Clival Incidentaloma

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CASE HISTORY

A 62-year-old male had experienced atypical dysuria and penile pain for 6 months. He had already seen a urologist and a neurologist. The urological and neurological evaluations had not identified the cause of his pain. He felt well and had no other complaints. His medical history was remarkable only for kidney stones for which he had undergone lithotripsy. The patient's physical and neurological examinations were completely normal. Magnetic resonance imaging (MRI) of the thoracic and lumbar spine, obtained previously, was also normal.

Computed tomography (CT) of the head showed a well-defined focus of bone erosion in the clivus slightly to the right of midline and opening into the midline through the posterior clivus. MRI of the brain showed a nonenhancing, sharply demarcated mass located in the midclivus, extending toward the prepontine cistern (Fig. 1). The mass was hypointense on T1-weighted images and hyperintense on T2-weighted images.

ACTUAL MANAGEMENT

The patient underwent a left retrosigmoid craniotomy for resection of the tumor. Intraoperative MRI-based image guidance (Stealth, Medtronic) was used. The cerebellopontine angle was accessed. The tumor was exposed by working between the

vestibulocochlear and cranial nerve IX, X, XI complex. The mass was grayish-white with a stalk extending from the clivus. It was firm and partially calcified. The mass was completely resected and a margin of adjacent tissue and bone was removed. Final pathological examination revealed a modestly cellular mass with clusters and strands of cells with myxoid stroma, which tested positive for keratin AE1/AE3 and S-100, consistent with a diagnosis of chordoma.

Postoperatively, the patient complained of bilaterally decreased hearing, which resolved by the time he returned to the office for suture removal. The patient was discharged to home on postoperative Day 6. Early follow-up MRI showed no residual mass. The patient was referred for gamma knife radiosurgery to the tumor bed.

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RESPONSES

This 62-year-old gentleman presented with a lytic clival lesion discovered during the workup for an

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Figure 1 (A) Axial CT (bone window) shows a focus of bone loss in the clivus that opens toward the pons. (B) Sagittal noncontrast T1-weighted MRI shows a well-circumscribed nodule in the clivus. (C) The mass, which is bright on this axial T2-weighted MRI, appears to extend into the subarachnoid space.

unexplained painful syndrome of the genitourinary area. On the available bone window CT scan, the lesion seems purely lytic without expansion of the adjacent clivus. Its borders are sclerotic and well defined except on its posterior aspect where the bony rim is interrupted and allows direct communication between the lesion and posterior fossa. Hounsfield unit measurements would be useful to determine whether the density within the lesion is identical to that of CSF. The lesion seems fully separated from the sphenoid sinus. No calcifications are visible. We assume that no other similar lesions are present anywhere else in the skull. The lesion is homogeneous on both T1- and T2-weighted MRIs and follows the intensity of CSF.

There is no contrast enhancement per report. Thoracic and lumbar spine MRIs were reported as negative; the cervical spine apparently was not imaged.

DIFFERENTIAL DIAGNOSIS

Several radiological characteristics of the lesion point to its probable benign nature: sharp and sclerotic margins, lack of expansive effect on the adjacent bone, absence of contrast enhancement, and absence of other lesions. The homogenous appearance of the lesion on MRI and the presence of a connection between the lesion and subarachnoid space raise the possibility that it could be an outpouching of the subarachnoid

space within the basioccipital portion of the clivus. Small meningoceles in the basiocciput are believed to present vestiges of the cephalic end of the notochordal canal (canalis chordae).¹ They have only been considered symptomatic in isolated cases in which the canal was responsible for a direct connection between the subarachnoid space and retropharyngeal area, causing multiple unexplained episodes of meningitis.² Other less likely diagnoses would be plasmocytoma/multiple myeloma (rendered unlikely by the isolated nature of the lesion), chondroma (very rare in the skull and usually associated with calcifications), and chordoma (very unlikely because of its usual heterogeneous signal, bone destruction, and frequent contrast enhancement). Our working diagnosis would therefore be a small basioccipital meningocele. As a matter of due diligence, we would evaluate the patient further to confirm that this lesion is isolated (bone scan). We would re-image it at 3 months to exclude a rapidly growing process such as a metastasis. If its size and appearance had not changed over 3 months, another clinical and radiological follow-up would be obtained at 1 year to plot the specific growth curve of the lesion.

The etiology of the genitourinary pain would still need to be determined, and a pelvic malignancy should be formally ruled out. This work-up would be deferred to urology.

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The author describes the case of a 62-year-old man who was found to have an incidental clival lesion after undergoing investigation of a urological problem. Even in retrospect, the lesion was totally asymptomatic.

The imaging is quite characteristic of a clival ecchordosis physaliphora. These are thought to be remnants of the primitive notochord and invariably remain asymptomatic. They are often in the midline, at the dorsal-most aspect of the clivus and closer to the foramen magnum than to the pituitary fossa. Extension into the subarachnoid space does not preclude the diagnosis. Indeed, ecchordoses physaliphorae that appeared to be hanging from the dorsal clivus by a thin stalk have been documented. Ecchordoses physaliphorae are radiolucent on CT and may have sclerotic edges. Their MRI features are similar to those of chordomas, and this tumor presents the greatest diagnostic challenge to the clinician. Ecchordoses physaliphorae are hypointense on MRIs and hyperintense on T2-weighted MRIs. They do not enhance, unlike the other notochord remnant, the chordoma, which certainly can enhance with gadolinium.

My recommended management for this patient is to repeat MRI after 6 months. If there is no clinical or radiological progression, these benign lesions can be left alone. If the patient becomes symptomatic or if enhancement or significant growth of the lesion is apparent, excision or biopsy is indicated. I would recommend the endoscopic transnasal approach. This technique provides access to the entire skull base from the anterior cranial fossa floor to the junction between the mid and lower third of the clivus. With the 30-degree angled endoscope, more laterally placed skull base lesions can also be accessed. This minimally invasive approach is well tolerated by patients who experience little postoperative discomfort and few complications.

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