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A Case of Sturge-Weber Syndrome

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In 1879 Sturge described a patient with hemiparesis, epilepsy and a facial naevus. He ascribed the neurological condition to a naevus of the brain similar to that on the patient's face. Weber gained the distinction of providing the other half of the eponym by describing the radiological appearances of a similar case. In 1955, however, he advanced the self-effacing descriptive term "encephalofacial angiomatosis". The essential pathological features of the disorder are facial naevus and leptomeningeal angiomatosis. It is generally agreed that buphthalmos (present in Sturge's original case) is not a necessary component.

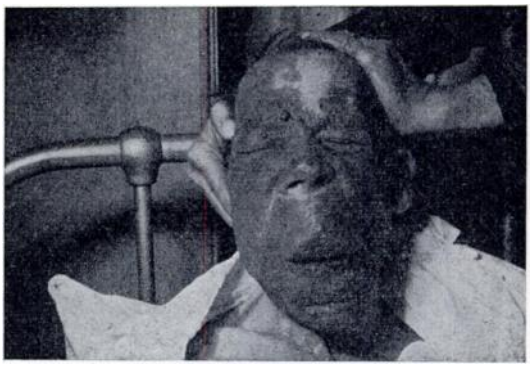
Case Report

The patient died at the age of 34, having spent six years in Dingleton Hospital. He had been under institutional care since the age of 13 and was certified insane at the age of 15 following an assault upon his parents. He suffered from epileptic fits and his intelligence was low. In hospital he was a difficult patient because of sly, trouble-making and aggressive behaviour.

He is said to have had a difficult birth; the cord was around the neck and caused severe asphyxia. In infancy the naevus presented as a bilateral port-wine stain over two-thirds of the head and neck, but the deformity became more marked at puberty with hypertrophy of the lips. The condition was investigated and he received "radium treatment" about the age of 8. In 1955 an attempt was made to improve his appearance by plastic repair. His fits were well controlled by phenobarbitone and primidone (Mysoline), but he remained emotionally labile. During the year before his death, the haemangioma darkened and the surface became easily abraded. The mucous membranes of the mouth and tongue were involved and constantly oozed blood, causing anaemia. The haemoglobin was 28 per cent. before death; the colour index was low and the white cell count around 4,000.

Necropsy showed involvement of both sides of the face and front of the neck by a capillary haemangioma which also extended to the anterior chest wall. The frontal bone showed dense bony thickening. The thyroid was atrophied and sections suggested angiomatous involvement. The brain after fixation weighed 1,340 g. There were diffuse angiomatous changes in the meninges. Cerebral atrophy was marked, on the right more than on the left. A small angiomatous malformation was present in the central white matter of the right frontal lobe and dilated vessels were seen elsewhere in the brain substance. There was also a tumour-like enlargement of the right glomus of the choroid plexus. The changes in the cortex were variable but most marked in the right hemisphere. They consisted mainly of gliosis and some loss of nerve cells. Calcification was notable in the right calcarine cortex and close to the walls of the abnormal vessels in the white matter. Pigment resembling thorotrast was found in the liver and spleen.

The pathological changes in the skin, mucous membranes, leptomeninges and brain were characteristic of Sturge-Weber disease. The immediate cause of death was cardiac failure caused by severe anaemia and a mild respiratory infection.



Comment

In a monograph, based largely on the detailed investigation of seven cases, Alexander and Norman (1960) make a comprehensive survey of the Sturge-Weber syndrome. They point out that meningeal angiomatosis is to be expected only when the facial naevus involves the upper eyelid or forehead and that the clinical diagnosis

may thus be inferred from inspection of the naevus alone. Most of their personal cases were children, and they emphasize the benefit of surgical extirpation of the pathological tissue to prevent further epilepsy and brain damage. The cerebral involvement of this patient was too widespread for surgical treatment, and over the years deterioration had occurred in personality and intellect.

The thorotrast found post mortem was injected during investigations in the patient's childhood. The late effects of this substance (thorium dioxide) have been investigated and are discussed by Looney (1960). He finds relatively few pathological states that can be attributed with confidence to thorotrast or its radio-active daughter elements, but cites evidence of possible deleterious effects, including tumour formation and aplastic anaemia. It is conjectural how significant the presence of the thorotrast was in this case and how far it affected the patient's terminal condition.

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