

CASE REPORT

The nerve of legal entrapment

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Abstract

Chordomas are a rare type of bone tumor that arises from the embryological remnant of the notochord. They originate at any point along the axial spine with the sacrum and the skull based region being the most commonly affected sites. Chordomas are slowly growing, indolent tumors, presenting insidiously, but also carry a high recurrence rate with a tendency to invade contiguous structure making their treatment challenging. The current standard of care for localized chordoma is aggressive cytoreductive surgery followed by high dose adjuvant radiotherapy. We present a unique case of a 72-year-old lawyer with a skull base chordoma invading into the hypoglossal canal and causing isolated hypoglossal nerve paralysis.

INTRODUCTION

Chordomas are usually slow-growing tumors, with a tendency to invade locally [1–7]. This cancer can arise at any point along the spinal cord with the sacrum being the most commonly affected (50–60%) followed by the base of the skull where the annual incidence is estimated to be 0.089 per 100,000 and whose overall median survival time is 9.2 years after appropriate therapy [1, 2, 5–7,8]. Given the subtle presentation, diagnosis can be challenging, and management of this exceedingly rare disease is still evolving. In February 2015, the European Society for Medical Oncology (ESMO) published the first guidelines for the diagnosis and treatment of chordoma in *Lancet Oncology* [6].

CASE REPORT

A 72-year-old male attorney presented to the hospital complaining of worsening headache, neck pain and difficulty with speaking. His prior medical history was significant for hypertension, dyslipidemia and well-controlled type 2 diabetes mellitus. In the patient's words:

I would hope that you would report my complaints preceding August 6, 2016, diagnosis relating to stiffness and pain in my

neck, headache, and pain in primarily my right shoulder. This began, as best I remember, about a year and a half prior to the diagnosis. These complaints were pronounced when I saw you, as I recall, in May 2016. They appear to be similar or the same as I have experienced since the diagnosis. While these are rather common ailments for a person my age and do not generally suggest a malignant tumor, it seems to me that my complaints might lead a medical provider to consider the possibility of a type of Sarcoma, including Chordoma, possibly leading to an early diagnosis. For a few weeks prior to August 6, I woke earlier than desired on a regular basis. On August 4, I realized that I had been unable to move my tongue all the way to the left in my mouth since the day before. On August 5, that continued and I felt a slight tremor in my tongue. I remembered the symptoms my wife had taught me based on her experience as a social worker at a rehabilitation hospital, I thought that it might indicate a stroke.

Examination showed a concerned but well-appearing septuagenarian, who was neither febrile nor hypertensive. He was alert, had no word finding or understanding difficulties and had a normal gait. His tongue had mild right sided atrophy without fasciculation and deviated to the right on extrusion. He had difficulty enunciating his labials and was unable to perform lingual gymnastics without slurring his words. Apart from a mild length dependent diabetic polyneuropathy affecting

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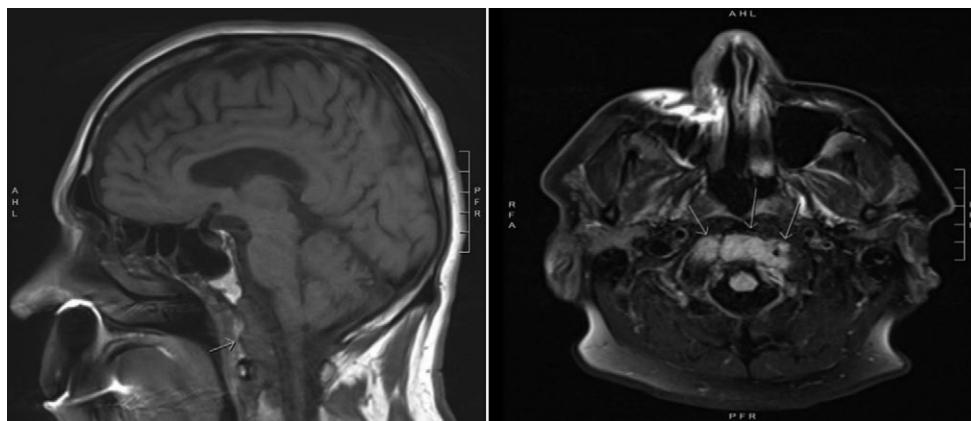


Figure 1: MRI brain demonstrating an expansible lesion at the central skull base involving the inferior aspect of the clivus, medial aspect of both occipital condyles, and the left hypoglossal canal raising the possibility of skull-base chordoma.

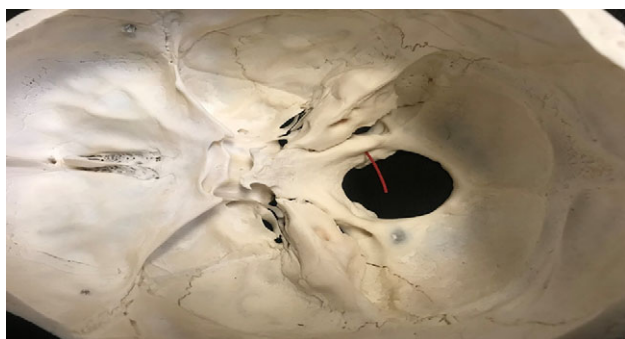


Figure 2: Marker showing right hypoglossal canal and its proximity to the clivus.

predominantly vibration, the rest of a detailed neurological examination was normal.

Our initial diagnosis was a medial medullary syndrome although he had no contralateral posterior column findings and the acute neck pain we misattributed to a vertebral dissection. MRI showed no restricted diffusion in his medulla but did show a clival mass invading his right hypoglossal canal (See Fig. 1). Subsequent neurosurgical debulking of his tumor confirmed chordoma.

DISCUSSION

The association of isolated hypoglossal nerve palsy with a chordoma is a double rarity since both conditions are infrequently seen. The anatomy of the hypoglossal canal is shown (see Fig. 2). As mentioned, chordomas arise along the axial skeleton, present at an average age of 60 years but occasionally occur in children [1, 2, 6]. Familial chordomas have also been reported, which theorize a genetic rule in the etiology of such tumors [9]. Metastases occur in 30% of these tumors, usually to the lung, liver, bone, sub-cutis and lymph nodes, but the poor prognosis is usually related to local destruction, accordingly headache and diplopia are the most frequently reported findings [1, 2, 5, 6].

MRI is the best diagnostic modality for chordoma. MRI also provides better differentiation of the soft tissues component of the tumor from surrounding structures aiding in primary tumor assessment [6]. Treatment is via cytoreductive surgery and the effectiveness of this is the most important determinant of patient survival [2, 3, 6, 7]. Postoperatively, either MRI or CT scan should be commenced to assess the residual tumor burden [6]. Adjuvant

radiotherapy is commonly used to prevent recurrence which if recurring carries a poor prognosis and a higher metastasis rate [1, 2, 6]. Chordomas are classified as radioresistant tumors requiring high doses (at least 60–70 Gy) [6, 7]. Adjuvant radiotherapy is currently being administered using proton therapy allowing high doses to be delivered to the tumor while sparing surrounding structures [2, 6]. Localized chordoma should be followed up with MRI every 6 months after resection for 5 years [6]. If no disease progression is noted MRI should be done every year for another 15 years [6]. Given the insidious disease presentation and potential for devastating neurological sequelae, early diagnosis is paramount and this requires the clinician to be aware of even vanishingly rare disease processes.

CONFLICT OF INTEREST STATEMENT

None declared.

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ETHICAL APPROVAL

No approval is required.

CONSENT

Patient permission was obtained prior to writing this report

GUARANTOR

Dr Mark Flemmer

CONFLICT OF INTEREST STATEMENT

None declared.

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