MISINTERPRETATION OF SPHENOIDAL RIDGE MENINGIOMATA*†

BY

ALY MORTADA

Department of Ophthalmology, Faculty of Medicine, Cairo University, Cairo, Egypt

SPHENOIDAL ridge meningiomata are a common cause of unilateral proptosis (Elsberg, Hare, and Duke, 1932). Ophthalmologists are more familiar with the classical picture of sphenoidal ridge meningioma usually occurring in adults and giving rise to unilateral proptosis (Fig. 1), optic atrophy, oculomotor palsies, anaesthesia in the distribution of the fifth nerve, and (rarely) pituitary disturbance and uncinatate fits.

FIG. 1.—Right gradual progressive proptosis of 4 years' duration due to a right sphenoidal ridge meningioma in a woman aged 40 years.

The characteristic *x* ray reveals involvement of the lesser and greater wings of the sphenoid bone by dense hyperostosis, which is usually diffuse (Figs 2 and 3) diminishing the size of the orbit, optic canal, and superior orbital fissure.

FIG. 2.—Postero-anterior *x* ray, showing typical diffuse hyperostosis of right lesser and greater wings of sphenoid bone due to sphenoidal ridge meningioma.

FIG. 3.—Lateral *x* ray, showing diffuse hyperostosis of lesser and greater wings extending to body of sphenoid bone, narrowing the sphenoidal sinus but not affecting the sella turcica, due to right sphenoidal ridge meningioma.

Swelling of the temple may also occur (Figs 4 and 5, overleaf).

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† Address for reprints: 18A 26th July St., Cairo, Egypt.

FIG. 4.—Left gradual progressive proptosis of 5 years' duration and swelling of temple due to left sphenoidal ridge meningioma affecting the pterion in a woman aged 42 years.

FIG. 5.—Postero-anterior x ray, showing hyperostosis in pterion region.

Cushing and Eisenhardt (1929) first pointed out the distinct clinical pictures due to meningiomata of the different sites of the sphenoidal ridge:

(1) Inner third meningioma usually causes unilateral optic atrophy, exophthalmos, and ocular nerve palsies.

(2) Middle third meningioma is asymptomatic when small, but may cause olfactory hallucinations and contralateral homonymous hemianopia. Unilateral optic atrophy, ocular nerve palsies, and proptosis are later manifestations.

(3) Outer third meningioma of the globular type is usually asymptomatic. When *en plaques* it produces unilateral exophthalmos, swelling of the temple, and optic atrophy.

The more laterally situated neoplasms are referred more frequently to the ophthalmologist because of unilateral proptosis from involvement of the orbital bones (Duke-Elder, 1949).

Among my 300 cases of unilateral proptosis there were twelve cases of sphenoidal ridge meningiomata. Although the x ray picture is usually characteristic, the following seven cases show how other orbital bony tumours may be suspected. These variants are of extreme interest to the ophthalmologist.

Case Reports

The seven cases described below occurred in adults. There was no significant family history. The height, weight, and general development of the patients were normal. There was no history of trauma or radiotherapy, no signs of endocrine disturbance, no enlargement of lymph glands or of thyroid gland, liver, or spleen. The chest and abdomen were normal. The skin did not show pigmentation, haemangiomas, or nodules. There were no other tumours in the body, and no signs of increased intracranial pressure. The nervous system was normal as well as the temperature, pulse, blood pressure, urine, faeces, and blood studies. The blood Wassermann reaction, tuberculin test, and Casoni's test for hydatid were negative. Basal metabolic rate and iodine¹³¹ uptake were normal. X ray films of the rest of the skeleton were normal. The sella turcica, cerebral angiographs, teeth, nasopharynx, and nasal sinuses were all normal.

There were no acute inflammatory local signs such as redness of the lids, or conjunctival

chemosis. The proptosis was not intermittent or pulsating. External palpation did not reveal orbital masses.

Case 1, a 32-year-old woman (Fig. 6), complained of gradual proptosis and deterioration of vision in the right eye of 4 years' duration. Prominence of the frontal and temporal bones on the right side gave an appearance of facial asymmetry.

The left eye was normal and the visual acuity 6/9.

The right eye showed forward and downward proptosis of 23 mm. (left 16 mm.) with slight limitation of ocular movements in all directions. The fundus showed post-papilloedemic optic atrophy. The visual acuity was hand movements.

FIG. 6.—Case 1. Right gradual proptosis of 4 years' duration in a woman aged 32 years. Skull *x* rays were suggestive of fibrous dysplasia of right orbital bones. Transcranial exploration proved presence of right sphenoidal ridge meningioma.

FIG. 7.—Case 1. Postero-anterior *x* ray. Thickening and expansion of right lesser and greater wings of sphenoid bone and orbital process of frontal bone were suggestive of fibrous dysplasia of orbital bones. The true diagnosis was sphenoidal ridge meningioma.

Skull *x* rays: Postero-anterior view (Fig. 7) showed thickening and expansion of the lesser and greater wings of the sphenoid bone on the right side leading to narrowing of the superior orbital fissure. The bony cortex was attenuated and the bone looked amorphous. There was also involvement of the orbital process of the frontal bone on the same side leading to a diffuse opacity superimposed upon the upper part of the orbital cavity. Lateral view showed involvement of the orbital roof and extension of the bony process in the body of the sphenoid bone with involvement of the region of the sphenoid sinus. Oblique views showed narrowing of the right optic canal.

These findings were suggestive of fibrous dysplasia of the right orbital bones, but orbital exploration did not reveal any orbital tumour.

3 years later right proptosis was 25 mm., and a postero-anterior *x* ray (Fig. 8) showed more marked density of the lesser and greater wings of the sphenoid bone and the orbital process of the frontal bone. The superior orbital fissure was not seen. Orbital exploration revealed irregular thickening at these sites.

FIG. 8.—Case 1. Postero-anterior *x* ray taken 3 years later, showing accentuation of previous findings. The right superior orbital fissure is not seen.

Case 2, a 38-year-old man (Fig. 9), complained of gradual protrusion of the right eye of 12 years' duration with deterioration of vision for the past 5 years.

The left eye was normal and the visual acuity 6/9.

FIG. 9.—Case 2. Right gradual proptosis of 12 years' duration in a man aged 38 years. *X* rays were suggestive of orbital osteoma arising from the right lateral orbital wall. Transcranial exploration proved presence of right pterion meningioma.

The right eye showed forward, downward, and medial proptosis of 25 mm. (left 15 mm.), with limitation of ocular movements upwards and outwards. The optic disc showed temporal pallor. The visual acuity was 1/60. Exploration of the orbit revealed a bony hard mass 1.5×1 cm. with a smooth surface involving the lateral wall of the orbit.

Skull x rays: Postero-anterior (Fig. 10) and lateral (Fig. 11) views showed a globular mass of thickened bony condensation involving the lateral sides of the greater wing of the sphenoid and the orbital plate of the frontal bones. Oblique views showed normal optic canals.

FIG. 10.—Case 2. Postero-anterior *x* ray, showing in the right orbit a globular bone condensation arising from the lateral orbital bony wall, which was wrongly suggestive of an osteoma.

FIG. 11.—Case 2. Lateral *x* ray, showing globular bony condensation arising from region of pterion.

The findings were suggestive of fibrous osteoma or ossifying fibroma at the junction of the lesser and greater wings of sphenoid and frontal bones.

Case 3, a 54-year-old man (Fig. 12), complained of gradual proptosis and diminution of vision of the left eye of 3 years' duration.

FIG. 12.—Case 3. Left gradual proptosis of 3 years' duration in a man aged 54 years. *X* rays were suggestive of osteolytic osteosarcoma of left lesser and greater wings of sphenoid bone. Transcranial exploration proved the presence of a left sphenoidal ring meningioma.

The right eye was normal, with visual acuity 6/6.

The left eye showed forward and downward proptosis of 23 mm. (right 15 mm.) with limitation of ocular movements upwards. The fundus showed optic atrophy. The visual acuity was 1/60 with no refractive error. Exploration of the orbit revealed a bony mass affecting the lateral orbital wall.

Skull x rays: The lesser and greater wings of the sphenoid bone on the left side showed evidence of destruction. The superior orbital fissure was not seen (Fig. 13). Oblique views showed a narrow optic canal.

FIG. 13.—Case 3. Postero-anterior x ray, showing osteolysis of left lesser and greater wings of the sphenoid bone. The left superior orbital fissure is not seen.

The findings were suggestive of an osteolytic osteosarcoma of the left side of the sphenoid bone.

Case 4, a 40-year old woman (Fig. 14), complained of proptosis and diminution of vision of the left eye of 3 years' duration.

The right eye was normal, with visual acuity 6/9.

The left eye showed forward, downward, and outward proptosis of 25 mm. (right 16 mm.) with limitation of ocular movements inwards and upwards. The fundus showed optic atrophy. There was no perception of light. Orbital exploration revealed a bony mass above and below the superior orbital fissure.

FIG. 14.—Case 4. Left gradual progressive proptosis of 3 years' duration in a woman aged 40 years. X rays were suggestive of osteolytic osteosarcoma of left lesser and greater wings of sphenoid bone. Transcranial exploration proved presence of left sphenoidal ridge meningioma.

FIG. 15.—Case 4. Postero-anterior x ray, showing osteolysis of left lesser and greater wings of sphenoid bone and an irregular margin of superior orbital fissure.

Skull x rays: Lateral and postero-anterior views (Fig. 15) showed irregular destruction of the lesser and greater wings of the sphenoid bone with subsequent poor definition of the bony contour of the superior orbital fissure. Oblique views showed a narrow optic canal.

The findings were suggestive of an osteolytic osteosarcoma of the left side of sphenoid bone.

Case 5, a 28-year-old man (Fig. 16) complained of gradual proptosis and diminution of vision of the right eye of 3 years' duration.

FIG. 16.—Case 5. Right gradual progressive proptosis of 3 years' duration in a man aged 28 years. X rays were suggestive of osteoblastic osteosarcoma of right lesser and greater wings of sphenoid bone. Transcranial exploration proved presence of right sphenoidal ridge meningioma.

The left eye was normal with visual acuity 6/12.

The right eye showed forward and downward proptosis of 24 mm. (left 15 mm.) with limitation of ocular movements upwards. The fundus showed optic atrophy. There was no perception of light. Orbital exploration revealed a hard mass 1 × 1.5 cm. involving the lateral orbital wall.

Skull x rays: Postero-anterior view (Fig. 17) showed that the lesser and greater wings of the sphenoid bone on the right were replaced by an area of irregular bone density with loss of contour of the right superior orbital fissure. Oblique views showed loss of contour of the right optic canal.

4 years later the postero-anterior view (Fig. 18) showed that this irregular bone density was now surrounded by a sun-burst appearance.

FIG. 17.—Case 5. Postero-anterior x ray, showing lesser and greater wings of sphenoid bone on right side replaced by irregular bone density with loss of contour of superior orbital fissure.

FIG. 18.—Case 5. Postero-anterior x ray taken 4 years later, showing replacement of right lesser and greater wings of sphenoid bone by an area of irregular bone density surrounded by sun-ray appearance.

The findings were suggestive of an osteoblastic osteosarcoma of the right side of the sphenoid bone.

Case 6, a 40-year-old woman (Fig. 19), complained of gradual proptosis of the right eye of 4 years' duration.

The left eye was normal with visual acuity 6/9.

FIG. 19.—Case 6. Right progressive proptosis of 4 years' duration in a woman aged 40 years. X rays were suggestive of a malignant tumour of the orbit destroying the right lesser wing of sphenoid bone. Transcranial exploration proved presence of right sphenoidal ridge meningioma.

The right eye showed forward and downward proptosis of 22 mm. (left 16 mm.) with limitation of ocular movements upwards. The fundus was normal. The visual acuity was 6/9. Orbital exploration revealed no abnormality.

Skull x rays: Postero-anterior view (Fig. 20) showed that the lesser wing of the sphenoid bone was thinned, and that the superior orbital fissure was abnormally wide due to irregular destruction of the bone on both sides. Oblique views showed normal optic canals.

FIG. 20.—Case 6. Postero-anterior x ray, showing thinned-out right lesser wing of sphenoid bone and irregular sides of superior orbital fissure.

The findings were suggestive of a malignant orbital tumour destroying the right lesser wing of the sphenoid bone.

Case 7, a 35-year-old man (Fig. 21), complained of gradual proptosis of the left eye of 18 months' duration.

Apart from divergent concomitant squint, the right eye was normal with visual acuity 6/18.

The left eye showed forward and downward proptosis of 23 mm. (right 16 mm.). The fundus was normal. The visual acuity was 6/9. Orbital exploration revealed no abnormality.

Skull x rays: Postero-anterior, lateral, and oblique views showed normal findings.

The case was diagnosed as one of proptosis of unexplained origin.

FIG. 21.—Case 7. Left gradual progressive proptosis of 18 months' duration in a man aged 35 years. Skull x rays showed no abnormality, and orbital exploration was negative, but one year later a left sphenoidal ridge meningioma was found on transcranial exploration.

FIG. 22.—Case 7. Postero-anterior x ray one year later, showing slight thickening of lesser and greater wings of left sphenoid bone.

One year later the postero-anterior view (Fig. 22) was interpreted by one radiologist as showing no abnormality, and by another as showing slight thickening of the lesser and greater wings of the left sphenoid bone and a wider left superior orbital fissure. Oblique views showed normal optic canals.

These seven cases were referred to the neurosurgeon, when transcranial exploration and biopsy examination showed that all were due to sphenoidal ridge meningiomata *en plaques*.

Discussion

Hartmann and Gilles (1959) described two types of change in the lesser and greater wings of sphenoid in sphenoidal ridge meningioma: erosion and hyperostosis.

(1) Osteolytic lesions, as in Cases 3 and 4, in which decalcification of the lesser wing extends to the greater wing so that the orbital landmarks are eradicated and appear as a uniform spongy, structureless shadow. At other times as in Case 6, there is true erosion caused by the invading tumour. The superior orbital fissure is changed in shape, and may be wide, irregular, or difficult to locate.

(2) Hyperostotic lesions may be limited to the lesser wing. More often the tumour covers the lesser wing entirely, encircling the optic canal and spreading to the greater wing and even to the body of the sphenoid bone. The superior orbital fissure is narrowed or even obliterated.

As a rule meningiomata of the olfactory groove produce proptosis at a late stage when the growth invades the orbit, while tumours of the sphenoid ridge produce proptosis at an earlier stage owing to the development of hyperostosis. This difference has a pathological explanation (Duke-Elder, 1952). The bones of the roof of the orbit are almost avascular and do not react to neoplastic infiltration by proliferation but are merely decalcified and absorbed by continued pressure, whereas the marrow containing bone around the pterion proliferates defensively to produce diffuse thickening at the apex of the orbit.

Reese (1963) stated that "x ray films are helpful in most cases. About 80 per cent. of these tumours will show bone destruction or hyperostosis or both."

According to Pfeiffer (1943), the hyperostosis may be located radiologically at any part along the sphenoid ridge. At first it may appear as a slight thickening of the bone with a smooth cortical surface and no visible spicule formation. Bone destruction or pressure atrophy also occurs. It must be remembered that overgrowth of bone may be produced by tumours other than meningiomata and also that meningiomata frequently fail to produce bone changes demonstrable radiologically.

X ray changes depend on whether the meningioma is psammomatous or osteoblastic, globular or *en plaques*, pressing or invading the bone, and also on whether the invaded bone is avascular or vascular. In skull x rays of the lesser and greater wings of the sphenoid bone the following appearances may be seen:

- (1) No demonstrable change, as in Case 7.
- (2) Faint bony thickening, usually due to slight pressure of the tumour. This may be misinterpreted as fibrous dysplasia of orbital bones as Case 1. It may also be regarded as due to obliquity in taking the film or to the superimposed shadow of an orbital soft tissue tumour.
- (3) Localized hyperostosis, which may be diagnosed as osteoma as in Case 2.
- (4) Osteolysis, which may be diagnosed as an osteolytic osteosarcoma as Cases 3 and 4. A similar case was described by Franceschetti (1959), showing an osteolytic appearance at the back of the orbit. A similar picture may be produced by osteolytic metastasis.
- (5) Destructive lesions due to invasion of bone as in Case 6. This must be differentiated from a malignant orbital tumour destroying the bone.
- (6) Irregular bone thickening with raised periosteum giving a sun-ray appearance simulating an osteoblastic osteosarcoma as in Case 5.

The superior orbital fissure may be narrower or wider than normal, or have an irregular margin according to the type of reaction involving its bony border, *i.e.* bone formation, osteolysis, or destruction.

The x ray appearances have also to be differentiated from those due to other causes, which include orbital hyperostosis, osteoma, metastasis, osteitis deformans, osteopetrosis, osteitis fibrosa cystica, and leontiasis ossea.

The commonest cause of hyperostosis along the lesser and greater wings of the sphenoid bone is a meningioma (Cushing and Eisenhardt, 1938; Alpers and Groff, 1934). Less common causes are infiltrating sarcomata (Dandy, 1941), haemangiomata, or neuro-fibromata of the orbit or cranium, epithelioma of the nasopharynx, or arterio-venous aneurysm. A similar neo-formation may arise after trauma. Orbital osteomata may arise from the nasal sinuses.

Bony metastasis usually occurs in the flat bones of the cranial vault; there is also a rare osteoblastic type (as in some cases of cancer of the prostate or bladder) and a commoner osteolytic type.

Osteopetrosis (Albers-Schönberg, 1904) usually begins in childhood and persists throughout life; the bones, including those of base of the skull, become dense and sclerosed like marble while the vault remains normal.

The localized and generalized osteitis fibrosa cystica described by von Recklinghausen is characterized by a combination of absorption and formative processes so that the bone is replaced by fibrous osteoid tissue which may contain cysts and form localized tumours. The condition is due to disease of the parathyroid with disturbance of calcium metabolism. The process may affect the orbit, particularly the frontal bone, when tumour-like masses may give rise to proptosis (Wright, 1938).

Osteitis deformans (Paget, 1876), a disease occurring in adults, is characterized by absorption and new formation of bone leading to enlargement of the skull and bowing of the limbs and clavicles. New bone may cause proptosis by diminishing the size of the orbit. The base of the skull may also be involved.

Leontiasis ossea (Virchow, 1896), which may occur at any age, is characterized by dense thickening of the cranial and facial bones, sometimes with the formation of large opaque bony tumours. It is rarely localized to one or more cranial bones (Reynolds, 1947) and usually causes an enlargement of the frontal region leading to downwards displacement of the orbits (Philps, 1939).

In cases of progressive proptosis of unexplained origin, when orbital exploration shows no abnormality, repeated skull x rays usually reveal a deep bony tumour of the orbital wall or a tumour or mucocele of an accessory nasal sinus (Mortada, 1963). In sphenoidal ridge meningioma the changes in the lesser and greater wings of the sphenoid may not be well seen radiologically. When the lateral part of the lesser wing of the sphenoid is affected the optic nerve is usually undisturbed for a long time. In this case the symptoms are cranial fullness with deformity in the temporal region and proptosis from involvement of the orbital bones (Stender, 1933; Knapp, 1938; Smith, 1939). If the swelling of the temple is not apparent the only sign is a unilateral exophthalmos. If investigations are negative as in Case 7, years may pass before the sphenoidal ridge meningioma becomes evident.

Orbital exploration in sphenoidal ridge meningioma usually reveals a bony mass affecting the posterior part of the lateral orbital wall. Sometimes the bony change is demonstrable on x ray but not felt in the orbit and *vice versa*.

Summary

(1) The x ray appearance of sphenoidal ridge meningioma may be confused with the following orbital lesions and tumours.

- (i) Fibrous dysplasia of orbital bones.
- (ii) Osteoblastic osteosarcoma.
- (iii) Osteolytic osteosarcoma.
- (iv) Ossifying fibroma or osteoma.
- (v) Malignant orbital tumour with destruction of lesser and greater wings of the sphenoid bone.

Transcranial exploration by a neurosurgeon will reveal the true nature of the tumour.

(2) Many cases of proptosis of unexplained origin in which investigations are negative are due to an early meningioma usually *en plaques* affecting the lateral side of the lesser wing of the sphenoid bone.

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