Juxtasellar hyperostosis of non-meningiomatous origin

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Seven patients are described who had juxtasellar hyperostosis with visual disturbance secondary to non-meningiomatous lesions. Two had chromophobe adenomas, one craniopharyngioma, one carcinoma of the sphenoid sinus, one a thrombosed aneurysm of the intracavernous portion of the internal carotid artery, one epidermoidoma of the orbit, and one chondroblastoma of the anterior clinoid process. The diagnosis of meningioma was entertained initially on the basis of hyperostosis plus visual impairment. Careful evaluation of hyperostosis is essential for correct diagnosis of meningioma, according to our experience. Suprasellar meningiomas almost invariably produce irregular hyperostosis of the planum sphenoidale, often associated with serration and blistering. Sphenoid meningioma, when it is sclerotic, always shows thickening or expansion of the sphenoid wings. Therefore, in the absence of typical meningiomatous hyperostosis, one can readily differentiate non-meningiomatous hyperostosis from true meningioma.

KEY WORDS • hyperostosis, non-meningiomatous • differential diagnosis • meningioma

During the past several years we have seen seven patients with non-meningiomatous juxtasellar hyperostosis that resembled meningioma radiographically. Initially, five of the seven cases (two chromophobe adenomas and one each of craniopharyngioma, thrombosed carotid aneurysm, and chondroblastoma) could not be distinguished from true meningioma even after clinical and laboratory investigations. The correct preoperative diagnosis was made in the remaining two cases, one of carcinoma of the sphenoid sinus and one of epidermoid tumor of the left orbit. As our experience with craniobasal meningioma increases, we have become more alert in recognizing subtle yet convincing diagnostic signs of meningiomatous hyperostosis. With the use of tomography, we have previously found that the differential diagnosis between meningiomas and non-meningiomatous conditions may be readily made when blistering and/or serration of the hyperostosis are demonstrated in the planum sphenoidale. Sphenoid meningiomas always demonstrate varying degrees of bone proliferation on tomograms when sclerosis is evident on plain film, and the finding of pure sclerosis without bone expansion of the sphenoid wings virtually eliminates the possibility of meningioma. It is our purpose in this paper to analyze seven cases of non-meningiomatous
hyperostosis and to emphasize the importance of recognizing meningiomatous hyperostosis for differential diagnosis.

Case Reports

A summary of the clinical and diagnostic findings and course for the patients in this series is given in Table 1.

Case I

This 56-year-old woman was admitted because of progressive visual disturbance in the left eye for 7 months. Temporal hemianopsia and optic atrophy were present in the left eye, and a superior temporal quadrantanopia in the right.

A skull film revealed an irregular hyperostosis of the planum sphenoidale. The pituitary fossa was slightly enlarged and demineralized. Brain scan showed an area of increased uptake in the suprasellar region. A left carotid arteriogram revealed slight elevation of the main trunk of the anterior cerebral artery without tumor blush. A pneumoencephalogram with a hanging-head midsagittal tomogram again showed hyperostosis associated with a sharp indentation at the rostral portion of the third ventricle (Fig. 1). A preoperative diagnosis of tuberculum sellae meningioma was made, and a frontal craniotomy was performed. At surgery a 5 × 3 × 2-cm encapsulated mass in the suprasellar region was subtotally removed. The pathological diagnosis was chromophobe adenoma.

Case 2

A 65-year-old woman was admitted because of visual impairment in both eyes. There was a total loss of vision in the right eye and 20/20 in the left. There was a definite temporal field defect in the left eye to the color red. An amaurotic pupil and extensive optic atrophy were present on the left.

A skull film revealed an irregular hyperostosis, confined to the tuberculum sellae. The pituitary fossa was slightly enlarged and had a double floor (Fig. 2 left). A carotid arteriogram showed elevation of the main trunk of the anterior cerebral artery in an arcuate fashion without tumor blush. A pneumoencephalogram with a hanging-head midsagittal tomogram revealed an indentation at the rostral end of the third ventricle, and a hyperostosis of the tuberculum sellae suggestive of a meningioma (Fig. 2 right). A preoperative diagnosis of tuberculum sellae meningioma with suprasellar extension was made, and a frontal craniotomy was performed. At surgery a suprasellar tumor measuring 2.5 × 2.0 × 2.5 cm was seen obscuring the sella turcica and posterior portion of the planum sphenoidale. The tumor was subtotally removed.

The pathological diagnosis was malignant pituitary adenoma. The patient died of cardiac disease 1 month after the craniotomy, and the autopsy specimen of the sphenoid bone revealed diffuse neoplastic infiltration of the tuberculum sellae, right anterior clinoid process, and pituitary fossa. A radiograph of the specimen confirmed the sellar hyperostosis (Fig. 2 right, inset).

Comment

Both patients in Cases 1 and 2 showed clinical evidence of progressive visual impairment secondary to chiasmal compression. Radiologically, both had anterior extension of the pituitary tumor and nonspecific
Non-meningiomatous hyperostosis

### TABLE 1

Summary of seven cases of non-meningiomatous hyperostosis

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age, Sex</th>
<th>Ocular Findings</th>
<th>Radiological Findings</th>
<th>Histological/Pathological Diagnosis; Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 56 F</td>
<td></td>
<td>lt temporal hemianopsia, optic atrophy; lt superior temporal quadratic anopsia</td>
<td>x-ray film: irregular hyperostosis in planum sphenoidale; slightly enlarged pituitary fossa; brain scan: positive; arteriogram: suprasellar mass with with no tumor blush</td>
<td>chromophobe adenoma (2 × 2 cm mass in anterior suprasellar region); surgery and radiation therapy</td>
</tr>
<tr>
<td>2 65 F</td>
<td></td>
<td>total blindness optic atrophy, amaurotic pupil on rt; temporal hemianopsia, history of open angle glaucoma on lt</td>
<td>x-ray film: hyperostosis in the tuberculum sellae with slightly enlarged pituitary fossa; arteriogram: suprasellar mass with no tumor stain</td>
<td>malignant chromophobe adenoma (2.5 × 2.0 cm); surgery</td>
</tr>
<tr>
<td>3 42 M</td>
<td></td>
<td>slowly progressive visual loss for 2 yrs; rt optic atrophy, massive temporal field defect; blindness, optic atrophy on lt</td>
<td>x-ray film: thick and sclerotic planum sphenoidale and floor of the sella; brain scan: positive; arteriogram: suprasellar mass with no tumor blush</td>
<td>craniopharyngioma (4.0 × 2.5 cm); surgery and radiation therapy</td>
</tr>
<tr>
<td>4 64 M</td>
<td></td>
<td>total blindness in 6 wks, optic atrophy on rt; progressive visual loss, inferior altitudinal scotoma, normal disc on lt</td>
<td>x-ray film: hyperostosis of tuberculum sellae and planum sphenoidale; tomogram: partial destruction of floor of sella with soft tissue mass in the sphenoid sinus; arteriogram: normal</td>
<td>adenocarcinoma of sphenoid sinus (trans-sphenoidal stereotaxic biopsy); radiation therapy</td>
</tr>
<tr>
<td>5 13 M</td>
<td></td>
<td>occipital headaches and diplopia; on rt progressive ptosis, dilated and fixed pupil, upward and downward gaze impairment, normal fundus and visual field; lt eye normal</td>
<td>x-ray film: faint calcific density in rt parasellar region adjacent to anterior clinoid process; tomogram: 1.5 cm calcific or bony mass attached to the right anterior clinoid, sclerotic planum sphenoidale; brain scan: positive; arteriogram: medial and posterior displacement of carotid siphon with no tumor blush</td>
<td>chondroblastoma (anterior clinoid); surgery</td>
</tr>
<tr>
<td>6 47 F</td>
<td></td>
<td>blurred vision and pain for 2 wks, pale, edematous disc with vascular engorgement, superior orbital fissure syndrome on rt; lt eye normal</td>
<td>x-ray film: sclerosis of right sphenoid wings; arteriogram: irregular segmental stenosis of right carotid siphon with proximal occlusion, elevation of M-1; orbital venogram: nonfilling of the right superior ophthalmic vein</td>
<td>thrombosed cavernous sinus aneurysm (2.5 × 3.5 cm); surgery</td>
</tr>
<tr>
<td>7 38 F</td>
<td></td>
<td>progressive exophthalmos for 6 mos on lt; rt eye normal</td>
<td>x-ray film: sclerosis of lt orbit; tomogram: dehiscence of sphenoid wing; pneumo-orbitotomography: cystic mass with dark brown fluid; arteriogram: normal</td>
<td>epidermoid tumor (3.0 × 4.0 cm) of the lt orbit; surgery twice</td>
</tr>
</tbody>
</table>

Enlargement of the pituitary fossa. The radiological diagnosis of meningioma was made on the basis of irregular hyperostosis involving the tuberculum sellae and planum sphenoidale; there was evidence of a suprasellar mass in both cases. Blistering and serration, which are considered to be pathognomonic of meningioma, were not present in either case. Neither case showed the vasculature typical of meningioma; chromophobe adenomas occasionally demonstrate meningioma-like tumor blush.
Case 2

Left: The pituitary fossa of the sella turcica is slightly enlarged, and has a double floor. An irregular hyperostosis is noted involving the tuberculum sellae and anterior margin of the pituitary fossa (arrows). Right: Hanging-head midsagittal tomographic section of pneumoencephalogram. The sellar hyperostosis can be seen (arrows). The rostral end of the third ventricle is amputated (arrowheads) by suprasellar extension of a mass. Inset: Radiograph of the specimen demonstrating the sellar hyperostosis secondary to bone invasion by malignant chromophobe adenoma.

Case 3

A 42-year-old man was admitted with a 2-year history of slowly progressive blurred vision. There was temporal hemianopsia of the left eye, and the right eye was blind. The fundi showed pallor of both optic discs.

Skull films and tomograms showed slight irregularity involving the tuberculum sellae, planum sphenoidale, and the floor of the pituitary fossa (Fig. 3). A right carotid arteriogram revealed marked elevation of the horizontal portion of the anterior cerebral artery in arcuate fashion, associated with draping of the orbital branches and indicative of an extra-axial tumor. No tumor blush was present. A preoperative diagnosis of tuberculum sellae meningioma was made, and a frontal craniotomy was performed. At surgery, a 4 x 3 x 2.5-cm encapsulated reddish-gray nodular mass with cystic components was totally resected.

Fig. 3. Case 3. Left: Lateral skull film reveals irregular hyperostosis along the undersurface of the planum sphenoidale (lower arrows). The tuberculum sellae is irregular in outline (upper posterior arrows). The distal end of the planum sphenoidale is thickened (anterior arrow). Right: Midsagittal tomogram confirmed the irregular hyperostosis of the tuberculum sellae and planum sphenoidale (arrowheads). An irregular notch is seen in the posterior aspect of the tuberculum sellae secondary to bone erosion which was confirmed at surgery.
Non-meningiomatous hyperostosis

The histological diagnosis was craniopharyngioma. The patient was given a course of radiation therapy, but he died from anaphylactic shock during an ACTH injection. Postmortem examination revealed necrosis of the hypothalamus and septum pellucidum. Residual craniopharyngioma was found in the sphenoid sinus and pituitary fossa.

Comment

Barnett reported 62 cases of craniopharyngioma, of which seven (11%) demonstrated an increase in the density of the sella turcica. No mention was made of the planum sphenoidale, however. The pathogenesis of hyperostosis of the tuberculum sellae, planum sphenoidale, and sellar floor is unknown. At autopsy, the craniopharyngioma in our Case 3 involved the area both above and below the sella turcica in a dumbbell fashion. A longstanding impairment of venous drainage secondary to craniopharyngioma may be a contributing cause of hyperostosis. In retrospect, there was no evidence of serration or blistering in the planum sphenoidale.

Case 4

A 64-year-old man was admitted because of sudden loss of vision in the right eye for 6 weeks. His visual acuity showed no light perception in the right eye; the left eye had 30/30 vision with +4 correction. There was pallor of the right optic disc. An inferior altitudinal scotoma of the left eye was present.

A routine skull x-ray film showed irregular hyperostosis extending from the tuberculum sellae to the planum sphenoidale with slight cortical irregularity of the pituitary fossa (Fig. 4 left). Midsagittal tomograms revealed a destructive lesion in the pituitary fossa associated with a soft tissue mass within the sphenoid sinus, in addition to the hyperostosis of the planum sphenoidale and tuberculum sellae (Fig. 4 right). Biopsy of the sphenoid mass revealed undifferentiated carcinoma, of either glandular or epidermoid origin.

Comment

The value of tomography is evident in this case. The tomograms clearly demonstrated a small destructive lesion in the pituitary fossa associated with a soft tissue mass in the sphenoid sinus, highly suggestive of malignant tumor originating in the sinus. The lesion proved to be poorly differentiated adenocarcinoma. The hyperostosis and the tuberculum sellae and planum sphenoidale remained unchanged throughout the entire course of the disease from 1968 to 1971, despite progressive tumor extension to the

![Fig. 4. Case 4. Left: Lateral skull film reveals a diffuse hyperostosis involving the tuberculum sellae and planum sphenoidale (arrows). There is bone destruction in the floor of the pituitary fossa. Right: Midsagittal tomogram showing erosion of the pituitary fossa (arrow) and a soft tissue mass (T) in the sphenoid sinus secondary to carcinoma with invasion of the pituitary fossa. Diffuse reactive hyperostosis of the planum sphenoidale and tuberculum sellae is again demonstrated (arrowheads).](image)

Fig. 5. Case 5. Left: The lateral tomogram of the sella turcica in the right parasagittal plane reveals an irregular hyperostosis in the anterior aspect of the sella with obliteration of the anterior clinoid process and tuberculum sellae (arrowheads). Right: Tomogram of coronal section reveals an egg-shell calcification (upper arrows) in the superior margin of the tumor, associated with erosion of the lateral aspect of the pituitary fossa and wall of the sphenoid sinus (lower arrow).

Case 5

This 13-year-old boy was admitted because of intermittent throbbing occipital headaches and progressive drooping of the right eyelid for 2 years. He also had diplopia and a ptosis on the right. The right pupil was dilated and fixed and did not react to light or accommodation directly or consensually. Impairment of upward and downward gaze was present on the same side.

A skull film showed a faint calcific density in the right parasellar region adjacent to the anterior clinoid process. Tomograms revealed a calcific or bony mass, measuring approximately 1.5 cm in the right parasellar region, arising from the anterior clinoid process, and associated with hyperostosis of the planum sphenoidale (Fig. 5). A right carotid arteriogram showed medial and posterior displacement of the carotid siphon without tumor blush.

A preoperative diagnosis of meningioma was made and right frontotemporal craniotomy was performed. At surgery, a firm cartilaginous mass 1.5 cm in diameter was found attached to the anterior and lateral aspect of the right anterior clinoid processes adjacent to the greater wing of the sphenoid. The tumor was covered with dura, and about 90% of the tumor was excised. The histological diagnosis was chondroblastoma.

Comment

Chondroblastoma is an uncommon cartilaginous tumor of the epiphysis which usually involves the long bone in the young age group. Only two cases of this tumor originating in the skull have been reported and the temporal squama was the site of involvement in both. Meningiomas, chordomas, and chordomas all may calcify. In our case, a calcified tumor was attached to the anterior clinoid process and adjacent planum sphenoidale. This is a frequent site for meningioma, but an unusual one for chondroma or chordroma. Chordomas frequently destroy the sphenoid bone and oc-
Non-meningiomatous hyperostosis

Fig. 6. Case 6. Radiograph showing postero-anterior view of the skull reveals homogeneous sclerotic density involving the greater wing of the right sphenoid bone, suggestive of meningioma. The sclerosis was confirmed by tomography. Occasionally calcify.\(^\text{16}\) Again, absence of the serration and blistering in the planum sphenoidale in this case suggests that it is less likely to be a meningioma.\(^\text{14}\)

Case 6

A 47-year-old woman was admitted because of blurred vision and pain in her right eye for 2 weeks. A pale edematous disc was demonstrated, and right lateral and medial rectus palsy with mild ptosis were present.

Skull films and tomograms showed homogeneous sclerosis in the right greater wing, suggestive of meningioma (Fig. 6). An orbital venogram revealed nonfilling of the superior ophthalmic vein on the same side. A right brachial arteriogram revealed an irregular, segmental narrowing and occlusion of the right carotid siphon with elevation of the main trunk of the middle cerebral artery, indicative of a mass lesion in the right parasellar region.

A preoperative diagnosis of sphenoid meningioma was made. At surgery, an oval-shaped intracavernous carotid aneurysm measuring 2.5 × 3.5 cm was found. A typical laminated blood clot was evacuated after the aneurysmal sac was opened. The bone sclerosis persisted after surgery.

Case 7

A 38-year-old woman was admitted with a 6-month history of progressive exophthalmos of the left eye.

A skull film showed diffuse sclerosis in the left orbital margin and sphenoid wings suggestive of meningioma (Fig. 7 left). On tomogram, however, a bony dehiscence in the roof of the orbit was clearly demonstrated suggesting an epidermoidoma or dermoid tumor.\(^\text{19}\) During air orbital-tomography, 6 cc of dark brownish, oily fluid was aspirated from the cystic mass, which was well delineated by air (Fig. 7 right). The aspirated

Fig. 7. Case 7. Left: Plain skull film shows diffuse hyperostosis of the right orbit. A coronal tomographic section through the superior orbital fissure reveals diffuse sclerosis involving the medial aspect of the greater wing and inferior aspect of the lesser wing suggestive of a sphenoid ridge meningioma (arrowheads). A dehiscence or bone defect is seen in the orbital roof posteriorly (arrows). Right: Lateral tomogram with orbital air insufflation reveals a soft tissue mass in the apex of the orbit (arrowheads) associated with a dehiscence in the orbital roof (arrows) secondary to an epidermoid.
fluid contained a large amount of cholesterol crystals.

At frontal craniotomy, a cystic reddish-blue mass was found extruding through the posterior wall of the orbit into the subfrontal region on the left side. The histological diagnosis was epidermoid tumor (cholesteatoma) originating in the left orbit.

Comment

In Case 6 there was evidence of a cavernous sinus superior orbital fissure syndrome secondary to a large thrombosed carotid aneurysm in the cavernous sinus. The pathogenesis of bone sclerosis of the right greater wing is uncertain. It is less likely to be a meningioma, in retrospect, due to the fact that there was no bone proliferation associated with sclerosis of the greater sphenoid wing. Similarly, Case 7 showed no evidence of bone proliferation of the sphenoid wings. This case showed evidence of dehiscence in the sphenoid wing, which is highly suggestive of epidermoid or dermoid tumor.15 Accidental aspiration of the fluid during orbital tomography confirmed the diagnosis. The sclerosis of the lesser and greater sphenoid wing and orbital margin is certainly reactive in nature, but its pathogenesis is again uncertain. In both cases, absence of bone proliferation of the sphenoid wing would eliminate the possibility of meningioma.

Discussion

It is well known that meningioma is the most common cause of hyperostosis in the juxtasellar region.4,11,14 "En plaque" type meningioma usually causes a sheath-like, smooth, homogeneous ivory sclerosis in the sphenoid ridge.4,25 Midline subfrontal meningiomas generally produce irregular hyperostosis in the planum sphenoidale, and/or tuberculum sellae.5,14 Most tuberculum sellae meningiomas actually originate from the planum sphenoidale. Blistering is a frequent sign of planum sphenoidale meningioma. This effect (a bubbly radiolucency within the hyperostosis) is due to herniation of air cells (usually from the ethmoid sinus but occasionally from sphenoid sinus) within the bone sclerosis, and is considered to be virtually pathognomonic of meningioma in this region.14,16 It does not occur in the calvaria or base of the skull other than at the midline anterior fossa.

We made retrospective and prospective studies of 66 patients with midline subfrontal meningioma. Over 90% of these patients showed varying degrees of hyperostosis in the planum sphenoidale and/or tuberculum sellae associated with visual disturbance.14

The pathogenesis of meningiomatous hyperostosis remains unclear. Histological sections of the hyperostotic bone demonstrated infiltrating meningioma cells in most cases; in some cases, however, no tumor cells were identified.4,12 Phemister,20 Cushing,4 and Penfield18 stressed that meningioma cells in bone spaces may stimulate osteoblasts to cause new bone production. Kolodny13 suggested that the hyperostosis of meningiomas is due to the stimulus of slowly progressing local dilatation of vascular channels in the bone, and is not directly related to the infiltrating tumor cells. He believed that bone proliferation usually precedes the tumor cell infiltration. Both theories suggest that new bone formation is due to reactive change, and is not an integral part of the tumor. Hyperostosis or bone sclerosis may occur with primary osteogenic sarcoma,6 chondroma,2,8 chondroblastoma,8,10 fibrous dysplasia,12 osteoblastic metastases from carcinomas23,24 (usually from prostate and breast), lymphomas (especially Hodgkin's disease), chronic infections,9 heavy metal poisoning, vascular stasis (chronic thrombophlebitis), or trauma.7,12 The pathogenesis of the sclerotic reaction demonstrated in our patients with craniopharyngioma, epidermoidoma, and thrombosed cavernous aneurysm may be explained on the basis of venous stasis.

Special caution should be exercised in evaluating juxtasellar hyperostosis since the correct diagnosis of meningioma is often established solely on the basis of typical hyperostosis, and the contrast studies may be nonspecific. Although hyperostosis of the planum sphenoidale accompanied by the chiasmal syndrome is highly suggestive of meningioma, a variety of non- meningiomatous lesions described in this paper can also produce similar hyperostosis. Blistering, nodular calcifications, and serration are frequently associated with midline subfrontal meningioma; these signs were absent, however, in the non-meningiomatous lesions that we have presented here.

Sphenoid meningioma should demonstrate
thickened sphenoidal wings when they are sclerotic. The absence of bone proliferation and the presence of sclerosis virtually eliminates the possibility of meningioma. Therefore these signs are considered to be extremely useful in differentiating true meningioma from non-meningiomatous conditions. Meticulous polytomographic study is required to demonstrate both salient and subtle bone changes. Typical meningioma blush with dural blood supply is virtually pathognomonic of meningioma; none of our cases with non-meningiomatous hyperostosis demonstrated meningiomatous tumor blushes or hypertrophic meningeal vessels.

References

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