Chondromyxoid Fibroma of the Skull Base Invading the Occipitocervical Junction: Report of a Unique Case and Discussion

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ABSTRACT

Chondromyxoid fibroma (CMF) is a rare bone tumor representing less than 1% of total bone neoplasms. It is a slow-growing, locally destructive tumor that usually affects the metaphyseal region of long bones. The occurrence of this tumor in the skull base or cervical spine is exceptionally rare. We present the first case of CMF originating in the clivus and extending into the atlas and discuss the surgical management of this case.

KEYWORDS: Chondromyxoid fibroma, skull base, cervical spine, stabilization

Chondromyxoid fibroma (CMF), first described by Jaffe and Lichtenstein¹ in 1948, is one of the least common bone tumors, representing less than 1% of total bone neoplasms. It is a slow-growing, locally destructive tumor with no known tendency to metastasize but a propensity for local recurrence in the context of incomplete resection. It typically affects males more than females with a preponderance for those in their second and third decade.² The occurrence of this tumor in the skull base or cervical spine is rare; it most commonly arises in the metaphyseal region of long bones.² The rarity of this tumor makes it seldomly suspected preoperatively and may also make histological confirmation challenging. We present the first case of CMF originating in the clivus and extending into the atlas, requiring combined resection and stabilization surgery. We discuss the novel surgical issues that resulted and the wider application of this to skull base tumor surgery.

CASE REPORT

A 22-year-old Maltese man presented with a 3-year history of slowly progressive bilateral tinnitus and no other symptoms. Physical examination was normal, including no facial weakness and normal hearing (confirmed pure tone audiometry).

A magnetic resonance imaging (MRI) scan was performed and showed a 30 × 11 × 22 mm homogeneously enhancing, well-defined mass arising within the clivus and extending inferiorly to the left occipital condyle and the lateral mass of C1. The radiological diagnosis was one of a locally invasive malignant tumor with chondrosarcoma being most likely (Fig. 1). Preoperative cerebral angiography demonstrated no significant tumor blush and patency of both vertebral arteries.

Complete resection of the tumor was achieved by combining a left-sided Fisch type B infratemporal fossa approach with a far lateral neck dissection to the C1 lateral mass. A C-shaped incision in the temporal area

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Figure 1 Preoperative postcontrast T1-weighted magnetic resonance images: (A) sagittal, (B) axial, (C) coronal sections.

was extended retroauricularly into the mastoid region and extended into the neck along the anterior border of the sternocleidomastoid muscle. The external auditory canal was incised, closed, and mobilized anteriorly. A subtotal petrosectomy allowed access to and mobilization and protection of the internal carotid artery within the foramen lacerum. The far lateral neck dissection provided access to C1 and hence the mobilization and protection of the vertebral artery. These superior and inferior approaches to the tumor allowed complete tumor removal.

Figure 2 Postoperative plain radiograph demonstrating occipitocervical fusion.

Postoperatively, the potential for occipitocervical instability was managed by keeping the patient in a hard collar. The patient also developed a mild left-sided lower-motor neuron facial nerve palsy. A second operation was performed 3 weeks later to evacuate a post-operative tumor-bed hematoma with subsequent resolution of the left facial weakness.

Two weeks after the second operation, an occipitocervical fixation was performed, from the occiput to C2 with Oasys instrumentation (Stryker, UK) because of the extensive removal of the atlas and condyle. Post-operative X-rays showed appropriate position of the construct (Fig. 2).

Intraoperative frozen section histology suggested the tumor was unlikely to be malignant. Histology showed a chondroid tumor forming lobular structures with the cellularity being increased at the periphery, where spindle or stellate tumor cells were admixed with occasional giant cells (Fig. 3). Myxoid degeneration was noted in the hypocellular areas. There was no mitotic activity. Most tumor cells were weakly positive for S100. Ultimately, the diagnosis of chondromyxoid fibroma was reached.

Figure 3 Histological analysis. Hematoxylin and eosin staining, original magnification 100 $\times$.

The patient made a full recovery following the fixation surgery with no new neurological deficit except left-sided hearing loss. MRI scans at 6 months and 2 years postoperatively have shown no evidence of residual or recurrent disease, and patient has returned to full-time employment.

DISCUSSION

CMFs are uncommon benign cartilaginous tumors² with involvement of skull or spine being exceptionally rare. There are 62 cases of CMF arising in the cranial bones³ and 42 cases arising in the spine.⁴ We found only six cases reported in the literature of a clivus origin⁵⁻⁷ and 10 cases affecting the cervical spine.^{4,8-16} We believe that the case presented here is the first to document CMF originating in the clivus with extension into the atlas that subsequently required stabilization surgery.

Keel et al reported a series of three CMF originating in the clivus but variably extending superiorly into sphenoid and ethmoid sinuses.⁵ Their treatment of surgery via local curettage and subsequent radiotherapy in one case resulted in effective control with one recurrence managed with further surgery and radiotherapy. That one patient received postoperative radiotherapy for residual disease reflects both the difficulty in achieving complete resection of a locally aggressive tumor in this area, as well as the tendency to underestimate this tumor as a more benign counterpart such as chondroma and hence perform less aggressive initial surgery.

The presentation of CMF arising in the skull base is usually insidious, with nonspecific symptoms such as pain or symptoms of cranial nerve or central nervous system compression that are, however, progressive and unremitting over time. This often leads to a delay in the diagnosis of this very rare disease. Physical examination aside from the presenting symptoms is typically normal.

Radiological findings may be characteristic. The computed tomographic findings¹⁷ consist of an intact overlying bone cortex expanded by the tumor. There is matrix calcification in 75% of CMF of the skull base.¹⁷ On MRI, CMF is usually hypointense on T1-weighted images and hyperintense on T2-weighted images. MRI allows confirmation of these lesions as being extra-axial, of bony origin with cortical expansion. There is usually only minimally diffuse central contrast enhancement.

Surgery is the mainstay of treatment. Of the six cases that arose in the clivus,⁵⁻⁷ the initial treatment in all cases was surgical resection. One patient⁵ had local progression postoperatively and received radiotherapy. At subsequent follow-up, all six patients remained clinically free of disease. CMF is associated with local recurrence in up to 30% of cases^{18,19}; no cases of metastasis have been reported. Adjuvant radiotherapy has

been shown to be successful in the skull base where complete resection was impossible.²⁰

CMF of the cervical spine is also treated by surgical resection. Of the 10 previous cases,^{4,8-16} with levels including C2, C3, C4, C5, and C7, instability has been recognized as a potential problem with interlevel fusion performed either anteriorly or posteriorly. There is one case of CMF involving C2 associated with occipitocervical instability, which was treated with transoral resection of the tumor and occipitocervical fusion performed as a single-stage procedure.¹²

In this case, we have demonstrated that resection surgery can be dealt with in isolation, without having to extend the operation to include the occipitocervical fusion. This allows postoperative recovery and management of complications before proceeding with the definitive fusion. We would only advocate fixation at the time of initial surgery if the instability would be impossible to manage conservatively in the interim.

The consideration of postoperative radiotherapy was made for this case, especially in view of the findings of local recurrence in several patients. However, these patients were then treated with further local resection and radiotherapy. We believe that routine postoperative radiotherapy after the primary surgery is not indicated where there is a reasonable prospect of surgical cure, and equally when metastasis has not been previously described. Additionally, a second surgical procedure in this patient would be of increased morbidity in the aftermath of radiotherapy.

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