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Three Cases of Juvenile General Paralysis.

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In the following brief notes nothing is attempted beyond placing on record three undoubted examples of this interesting disease, and thus adding to an ever increasing list of cases.

CASE 1.—F. S—, æt. 20, general servant.

History.—Father and mother drunkards; in poor circumstances. Patient fourth child of family of five; elder children reported healthy, but younger sister mentally deficient. Patient was undoubtedly syphilitic in early childhood; had snuffles. Was always delicate; of average intelligence at school.

On admission, September 29th, 1905, she measured 4 ft. 9 in. Bridge of nose slightly depressed; marked cicatrices running at right angles to circumference of mouth. Teeth irregular but not characteristically syphilitic. Mammæ rudimentary.

The mental condition on admission was one of slight melancholia patient emotional, lachrymose; answers to questions irrelevant. Speech distinctly affected; some words stopped short and some run together; fibrillar twitching of facial muscles; tongue tremulous. Knee-jerks very active; plantar reflexes normal and equal. Pupils equal and react normally to light and accommodation.

Patient speedily recovered from depression and became irritable and peevish; did ward work; clean and tidy in her habits. After six months she became less observant, and was listless, apathetic, and idle. Gait was now affected; patient walked in an ataxic, swaying manner; legs dragged after her and fell with feet extended and wide apart. Mental condition one of increasing dementia; ultimately unable to answer the simplest questions, and during the last three months of life

https://doi.org/10.1192/bjp.54.224.112 Published online by Cambridge University Press

she was too demented and paralysed to speak at all. Fourteen days before death, and seventeen months after admission, she became convulsed; remained unconscious; had thirty-six seizures in all. Condition after first convulsion: pupils dilated, equal, no reaction to light. Patellar reflexes completely absent; well-marked bilateral Babinski sign. Shortly before death the legs became œdematous, and there were indications of gangrene of lungs.

Post-mortem examination.—Body emaciated. Hair poorly developed over pubis and in axillæ. Skull-cap adherent to dura mater, which is dense and thickened; many Pacchionian bodies, especially along the sides of the longitudinal fissure. Pia mater opaque; excess of subarachnoid fluid. On stripping off the pia the surface of the convolutions appeared markedly eroded, especially over parietal and temporal lobes. Lateral ventricles much dilated with clear fluid; no granulation of lining membrane, but abundant granulations in fourth ventricle. Convolutions atrophied; the grey matter markedly diminished. Cerebral substance generally soft. Basal arteries sclerosed. Liver fatty; capsule thickened; old cicatrix on under surface of left lobe. Deep notch in anterior border of right lobe. Kidneys show cicatrices on surface. Heart normal. Lungs contain scattered tubercular foci with large gangrenous abscess; cavity in right lower lobe. Uterus rudimentary. Encephalon = 1030 grms. Right hemisphere (undissected) 400 grms.; left (dissected) 410 grms. Cerebellum = 110 grms. Pons = 15 grms.

Case 2.—E. M—, æt. 15, of no occupation; admitted October 27th, 1899, from a home for waifs. Her mother reported to be a prostitute. No further history obtainable.

On admission.—She has the appearance of a child of ten; bridge of nose depressed, nostrils wide, head large, forehead prominent. Typical cicatrices at angles of mouth. Teeth irregular, not typically syphilitic. Palate highly arched. Speech and gait normal. Patellar reflexes increased; active plantar response. Pupils dilated, unequal; right larger than left; reaction to light and accommodation normal. Mental state one of extreme hilarity, mild excitement and restlessness with continuous laughter and incoherent chattering. Ten days after admission she lost power of legs; quite unable to walk; no seizure noticed. The reflexes remain unaltered, but speech distinctly affected; she cannot pronounce "artillery" and similar test words; distinct hesitancy and slurring. Tongue tremulous, marked twitching in lips and facial muscles. After a short stage of what resembled stupor, she again became happy and intelligent; remained in that condition for the next five years. During this period she once had retention of urine. In February, 1900, she had an acute attack of broncho-pneumonia. Had an attack of interstitial keratitis, which left permanent adhesions to lens in each eye. Menstruated regularly from admission till three months before death; copious losses. Body-weight increased, but stature remained unchanged. During last two years of life she gradually became more and more demented; not so hilarious, but quiet, reserved, demented, and worked but little and in a listless manner. Three months before death she again lost power in legs. Speech, which had gradually become

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typical of ordinary general paralysis, now entirely lost, and patient quite demented, understanding very little of what was said to her. Patellar reflex increased. Marked bilateral Babinski phenomenon; Argyll-Robertson pupils; no ankle clonus. Required catheterisation. Legs became œdematous. Occasionally vomited bilious matter. Developed a bulla over sacrum which disappeared before death.

Post-mortem examination.—Body well nourished. Hair abundant over pubes, but scanty in axillæ. Skull cap adherent to dura mater, which is thickened and dense. Pia opaque. Marked excess of sub-arachnoid fluid, also of cerebro-spinal fluid. Erosions of middle temporal and middle frontal convolutions on each side. Lateral ventricles enormously dilated. Abundant fine granulations on floor of ventricles. Cerebellum atrophied. Floor of fourth ventricle covered by coarse granulations. Degenerated gummatous mass attached to pia in Sylvian fissure. Arteries, especially basal, thickened. Liver shows cicatrix on under surface of right lobe; capsule thickened throughout. Uterus small, as are all the abdominal organs. Encephalon = 1075 grms.; right hemisphere = 430 grms.; left = 400 grms.; cerebellum = 56 grms.; pons = 14 grms.

Case 3.—T. M—, æt. 14, of no occupation. Third child of parents in good circumstances. Other members of family healthy. A reliable history difficult to obtain. Patient stated to have been a bright, intelligent child until about twelve months before admission. She then became irritable, peevish, erotic.

On admission, November 4th, 1905, she was passionate, declamatory; habits faulty. Height 5 ft. 4 in. Weight 4 st. 11 lb. Bridge of nose depressed. Teeth very irregular. Gait normal. Speech hesitating and indistinct, complete words being occasionally dropped out. Plantar and patellar reflexes normal. Pupil reflex not noted. Patient continued noisy and irritable for two months when progressive dementia developed. Bodily condition degenerated. Gait ataxic. Body bent forward, head extended. Menstruation regular; the loss very small. About middle of November (?) she began to have attacks of vomiting; bilious and liquid material ejected without effort. Makes inco-ordinated attempts to scratch when interfered with. Cannot articulate at all; points at articles and vainly attempts the pronunciation of their names. October 29th, 1906. General condition very feeble; extremities cold, livid. Pupils unequal; right dilated. Active patellar response. Marked bilateral Babinski's sign with flexion at knee-joint. Urine retained. On November 3rd patient died after twenty-four hours of unconsciousness, but no kind of "seizure" was observed.

Post-mortem examination.—Body well nourished. Legs œdematous. Hair on pubis and in axillæ poorly developed. Skull cap adherent to dura. Pia thickened, opaque, and adherent to tips of convolutions except throughout the posterior portion of occipital lobe on each side. Lateral ventricles dilated. Small circular sclerotic patch in outer wall of left ventricle. No granulations on lining membrane of lateral ventricles. Grey matter diminished, soft and dark. White matter pultaceous. Cerebellum normal. Floor of fourth ventricle covered by exuberant granulations—a typical example of "cat's tongue." Excess of cerebro-

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spinal fluid. Encephalon = 970 grms.; right hemisphere = 395 grms.; left = 370 grms. Cerebellum = 150 grms. Pons = 10 grms. Other organs showed no pathological change.

Remarks.—When Clouston, Mott and others recorded their cases of adolescent general paralysis, they drew attention to all the points that receive attention from us as practical physicians. Anyone acquainted with the literature of the subject must at once perceive how the cases now detailed emphasise the conclusions arrived at by former observers. In relation to the age at which the disease usually appears, there is one point which has attracted some attention, but about which nothing definite has been decided; I refer to the age limit beyond which inherited syphilis does not appear to be able to produce general paralysis. Yet this is really an important question, which when solved may account for those cases of the disease in which a history of primary syphilitic infection cannot be obtained.

In two of the cases the stature was markedly under the average; the patients appeared more like children, and this juvenile appearance was retained to the end; more especially was this true as regards the facial expression. Though some of the sexual characteristics were poorly developed, this was not so in Case 2, who was a developed woman, though in miniature, and in her the monthly periods were quite normal.

As already stated it is not my intention to use these three cases as a text and to proceed to discourse on all the points of interest arising from a consideration of general paralysis. Still a word may be said as to the diagnosis of these cases. Until Clouston recognised their true character they were considered to be instances of dementia, but now asylum physicians have no difficulty in diagnosing them. It is otherwise with the ordinary practitioner. It is only rarely that he arrives at a correct diagnosis. Sometimes the mistakes are quite remarkable, but only one need be referred to—that of a girl, æt. 18, in a refuge. Although the history of the case was defective, it would surely have been possible to avoid the error of considering her a deaf and dumb idiot, when she really was a case of far advanced general paralysis.

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