Cylindromas of the base of the skull

Report of four cases

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Cylindromas are rarely encountered in the neurosurgical field. Four cases of this rare tumor are presented. Conventional and computerized tomography scanning were most useful in establishing the diagnosis; angiography did not provide any further information. Because these tumors are well demarcated from surrounding structures, even in cases of recurrence, surgery is the treatment of choice. Radiation therapy is useful in the postoperative management. Long survival times with multiple recurrences are the rule.

KEY WORDS • cylindroma • adenoid cystic carcinoma • skull tumor

Cylindromas are well known to otolaryngologists since they represent a significant percentage of the malignant tumors that they encounter. Neurosurgeons are less familiar with them and cylindromas have been the subject of only isolated case reports in the neurosurgical literature.

Originating from glandular tissue of epiblastic origin, cylindromas have been called by many different names, the most popular being “adenoid cystic carcinoma.” To conform to the definition of cylindroma, the tumor must be formed entirely of cylindromatous tissue and must be distinguished from islands of cylindromatous changes that may occur particularly in salivary glands. Histologically malignant, these tumors are characterized by a long clinical course with a delay as long as 20 years in some cases between the first clinical signs and therapy.

Despite combined surgery and radiation therapy, recurrence is usual but may be long delayed (5 to 10 years). Local recurrence is frequent since these tumors are locally invasive but distant metastases can occur, most often pulmonary but in rare cases cerebral. Four cases involving the sphenoid sinus are reported.

Case Reports

Case 1

This 35-year-old woman complained of orbital pain 2 years prior to admission. Horizontal diplopia ensued, followed by right-sided loss of visual acuity. When she was evaluated, blindness was total on the right side associated with severe supra- and intraorbital pain, and she complained of hearing loss on the right. The right-sided exophthalmos was not reducible and the right sixth nerve was completely paralyzed. Homolateral hearing loss was present. X-ray films showed erosion of the sella turcica with enlargement of the optic canal (twice the normal size). Carotid angiography showed forward displacement of the siphon. No neovascularity was identified.

At surgery, the tumor was found to involve the sphenoidal fissure and was extradural in nature. The cavernous sinus was invaded as well as the temporal fossa. The gross appearance was that of a neurinoma or meningioma. Histological study established the diagnosis of mucoid cylindroma. The patient received postoperative radiation therapy (60 Gy) resulting in a total neurological recovery. After a 4-year remission, local recurrence was deemed inoperable and the patient died 41/2 years after the original intervention.

Case 2

This 53-year-old man was operated on for a bronchial tumor in 1978. Histological study revealed that it was a cylindroma. After a 3-year remission, a local recurrence was treated successfully with laser reduction. At the time of the second surgery, the patient complained
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FIG. 1. Case 2. A: Skull plain film showing dense tumor in the anterior part of the sphenoid sinus (arrows). B: Orbital computerized tomography scan with contrast injection showing an enhancing tumor in the sphenoid sinus (arrow).

of decreased vision in the right eye. Tomography of the skull revealed a well-defined opacity in the right sphenoid sinus with involvement of the superior, medial portion of the orbit (Fig. 1A). Computerized tomography (CT) confirmed the presence of a well-defined spheno-orbital soft-tissue lesion (Fig. 1B).

At surgery in 1981, 3 years after the initial diagnosis, the tumor was easily accessible through a unilateral nasoethmoidosphenoidal approach. Tumor removal appeared total including ablation of the sphenoid mucosa. The lesion was poorly vascularized. The histological results were comparable to those of the tumor removed 3 years before. Postoperative radiation therapy was given. Following surgery and radiation treatment, the patient presented with a normal neurological examination. Four years following therapy, the patient remains in remission.

Case 3

In 1971, this 57-year-old man presented with nose bleeds, unilateral left-sided exophthalmos, trigeminal pain, and loss of vision in the left eye. Tomography revealed a lesion occupying the sphenoid sinus. A transsphenoidal operation was carried out with removal of a tumor that was white in color with well-defined margins. Ablation of the sphenoid mucosa was performed. The pathological specimen was typical for mucoid cylindroma. Postoperatively, radiation therapy was given with satisfactory clinical results.

After a 4-year remission, the patient presented with pain in the territory of the left trigeminal nerve. Tomography revealed tumor recurrence in the sphenoid sinus with scans similar to those obtained 4 years earlier. This time, however, ethmoid involvement was present with destruction on the left side and invasion of the cribriform plate. The tip of the petrous bone was also destroyed on the left. Angiography was negative for neovascularization. A combined transsphenoidal and frontal approach was used: again, cleavage planes were well defined and an impression of total tumor removal was obtained. Additional radiation therapy (50 Gy) was given. Clinically, the outcome was quite satisfactory with disappearance of pain. Unfortunately, recurrence occurred 2 years later and the patient died after a trial of chemotherapy which had no significant effect on the tumor.

Case 4

In 1972, this 29-year-old woman complained of nose bleeds and bifrontal headaches. On examination she presented with a space-occupying lesion of the sphenoid sinus with extension into Meckel's cave. Following biopsy, radiation therapy was initiated (50 Gy), resulting in shrinkage of the tumor and clinical improvement. In 1977, the tumor recurred, with more nose bleeds, amenorrhea-galactorrhea syndrome, and headaches. Tomography revealed a round mass filling the sphenoid sinus, with erosion of the sellar floor and the upper half of the clivus. At transsphenoidal surgery, the tumor appeared to have a well-outlined plane of cleavage but local invasion of the posterior superior aspect of the sphenoid bone made total removal unlikely. Histological study (Fig. 2) confirmed the diagnosis of cylindroma.

Following surgery, a total radiation dose of 70 Gy was delivered to the tumor site. The patient enjoyed a 2-year remission which was followed in 1979 by massive recurrence with involvement of the third through eighth cranial nerves. A frontotemporal craniotomy was then performed with subtotal removal of the tumor. Two
years later, in 1981, a second craniotomy was performed with partial removal of the lesion. Again, the tumor presented with a well-defined plane of cleavage, making its removal feasible except for the area of insertion on the sphenoid bone. The patient died 1 year after this third operation.

Discussion

Occurrence of cylindroma in the craniofacial area is not exceptional; 166 cases of orbital cylindromas have been reported in the literature. These lesions are usually located laterally (originating from the lacrimal glands), are diagnosed early because of the presence of exophthalmos combined with oculomotor deficits, and can be differentiated from posterior orbital involvement with the origin in the sphenoid sinus. An ethmoid localization is also well known, with 100 cases being published since the series of Oppenheim, et al. This type of cylindroma has been linked to woodworking.

On rare occasions, cylindromas can be found in the sphenoid sinus. We have been able to find reports of 10 such cases in the literature, to which we add our four cases. Most of these are characterized by headache, facial pain, and ophthalmological deficits. Some patients may present with epistaxis. The neurological complaints are related to the area of major involvement, such as the orbital apex, sphenoid fissure, cavernous sinus, and the tip of the petrous bone. The interval between onset and surgery was reported as 2.7 years in the literature as opposed to 1.3 years in our series. This delay is much shorter than in the case of salivary-gland cylindromas, which had an average delay of 7 years and some cases had a 20-year interval. These lesions are best outlined by radioisotope tomography and CT scanning, and combining both techniques will provide an anatomical delineation of tumor involvement. Angiography has never revealed a tumor blush and does not help in the diagnosis except in relation to indirect vessel displacement.

Chemotherapy (used in only one of our cases) has not proven to be an effective treatment. Reports regarding its use lack long follow-up review and only partial results have been reported. Radiation therapy alone, although proposed by some authors, does not appear to be curative; however, several reports mention the efficacy of this treatment modality as a palliative measure, and also in combination with surgical removal. Some cylindromas have been found resistant to radiation therapy.

Surgery remains the treatment of choice for these tumors. Excision, which should be as radical as possible, is facilitated by the fact that these tumors have a well-defined cleavage plane and are not highly vascular. The surgeon’s impression of “total” tumor removal must be tempered, however, by the high incidence of local recurrences. The frequency of local recurrence is difficult to establish from the literature because of differences in tumor size and location. Half of the recurrences occurred after an interval of 1 to 3 years. In our series, three patients presented with a local recurrence after a delay averaging 4 years; they were the cases with the largest lesions. Histological studies performed on bone insertions revealed tumor infiltration beyond the limits of the surgical excision. Studies on surrounding neurological structures revealed similar findings. Local recurrence does not contraindicate additional surgery since it provides further clinical remission and prolonged survival. Indeed, this kind of tumor is characterized by the frequency of recurrences (up to 10 in 22 years according to Debain, et al.). Our Cases 3 and 4 had satisfactory clinical improvement following recurrences, with survival times of 6 and 10 years despite the volume of the tumors and their invasive nature. As the disease progresses, however, the tumor recurs within shorter periods of time with wider invasion, and the repeated surgical procedure becomes less palliative.

The natural history of cylindromas is a prolonged course with distant metastases appearing unpredictably most often in the lungs (80% to 90% of cases). In our Case 2, the spread of the disease appears unusual. Indeed, the primary bronchial tumor occurred 3 years prior to sphenoid involvement. This case presented originally as a classical bronchial cylindroma. We think that the bronchial recurrence occurring at the same time as the sphenoid tumor must be considered as a local recurrence rather than metastatic disease from the sphenoid sinus. It seems that this patient presented either with two primary cylindromas or with sphenoid metastasis from a bronchial cylindroma. Neither occurrence has been reported previously in the literature.

In conclusion, cylindromas are best treated with surgical resection, at least at the time of first diagnosis. The resection should be as radical as feasible. This approach is aided by the easy cleavage plane of the
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tumor from surrounding tissue despite their invasive nature. Because of this local invasive characteristic, surgery should be followed by radiation therapy. Since these tumors remain amenable to surgical excision when they recur, repeated surgery is indicated to provide the patient with further clinical remission. The long survival times associated with multiple recurrences and repeated surgery should encourage the neurosurgeon to remain aggressive with this type of tumor.

References

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