GLAUCOMA SECONDARY TO FIBROCYSTIC DISEASE OF BONE

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W.G.W., a farmer aged 58 years, presented himself on December 3, 1946, complaining of loss of vision and discharge from his left eye. He had always suffered from frontal headache on the left side, and recalled as a youth it was often so severe that he had to cool his head on cold metal objects. He had a swelling over the left temporal region, which an old doctor had remarked upon some forty years ago.

On examination there appeared to be some proptosis of the left eye (3—4 mm.) and displacement outwards in the orbit and a bony

FIG. 1.

hard prominence of the fronto-temporal region on the left side. The tension of the left eye was raised. R.V. 6/9 with -0.5 cyl. 90° 6/6. L.V. 6/60 not improved by glasses. No discharge was seen and cultures from the eyes were sterile.

Physical examination.—There was a bony hard swelling of the left fronto-temporal region and a hard swelling over the occiput and in the left side of the jaw. The skin was normal and no fibromata were found.

His cardio-vascular system was normal, but blood pressure was 190 mm. Hg systolic and 85 mm. Hg diastolic and there was some fibrosis at the base of the left lung. Urine was normal to clinical tests.

Special investigations.—The tension of the eyes was right 25.75 mm. Hg and the left 33.75 mm. Hg average. (Schiotz tonometer with weights.) Field of vision in the right eye was full and the left restricted to a small field in the upper and outer quadrant.

LEFT 25-3-47

Ophthalmoscopy.—Right fundus was normal—the left showed a deeply cupped disc of some 6 dioptres and considerable atrophy.

Pathological Investigations.—Urine acid. Specific gravity 1022. Albumen nil. Sugar nil. Acetone nil. Centrifuged deposit nothing abnormal seen. Culture sterile. No Bence Jones proteose present.

Blood Count.—R.B.C. 5,660,000. Haemoglobin 100 per cent. Colour index 0.9. White blood corpuscles 13,400—polymorphonuclear neutrophils 73 per cent. Eosinophils 3 per cent. Lymphocytes 15 per cent. Monocytes 9 per cent. Serum calcium 9.6 mgms. per cent. Inorganic phosphate 5.0 mgms. per cent. Normal 2—3.7 mgms. per cent.

X-ray examinations showed a honeycombed condition of bone in the left frontal, temporal and occipital bones of the skull. The

FIG 2.

sphenoid was fully involved, and although the orbital fissure could be demonstrated on the right side, it could not be so demonstrated on the left side. The frontal and maxillary sinuses were obliterated on the left side although clearly seen on the right side. A complete radiological examination of the skeleton was made and the only other bones affected were the left side of the mandible and the lower third of the right humerus. Oto-rhinological examination by Mr. Ware revealed nothing of added interest.

Operation

On December 17, I performed a corneo-scleral 1.5 mm. trephine operation and his convalescence was uneventful. Subsequent tension readings were within normal limits: average 23.0 mm. Hg (Schiötz).

I think that there is little doubt that the ophthalmic condition was caused by the abnormal bony formation because it was associated with proptosis displacement and increased tension of the left eye.

FIG. 3.

FIG. 4.

FIG. 5.

The bone condition is not a simple one to explain—at first one considered leontiasis ossium, but the fully developed condition is characterised by the leonine appearance which the name suggests and of this he has none. A malignant condition is excluded by the long history borne out by earlier photographs.

An infective condition arising from the frontal sinus in adolescence would be a feasible one, except that the condition rides rough-shod over suture lines and appears in other parts such as the left side of the mandible and the right humerus.

A diagnosis of fibrocystic disease of bone is more probable when the cystic condition of the lower end of the right humerus is examined, but until this was discovered xanthomatosis seemed a likely diagnosis to explain the unilateral involvement of the skull. The full Hans Schüller-Christian syndrome was not present.

I submitted the films to Sir Thomas Fairbank, who discussed them with Dr. Coldwell. Dr. Coldwell is inclined to consider that it is a mixture of xanthoma and fibrocystic disease, but Sir Thomas thinks that the humerus gives the key to the situation and that fibrocystic disease of the bone is the correct diagnosis. He thinks it is atypical and that there is no evidence of parathyroidism.

The ophthalmic condition suggests that the globe is displaced outwards and forwards by pressure within the orbit and that this has prevented the return venous drainage. The tension was undoubtedly raised, but it is a matter of conjecture whether the optic atrophy was the result of glaucomatous rise of tension within the eye or whether it was due to tension on the optic nerve as the result of stretching. It is true that I have been unable to demonstrate a narrowing of the orbital fissure, but the fact that it is demonstrated on the right and cannot be demonstrated on the left suggests that the fibrocystic disease of the sphenoid has so narrowed the fissure that it cannot be seen.

I suggest, therefore, that this is a case of diffuse fibrocystic disease of bone mainly unilateral, which has caused a secondary glaucoma of the left eye and optic atrophy. It is of double interest from the bone and from the ophthalmic viewpoint.

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