which act she was just capable of performing, a coarse

indefinite tremor was observed to affect the trunk and head. Micturition and defæcation were performed without calling for the nurse. The knee phenomena were present and equal, and easily obtained. Nothing definite could be learnt from the superficial reflexes, the patient became so restless under examination. Nothing abnormal made out in the heart or lungs.

December 23rd.—Pulse 108 and regular.

26th.—The temperature was 100.6° on the night of the 21st, otherwise it had not been above the normal. The notes then say that patient got quite excited at times, and that she had narrated her experiences of the performances of the Salvation Army; that, although drowsy and quiet for the most part, she was communicative at times, the speech being jerky and peculiar. There had been no fresh vomiting.

29th.—All the symptoms have continued; the edges of the optic discs are obscure, nothing wrong noticed in the vessels of the fundus oculi. The knee phenomena are certainly excessive this morning, but no ankle clonus could be got. There has been vomiting the last three days.

30th.—The patient got very drowsy about 4 p.m. The respiration began to be very slow, whilst the heart was going fairly well. The pulse was 84, and regular. Much vomiting. She had to be fed through the nose. The coma endured and the breathing failed at 1 a.m., December 31st.

Autopsy (twelve hours after death).—Rigor mortis everywhere present. There were no signs of rickets, syphilis, or struma. The meninges and large vessels were natural. The pons Varolii was enlarged in all its dimensions, the left side perhaps more than the right, the cerebellar peduncles were enlarged, especially the left one; the basilar artery lies in the valley formed by the overgrowth of each half of the pons.

The greatest width	was	$2\frac{1}{4}$	inches.
„	length	„ $1\frac{1}{2}$	„
„	thickness	„ $1\frac{1}{4}$	„

The left cerebral peduncle was perhaps a little enlarged. Both the third nerves were pressed upon. The fifth left nerve was enlarged at its superficial root. The medulla oblongata seemed swollen, but its parts were recognisable. The floor of the third ventricle was convex and too prominent. The sixth and seventh nerves were involved at their superficial origins. The other nerves were apparently free from disease at their superficial roots. Sections show the pons to be diseased right through its substance, and in the centre there was a grey-red area. The whole of the floor of the fourth ventricle seemed to be affected with the disease. There were three flame-shaped hæmorrhages in the right retina, disposed in a radiating manner in the neighbourhood of the optic disc; they were not more than three lines long. Both optic discs had a swollen look, the vessels were not abnormal. There were no hæmorrhages into the left retina. Both internal ears were natural. There were genuine ecchymoses, not mere hyperæmia, in the gastric and intestinal mucous membranes, a few patches in the small bowel and one in the cæcum. The kidneys were sanguinolent, and their consistence was increased, they weighed $4\frac{1}{2}$ oz. together. The spleen was flabby, weighed $3\frac{1}{4}$ oz., had very abundant white areas, "Malpighian corpuscles," so that the intervening red tissue formed only a fine network as seen on section. The liver weighed 21 oz., was full of blood, and had its consistence somewhat increased. None of the above-mentioned organs gave any reaction with iodine. No tubercle was found anywhere. The other organs, including the lymphatic glands, were all natural. Some urine, tested from the bladder, was acid, had no albumin, and no sugar.

Microscopical characters.—The most obvious change is a small-celled infiltration; sections from the thick part of the tumour consist of very little else than cells. The shape of these units is various, they are mostly round or oval, sometimes reniform. The alteration of the brain at this part is so great that it is impossible to recognise any

normal structure. A stained section examined with the naked eye by transmitted light shows certain markings, which in their general arrangement recalls the naked-eye appearance of the pons. Such a recognition of normal configuration is very striking. This replacement of healthy structure by diseased tissues without much alteration of the macroscopic appearance is a fact of not unfrequent occurrence, not only in the brain but in other organs. The normal anatomy draws the lines along which future disease must be built. This may be called the "principle of the preservation of pattern." The neuroglia and natural nerve-fibres are not recognisable. There is an intercellular substance of homogeneous appearance which does not become black when treated with osmic acid. I conclude from this that normal medullated fibres are not present; though the presence of axis cylinders is of course not denied. No nerve-cells are to be detected where the growth is most abundant, but in places a ganglion cell very little changed from the healthy appearance may be discovered. There is nothing to be said of the state of the blood-vessels. There are no extravasations.

My thanks are due to Dr. Gowers for having examined my sections. He fully agreed with my interpretation of them.

Remarks.—It seems certain that till now little or nothing has been known of such a disease as nearly symmetrical gliomatous enlargement of the pons Varolii by the majority of physicians. The evidence for that opinion consists in the facts that Dr. Dickinson described his example as one of "peculiar hypertrophy," &c., and that Dr. Gee recorded his cases and wrote of them in the sentence which has already been quoted. Further, compared with my own sections, Dr. Percy Kidd's specimens, which he has kindly given me the opportunity of examining, are undoubted samples of glioma, and thus there should be no necessity for the continuance of the name 'gelatiniform enlargement of the pons.' That phrase has served its purpose

and may now be eliminated from medical nomenclature. The macroscopical aspect of the affection is by far the most striking and interesting feature, and for this reason a plate taken from a drawing by Dr. Westmacott has been added to this paper (see Plate IX). With regard even to the coarse etiology, we are very much in the dark. It would appear that traumatism has been observed to precede the appearance of the disease in the pons as of other parts of the brain, but the influence of injury in the causation must be a very difficult point to settle. The disease seems to have its starting-point in the neuroglia, and hence we can partly understand the insidious mode of its clinical commencement and progress. The signs during life are frequently more marked on one side of the body; this was so in the first case here recorded. Gee, Kümmell, and Leyden have reported similar cases ('St. Barth. Hosp. Rep,' xvii, and 'Zeitschrift für Klin. Mediz.,' Band. ii, S. 282, 1881).

I have nothing to add to what is already known about the pathological and regional diagnosis. Polyuria has been observed but never albuminuria or glycosuria, nor has any sudden elevation of the temperature of the body been recorded, such changes are more likely to occur in acute affections of the pons (see Gowers in second volume of 'Brain,' p. 466). Though it is conceivable that a hæmorrhage might cause sudden pyrexia or glycosuria. And hæmorrhages are liable to occur in such gliomata.

After writing the above, my attention was directed by Dr. Gowers to a paper by Schulz in the 'Neurologisches Centralblatt' for January, 1883. This article narrates a case of the disease in a man, æt. 32, which is precisely similar to my cases in the symmetrical enlargement of the pons and other characters. Schulz has also collected all the cases recorded of glioma of the pons, whether localised or diffuse; the majority occurred in children not over the age of twelve. Lastly, Schulz boldly records Kidd's

case (' St. Barth. Hosp. Rep.,' xiii) as a sample of glioma, though it is probable that he has never seen Dr. Kidd's specimens.

DESCRIPTION OF PLATE IX.

(Gliomatous Enlargement of the Pons Varolii, ANGEL MONEY, M.D.)