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Case Report

Primary Intraosseous Xanthogranuloma in Adult Cervical Spine: A Case Report of Benign Cause of Lytic Bone Lesion

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Abstract

Lytic lesions in adult spine are a common manifestation of aggressive disease such as primary bone tumor, metastasis, myeloma, or infectious pathology. Xanthoma arising in the spine with purely intraosseous component is an extremely rare occurrence with only six cases reported in the adult population, none in the cervical region. We report the first case of primary xanthoma of the cervical spine in a 50-year-old male solely confined to osseous compartment. The imaging mimics of lytic lesion with expansile mass in adult spine are reiterated.

Keywords: *Adult, bone xanthoma, cervical, lytic, spine*

Introduction

Bone xanthoma is a known entity with predilection for the appendicular skeleton, but involvement of the axial skeleton is also well mentioned in the literature.[1] The spine is an atypical site for xanthoma and can be divided into osseous, extradural, intradural, and intramedullary types.[2-8] Intraosseous involvement is rare, and only six cases have been reported in the adult population.[9-14] Here, we report a case of primary xanthoma occurring in the cervical spine in a 50-year-old male, presenting as lytic expansile lesion which was presumed to be aggressive lesion such as mets, myeloma, or tuberculous granuloma. This is the seventh case among adult spinal intraosseous xanthoma. Cervical involvement has been reported in seven cases till date,[2,3,5-7,15,16] with only one case of intraosseous involvement in the pediatric age group.[16] To the best of our knowledge, this is the first case of cervical spine involvement as primary intraosseous lesion in the adult age group.

Case Report

A 50-year-old male patient was referred to our hospital, with a 4-month history of neck pain and gradually progressive weakness of all four limbs. Over the past 2 weeks, the patient noticed significant deterioration and became bed bound. The patient denies any previous history of trauma, malignancy, or medical history of note. Clinical examination revealed spastic quadriparesis of 3/5 with 80% decreased sensation of all modalities below C5. Sphincters were found to be intact.

Imaging

Plain X-ray cervical spine on anterior-posterior (AP) view showed a large lucent lesion replacing the C5 vertebra with loss of the left pedicle [Figure 1a]. The magnetic resonance imaging (MRI) of the cervical spine demonstrated a T1 and T2 isointense, expansile mass lesion involving the C5 vertebral body extending into the left pedicle but sparing neural foramina. On T2, small hyperintense intralesional cavity was noted in the anterior part. The lesion extended across the C5/6 intervertebral disc and into the anteroinferior part of the C4 vertebra. Significant cord compression with absent thecal sac against C5 was noted. The lesion enhanced homogeneously with gadolinium and appeared completely extradural [Figure 1b-d]. Based on imaging, metastasis, myeloma, and tuberculous granuloma were suspected.

Treatment

The patient underwent standard anterior cervical decompression and fixation. Peroperatively, tumor appeared as a well-encapsulated, firm, multinodular

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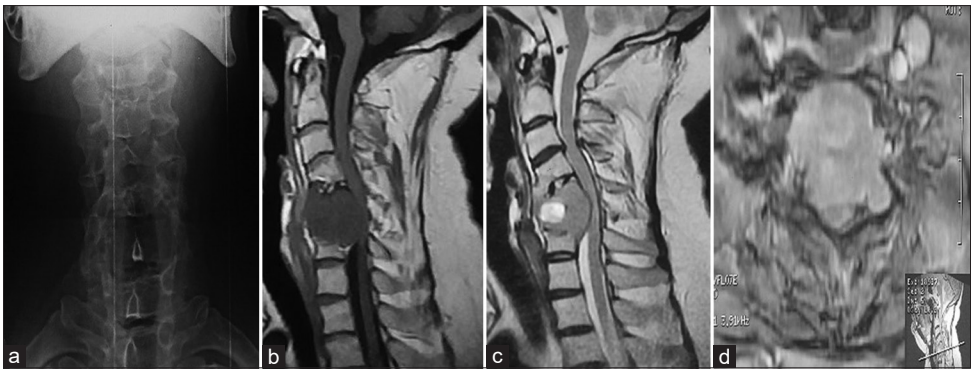

Figure 1: (a) Interpedicular and interspinous widening at C5, implementing large expansile lytic lesion replacing C5 vertebral body. Left C5 pedicle absent while right C5 pedicle displaced and thinned out. (b and c) Sagittal cervical spine magnetic resonance imaging showing soft-tissue mass isointense on T1 and T2, replacing C5 body, with T2 hyperintense cavity in the anterior part. (d) Axial section at C5 level, showing homogeneous enhancement of lesion, extension to the left pedicle, and thecal sac compression. Posterior elements are well spared

mass, yellow tan in color, 3 cm × 3 cm × 2 cm in size. There was a small necrotic part corresponding to the T2 hyperintensity. The lesion was excised in piecemeal fashion [Figure 2a] to completely decompress the cord. No clue of bone was found within the mass. At this point, differential was further narrowed to granulomatous lesion like tuberculosis. Histology revealed sheets of histiocytes with foamy cytoplasm and bland nuclei. Focal cholesterol clefts and multinucleated giant cells were also noticed. There was no cellular atypia. CD68 was positive and S100 was found to be negative [Figure 2b and c].

The serum calcium was normal, and lipid profile showed normolipemia. Clinical examination did not reveal any soft-tissue xanthomas. The patient was able to return to his normal activities 4 weeks after the operation. Postoperative imaging as per routine following anterior decompression and fixation consisted of plain X-ray cervical spine AP and lateral views [Figure 3a and b]. Postoperative computed tomography (CT) of the same area was planned at the 6-month follow-up both to see the integrity of fixation as well as to rule out the tumor recurrence, but the patient was not able to reach out for imaging because of pandemic restrictions. Currently, he is 1 year out of his surgery, with no reported complaint as per telephonic follow-up.

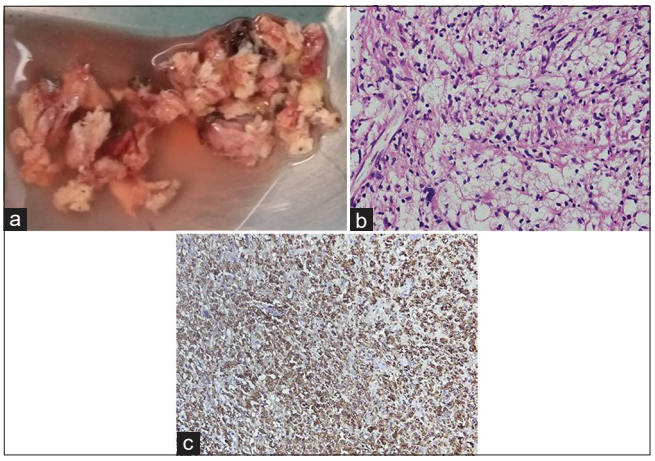

Figure 2: (a) Biopsy specimen as yellow soft-to-firm mass removed piecemeal. (b) Histological findings (hematoxylin and eosin stain): Numerous foamy cells, cholesterol clefts, and multinucleated giant cells. (c) Immunohistochemical findings (streptavidin-biotin-peroxidase method): Cells showed strong positivity for CD68

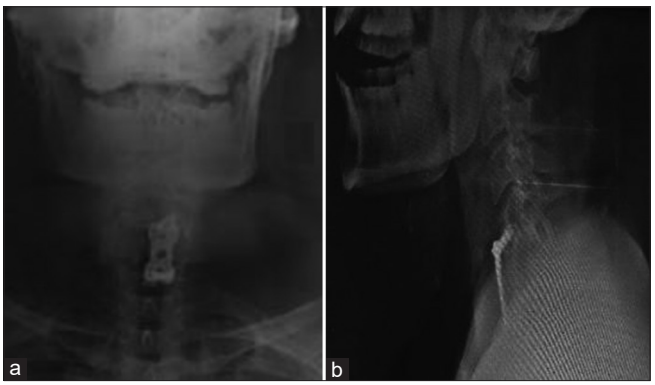

Figure 3: (a) Postoperative X-ray cervical spine, anterior-posterior view. (b) Lateral view, due to very short neck, lower screws are not visible

Discussion

Abnormal cholesterol deposition within various tissues such as skin, subcutaneous tissue, tendons, and bones constitutes the term "xanthoma." Bone xanthomas are common and have a predilection for the appendicular skeleton, although skull, ribs, and pelvis are also well mentioned among the axial skeleton.[17] Xanthoma arising within the vertebra is a rare occurrence with six cases reported so far in the adult population [Table 1].

As evident, the term "secondary" means occurring in the setting of endocrine or metabolic diseases with increased cholesterol levels, or a preexisting bony lesion, whereas the term "primary" is reserved for cases with no preexisting lesion as well as normal lipid profile.[1] However, it has been reported that bone xanthoma can precede the onset of hyperlipidemia as long as 15 years and can be the first sign of dyslipidemia.[18]

The usual reported age of bone xanthoma is after the second decade with male predominance.[1] The same

Table 1: Summary of the reported cases of intraosseous xanthoma in adult spine

Reported cases authors/year	Age/ gender	Level involved	Extent of bony involvement	Canal/ paravertebral extension	Hyperlipidemia/ cutaneous stigmata	Surgery
Robertson *et al*., 1995[9]	39 years/ male	T2	Right lamina/pedicle/dorsal part of the body of T2	+/+	−/−	Laminectomy, transpedicular curettage, and rectangular loop fixation
Chung *et al*., 1999[10]	22 years/ male	L5-S1	Left lamina L5-S1, left L5-S1 facet, Sacral ala	+/+	−/−	Left L5-S1 laminectomy, excision of soft-tissue mass, and curettage of body. Orthosis no fix
Huang *et al*., 2004[11]	58 years/ male	S1	Body of S1, left SI joint, left iliac blade	+/+	+/+	Curettage of intraosseous lesion. No fixation
Jain *et al*., 2011[12]	22 years/ female	T7	Body, left pedicle	+/+	Not mentioned	Laminectomy and rectangular loop fixation
Agabegi *et al*., 2011[13]	47/ female	L2	Body, right psoas, retroperitoneum	−/−	Not mentioned	Laminectomy, debulking, and T10-L4 transpedicular fixation
Mcloughlin *et al*., 2019[14]	75 years/ male	T7	Body, right pedicle/TP	−/−	+/−	Fluoroscopic open biopsy
Present case	50 years/ male	C5	Body, left pedicle	+/−	−/−	C5 corpectomy and anterior plate fixation

demographic distribution has been observed in spinal intraosseous xanthomas reported so far. Regarding systemic associations, hyperlipidemia and cutaneous xanthoma were found in 69% and 53% of patients of bone xanthomas, respectively.[11] Among vertebral xanthomas, only two cases have reported hyperlipidemia,[11,14] and out of these, only one had documented cutaneous stigmata.[11] A case closely resembling to ours in the pediatric age group has been documented.[16]

Notably, all reported cases presented as lytic lesion and suspected to be aggressive pathology, and none was predicted on clinical or radiological grounds. The diagnosis was made purely on histological features. On X-ray and/ or CT, a large lucent lesion occupying the vertebral body with extension into the pedicles, transgressing the adjacent disc spaces was a consistent finding in all cases. However, in comparison to xanthomas occurring in long bones, no evidence of reactive bone or sclerotic margin was found.[17]

So far, no specific MRI features have been demonstrated, making radiological diagnosis ever challenging. The MRI signal depends on the content of lesion. More fibrous spindle cells would give low T2, whereas more foamy cells and cholesterol clefts appear low on fat-saturated sequence.[19]

The differential diagnosis of a lytic lesion in adult spine is wide, and common suspects after the fourth decade are myeloma and bone metastases. In Pakistan, tuberculosis is endemic and commonly encountered as lytic lesion with soft-tissue mass, so it was in our top differential. Among the reported cases, one sacral lesion was presumed to be tuberculous granuloma.[11]

Myeloma is low on T1 and high on T2. The MRI features of metastasis can be variable, but most of these are low on T1 and mixed to high signal on T2. Although bilateral pedicle involvement is a well-known sign for malignant disease, expansile lesion in contrast to vertebral collapse favors benign pathology.[20,21] Cortical disruption and expansion both are documented with bone xanthoma.[22] Epidural and paravertebral extension was found in all but one case in the reported literature. Epidural but not paravertebral extension was found in our case.

Although international classification of histiocytic disorders has grouped xanthogranuloma and Langerhans cell histiocytosis together as "dendritic cell-related" histiocytoses, xanthoma is clearly described as non-Langerhans cell histiocytosis and can be differentiated by immunohistochemical stains[19] [Table 2].

Despite being a benign pathology, treatment depends on the area of bone involved, i.e., anterior or posterior elements and neural compromise. In case of posterior element disease, simple curettage or laminectomy achieves neural decompression, with or without fixation. Anterior disease is dealt anteriorly through corpectomy and graft fixation. More radical approaches such as total spondylectomy are not favored for benign disease in mobile segment of the spine.[23] Surgical morbidity of en bloc resections in the spine has been reported to be as high as 35%.[24] Subtotal removal is followed by radiotherapy while total removal is considered curative.[13,16] Lipid-lowering medical therapy is added as an adjunct in cases of secondary disease. No recurrences have been reported in the literature following appropriate treatment.[1]

Our patient was treated with near-total excision of the tumor with C5 corpectomy using standard anterior approach with bone grafting and plate fixation. Postoperatively, the patient had a significant improvement in spasticity and power in all four limbs.

To the best of our knowledge, our case is the first case in the adult age group affecting the cervical spine. Considering

Table 2: Hallmark features differentiating from similar histological entities

Pathology	Key HP feature	Xanthoma
Benign fibrous histiocytoma	No E/C cholesterol deposits	Large E/C Cholesterol deposits
Langerhans cell histiocytosis	CD68+CD1a+More eosinophils	CD 68+, CD1a−Few eosinophils
Rosai–Dorfman disease	S100+	S100−

CD68 (a histiocytic marker), CD1a (excludes LCH), and S-100 (excludes RDD)

both children and adults, this is the second case to be reported in the cervical spine as intraosseous lesion. Intraosseous xanthoma should be added to the differential diagnosis of lytic bone lesion in adult spine.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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