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Surgical Neurology International

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Case Report

Eosinophilic granuloma of the cervical spine in a young adult: A rare case report

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Received: 06 April 2024
Accepted: 09 May 2024
Published: 14 June 2024

DOI
10.25259/SNI_262_2024

Quick Response Code:

ABSTRACT

Background: Spinal eosinophilic granulomas (EG) are rare tumors, mostly reported in the pediatric age group. They constitute <1% of primary bone neoplasms, and cervical spine involvement is uncommon.

Case Description: A 20-year-old male presented with neck pain for a 4-month duration. Six years previously, he had received six cycles of vinblastine for biopsy-proven histiocytosis of an axillary lymph node; this resulted in incomplete remission. Present magnetic resonance/computed tomography (CT) imaging revealed a lytic C2 body lesion with atlantoaxial instability. When the CT-guided biopsy was suggestive of EG, he was managed with definitive surgery and adjuvant radiotherapy.

Conclusion: Cervical spine EG is rare in adults. CT-guided biopsy should confirm the diagnosis and should be followed by definitive surgery and adjuvant radiotherapy.

Keywords: Adults, Cervical spine, Eosinophilic granuloma, Spine

INTRODUCTION

Eosinophilic granulomas (EG) constitute <1% of primary bone neoplasms and typically involve the thoracic spine but rarely the cervical spine.[1,6,7,10] These lesions rarely occur in adults, and may be treated by immobilization, steroids, surgery, radiotherapy (RT), and/or chemotherapy.[1,3,5,10] Here, we report a 20-year-old male who presented with a C2 vertebral EG that was successfully managed with surgery and adjuvant RT.

CASE ILLUSTRATION

A 20-year-old male presented with neck pain for a 4-month duration. He had 4+/5 quadriparesis with hypertonia but no sensory deficits. Six years previously, he received 6 cycles of vinblastine for biopsy-proven histiocytosis of an axillary lymph node, resulting in complete remission.

- X-ray, magnetic resonance (MR), computed tomography (CT) evaluation of EG
- X-rays, MR, and CT studies were consistent with the diagnosis of a C2 EG.

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Cervical spine X-rays revealed osteolytic destruction of the C2 body and atlantoaxial instability (AAI), while CT showed osteolytic destruction of the body/pedicles of C2 [Figures 1a-c]. MR imaging (MRI) showed that the marrow of C2 was replaced by a lesion which appeared isointense on T1 and iso-hypointense on T2 sequences that resulted in thecal sac compression [Figure 1d]. Imaging also revealed vertebra plana of the T9 with osteolytic destruction of the left femoral head [Figures 1e and f]. Further, the technetium bone scan showed increased uptake in the C2 and T9 vertebrae and left femur. Following a CT-guided biopsy of the C2 lesion, which was suggestive of histiocytosis, and X-rays showing AAI, the patient underwent an anterior transoral decompression with occiput to C3–4 fusion.

Pathology

Histopathological examination showed clusters and sheets of cells with abundant cytoplasm, grooved nuclei, delicate chromatin, and inconspicuous nucleoli in a background of inflammatory infiltrate rich in lymphocytes, plasma cells, and eosinophils [Figure 2]. These cells were immunopositive for S100 and CD 1a, confirming the diagnosis of Langerhans cell histiocytosis (LCH).

Follow-up treatment

He later received adjuvant low-dose radiation therapy in view of his positive history. At the follow-up of 2 years, the patient was doing well and there was no deformity or recurrence noted.

DISCUSSION

Clinical and radiographic presentation of EG

EG is considered to be the most benign and localized form of LCH and accounts for around three-fourths of those cases.[3,6] EG most commonly involves the skull, mandible, clavicle, pelvis, femur, ribs, and long bones and is a predominantly pediatric disease. Neck pain is the most

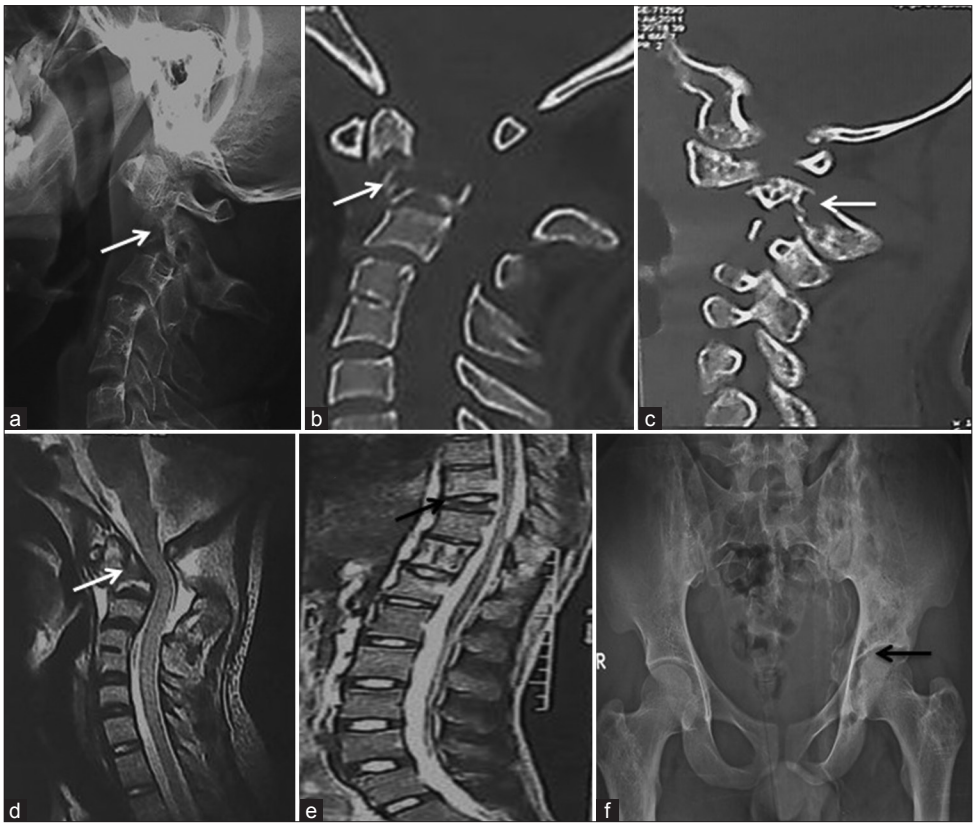

Figure 1: (a) Plain radiographs of the cervical spine showing an osteolytic destruction of the C2 vertebral body with atlanto-axial instability (white arrow). (b,c) Sagital CT-median and paramedian images showing destruction of the body and pedicles of C2 vertebra (white arrows). (d) Sagital T2 MRI showed iso to hypointense signal at the site of bony defect with evidence of thecal sac compression (white arrow). (e,f) Note that the disc spaces are not involved. Dorsal spine sagittal T2 MRI sequence showing features of vertebra plana of the T9 vertebral body (black arrow) and lytic destruction of the left femoral head (black arrow).

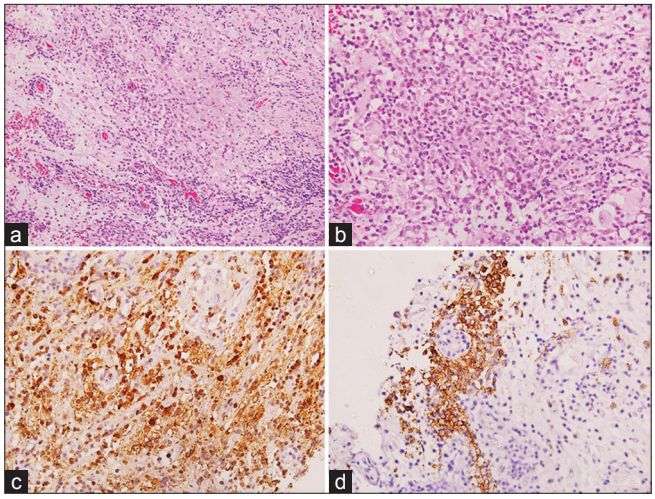

Figure 2: (a,b) Photomicrograph showing clusters of large cells with abundant cytoplasm in a background of inflammatory cells (H&E, x200), which have abundant cytoplasm and indented/grooved nuclei (H&E, x400). (c,d) These cells are immunopositive for S100 and CD 1a (IHC, x400). IHC:immunohistochemistry, H&E:Haematoxylin and Eosin.

common symptom, and neurological deficits with cervical lesions usually reflect vertebral collapse and/or epidural cord/root compression.[3,5,10] MRI is the imaging modality of choice; the lesions are mostly iso to hypointense on T1 hyperintense on T2 MRI sequences and show enhancement. CT and X-rays complement MRI, and a complete skeletal survey identifies multifocal lesions.[1,4,9]

Role of surgery

The role of surgery is controversial and should be limited to situations where there is a question regarding the diagnosis, bony instability, and/or severe neurological deficits.[1,3] Surgical options include curettage and biopsy, corpectomy with fusion, or posterior fusion procedures.[1,3,9]

Role of RT

The role of adjuvant RT has not been clearly defined in the literature.[8,10] Low-dose RT is the most commonly used as it facilitates sclerosis and tissue healing.[3,6] The usual indications for adjuvant RT are persistent painful lesions, neurological deficits, and progressive and recurrent lesions.[3,10]

Role of chemotherapy

Chemotherapy is indicated only in cases of multifocal/systemic disease or as first-line therapy in the pediatric age group with solitary EG in locations that preclude safe and complete resection.[6] Other treatment options include corticosteroids, radiofrequency ablation, and percutaneous poly-methyl-methacrylate cement vertebroplasty.[6,10] Recurrence or evolution of new lesions is rare.[6]

Pathology

On gross pathological examination, EG appears reddish-brown to yellow with intervening bony spicules. On microscopy, histiocytes are predominant cells which contain oval nuclei, eosinophilic cytoplasm, and CD1a positivity. Birbeck's granules are characteristically seen on electron microscopy, and S-100 protein immunohistochemical staining is diagnostic. Nuclear atypia and mitosis are rare features, and differential diagnosis includes Erdheim-Chester disease, Rosai-Dorfman disease, and other dendritic neoplasms.[2,6,10]

CONCLUSION

Cervical spine EG is rare in adults. CT-guided biopsy should be used to confirm the diagnosis and be followed by definitive surgery and adjuvant RT.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

Financial support and sponsorship

No financial support.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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How to cite this article: Prasad G, Krishna G, Kale A, Rajeshwari M, Kale SS. Eosinophilic granuloma of the cervical spine in a young adult: A rare case report. Surg Neurol Int. 2024;15:198. doi: 10.25259/SNI_262_2024

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