CASE NOTES

UNILATERAL EXOPHTHALMOS RESULTING FROM SCLEROSIS OF THE SPHENOID BONE*

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Case Report

A woman aged 53 was sent by her doctor to the West Suffolk General Hospital in July, 1955. She said that her right eye had become more prominent during the last year, but a snapshot taken 2 years previously suggested a prominence of the right eye at that time. She made no complaint of any symptoms referable to the exophthalmos. Her weight was constant and she preferred hot weather to cold. There was no past history of a serious illness except deafness resulting from measles 30 years previously. Her father was alive and well at the age of 80 and her mother had died at 81, having only suffered from some osteo-arthritis previous to her last illness. Two brothers and two sisters were alive and well. The patient was married and had a healthy son and daughter.

Examination.—She was a healthy-looking woman with 8 mm. proptosis of the right eye (Figs 1 and 2). Visual acuity (uncorrected) was 6/12 in each eye, with physiologically cupped discs and normal visual fields.

FIGS 1 and 2.—8-mm. proptosis of right eye.

X-ray examination of the skull showed a uniform sclerosis of bone in the right frontal and orbital region which was fairly well defined. The whole of the lesser and greater wing of the sphenoid and the lateral wall of the orbit and zygoma were sclerosed (Figs 3 and 4, opposite).

Serial *x* rays of the head in a gradually forward-tilting position to exclude the petrous temporal bone showed that both wings of the sphenoid were involved. Lateral *x* rays showed a normal sella turcica. Further *x* rays to cover the skeleton did not disclose any other bony lesion. The opaque left antrum would appear to be a simple infective condition while the sinus, ethmoids, and antrum of the affected side appeared to be normal.

The following pathological investigations revealed nothing abnormal:

Blood.—Red blood corpuscles 4,120,000; haemoglobin 93 per cent.; white blood corpuscles 7,800, polymorphs 62 per cent., lymphocytes 36 per cent., eosinophils 2 per cent.; erythrocyte sedimentation rate 19 mm./1 hr. Wassermann reaction and Kahn test negative. Alkaline phosphatase 5.6 units/100 ml.; total protein 6.6 g/100 ml., albumin 4.4, globulin 2.2, A/G ratio 2-1; serum cholesterol 130 mg./100 ml., blood phosphorus 2.6 mg./100 ml., serum calcium 10 mg./100 ml.

Urine.—No Bence-Jones protein.

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Lumbar Puncture.—Cerebro-spinal fluid clear and colourless, pressure 120 mm; Queckenstedt positive, protein 28 mg. per cent., cells none, no excess globulin, glucose normal.

FIG. 3.—Sclerosis of sphenoid extending to zygoma.

FIG. 4.—Antero-posterior view showing involvement of both wings of sphenoid.

Discussion

Duke-Elder (1952) discusses unilateral proptosis and gives a very full classification of causes; he goes on to discuss causes of the relatively rarer bilateral proptosis and gives a list of osteopathies. It would appear that a condition akin to osteitis fibrosa would be more likely to give rise to a

bilateral condition. Cory (1947) reported a case of fibrocystic disease of bone affecting the left orbit associated with glaucoma.

Sir Thomas Fairbank, who was good enough to see the films, writes:

If it is leontiasis (fibrosis) it is quite unusually dense and uniformly dense in parts at least. I've not seen such intense uniform density in polyosteitic fibrous dysplasia.

Meningioma was considered until subsequent *x* rays showed that the outer table of the skull was mainly involved and that the sclerosis first noticed in the sphenoid extended to the zygoma and frontal bone. Oblique views showed the optic foramen and sphenoidal fissure to be of normal size and that in this position both tables were affected (Figs 4 and 5).

FIG. 5.—Oblique view, showing optic foramen and sphenoidal fissure of right side.

Benedict (1941) used the term hyperostoses to cover a variety of pathological lesions many of which arose from the soft structures, and said that thickening of the great wing of the sphenoid was frequently brought about by a meningioma spreading along the surface of the cranial vault. This may penetrate the bone and enter the orbit and cause exophthalmos, often with paralysis of ocular muscles.

In the case under discussion meningioma seemed to be ruled out by the involvement of the outer table, the spread to the zygoma, and the relatively unchanged condition of the patient after 18 months.

Harrison (1942) reported a case of osteoma of the orbit with resulting bilateral optic atrophy.

Newell (1948) reported a case of involvement of the right orbit with sphenoidal osteoma causing diplopia; the case was observed for 2 years, in which time no further symptoms appeared and there was no increase in the size of the tumour.

The earliest reports I have read are those of Hewett (1867), writing on exostosis of the skull. He appears to have travelled widely both in Great Britain and in Europe observing specimens in museums. No similar condition is described but he, along with other observers, notes the tendency of osteomata to occur near the suture lines, particularly those of the frontal bone.

Glenn (1950) discusses osteomata at length and describes circumscribed tumours, which may involve one or both tables of the skull. These may appear as diffuse tumours arising from the sphenoid or temporal bones or in the orbito-ethmoid or orbito-maxillary areas. The diffuse lesions produce tremendous thickening of the involved bones and are usually the type which give rise to most symptoms. Circumscribed osteomata are not difficult to recognize, as they appear as mould-like tumours from the surrounding bone or in the accessory air sinuses.

Summary

An unusual case of sclerosis of bone is discussed, which mainly affects the sphenoid bone, but extends to the zygoma and frontal bones. It would appear to belong to the osteomata, and, as Sir Thomas Fairbank writes, is too uniformly dense to belong to the leontiasis ossea or fibrocystic group of bone diseases. The condition would not appear to be rapidly progressive and no active treatment is considered advisable.

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REFERENCES

- BENEDICT, W. L. (1941). *Amer. J. Ophthal.*, **24**, 1005.
CORY, J. W. E. (1947). *British Journal of Ophthalmology*, **31**, 731.
DUKE-ELDER, S. (1952). "Text-book of Ophthalmology", vol. 5, p. 5629 *et seq.* Kimpton, London.
GLENN, J. C. (1950). In "Diagnosis and Treatment of Tumors of the Head and Neck", ed. G. E. Ward and J. W. Hendrick, p. 756. Williams and Wilkins, Baltimore.
HARRISON, W. J. (1942). *Amer. J. Ophthal.*, **25**, 1233.
HEWETT, P. G. (1867). *St George's Hosp. Rep.*, **2**, 1.
NEWELL, F. W. (1948). *Amer. J. Ophthal.*, **31**, 1281.