Endoscopic endonasal resection of a primary intraosseous clival myxoma: illustrative case

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BACKGROUND An intraosseous myxoma is a rare, benign mesenchymal tumor that penetrates the bone. The occurrence of an intraosseous myxoma in the clivus is a unique presentation of the disease.

OBSERVATIONS The authors discuss the case of a 15-year-old male with a new diagnosis of a primary clival intraosseous myxoma presenting with cranial nerve VI palsy. This is the third documented case of this pathology occurring in the clivus. This patient was successfully treated with endoscopic endonasal resection of the tumor.

LESSONS Primary clival intraosseous myxomas are extremely rare, but nonetheless it is important to add it to the differential diagnosis of clival masses. This mass has a high risk of recurrence, and prior literature suggests gross total resection may improve chances of progression-free survival. However, further larger studies are needed to provide guidelines regarding proper management of this pathology.

Primary intracranial myxomas (PIMs) are extremely rare, benign infiltrative tumors that grow from embryonal mesenchymal cells. Myxomas are most commonly found in the atrium of the heart, but they may also be found in skeletal muscle, skin, subcutaneous tissue, bone, and the mandible.1 When found intracranially, myxomas can be differentiated into primary and secondary myxomas.2 PIMs are most frequently derived from the skull base, with case reports previously citing them occurring in the sellar fossa, temporal bone, and orbit.3-5 Secondary myxomas are usually due to metastatic emboli from cardiac myxomas.6 Although these tumors have been known to recur in the setting of cardiac myxomas, it is unclear what the recurrence rate is of PIMs. In addition, given the rarity of this tumor, there is little evidence supporting proper management of intracranial myxomas. In our present report, we illustrate a case of a pediatric patient with a large intracranial myxoma involving the clivus and left cavernous sinus that was treated via an endoscopic endonasal approach. The purpose of this article is to illustrate the variable presentation of this phenomenon and to provide possible management strategies for this pathology.

Illustrative Case

A 15-year-old male presented with headaches and blurred vision after he had been struck in the head by a baseball during practice. The patient initially saw his pediatrician, who diagnosed him with a migraine and referred him to ophthalmology and traumatic brain injury sports medicine for postconcussive symptoms. The finding of initial ophthalmological examination including visual fields and dilated fundoscopic examination was unremarkable. Two weeks after initial presentation, the patient went to see a sports medicine doctor, who noticed that the patient had impaired lateral gaze in the left eye and questionable ptosis. Magnetic resonance imaging (MRI) of the brain and orbits with and without contrast was completed, which showed a large clival mass extending into the left cavernous sinus (Fig. 1). The patient was subsequently referred to our institution for further neurosurgical evaluation. Initial neurological examination on presentation showed a left cranial nerve VI palsy and grossly normal visual acuity, without any other signs of cranial nerve dysfunction or focal neurological deficits. The patient was subsequently admitted to the

ABBREVIATIONS CTA = computed tomography angiography; MRI = magnetic resonance imaging; PIM = primary intracranial myxoma.

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floor and started on Decadron (Merck & Co.) 4 mg every 6 hours. Given the patient’s symptoms and the effect of the mass on the sella, endocrinology and ophthalmology were consulted. Endocrinological evaluation showed no signs of endocrinopathy. Ophthalmological assessment showed 20/20 vision in both eyes, with equal reactive pupils and evidence of a left cranial nerve VI palsy. The finding of a dilated fundoscopic examination was negative for papilledema. Given these findings, resection was offered via an endoscopic endonasal approach due to the medial location and extension of the tumor inferiorly into the clival recess. Computed tomography angiography (CTA) of the head and neck were completed for preoperative planning purposes. CTA showed close proximity of the mass to the bilateral internal carotid arteries and basilar artery (Fig. 2). Chest CT with contrast was also completed, the finding of which was negative for signs of metastatic disease.

Five days after initial presentation, the patient underwent endoscopic endonasal resection of his clival mass. The patient was positioned supine, and stereotactic neuronavigation was used as a surgical adjuvant. The right middle turbinate was infiltrated with 1% lidocaine with 1:100,000 epinephrine and then excised. The inferior aspect of the right superior turbinate was then incised, exposing the sphenoid ostium. After the sphenoid ostium was exposed, a right-sided nasoseptal flap was elevated. Subsequently, the sphenoid rostrum was drilled off in its entirety, exposing the tumor, which appeared to be eroding through the clivus. The tumor was internally debulked with dissection proceeding posteriorly down to the posterior fossa dura and then anteriorly to the pituitary gland dura. The lateral remnants of the tumor were then dissected bilaterally along the carotid arteries, leaving a thin layer of residual tumor along the internal carotids. Intraoperatively, there appeared to be abnormal infiltration of the surrounding sphenoid bone. This bone was resected until grossly normal-appearing bone was identified. Frozen pathology showed a low-grade myxoid neoplasm and no signs of chordoma. Given the frozen pathology, the decision was made to close. The procedural defect was closed with a nasoseptal flap. Permanent pathology was consistent with lesioned cells positive for epithelial membrane antigen and negative for cytokeratin, S-100, SOX10, smooth muscle actin, desmin, and progesterone receptor. The final pathological diagnosis was an intraosseous myxoma. The patient was discharged on postoperative day 1, and he was weaned off steroids over the course of 2 weeks. One week postoperatively, a multidisciplinary tumor board was held, and it recommended postoperative surveillance imaging without any adjuvant therapy. At the patient’s 2-week follow-up, his cranial nerve VI palsy had resolved. Repeat MRI at that time showed an area of possible residual tumor in the parasellar region close to the left carotid artery (Fig. 3). Currently, the patient is otherwise asymptomatic and will undergo repeat brain MRI in 6 months for radiological surveillance.

Discussion

Observations

Although primary myxomas most commonly occur in the heart, they can rarely also originate in the head and neck, typically in the mastoid, maxilla, and oral cavity.8 Rarely, these lesions can be primarily intracranial,
with 68 reported cases presented in the literature. When intracranial myxomas infiltrate the bone, they are defined as intraosseous, and they are most often found within the temporal bone. There are only three reported cases of this lesion occurring within the clivus.

Our case differs from those in the current literature due to the myxoma’s location in the clivus, as well as our use of an endoscopic endonasal technique for resection of a PIM in this area. There has only been one other documented case of an endoscopic endonasal approach to cranial myxoma resection, which was an extracranial myxoma located within the sphenoid sinus. This tumor’s presentation within the clivus is significant, given that chordomas classically also involve this area of the skull. In contrast to myxomas, chordoma management classically involves maximally safe resection and adjuvant proton beam therapy for optimization of survival. Therefore, understanding that a myxoma can present similarly to a chordoma is critical, because it can greatly impact clinical decision making and follow-up.

The diagnosis of an intracranial intraosseous myxoma is difficult, given its nonspecific clinical presentation. Clinically, myxomas can mimic other skull base lesions, such as chordomas, chondrosarcomas, and meningiomas. Radiographically, intracranial myxomas may present as a hypodense or isodense mass on CT, with bone destruction and varying enhancement patterns. On MRI, these lesions are often hypointense on T1-weighted images and hyperintense in T2-weighted images; however, they may also be heterogeneous in presentation. Histologically, myxomas classically present with hypocellular areas of satellite and spindle-shaped cells in a myxoid matrix, staining positive for vimentin and negative for cytokeratin and S-100.

Given the rarity of PIMs, there is little data regarding guidelines for management of these lesions. The largest cohort reported to date was completed by Weng et al. in 2019. In their study, they discussed the outcomes of 30 PIMs at their institution and systematically reviewed 35 other cases as well. Of the cases of PIM reported, only two patients had lesions located within the clivus. Of note, both of these patients died shortly after surgery of either intracranial hematoma or respiratory failure, suggesting that intracranial clival myxomas may be more surgically high risk. This makes management difficult, given that gross total resection is the only identified variable associated with increased progression-free survival in these patients. With regard to tumor recurrence, the reported rate of local recurrence of PIMs is roughly 25%, usually occurring within the first 2 years after treatment.

This high rate of recurrence has been attributed to the difficulty of gross total resection of these lesions due to their infiltration of the skull base. Given this high risk of recurrence, close radiographic follow-up is recommended for PIMs.

Although maximal surgical management has been shown to lead to decreased recurrence risk for primarily intracranial myxomas, the role of radiotherapy in this pathology is somewhat controversial. Multiple studies have shown that myxoid tumors are primarily insensitive to radiation therapy. Furthermore, some authors have suggested that radiation therapy may actually predispose these tumors to becoming secondarily neoplastic. However, although there is evidence of brain metastasis from cardiac myxomas, there are no documented reports of secondary malignancies from PIMs. Conversely, other studies have suggested that radiation therapy may be of benefit; however, these authors suggest that larger sample sizes are needed to accurately evaluate this treatment’s effect on tumor recurrence. Currently, there are no specific guidelines for the role of radiotherapy in managing primarily intracranial myxomas.

**Lessons**
We believe this is the third occurrence of a primary intraosseous clival myxoma. This case shows that intraosseous myxomas can mimic more commonly encountered lesions of the clivus. Despite its rarity, a clival myxoma should be included in the differential diagnosis of clival masses, because the management and treatment of this tumor can differ greatly from other neoplasms of the skull base, and it is associated with a high risk of recurrence. Prior data suggest that maximal safe gross total resection is associated with progression-free survival, whereas the role of radiotherapy remains controversial.

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Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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