CHONDROMYXOID FIBROMA OF C1: FIRST CASE REPORT

INTRODUCTION

Chondromyxoid fibroma (CMF) is a rare benign tumor of the bone derived from cartilage precursor elements. The spine is an uncommon primary site with only 10 cases of CMF in the cervical spine reported to date. However, there are no reports of CMF involving the first-cervical (C1) vertebra. We report the first case of CMF in C1, treated with a postero-lateral and posterior surgical approach.

CASE REPORT

A 25-year-old monozygotic female twin presented with upper cervical pain and impaired mobility, especially when turning her head to the right side. On physical examination, lateral neck rotation and range of motion was restricted but there was no tenderness or mass on palpation. Neurological examination revealed no significant findings. Computed tomography (CT) scan and magnetic resonance imaging (MRI) both demonstrated a contrast-enhancing lytic lesion of the insufflated type, with a punched-out appearance, located at the right lateral aspect of C1 and extending to the anterior arch. A CT-guided needle biopsy was performed using a postero-lateral approach but the tumor material was insufficient for the pathological analysis.

ABSTRACT

Background: Chondromyxoid fibroma (CMF) is a rare, benign primary bone tumor. The cervical spine is an uncommon site for this tumor, with only 10 reported cases to date and none involving the first cervical vertebra (C1). Case Report: Female patient, 25-year-old monozygotic female twin, presented with cervical pain. Radiographic imaging demonstrated a contrast-enhanced, right-sided lytic lesion of the insufflated type in C1, with a punched-out appearance and extending to the anterior arch. A postero-lateral and a posterior approach were performed in two steps to resect the tumor followed by occipitocervical fixation. Pathology confirmed the diagnosis of CMF. At one year, the patient remains disease free with excellent spinal stability. Conclusion: Spinal surgeons may need to treat rare spinal tumors. Despite the proximity to neural and vascular structures, the goal of surgery is always a radical resection due to high recurrence rates.

Keywords: Cervical spine; Chondromyxoid fibroma; Bone tumor.
examination. Based upon the location, size and extent of the lesion, a postero-lateral surgery was planned to achieve radical resection and fixation. The need for posterior fixation was anticipated.

A postero-lateral approach was performed to expose the C1 vertebra (procedure described previously). The lesion was soft, in some parts containing liquid, tan-colored, and well-vascularized. The anterior and lateral portion of the mass of C1 and the anterior arch through the midline could be resected in block, but the posterior portion of the lateral mass of the C1 was partially resected by aspiration and curettage. The technical difficulties to remove the posterior portion of the tumor completely changed the surgical plan and it was found necessary to use a posterior approach. An iliac bone graft was implanted in the resection bed to achieve initial stability. Blood loss was 400 ml and operative time was 3 hours. Pathology showed stellar neoplastic cells with an increased number of pleomorphic nuclei and myxoid matrix that were diagnostic of CMF (Figure 2).

The residual posterior tumor was resected one week later through a posterior approach. An occipitocervical fixation was performed using the Occifix® system, GMReis™, and iliac bone graft. The C2 pedicle screw was not firm so it was decided to extend the fixation to C4 (Figure 3B). Blood loss in the second surgery was 300 ml and operative time was 2.5 hours. A 30-day postoperative CT demonstrated complete resection of the lesion with stable occipitocervical fixation (Figure 3A). At 1-year follow-up there was a calcified region between the lateral mass of C1 and the odontoid process (Figure 3C). Clinically, preservation of 50% lateral neck rotation bilaterally was achieved and the patient was pain-free without compromising the neurological aspects at 1-year follow-up.

**DISCUSSION**

The first case report published on CMF was in 1948 by Jaffe and Lichtenstein. CMF is a benign tumor characterized by lobulated areas, spindle-shaped or stellate cells, and abundant chondroid or myxoid material. Central portions of the tumor have greater cellularity and heterogeneous population of multinuclear giant cells. The metaphysis of the long bones is the most common primary site; vertebral involvement is uncommon and occurs in only 8% of cases. Radiologically CMF appears as a well-defined lytic lesion with lobulated margins, a sclerotic ring, and septations. Occasionally, periosteal extension may occur due to fusiform expansion of the bone and calcifications are uncommon.

Only 10 cases involving the cervical spine have been reported, none in C1 (Table 1). A female preponderance was observed in patients with cervical spinal CMF, with C2 being most commonly affected. The most common clinical manifestation reported was neck pain. Radiologically CMF appears as a well-defined lytic lesion with lobulated margins, a sclerotic ring, and septations. Occasionally, periosteal extension may occur due to fusiform expansion of the bone and calcifications are uncommon.
The radiological exam can be useful to try to differentiate benign chordoid tumors from malignant chordoid tumors in the preoperative period. The radiography has been the most frequently used imaging modality in the studies. The typical radiographic appearance of a benign chordoma is described as saucerization of the adjacent cortex, with a well-formed sclerotic periosteal reaction. A soft-tissue mass is not frequently shown, and matrix calcification may appear as much as possible.

### Table 1. Reported cases of cervical spinal chondromyxoid fibroma

<table>
<thead>
<tr>
<th>Cases</th>
<th>Age/Gender</th>
<th>Level</th>
<th>Management</th>
<th>Follow-up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Schajowicz et al, (1971)</td>
<td>6/F</td>
<td>C3</td>
<td>Resection</td>
<td>48, NR</td>
</tr>
<tr>
<td>Standefor et al, (1982)</td>
<td>20/F</td>
<td>C7</td>
<td>Irradiation, posterior decompression, anterior resection</td>
<td>15, NR</td>
</tr>
<tr>
<td>Provelegios et al, (1988)</td>
<td>32/M</td>
<td>C4</td>
<td>Anterior curettage, bone graft</td>
<td>10, NR</td>
</tr>
<tr>
<td>Zilmer et al, (1989)</td>
<td>20/F</td>
<td>C7</td>
<td>Resection, radiotherapy</td>
<td>84, MR</td>
</tr>
<tr>
<td>Rivieraz et al, (1991)</td>
<td>41/F</td>
<td>C5</td>
<td>Posterior decompression, anterior curettage and bone graft</td>
<td>30, NR</td>
</tr>
<tr>
<td>Wu et al, (1998)</td>
<td>Not reported</td>
<td></td>
<td>Curettage/excision</td>
<td></td>
</tr>
<tr>
<td>Lopez-Ben et al, (2002)</td>
<td>20/M</td>
<td>C2</td>
<td>Transoral vertebrectomy, posterior fusion</td>
<td>24, NR</td>
</tr>
<tr>
<td>Bala et al, (2006)</td>
<td>36/M</td>
<td>C2</td>
<td>Transoral curettage, anterior fixation</td>
<td>6, NR</td>
</tr>
<tr>
<td>Subach et al, (2010)</td>
<td>27/F</td>
<td>C6</td>
<td>Laminctomy, resection, posterolateral fusion at C5-C7</td>
<td>12, NR</td>
</tr>
<tr>
<td>Our case</td>
<td>25/F</td>
<td>C1</td>
<td>Far-lateral curettage, posterior curettage and occipitocervical fixation</td>
<td></td>
</tr>
</tbody>
</table>

NR – No Recurrence; MR – Malignant Recurrence

### REFERENCES

15. Nojima T, Unni KK, McLeod RA, Pritchard DJ. Periosteal chondroma and periosteal chondroblastoma, aneurysmal bone cyst, giant cell tumor, osteoblastoma, and fibrous dysplasia (FD). CMF is difficult to distinguish from chondrosarcoma histologically, because peripheral nuclear condensation and central hypercellularity can be seen in both. While diagnostic distinction from chondrosarcoma is important, resection remains the mainstay of treatment in both. CMF may also be confused with FD due to the presence of myxoid elements, especially if no lobulations are present. One of the challenges in treating CMF is that a symptomatic spinal mass is approached as malignant, with the goal of being complete resected and fusion when necessary, as in this case. Sometimes complete tumor resection is not possible because of the proximity to a neural or vascular structure and in such cases surgeons should perform other types of surgical approach when possible, as in this case, in order to try to remove as much as possible.

**CONCLUSION**

CMF is a rare benign tumor derived from cartilage precursor elements. Despite the proximity to neural and vascular structures a radical resection is preferred due to high postoperative recurrence.