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Case Report

Unusual Occurrence of Spinal Chondromyxoid Fibroma of the C2 Odontoid: A Case Report of a Rare Tumor at an Extremely Uncommon Site

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Abstract

Chondromyxoid fibroma is a rare benign cartilaginous bone tumor that is uncommonly found in the spine. To date, among 13 cases reported in the cervical spine, only two cases involve the C2 vertebra and all arise from the body. We report the first case occurring in the odontoid process of a 26-year-old woman. Firstly, because of the craniocervical instability, fusion from the occipital condyles to the posterior elements of C1 through C3 was performed. Secondarily, an expansile tumor of the C2 odontoid process was resected along with phenolization and synthetic processed bone grafting via a transoral approach. The postoperative course was uneventful without recurrence at 3-year-follow up. Then, she has returned to full-time employment.

Keywords: Chondromyxoid Fibroma; Cervical Spine; Odontoid Process; Recurrence

Introduction

Chondromyxoid fibroma (CMF) is a relatively rare, benign cartilaginous bone tumor accounting for < 1% of primary bone neoplasms, usually involving bones of the lower extremity during the second or third decades of life [1-5]. About 700 cases of CMF have been previously mentioned in the modern English literature and less than 50 cases involve the spine [5-11]. To the best of our knowledge, only 13 cases have been previously mentioned in the cervical spine with only 2 cases involving the axis (C2) and all arising from the body (Table 1). We report a 26-year-old woman who presented with CMF arising from the odontoid process associated with C1-C2 instability. This is an additional third reported case of this rare tumor in this location and first case involving the odontoid process.

Case Report

A 26-year-old right-handed woman presented with seven months of upper cervical pain and impaired mobility of her left upper limb. Because the pain increased in intensity and become associated with paresthesia in the right upper extremity approximately one week ago; she decided to consult the Surgical Clinic. On physical examination, the patient had a reduced range of motion of the neck, particularly with rotation and lateral bending. However, there was no tenderness or mass on palpation. Neurological examination revealed left upper limb monoparesis.

All laboratory tests including full blood count, electrolytes, erythrocyte sedimentation rate, C-reactive protein, glycemia and rheumatoid factors were within normal limits. Radiographs of the cervical spine demonstrated an expanded radiolucent osteolytic le-

sion within the odontoid process with well defined margins as well as instability at C1-C2 (Figure 1). Reconstructed sagittal, axial and coronal Computed Tomography (CT) images showed a contrast-enhancing lytic lesion of the insufflated type with a punched-out appearance, thin sclerotic rim and endosteal scalloping measuring 14 mm x 21 mm located in the C2 odontoid process. The margins of the lesion were preserved without erosion. No calcification or associated soft tissue mass were present (Figure 2).

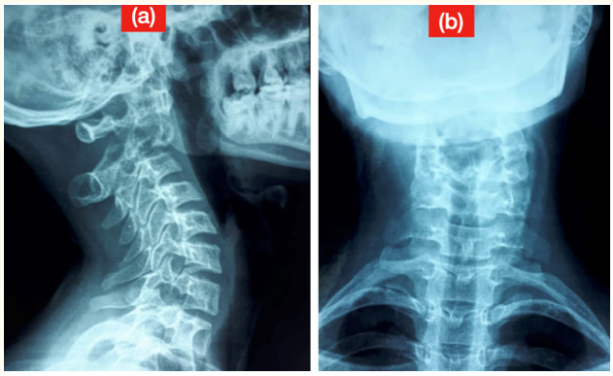

Figure 1: Lateral (a) and anteroposterior (b) radiographs of the cervical spine showing an expanded radiolucent osteolytic lesion within the odontoid process with well defined margins.

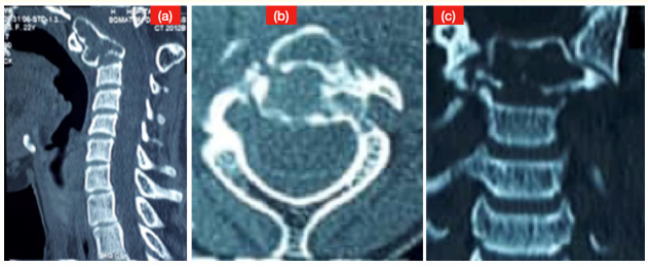

Figure 2: Reconstructed sagittal (a), axial (b) and coronal (c) CT images showing a contrast-enhancing lytic lesion of the insufflated type with a punched-out appearance, thin sclerotic rim and endosteal scalloping measuring 14 mm x 21 mm located in the C2 odontoid process. The margins of the lesion were preserved without erosion. No calcification or associated soft tissue mass were present.

Based upon the location, size and extent of the lesion, a CT-guided needle biopsy was performed using a transoral approach for the pathological examination. Histological sections 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. Myxoid degeneration was noted in the hypocellular areas. Secondary changes, such as necrosis or hemorrhage, psammoma bodies, cellular atypia or mitotic activity were not seen. Ultimately, the diagnosis of chondromyxoid fibroma was reached (Figure 3).

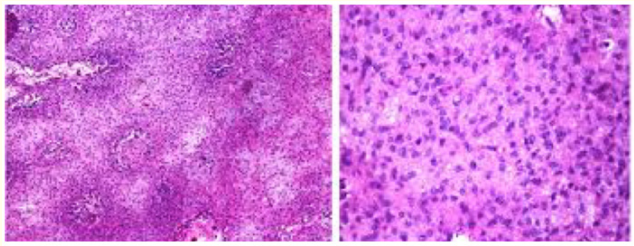

Figure 3: Photomicrograph slides of the excised chondromyxoid fibroma, at 40x magnification, showing cellular neoplasm which is well demarcated from the adjacent osseous tissue arranged as vague nodules in chondroid pattern. The cells are round to oval with vesicular nuclei and abundant eosinophilic cytoplasm and display well delineated cell borders. Neither cell atypia nor mitosis is detected. Giant cells are noted in the periphery of the lesion. There is no evidence of calcification.

Because of the craniocervical instability, the patient consented for a 2-phase-surgical approach. Firstly, in prone position on a translucent orthopedic table under general anesthesia, instrumented segmental fusion of the occipital condyles through the posterior elements of C1, C2, C3 was performed under fluoroscopic guidance to maintain the sagittal balance of the spine and achieve initial stability. Secondarily, in supine position, a midline oral mucosal incision from the mid-portion of C1 to the top of C3 was used, with soft tissue dissection carried down to the anterior arch of C1 and to the C2-C3 interspace. On inspection during the operation, the ventral cortical margin of C2 odontoid was thinned and expanded. The tumor had caused centripetal expansion and was not friable or hemorrhagic. It was therefore completely removed by curettage leaving a thin shell of cortical bone between the resection cavity and the thecal sac. Thereafter, phenolization and synthetic processed bone grafting were performed. Blood loss was 600 ml and operative time was 6 hours. Postoperatively, the patient was immobilized with a hard cervical collar for three

months. She therefore underwent rehabilitation and recovered all neurological functions.

At 3-year-follow up, the patient had no neurological symptoms. Dynamic radiographs of the cervical spine showed no evidence of residual or recurrent disease with union of the graft at the site of the complete resection of the lesion and satisfactory alignment of the cervical spine with stable occipitocervical fixation (Figure 4). Clinically, lateral neck rotation bilaterally was achieved and the patient was pain-free without compromising the neurological aspects. Then, she has returned to full-time employment.

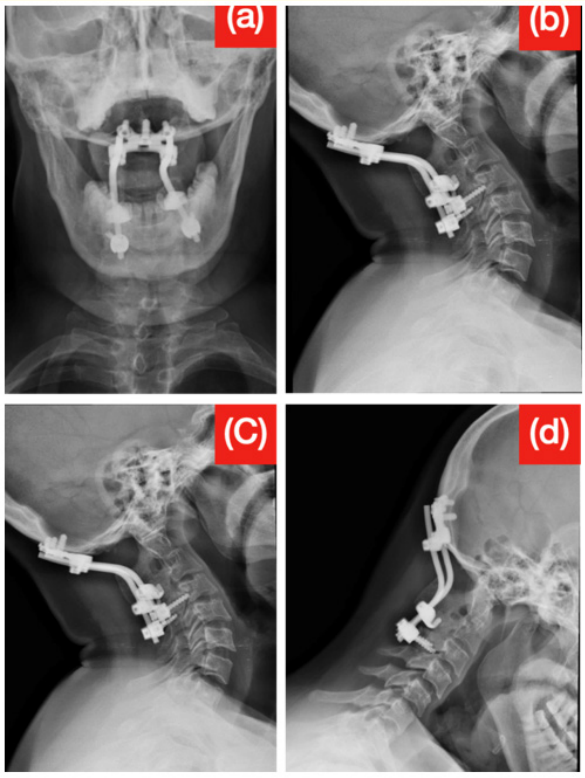

Figure 4: At 3-year-follow up, anteroposterior (a), lateral (b) and dynamic radiographs in hyperextension (c) and hyperflexion (d) of the cervical spine showing union of the graft at the site of the complete resection of the lesion and satisfactory alignment of the cervical spine with stable occipitocervical fixation. No evidence of residual or recurrent disease is detected.

Discussion

Chondromyxoid fibroma (CMF) was first described as a distinctive clinical entity by Jaffe and Lichtenstein in 1948; formerly it was classified as myxoma or a myxomatous variant of giant-cell tumour, or mistaken for a malignant lesion, especially chondrosarcoma, chondromyxosarcoma or myxosarcoma [9-22]. The 2002 World Health Organization (WHO) classification of bone and soft tissue tumors defines CMF as "a benign tumor characterized by lobulated areas of spindle-shaped or stellate cells with abundant myxoid or chondroid intercellular material separated by zones of more cellular tissue rich in spindle-shaped or round cells with a varying number of multinucleated giant cells of different sizes" [5-9,23]. It is a rare tumor accounting for < 1% of all benign bone tumors, and the least common benign cartilaginous bone tumor [3,9-24]. The most common locations of CMF are the bones of the lower extremities, with a tendency for the metaphyseal region of the distal femur and proximal tibia, followed by the foot [24-29]. Rare sites are the pelvis and sacrum, fibula, spine, sternum and ribs, clavicle, radius, skull base and calvarium [9-34]. More than 700 cases of CMF have been previously mentioned in the modern English language literature, and < 50 cases involved the Spine [5-13,20,25,26]. Benson and Bass were the first authors to report CMF of the spine [7]. In CMF involving the spine the most common site of involvement is the thoracic vertebrae [5-11,19]. The posterior elements and the posterior part of the vertebral bodies are most commonly involved [21]. Involvement of the cervical vertebrae is rare, and was reported in 13 cases with only two involving the C2 vertebra [3-21]. For the two cases involving the C2 vertebra, the anatomical location was the body [13,14]. We report the first case involving the C2 odontoid process.

The etiology of the development of CMF is unknown. Recent studies suggest that pericentromeric inversion; inv (6) (p25q13) is a diagnostic marker for this tumor [7-14,35]. It occurs with an approximately equal sex ratio [2-4,5,10]. The patient age is variable, ranging from 6 to 87 years. However, most occur in the second or third decade of life with mean age of 31.1 years. A second peak is in the fifth to seventh decades [4-11,24,25,27,28]. A case of congenital chondromyxoid fibroma has been reported by Mendoza., *et al* [28]. A female preponderance was observed in patients with cervical spinal CMF (67%) and the mean age was 26.6 years (interval; 6 - 41 years). The two patients with C2 CMF were male with respectively 20 and 36 years (Table 1). Our case is a 26-year-old woman. Her age is within the common peak incidence of cervical spinal CMF.

No	Cases	Age/Gender	Level	Management	Follow-up (months)
1.	Schajowicz., *et al*. (1971) [3]	6/F	C3	Resection	48, NR
2.	Standefer., *et al*. (1982) [15]	20/F	C7	Irradiation, posterior decompression, anterior resection	15, NR
3.	Provelegios., *et al*. (1988) [16]	32/M	C4	Anterior curettage, bone graft	10, NR
4.	Zillmer., *et al*. (1989) [11]	20/F	C7	Resection, radiotherapy	84, MR
5.	Rivierez., *et al*. (1991) [17]	41/F	C5	Posterior decompression, anterior curettage and bone graft	30, NR
6.	Wu., *et al*. (1998) [5]	NI		Curettage/excision	NI
7.	Lopez-Ben., *et al*. (2002) [14]	20/M	C2 (Body)	Transoral vertebrectomy, posterior fusion	24, NR
8.	Bala., *et al*. (2006) [13]	36/M	C2 (Body)	Transoral curettage, anterior fixation	6, NR
9.	Jonathan., *et al*. (2008) [19]	35/M	C7	C7 central corpectomy, incomplete intralesional curettage with iliac bone grafting and C6 to T1 interspinous wiring	96, NR
10.	Subach., *et al*. (2010) [18]	27/F	C6	Laminectomy, resection, postero-lateral fusion at C5-C7.	NI
11.	Sfreddo., *et al*. (2012) [12]	25/F	C1	Lateral curettage, posterior curettage and occipitocervical fixation	12, NR
12.	Crocker., *et al*. (2012) [20]	22/F	C1	Anterior resection, occipitocervical fixation	24, NR
13.	Zahir., *et al*. (2015) [21]	36/F	C3, C4	Resection	24, NR
14.	Our case, Hope *et al* (2020)	26/F	C2 (Odontoid)	Occipital cervical fusion, transoral resection with phenolization and synthetic processed bone grafting	36, NR

Table 1: Reported cases of cervical spinal chondromyxoid fibroma in the literature.

NR: No Recurrence; MR: Malignant Recurrence; NI: Not Indicated; M: Male; F: Female.

The most common clinical manifestation of cervical spinal CMF reported was progressive longstanding neck pain and motion restriction [12-15,21]. Radiologically CMF appears as a well-defined radiolucent lytic lesion with lobulated margins, a sclerotic ring, and uncommonly contain visible calcification or trabeculation. Occasionally, periosteal extension may occur due to fusiform expansion of the bone. CT scans display the sclerotic margin calcifications and trabeculation better than conventional radiographs. On MRI, CMFs have a heterogeneous appearance due to their different tissue components. The chondroid and myxoid tissues have an intermediate to high signal on proton-density and T2-weighted images and a low signal on T1-weighted images. CMFs show a high accumulation of fluorodeoxyglucose in positron emission tomography (PET) scan. Radiological differential diagnosis includes chondrosarcoma, osteoblastoma, chondroblastoma, enchondroma, fibrous dysplasia (FD), aneurysmal bone cysts (ABC), giant cell tumor, non-ossifying fibroma, metastasis, multiple myeloma and collapsed hemangioma of the vertebral body [7-12,19,21]. Histologically, CMF is a benign cartilaginous neoplasm characterized by lobulated areas, of spindle-shaped or stellate cells with abundant myxoid or chondroid intercellular material. The lobules are more cellular at the periphery. Central portions of the tumor have greater cellularity and heterogeneous population of multinuclear giant cells. Calcification is found microscopically in approximately one-fourth of the patients [7-12]. Our case was an adult female in her second decade of life who had suffered from a long history of neck pain and right upper limb monoparesis, with radiological features of an eccentric radiolucent lytic lesion with thinned sclerotic margin and punched-out appearance of the whole odontoid process. No calcifications were seen. But, we did not perform MRI and PET for our patient. Histological results confirmed the diagnosis of CMF of C2 odontoid process.

Due to the rarity of CMF in spine, there is no well-established management protocol. Various treatment options have been used by different investigators. En bloc excision is relevant for CMF in the long bones but such strategies in the cervical spine are practically not possible because of the proximity to a neural or vascular structure and may produce instability. Hence less radical procedures such as aspiration, curettage with or without bone grafting are employed to preserve mechanical stability. This, however, significantly increases the chance of recurrence to 80% in the case of curettage alone and 7% in the case of curettage with bone grafting [12-19,36]. Adjuvant radiation is not recommended because of the potential of malignant transformation. Zillmer and Dorfman report a case of a woman with a CMF involving C7 that was wrongly reported as a giant cell tumor and was treated with radiation therapy following intralesional excision. She presented seven years later with recurrent CMF and malignant fibrous histiocytoma [11]. Dahlin and Unni also have reported a case with development of a fibrosarcoma at the site of a tibial CMF that had received radiation therapy [37]. Other reported complications of radiotherapy are radionecrosis, osteomyelitis and sarcomatous transformation [3-5,12,38]. Tumor recurrence after surgical treatment may occur in 4 to 20% of cases, more commonly in younger patients. The age of diagnosis was proposed as a factor for increased recurrence rates, with the suggestion that the reduced resistance of the pediatric thin cortices and spongiosa contributes to the aggressive behavior of the lesion [1-3,12,25,38,39]. Most recurrences, up to 80% with a 30-year follow-up, occur when curettage alone and not en bloc resection is performed [3-11,12,36]. Wu., *et al*. in a series with 278 patients, have found that the possibility of recurrence after surgery may arise up to 11% [5-19]. Known cases of spinal recurrence have been described [11-13,40,41]. In the management of our case we decided in favor of curettage with synthetic processed bone grafting, phenol treatment and posterior occiput to C3 fixation. No radiation therapy was given. On a 3-year follow up, no local recurrence is seen. However, a careful follow-up is required because of the possibility of recurrence as our patient was 26 years at the time of diagnosis. The patient had resolution of his neurological deficits postoperatively and had maintained this complete neurological recovery over a significantly long period. She was then allowed to return to her full-time employment.

Conclusion

Chondromyxoid fibroma is a rare benign cartilaginous bone tumor that is uncommonly found in the spine, especially the cervical spine. Histological diagnosis is essential in order to provide the patient with an accurate management of the pathology. Despite the proximity to neural and vascular structures complete resection is the mainstay of treatment due to high postoperative recurrence. When complete resection is not possible, partial resection with bone grafting yields good results. Thus, a close long-term follow up is warranted, more so in young patients. We have reported the clinical, imaging and pathological findings as well as treatment and outcome of a 26-year-old woman diagnosed with a chondromyxoid fibroma involving the C2 odontoid process treated successfully.

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Conflicts of Interest

None of the authors has any potential conflict of interest.

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Ethical Approval

This article does not contain any studies with human participants or animals performed by any of the authors.

Informed Consent

Written informed consent was obtained from the patient to publish the information, including her photographs.

Authors' Contributions

All authors have participated equally in the present study.

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