Short Notes and Clinical Cases.

A NOTE ON THE PRESENCE OF ENDARTERITIS OBLITERANS IN THE BRAIN OF A MONGOLIAN IMBECILE.

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THE hypothesis that mongolism is a special form of congenital syphilis appears to be gaining ground, particularly in France, but it must be admitted that the pathological evidence for such a view remains extremely scanty. Indeed, with the solitary exception of a report by Babonneix¹ syphilitic lesions in the brain of the mongol do not appear to have been recorded. In the case of Babonneix, in addition to vascular and meningeal changes, a small tumour having the histological features of a gumma was found in the sulcus separating the right prefrontal and postfrontal gyri. To this case I am now able to add a second in which the outstanding feature was a syphilitic affection of the cerebral vessels.

CASE HISTORY.

The patient, an imbecile, age 14, was admitted to Leavesden Mental Hospital on April 28, 1923.

Family History: Both parents had always enjoyed good health and the mother was unable to assign any cause for the birth of a mongol. She had no worries or frights during pregnancy, and no miscarriages or still-births. The family consisted of four children, of whom the patient was the youngest. At the time of his conception the mother was 41 years of age, the father 39 years. The mother came of a long-lived family and on neither side was there any history of insanity, mental deficiency, or epilepsy.

State on admission: A restless mischievous imbecile, unable to dress or wash himself without assistance. Mental age 4 years. Height 4 ft. 2½ in. Weight 3 st. 12½ lbs.

Stated briefly, the signs of mongolism shown by this patient were as follows:—

Head: circumference 19½ in.; small, rounded, and bradycephalic. Palpebral fissures oblique: long eyelashes. Eyes brown: pupils slightly eccentric with normal reactions: fixation-nystagmus.

Ears: small, rounded and hollowed-out.

Nose: short, squat, with triangular-shaped nostrils directed forward.

Cheeks: bright red flush. Well-formed rectilinear eyebrows.

Tongue: not unduly large but fissured in an irregular manner: hypertrophied circumvallate papillæ. Mouth habitually open. Articulation indistinct.

Teeth: carious, irregular and crowded together. Palate high and narrow. Tonsils enlarged. Hair on scalp dry and scanty.

Hands : soft and flabby ; incurring of right minimus. Left little finger short and thick with rudimentary nail.

Feet : broad and flat. Large cleft between first and second toes. Plantar line.

Abdomen : large and tumid ; umbilical hernia.

Circulation : blueness and coldness of extremities with cuticular mottling ; chilblains ? No valvular disease of heart. Blood pressure : systolic, 84 mm. ; diastolic, 60 mm.

General muscular hypotonia. Sexual development, infantile. Reflexes, normal. Habits, occasionally faulty. Wassermann reaction in the blood, negative. Spinal fluid not examined.

Subsequent History : On the afternoon of February 21, 1926, the patient had a seizure, unattended by loss of consciousness, and within a few hours the right face, arm, and leg became completely paralysed. There was also loss of speech. Temperature, 97° F. ; pulse 68 ; respiration 18.

When seen by me on February 23 there was no facial paralysis but the tongue deviated slightly to the right. There was marked hippus, and complete flaccid paralysis of the right upper limb ; the right lower limb was markedly paresed and jerking of the tendon of peroneus tertius was observed. The patient moved the left upper limb freely but with a certain amount of coarse tremor. The left lower limb was unaffected. Sensory functions could not be tested. Reflexes : right abdominal diminished ; left, brisk. Plantars : right, extensor response ; left, absent. Knee and ankle jerks exaggerated ; ankle clonus on right side. No loss of sphincter control.

Voluntary movement returned gradually in the paralysed limb and after the lapse of two months the patient became able to grasp objects very feebly with the right hand and to hold the limb off the bed for a few seconds. The right lower limb showed a considerable return of power, but was unable to support the weight of his body. When examined at the end of three months both extremities showed considerable increase of tone and the right upper limb had assumed the ordinary hemiplegic attitude of adduction, flexion at the elbow and pronation. There was no return of motor speech.

On June 22 the patient acquired bacillary dysentery, to which he succumbed on the eleventh day.

Post-mortem Examination : An autopsy was held eleven hours after death.

The body was that of an undeveloped youth in a poor state of nutrition. Rigor mortis and post-mortem lividity were well marked.

The right upper limb showed contracture at the elbow and the lower limb slight flexure at the knee.

The skull cap was thin and the dura mater adherent to its inner table. The brain weighed 36 ozs. and showed an extensive area of softening in the fore part of the left cerebral hemisphere.

The heart weighed 5 ozs. Myocardium firm and dark red in colour. The mitral valve segments showed a moderate degree of thickening at their free edges but no narrowing of the orifice or recent vegetations. The aorta and larger arteries appeared normal.

Lungs : right, 11 ozs. ; left, 13 ozs. No adhesions or evidence of tubercle ; congestion of both lower lobes. Peritoneal cavity distended by free gas. Liver 31 ozs. Reduced in size with slight fatty infiltration.

Spleen : 5 ozs. Large, tough and fibrous ; dark red in colour.

Kidneys : right, 2½ ozs. ; left, 3 ozs. Both reduced in size ; capsule thickened and adherent, leaving a slightly granular surface when removed. Cortex narrowed and irregular.

Small intestines : matted together.

Large intestine : much thickening and congestion of mucous membrane at its lower end with numerous large ulcers of irregular shape. One ulcer showed a small perforation.

Examination of Brain : The pia mater was thickened and firmly adherent to the softened convolutions. The cerebral hemispheres were rather rounded, the convolution pattern simple, and the superior temporal convolutions unduly small. The brainstem and cerebellum appeared normal.

The left cerebral hemisphere showed an extensive softening involving all the prefrontal convolutions, the superior, middle, and inferior frontal gyri, the pre- and post-central gyri, except in their lower thirds, part of the superior parietal gyrus, and the gyrus cinguli in its anterior half. The affected convolutions were shrunken, angular, and of varying consistence, some being firmer and some softer than normal brain tissue.

On cutting through the middle of the softened area in the vertical coronal plane it was seen that while the grey matter of the affected convolutions was completely destroyed, the underlying white matter had largely escaped. A small cavity was present beneath the upper end of the postcentral convolution; elsewhere in the affected region the medullary substance was firm and greyish white in colour.

The basal ganglia were normal.

With the exception of a very small cortical softening situated at the upper extremity of the right postcentral gyrus, the right cerebral hemisphere showed no gross lesion.

Vessels : the basilar and vertebral arteries were both a little dilated and thickened, but the most obvious departure from the normal concerned the anterior and middle cerebral arteries. These were tortuous, irregular in their diameter and of an almost pearly white colour (Fig. 1). They felt extremely firm and when cut across showed a marked narrowing of the lumen. The middle cerebrals were most affected and that on the right side had its lumen almost completely blocked.

Microscopic Examination : A study of the transverse sections of the left middle cerebral artery shows the presence of a marked degree of endarteritis proliferans (Fig. 2.) The inner coat is much thickened by numerous concentric laminae of fibrous tissue and has narrowed the lumen of the vessel to a considerable extent. In this tissue, which has a fenestrated appearance in certain areas, thin-walled capillaries can be seen. With toluidin blue and azur II the intima stains metachromatically. The internal elastic lamina has undergone new formation and splitting. The middle coat shows little abnormality and proliferative changes in the outer coat are slight. There is little evidence of cellular infiltration in any part of the vessel, the general appearance of the vessel suggesting a slowly developed degeneration of some duration.

The same appearances can also be seen in the minute terminal branches of this artery.

Brain Cortex : At the moment of writing it has only been found possible to examine the cerebral cortex of one area, namely, the right prefrontal region. No changes of a syphilitic character can be found. The pia mater is slightly thickened and oedematous, the nerve-cells show numerical deficiency and immaturity and the larger vessels a slight degree of hyaline-fibroid degeneration.

DISCUSSION.

As malformations of the heart are not uncommon in the mongol it was at first thought that the paralysis in this case might be due to an embolism of the left middle cerebral artery, but the mode of onset of the hemiplegia and the absence of signs of valvular disease militated against the acceptance of this view. The gradual development of paralysis without loss of consciousness was much more in favour of a diagnosis of cerebral thrombosis; on the other hand, the age of the patient and the negative Wassermann reaction suggested the operation of some other cause, such as encephalitis or brain tumour. Post-mortem examination revealed the presence of some old-standing thickening

FIG. 1. Brain of mongolian imbecile viewed from the front. The temporal and frontal poles have been separated to expose the diseased arteries. Note softening of the convolutions of the left frontal pole.

FIG. 2. Transverse section of left middle cerebral artery, showing endarteritis proliferans.

of the mitral valve segments but no evidence of recent endocarditis, and these changes were insignificant in comparison with the marked arterial degeneration found in the vessels at the base of the brain.

The lumen of both the left anterior and middle cerebral arteries was almost completely excluded and there can be little doubt that this condition was the immediate cause of the extensive softening in the left hemisphere.

The histological appearances of the diseased arteries were indistinguishable from those seen in syphilitic endarteritis, and the absence of all evidence of tubercle, which is capable of giving rise to a somewhat similar picture, makes it almost certain that we are here dealing with a proliferative endarteritis of syphilitic origin. The site of the vascular degeneration and the presence of cirrhotic changes in the kidneys are consistent with this supposition, while on the other hand the negative Wassermann reaction cannot be regarded as an objection to this view, for it frequently fails to confirm the presence of vascular syphilis in the adult and in young mental defectives displaying the typical physiognomy of congenital syphilis negative reactions are by no means uncommon.

Finally, a word may be said regarding the significance of these changes in relation to the etiology of mongolism. For the view that syphilis in the parents may give rise to mongolism in the offspring there is a good deal of support on clinical grounds, but the fact that syphilitic lesions are so seldom encountered in post-mortem material is decidedly against this view. In the organs of a cretin which I lately had the opportunity of examining there was abundant evidence of a syphilitic infection, but it would be rash to conclude from their presence that cretinism is therefore a form of congenital syphilis.

All that it is legitimate to say is that by attacking the endocrine organs syphilis may in some cases give rise to those signs of thyroid insufficiency which make up the clinical picture of cretinism, and I am inclined to believe that the syphilitic poison, operating in a similar manner, may occasionally be held responsible for the development of mongolism.

SUMMARY.

A young mongolian imbecile with a negative Wassermann reaction died some months after the development of a right-sided hemiplegia. Examination of his brain showed the presence of a widespread cortical softening in the left cerebral hemisphere, and an obliterative endarteritis of the cerebral arteries, indistinguishable from that caused by syphilis.

REFERENCE.

BABONNEIX, *Arch. de méd. des enf.*, 1909, xii, 499.