A CASE OF IDIOCY WITH UNIVERSAL RIGIDITY, THE RESULT OF SYPHILITIC DISEASE OF THE CENTRAL NERVOUS SYSTEM.

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A. R. H. a female child, aged $3\frac{1}{2}$ years, was admitted into the Hospital for Sick Children, under the care of Dr. Dickinson, on April 3, 1883. She was an illegitimate child; her mother died of consumption, aged 31. The child had been bottle-fed all through. She was in good health till two years old, when she had two "fits" (? bilateral) in one afternoon, which were attributed to the teething. Three days after these fits, the child is said to have become unconscious. Since then she had not recognised friends, had not spoken, nor walked; though before the fits she was intelligent, could speak and had walked well. The bowels had been constipated. There was no vomiting.

On admission, the gums were spongy and swollen; the bowels were freely opened after a purgative; the patient vomited three times without apparent cause; the temperature was 104 at night, and 101 in the morning; the corneæ were clear; the discoloured teeth were ground down level with the gums in the upper jaw, and projected a little beyond the gums in the lower jaw. The upper part of the right external ear stood away from the side of the head. Unless the optic discs were too pale, ophthalmoscopic examination revealed nothing abnormal. The eyeballs were moved about thoroughly in every direction; they had no characteristic position; there was no facial palsy in crying; the tongue seemed to move naturally and equally. There was practically no intellect. The child seemed to observe a bright lamp, cried when food was put into the mouth; deglutition was natural. The mouth was very large, the face seemed ill-developed; the head was brachycephalic and small; its circumference measured $18\frac{1}{2}$ in.; the measurement from ear to ear over the cranial vault was $12\frac{1}{2}$ in. The legs, arms and trunk, when left alone and at rest,

were not stiff, or but very little so; the slightest disturbance however (sometimes the same thing seemed to occur apparently spontaneously) developed a marked tonic spasmodic condition of all the limbs, with opisthotonos of all parts of the spine, cervical, dorsal, but especially lumbar. The legs were straight at all the joints. The arms were rotated in and adducted at the shoulder; were extended at the elbow, but not fully; pronated in the forearm; the thumb was turned into the palm and the fingers flexed thereon, the wrists were semi-flexed. This posture of parts was constant, fixation occurring when spasm came on; the onset did not recall a spasm the result of strychnia poisoning or tetanus, principally in the want of suddenness, but perhaps there might be less difference between them than was supposed. The knee-phenomenon could be got on each side without difficulty; the ankle clonus could not be obtained, on account of the rigidity.

April 13th.—The patient was in much the same state, but the gums were practically well. The temperature had been below the normal since April 8th.

April 16th.—The patient swallowed badly, and sometimes did not attempt to swallow at all. The food regurgitated through the nose. Vomiting had been frequent.

April 17th.—The patient swallowed better in the night, but this morning she was worse again, and a muco-purulent discharge was flowing from the mouth constantly; there was no cough nor vomiting; the muco-pus bubbled up in the mouth. The bowels were constipated; a nutrient enema was given, which returned in half an hour. The voice became very weak last evening, so that she had not screamed when disturbed; this was a new feature. The back and neck were much more arched.

April 18th.—Excessive pallor appeared at 6 A.M., and was universal and ghastly. There was marked collapse. The patient was pulseless, the surface cold, the breathing shallow and actually ceased at times; the lips were bluish. The rigidity had more or less disappeared when the patient was disturbed. The hands were clenched, but could be undone; the fingers were flexed, the thumbs turned, the arms adducted, the elbows gently flexed, the forearms over-pronated; the legs were extended at all the joints; there was conjugate deviation of both eyes to the right. Death took place at 7 P.M.

Autopsy.—The weight of the body was 17½ lbs. The limbs were stiffened in the posture described during life. The cranium was asymmetrical, the left fronto-temporal region sloping, and not so round and full as the right. The frontals and parietals were 7 mm. thick at the thickest parts; the diploë was dense; the fontanelles were closed, and the sutures were united, but

there were no bosses. The dura mater was decidedly thickened, especially about the walls of the sinuses; there was nothing abnormal in the venous sinuses; there was much oedema of the subarachnoid space; irregular opacity and thickening of the pia mater were observed chiefly over the sulci; the pacchionian bodies were fairly marked; the brain was atrophied, especially its left hemisphere; its walls in the lateral and posterior parts were quite thin; the convolutions for the most part were picked out and well defined; many of them had a yellowish hue and were quite tough. The walls of the basilar artery were opaque white, thickened, and contained red tough thrombus. Nearly all the pia-matral vessels were thrombosed and their walls diseased. Nearly all the large arteries had thickened and opaque walls; long lengths of vessels were thus diseased; the whole of the basilar and the terminations of the vertebrals were very conspicuously diseased, and by far the greatest part of the length of the vessels forming the circle of Willis was diseased. The orbital plates crushed rather than fractured with the chisel, their diploë was thick and vascular. There was nothing abnormal in the choroids or optic discs. The orbital fat on both sides was dotted with points of hæmorrhage. The middle ears contained muco-pus, but the membranes were not perforated.

The pons Varolii was very hard. The cerebellum was perhaps a little tougher than natural; each half was equal, and of about the same size and shape. The medulla and part of the cervical cord stand out like a stick, before being put into spirit. All the spinal cord felt hard; on section, the anterior horns in places looked puckered and indented.

The heart weighed 2 oz., was healthy, as was also the pericardium. The larynx had a slight shaggy appearance, and red specks were dotted about, but nothing more definite was seen.

The left lung was adherent nearly all over, its weight was $3\frac{1}{2}$ oz. An area of consolidation, of ill-defined outline, of a dark dirty-red colour mottled with white, and of diminished consistence, was situated in the middle of the left upper lobe.

The right lung weighed $4\frac{1}{2}$ oz.; the upper posterior part of the right lower lobe, and the back half of the upper lobe were consolidated, of dirty dark-red colour, mottled with white, of ill-defined outline, and of diminished consistence. The bronchial tubes were all dilated, in one place having the finger-glove look; there was much mucus in the tubes. In the large intestines only the first six inches showed the lymphoid structures to be of a yellow tint, of a circular or oval well-defined outline, with a central dimple. None measured more than $3\frac{1}{2}$ millimetres across, many were smaller.

The liver weighed 12 oz., was of a dark red colour, and of

diminished consistence; there were no gummata. The pancreas was healthy. The spleen was big, of a dark red colour, weighed 2 oz., its consistence was not increased. The bronchial glands were enlarged; there were no cheesy glands anywhere. The kidneys weighed $2\frac{1}{2}$ oz. together. The bladder was healthy. The muscles appeared to be a little pale.

Microscopical Examination.—Sections of the spinal cord show a diffuse form of sclerosis throughout the whole of its length. The cerebral substance has not been microscopically examined, but it may be assumed that extensive sclerosis is present in those convolutions which felt hard at the autopsy. Many sections made from the basilar and middle cerebral arteries exhibit well-marked arteritis, chiefly of the inner coat; in some sections the chief thickening is between the corrugate elastic lamina and the outer coat. Complete microscopical examination has not been made, owing to the fact that the specimen was kept in spirit to preserve its naked-eye appearances, and the spirit has now so altered the texture of the brain and cord as to render a complete examination impossible.

Remarks.—The case is of interest, as an example of the production of idiocy by coarse syphilitic lesions of the brain. In my own experience, which is limited to four post-mortem examinations of children idiotic from or soon after birth, some gross organic lesion has been present, and was in all probability the cause of the amentia. In one case of microcephalus I assisted at the autopsy with Dr. T. Barlow, and the cerebral hemispheres were, roughly speaking, converted into thin walled cysts over the greater part of their extent. In my second case there was a tumour, apparently gummatous, the size of a Tangerine orange in the posterior region of the right hemisphere. The third example was one in which there was great atrophy of the right hemisphere, which was probably due to hæmorrhage, the result of instrumental delivery.¹ The last instance is the one here recorded. All the specimens are in the museum of the Hospital for Sick Children, Great Ormond Street.

My clinical experience at the hospital last mentioned leads me to believe that mental changes are by no means uncommonly produced by hereditary syphilis. I have met with children in whom the evidences of past syphilis were unmistakable, and in whom the mental condition was obviously not up to the average. This subject has been drawn attention to by an excellent contribution to 'BRAIN,' from the hand of Mr. Judson Sykes Bury, of Manchester.

¹ 'Transactions of Clinical Society,' 1888.