A CASE OF BILATERAL GANGLIONIC NEURO-GLIOMATA OF THE FACE

BY

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THIS case was reported in March, 1900, in the *Medical Chronicle*, a journal formerly published in Manchester, but now extinct.

The condition described is so rare that the article is worth recording again.

The patient, a baby aged nine months, was admitted under the care of the late Dr. David Little, but was handed over to Dr. Clegg. Two separate and distinct large swellings, each the size of the lower half of the nose, were present, and occupied the position of the lacrymal sac. The overlying skin was slightly discoloured, but movable over the swelling. No pain was produced on palpation, and each mass gave a semi-fluctuant feel. Both tumours were distinctly separated from the frontal bone by an interval of about 5 mm., but in the middle line over the nasal bone the thickening of the skin caused a slight swelling between the tumours. The masses could be moved somewhat laterally and vertically. The conjunctival sacs were both normal, but a small bead of semi-dried muco-purulent matter was always present at the inner canthi. There was no nasal obstruction, the head was of normal shape, and the fontanelles in normal condition for the age of the child, who was perfectly healthy.

At birth a small swelling had been noted on the left side of the nose, and the right swelling appeared a few weeks later. Both gradually increased in size until admission. There had always been a slight discharge at each inner canthus.

The family history was excellent, and the seven other sisters of the patient were all perfectly healthy.

It was assumed that the swellings had some relation to the lacrymal sac, but the tissues seemed too hard to be cut by the Weber's knife, and so a small portion of one tumour was removed for microscopic examination.

Later both tumours were excised; in the right the outer half was distinctly encapsuled, but at the inner side the capsule was adherent to the nasal process of the superior maxilla and to the nasal bone. The left mass was removed through a separate incision, and was found to be smaller and more distinctly encapsuled at the outer side, but here again at the inner side the capsule was adherent to the bone and skin. The wounds healed perfectly.

The child was seen five months later, and was still in perfect health.

The tumours appeared to the naked eye as two ovoid masses, the larger was three quarters of an inch long by half an inch in diameter, and the smaller five-eighths of an inch long by three-eighths of an inch in diameter; they were firm and somewhat elastic in consistency; the surface of each was ragged, of a fleshy appearance, and marked with patches of congestion and haemorrhage. The cut surface presented a uniform grayish pink, somewhat translucent, appearance, and a few small vessels.

Microscopically both tumours presented the same structure; they consisted of a sparsely cellular tissue enclosed within an ill-defined capsule derived from the adjacent muscular and connective tissues. In the former the most conspicuous elements were large ovoid cells, some provided with definite processes, and composed of finely granular protoplasm, in which were one or two large oval vesicular, lightly staining, nuclei. These ganglionic cells were lying in the midst of a peculiar reticulum consisting of delicate fibres, among which were embedded small oval nuclei surrounded by a very scanty cell-protoplasm. Throughout the neuroglial basis there were many thin-walled capillary vessels, and here and there a larger vessel with well-defined walls, coming apparently from the surrounding tissues.

The peripheral parts of the tumours formed, as already mentioned, a kind of ill-defined capsule derived from the surrounding tissues, and served sharply to define the limitations of the growth. Among the fibrous tissue, muscle fibres and blood vessels of which this peripheral zone was composed, there was no indication of any invasion by the tumour tissue.

As regards the nature of the tumours, Professor Delépine expressed as his opinion that they were ganglionic neurogliomata, a view in complete accordance with their minute structure. The existence of the large ganglion-like cells, many with distinct processes embedded in a tissue the peculiarities of which at once indicated its neuroglial nature, and the presence of doubtful nerve fibres seen in a few places, rendered the nature of the tumours beyond doubt. Concerning the pathogenesis of such tumours, it is probable that the majority must be regarded as originating from some part of the central nervous system, generally the brain, and as a result of some developmental disturbance. Axel Key some years ago described a case in which a ganglionic neuroglioma appeared on the left ala nasi of a man aged 31 years; the tumour, which had been noticed for one year only, was about the size of a plum, and distinctly encapsuled. He regarded this case as proving the possibility of the development of a ganglionic tumour from a peripheral nerve independently of preceding ganglion formation. Such a case is, however, we believe unique. The great majority of tumours of this nature which have been described have arisen in direct relation with the brain.

In the present case it seems not improbable, from a consideration of the position of the tumours, and their existence since birth, that they might have arisen in connection with the olfactory lobes, which possibly, as a result of some localised hydrocephalic condition, had protruded through congenital fissures of the skull, and gradually become constricted off with the closure of such defects.

Such a view at least provides the most rational explanation of the various features which have been detailed.

"INFANTILE AND CONGENITAL RETINAL FOLD"*

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RECENTLY, a number of cases of congenital retinal fold have been noticed. Some of them also showed anterior displacement of the optic papilla into the interior of the globe. Ancona and Kieuwe thought that this condition is caused by persistence of the connecting portion of von Szily's primitive epithelial papilla. Mann suggested that it is due to adhesion of the primary vitreous to the inner layer of the optic cup. Weve considered that the anomaly must arise through contraction of a mesodermal mass of connective tissue and that probably it is hereditary.

None of these writers found signs of inflammation, but Weve observed remnants of haemorrhage in one case. The essential cause of this curious abnormality remained a matter of conjecture and for this reason the following case is interesting.

The parents of a child three-and-a-half years old complained that 5 months before their child had developed a slight divergent squint and that the (right) eye looked strange.

On examination the eye showed no vascular injection, no exudates, the cornea was clear and the iris normal. The lens was turbid in its deep layers, especially at the centre. Behind the lens, temporally and below, a fairly well defined mass was seen against the retina. With dia-scleral illumination no shadow was observed. The intra-ocular tension was +1. Diagnosis uncertain. It was thought best to excise the globe.

Microscopical examination.—The globe measured 22 by 21.5 millimetres. These are normal figures (Fig. 1).

The cornea, anterior chamber and the angle of the anterior chamber were normal. The iris seemed a little more compact

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