Meningiomas in the cranio-orbital junction

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The author analyzes 20 cases of meningioma in the cranio-orbital junction and concludes that they represent an anatomical continuum of neoplasia rather than a single specific syndrome. Different manifestations of this continuum are identified and characteristic cases presented to illustrate the surgical management of each type of tumor. Some lend themselves to total excision, but unilateral blindness postoperatively may be the price. Unless radical operative procedures are curative they cannot be justified on the basis of short-term evaluation, since many of these tumors grow very slowly.

Key Words: meningioma □ cranio-orbital junction □ optic canal □ anterior clinoid process □ blindness □ exophthalmos

Meningiomatous growths may be found anywhere in the anatomical complex that comprises the cranio-orbital junction. Meningiomas in this region traditionally have been classified by anatomical boundaries or according to a limited clinical syndrome. This paper introduces the concept of an anatomical continuum of lesions delineated by three groups of tumors based on their location. Characteristic specific and transitional cases illustrate the groups. The not-too-rigidly defined groupings have important diagnostic and therapeutic considerations.

Analysis of Cases

Tumors of the Periforaminal Anterior Clinoidal Area (Group A)

The first group is illustrated by three cases, each involving a female (aged 15 to 29 years), with a history of painless, unilateral, progressive visual failure of 6 to 30 months' duration. Visual acuity in the affected eye ranged from 20/100 and J-14 to perception of finger movement only; in each the optic nerve was pale. Plain radiological skull series, foraminal views, and laminographic studies all were abnormal (Figs. 1-5), but the more sophisticated contrast studies of pneumoencephalography and carotid angiography were, in each instance, quite normal.

Operative findings were similar in each patient (Fig. 6): the anterior clinoid process was swollen and bulbous and, together with the adjacent region of the tuberculum sellae, was covered with abnormally vascular dura mater. Under the tumor-swollen overhang of the clinoid process was a suffused, reddish-purple optic nerve. The nerve was cuffed with tumor at the foramen and, in one patient, tumor involved the intimate epicarotid tissue. Tumor occupied the proximal optic canal and, together with the abnormal bone, some of which was invaded by tumor, was responsible for compression of the nerve.
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Operative Considerations. The dura mater was circumcised around the bulbous clinoid process and, together with the abnormal bone, was removed close to the superior orbital fissure where the oculomotor nerve was visualized. The optic canal was unroofed, and tumor was removed from within and from the periforaminal area. In two of the three cases, the nerve was sacrificed in favor of what appeared to be, grossly, a total excision; in these cases, postoperative ptosis abated within 4 months. In the one patient in whom tumor was known to have been left behind in the canal, there was no clinical change in the remaining nasal vision 5 years later, but a persistent hyperostosis of the medial wall of the optic canal was seen on radiological study.

Transitional Cases. The first group is comprised of tumors limited to the periforaminal area, the anterior clinoid process, and the proximal optic canal. Two patients illustrate the concept of a continuum of neoplastic growth extending beyond this region.

The first patient was a 35-year-old woman who presented, as did the others, progressive loss of vision in one eye of 8 months' duration. She had no light perception and showed pallor and edema of the optic nerve head and 2 mm of exophthalmos (Hertel). The optic foramen on the affected side was 1 mm larger than that on the opposite side; plain skull films were otherwise normal. Tumor was encountered about the optic foramen, entirely surrounding the nerve and carotid artery. It extended over the lesser wing of the sphenoid bone into the anterior part of the middle fossa and into the sella turcica as well as into the orbit within the sheath of Schwalbe (Fig. 7). Despite incomplete excision, this patient is...
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working full time 14 years later with no evidence of advancing tumor growth. Such a patient is best served by an intracranial procedure before an enucleation, for it is the intracranial portion of the tumor that governs curative excision. In the absence of total intradural removal, the globe could be spared and, failing prominent exophthalmos, the patient would have a better cosmetic result.

The second transitional case, again with clinoidal and periforaminal attachment as the common denominator, showed an entirely different direction of growth. This 43-year-old man had a 6-year history of unilateral deteriorating vision and a 1-year history of convulsive seizures. He was blind in the affected eye with an accompanying optic atrophy. His cerebrospinal fluid (CSF) protein was 120 mg%, and he showed a left anterior temporal electroencephalographic (EEG) slow-wave focus. Total excision of the meningioma, which had grown into the temporal lobe, involved the anterior clinoid process and the medial portion of the sphenoid wing. The patient is well, although not working, 13 years later.

Discussion. By 1929 Cushing and Eisenhardt\(^9\) recognized that meningiomas can arise from the sphenoid ridge lateral to the sella turcica and implicate the adjacent optic and oculomotor nerves. In 1934 Alpers and Groff\(^2\) described a syndrome of such involvement of the sphenoid ridge, which
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was amplified by Groff in 1936.14 The ophthalmological syndrome of large meningiomas of the lesser wing of the sphenoid bone was presented by David and Hartmann in 1935.11 In their comprehensive treatment of the subject of meningiomas in 1938, Cushing and Eisenhardt9 reported 13 examples of tumors of what they termed the deep or clinoidal third of the sphenoid ridge. The smallest tumor in their series presented clinically with the now-familiar complex of slight exophthalmos, early extraocular muscle paresis, nasal hemianopsia, and hyperostosis of the anterior clinoid encroaching on the optic foramen.

In 1953 Uihlein and Weyland22 reviewed 310 cases with unilateral loss of vision and found that, in 108, lesions of the optic nerves or visual pathways were the cause of blindness; 15 of these were due to meningiomas. Over a 20-year span, they collected 52 cases of meningiomas arising from the region of the anterior clinoid process; unilateral loss of vision was the initial symptom in 30 and was also total in 30. In 14 of their cases an osteoma involved the region of the anterior clinoid process.

These tumors, as described in the reports cited, appear to warrant a place within the spectrum of meningiomas involving the area

Fig. 5. Group A. Radiographic views of optic foramina. Note the sclerosis about the right foramen.

Fig. 6. Group A. Sketch of operative exposure of right tuberculum and clinoidal area. Tumor surrounds the right optic nerve and extends to the midline. Note abnormal vascularity of dura mater.

Fig. 7. Group A. Transitional Case 1. Equatorial section of optic nerve and globe. Tumor is seen within the sheath of Schwalbe surrounding the nerve.
of the optic canal but represent cases of much greater intracranial magnitude and, therefore, have different clinical implications. None was as focally delimited as those of our first group. However, Elsberg and Dyke\textsuperscript{13} described cases strikingly similar to the three in our first group. Although their patients had unilateral visual impairment and optic nerve atrophy, the radiological features were different. Instead of a hyperostosing reaction in the clinoidal area, atrophy or shortening of the anterior clinoid process was noted, together with encroachment on the interpeduncular cistern as demonstrated on pneumoencephalographic studies. It is not clear why Craig and Gogela\textsuperscript{2} wished to distinguish the foraminal meningiomas from those described by Elsberg and Dyke,\textsuperscript{13} for, except for the bilaterality of certain of the former tumors, the two reports seem to support the concept of a continuous spectrum of meningiomatous growth in this area.

When Dandy\textsuperscript{10} discussed orbital tumors in 1941, he presented the case of a 13-year-old girl with bilateral optic nerve involvement from meningiomas cuffed about each nerve and extending into the orbits. Blindness and roentgenographic signs were the important diagnostic features. Despite incomplete excision of the tumors, this patient was reported in good health, apart from her blindness, 18 years later. This report, as well as that by Craig and Gogela,\textsuperscript{2} provides evidence of the slow growth of these lesions.

"En Plaque" and Sclerosing Tumors of the Sphenoid Wing Region (Group B)

Eleven patients in this second group were traced and reviewed for this study; nine had had surgical confirmation of the meningiomatous nature of the lesion. Tumors categorized in Groups A and B had the common denominators of clinoidal or foraminal attachment and devastating involvement of the optic nerve at the cranio-orbital junction. Focal bone reaction was a prominent feature in some, but not all, and encroachment upon the orbital volume with resulting exophthalmos was dependent upon orbital extension of the tumor through the canal via the nerve sheath. The major distinguishing features of Group B are the following: the clinoidal and foraminal areas represent only a fraction of the involved site; progressive exophthalmos is the rule and often constitutes the primary complaint; and disturbances in visual acuity may be early in a few but are late in most. Distortion of the temporal bone, headache, and, occasionally, seizures complete the familiar syndrome. Bone reaction in this group is one of the hallmarks of the disease. Tomographic examination and radioisotope scanning have proved helpful in studying the extent of bone involvement but do not regularly distinguish reactive bone from tumor-infiltrated bone. The dura mater is invaded by a low-growing carpet of tumor similar to but of much wider extent than the low profile lesions encountered in the first group. Differential internal and external carotid angiography are especially helpful in delineating the extent of any intradural globular tumor mass as well as the vascular pattern of the hyperostotic portion of the process (Fig. 8).

Among the nine patients operated on in this group, there were no surgical deaths. However, except for improvement in the exophthalmos in certain cases, it is questionable whether operative therapy changed the course of the disease, and presumably no cures have been effected (Table 1).

The extent of hyperostotic bone reaction in this group is disproportionate to the amount of tumor present, as judged by meningiomas in most other locations, and the distribution of this bone response, as
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TABLE 1
Summary of findings in 11 cases of sclerosing en plaque sphenoid wing meningiomas

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Duration of Symptoms Before 1st Definitive Operation (yrs)</th>
<th>Date of 1st Definitive Operation</th>
<th>Dates of Subsequent Operations</th>
<th>Results</th>
<th>Follow-up from 1st Procedure (yrs)</th>
</tr>
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<tr>
<td>1</td>
<td>&lt;1</td>
<td>1935</td>
<td>1935, 1943</td>
<td>slight progression in last 16 yrs</td>
<td>36</td>
</tr>
<tr>
<td>3</td>
<td>10</td>
<td>1951</td>
<td>1955, 1960</td>
<td>progression, visual loss</td>
<td>&gt;10</td>
</tr>
<tr>
<td>4</td>
<td>2</td>
<td>1955</td>
<td>none</td>
<td>mild progression</td>
<td>20</td>
</tr>
<tr>
<td>5</td>
<td>2</td>
<td>1961</td>
<td>none</td>
<td>visual loss</td>
<td>10</td>
</tr>
<tr>
<td>6</td>
<td>2</td>
<td>1963</td>
<td>none</td>
<td>exophthalmos improved</td>
<td>8</td>
</tr>
<tr>
<td>7</td>
<td>2</td>
<td>1964</td>
<td>none</td>
<td>progression</td>
<td>7 (died)*</td>
</tr>
<tr>
<td>8</td>
<td>&lt;1</td>
<td>1968</td>
<td>none</td>
<td>cosmetic improvement</td>
<td>3</td>
</tr>
<tr>
<td>9</td>
<td>2</td>
<td>none</td>
<td>none</td>
<td>unchanged</td>
<td>&lt;1</td>
</tr>
<tr>
<td>10</td>
<td>6</td>
<td>1971</td>
<td>none</td>
<td>exophthalmos improved</td>
<td>&lt;1</td>
</tr>
<tr>
<td>11</td>
<td>15</td>
<td>none</td>
<td>none</td>
<td>unchanged</td>
<td>&lt;1</td>
</tr>
</tbody>
</table>

*Death due to unrelated causes.

seen roentgenographically, may be quite misleading if considered as indicative of the presence of tumor. Although tumor excision does not seem to reverse the bone reaction, the disparity between tumor mass and bone response may be part of the explanation for the protracted, and relatively innocent, clinical course. Irrespective of the osseous response, the location of these tumors in the cranio-orbital junction places them within our continuum of meningeal neoplasia.

Operative Considerations. That mass effects do justify operation and may occur late in the course of the disease is suggested from the report by Abbott and Glass who operated on their patient after a 36-year history of tumor. The evidence suggests that if a clinically significant intradural mass lesion can be excluded, operative treatment or orbital decompression is primarily cosmetic. As emphasized by Guiot and Derome it seems likely that complete excision and therefore cure can only be accomplished with lesions lying laterally in the pterional region which are diagnosed early. There is inadequate evidence on this point, however, just as there are insufficient follow-up data to validate the concept that the proportion of tumor removed determines recurrence.

When operative treatment is undertaken, a low-lying frontotemporal osteoplastic flap is turned, and both extradural and intradural attacks mounted. Intradurally, the carpet of tumor may be found surrounding the carotid artery and optic nerve. In such a case the bulk of soft tumor is reduced by the electrosurgical loop, and the operation completed extradurally. On only one occasion has our team found the dural tumor sufficiently limited in extent to hope for a cure by excision and dural grafting with freeze-dried homologous dura mater; even then, subsequent marginal examination of the specimen proved our hopes to be unfounded. The extradural procedure is similar, irrespective of the amount of intradural manipulation. It consists of
removal of the hyperostotic bone of the pterion, the sphenoid ridge medially to the superior orbital fissure, the roof, and, especially, the lateral wall of the orbit after the technique of Naffziger.\textsuperscript{18} The high-speed drill is of great value in scalloping away hyperostotic, vascular bone that may or may not be invaded by tumor. The value of unroofing the optic canal and removing any periorbital tumor plaque to aid in the control of exophthalmos has been emphasized by Guiot and Derome.\textsuperscript{15}

**Transitional Case.** The case of a 25-year-old woman with a family history of multiple neurilemmomatous and meningiomatous growths illustrates the anatomical continuum of meningeal neoplasia across the cranio-orbital junction as well as a biological variant of the en plaque tumors. In this case, the tumor failed to evoke the exuberant bone response characteristic of the group just discussed. The patient presented with unilateral ptosis, paresis of upward gaze, visual acuity reduced to finger counting only, and 4 mm of exophthalmos (Hertel) (Fig. 9). The nerve head was elevated, pale, and covered with engorged veins. On the affected side, the retrobulbar tension was increased and the corneal reflex severely impaired. Orbital venography disclosed upward displacement of the superior ophthalmic vein, and retrobulbar contrast studies showed an absence of filling in the orbital apex. Further investigation disclosed

![Fig. 9. Group B. Transitional case. Illustration of the exophthalmos and ptosis accompanying extensive intraorbital as well as intracranial meningiomas.](image)

an unsuspected ipsilateral frontoparietal globular tumor. At operation for the latter, an en plaque, 3 mm carpet of meningioma was also found that seemed to involve most of the right hemicranial dura mater. It extended into the floor of the middle cranial fossa and incorporated the lateral two-thirds of the sphenoid wing and the floor of the anterior fossa. One month later the orbital tumor was approached through an intracranial exposure. Tumor was found outside as well as within the sheath of Schwalbe, and intradural meningioma was encountered in the periforaminal area as expected. The patient's condition remained unchanged after these procedures. As with en plaque meningiomas, the precise site of origin for this patient's tumor(s) defies analysis.

Basofrontal meningiomas and those of the tuberculum sellae may secondarily involve the cranio-orbital interface. Cushing and Eisenhardt\textsuperscript{9} illustrated the extension of tumor within the optic nerve sheath intraorbitally from an olfactory groove meningioma. They posed the question of whether meningiomas of the sheath of Schwalbe arose there primarily or as an extension from an intracranial source; they proposed a foraminal site as most likely for such sheath tumors but presumed that primary intraorbital sheath tumors could occur without extension posteriorly to the optic canal. Their conclusion leads us to the last logical grouping in our spectrum.

**Discussion.** Cushing and Eisenhardt\textsuperscript{9} discussed 19 patients with en plaque tumors, in 16 of whom the duration of symptoms ranged from 1½ to 13 years and the neoplasms were verified surgically. Visual acuity was greatly reduced in three patients. There was one operative death, and reoperation was performed in three patients from 5 to 16 years later. Whereas exophthalmos was frequently reduced, seizures, if present, were not modified and little visual improvement occurred in the few patients in whom this was reported postoperatively.

Poppen and Horrax\textsuperscript{19} in 1940 reported their surgical experience with 10 such patients. (Nine of these were women; and all of Cushing's patients were women; however, cases of males are on record, and there is one among our group.) Poppen and Horrax reasoned that since, in advanced cases, these
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tumors are hazardous to excise, wide excision should be undertaken as early as possible. They assumed that the more radical procedure would give better results than would limited excision. Although their 10 patients had histories ranging from 3 months to 11 years, the longest postoperative follow-up period was just over 1 year, an inadequate time upon which to base such assumptions.

A less aggressive surgical attitude than that of Poppen and Horrax was recorded in 1952 when Castellano, et al., reported 25 patients, 15 of whom had the lesion verified surgically. There were two operative deaths and one late death; of the 12 survivors, seven were well without serious defects and five were unimproved or worse. Seven of their patients survived at least 10 years. Once again, the exophthalmos was the one disorder that improved (six patients) after operation. From comparative radiological studies, these authors reported that the hyperostoses grew extremely slowly, almost imperceptibly. A combination of cases from these three reports reveals an operative mortality of about 10%, a not insignificant figure to consider as one weighs the choices of therapy.

**Primarily Intraorbital Meningiomas**

*Usually of the Sheath of Schwalbe*

*(Group C)*

The notable features of the three patients who comprise this group are presented in Table 2. All three had ophthalmoscopic evidence of involvement of the nerve head, probably by tumor; in two, this was confirmed by pathological examination. A disturbing feature of one case was the surprisingly well-preserved visual acuity (also encountered in the case of Watson and Greenwood) that was necessarily sacrificed in the interests of tumor eradication.

**Operative Considerations.** Lesions in this group are best approached through a frontal craniotomy, for two reasons: 1) intradural exploration and the canalicular portion of the procedure can be accomplished, together with removal of the proximal portion of the lesion; 2) the intraorbital extent of the tumor, if any, can be well delineated and an excellent decompression afforded. When the globe is involved with tumor, the question of whether subsequent enucleation or exenteration is needed has not yet been resolved. One’s inclination is to eradicate the tumor, but long-term follow-up data may show this to be unnecessary. As a case in point, the patient in our Group C who had a subtotal excision may become a candidate for exenteration and/or further intracranial excision of her canalicular nerve and sheath.

**Orbital meningiomas** cause their mischief by pressure and, as is well recognized by most who have written on the subject, are characteristically slow in their evolution. The 30-year history of the case presented by Jensen is an example. In addition, they often remain undetectable, even many years after incomplete removal, a point to reflect on when visual acuity remains useful.

**Discussion.** The isolated cases reported by Coston, Dunn and Walsh, and Als were characterized by involvement of the optic nerve head. The frequent radiological development of normal optic foramina offers no guarantee that there may not be extension within the dural sheath into the optic canal. This point is clearly made by Dunn and Walsh, Reese, and Als, and is underscored by one of our cases.

Cushing and Eisenhardt painted what is recognized as the common clinical picture of a meningioma of the sheath of Schwalbe: a slow, painless, evenly progressive, unilateral protrusion of the globe, early blindness (although not always, as seen in our group), primary optic nerve atrophy, and retention of normal globe mobility (an observation not always confirmed). They noted the high frequency of preceding trauma and the large number of patients whose histories dated from preadolescence. In this regard it is of interest that Jackson considered these to be tumors of adult life and noted that the mass tended to be on the distal part of the intraorbital portion of the sheath, a point not in keeping with our experience (Fig. 10).

It should be clear that meningiomas may occur anywhere along the nerve sheath but also that intraorbital meningiomas may arise from other sources and have been reported as occurring bilaterally. They have not usually produced a focal radiological signature. However, current radiological techniques require us to qualify any comment as
TABLE 2

<table>
<thead>
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<th>Clinical Data</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
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<tr>
<td>age, sex</td>
<td>38 M</td>
<td>43 M</td>
<td>43 F</td>
</tr>
<tr>
<td>duration of symptoms</td>
<td>18 mos</td>
<td>18 mos</td>
<td>10 yrs</td>
</tr>
<tr>
<td>visual acuity</td>
<td>20/30</td>
<td>blind</td>
<td>blind</td>
</tr>
<tr>
<td>appearance of nerve head</td>
<td>blurred</td>
<td>pale plus abnormal mass</td>
<td>pale, elevated abnormal vessels</td>
</tr>
<tr>
<td>limitation of globe movement</td>
<td>no</td>
<td>no</td>
<td>yes</td>
</tr>
<tr>
<td>amount of exophthalmos</td>
<td>14 mm</td>
<td>not stated</td>
<td>16.5 mm</td>
</tr>
<tr>
<td>radiological studies of optic foramina</td>
<td>normal</td>
<td>normal</td>
<td>normal</td>
</tr>
<tr>
<td>tumor found in apical section of optic nerve</td>
<td>no</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td>tumor outside sheath in addition to within it</td>
<td>yes</td>
<td>yes</td>
<td>no</td>
</tr>
<tr>
<td>&quot;total&quot; excision</td>
<td>yes</td>
<td>yes</td>
<td>no</td>
</tr>
<tr>
<td>follow-up</td>
<td>8 yrs</td>
<td>8 yrs</td>
<td>2 yrs</td>
</tr>
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</table>

Conclusions

Analysis of the material of this survey suggests the concept that benign meningeal neoplasias in the cranio-orbital junctional area present as an anatomical continuum or spectrum. Such a concept will enable the clinician to recognize the potential areas from which and into which tumor may grow, to consider them in his preoperative examination, and to prepare for them in planning the surgical exposures.

In the past, convenient excision of artificial segments of the spectrum has permitted the description of various syndromes that have had their place in generating clinical recognition of the presence of tumors. However, these syndromes may be misleading when accepted as correlating closely with anatomical boundaries of tumor involvement. In truth, a given syndrome often is clinically "blind" to tumor involvement in another direction of the continuum. Depending upon the direction one moves along the continuum, another sign, symptom, or constellation of symptoms may be encountered to a greater or lesser degree.

The more we fill in the "blind" areas of our awareness, the more accurately we will be able to define each patient's disease, rather than forcing each case to conform to artificial compartmentalization.

Fig. 10. Group C. Equatorial section of optic nerve and globe. Tumor is seen distending the sheath and completely encircling the nerve. It extended into the optic canal.
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