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Case Report

A case report of eosinophilic granuloma in axis (C2) vertebra causing spinal cord compression

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ABSTRACT

Eosinophilic granuloma (EG), a benign osseous variant of Langerhans cell histiocytosis, is rarely observed in cervical spine and being in axis (C2) is even more infrequent. We present here an unusual case of EG of the axis vertebra with neurological impairment due to cervical cord compression, where the patient underwent decompression, C1–C2 spinal stabilization and steroid therapy. Cervical vertebrae involvement of EG with cord compression and neurological deficit is very unlikely, and this case report contributes to the clinical experience of this rare but highly prodigious disease. The benefit of surgical treatment with total removal of the tumor and spinal stabilization to prevent irreversible neurological deficit is emphasized.

Key words: *Cervical eosinophilic granuloma, Spinal cord compression, Spinal stabilization*

Eosinophilic granuloma (EG) is a solitary, nonneoplastic proliferation of histiocytes localized in bone or lung. It comes under the spectrum of the autoimmune disorder, Langerhans's cell histiocytosis (LCH) [1]. Despite EG's high percentage of bone involvement, the incidence of vertebral involvement ranges from only 7.8% to 25%. EG involving the vertebral bodies show a predilection for the thoracic spine followed by the lumbar spine [2]. The location at the level of the cervical spine is very rare, and even that correlated to symptoms of spinal cord compression is still rarer [3]. The diverse treatments of EG of cervical spine include immobilization, chemotherapy, radiation therapy, steroid injection, and surgery depending on the disease symptoms [3-5]. However, the adequate disease management modality is still a subject of controversy. We report the case of axis vertebra EG with neurologic deficit from an 11-year-old boy, issuing a displacement of the spinal cervical cord and compression, whose diagnosis was established after the radiological investigation and also by the lesion histopathology. We underline the rarity of the case, the excellent response to the surgical treatment, and the disease's good prognosis after 2 years of follow-up.

CASE REPORT

An 11-year-old male was presented to us with a history of neck pain and stiffness of a month's duration. The patient had restricted range of neck movements with torticollis toward the right side. Neurologic examination showed limb weakness of Grade 2. His laboratory data were unremarkable. Radiography of the cervical spine showed focal osteolysis of the axis (C2) vertebra (Fig. 1). Magnetic resonance imaging (MRI) (T1 contrast) of the cervical spine showed C1–C2 enhancement and destruction. Lateral mass with associated soft tissue was evident. MRI (T2 contrast) revealed cord compression and edema (Fig. 2). Computed tomography (CT) scans of the craniovertebral junction (CVJ) revealed destruction of the dens, body, bilateral pedicles, and the left lamina of axis vertebra with significant left lateral subluxation (Fig. 3).

To avert spinal deformity and permanent neurologic deficits, surgical correction through posterior approach was performed. In prone position, the skin and soft tissues were incised in the midline at the region of CVJ. It was found that the posterior elements of C2 were destroyed significantly including the pars. A biopsy was taken from the destroyed site. C1–C2 posterior spinal fusion was carried out with the cervical polyaxial screws and interfacetal C1–C2 arthrodesis with the iliac autograft. The surgical wound was closed over a closed draining tube in anatomical layers. Post-operative course was uneventful with resolution of pain and neurological symptoms. Post-surgery, treatment with oral steroid was commenced, and the patient continued on the steroid drug prednisolone, for 6 months. The follow-up investigations showed resolution of the C2 body lesion, no vertebral body collapse, and C1–C2 fusion. The bone scan showed no uptake at C1–C2 levels and in other sites (Fig. 4). At 2 years, the patient remains asymptomatic with no signs of recurrence.

Histopathologically, the section appeared as a soft tissue with osseous fragments composed of proliferated histiocytes mixed with infiltrates of eosinophils. The histiocytes had vesicular,

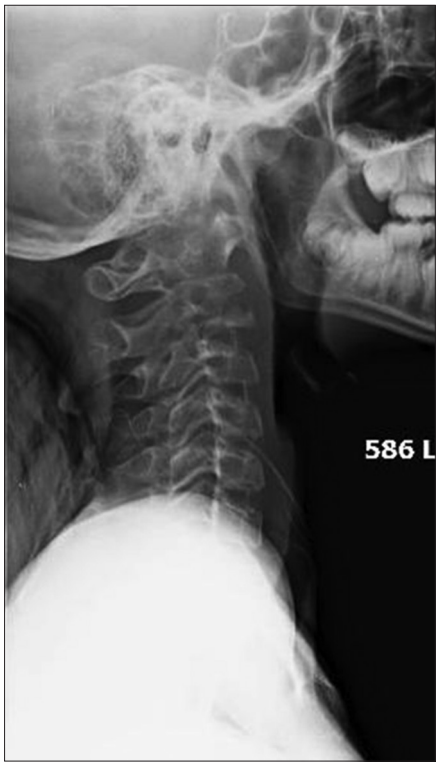

Figure 1: Osteolysis of the axis vertebra

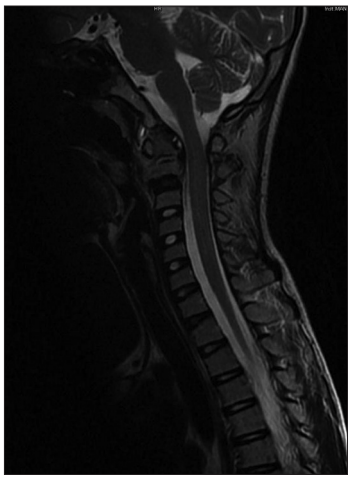

Figure 2: Cord compression and edema

grooved nuclei, and moderate to abundant cytoplasm. The immunohistochemistry study stained positive for S100. The findings were suggestive of LCH. Lack of nuclear atypia and atypical mitoses confirmed the pathology as EG and not the other variants of LCH.

DISCUSSION

EG, the benign form of LCH is characterized by unifocal involvement of bone or lung. The pathogenesis of EG remains inconclusive and it has been proposed that basic immune or genetic defects elicit proliferation of Langerhans cells. EG is identified by the presence of Langerhans cells in a characteristic milieu which includes histiocytes, eosinophiles, and neutrophiles. EG is differentiated from the other variants of LCH by

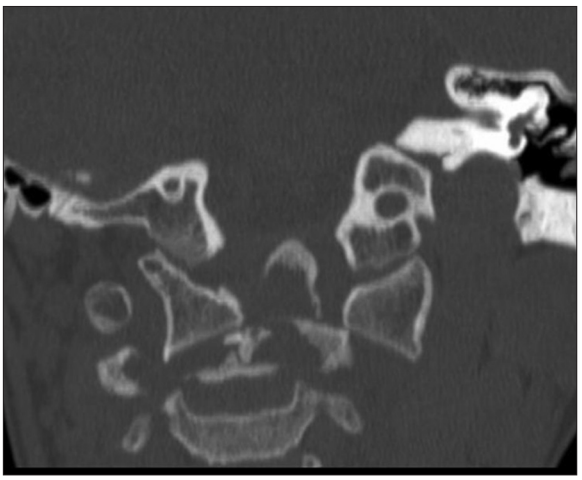

Figure 3: Destruction of vertebral body with significant lateral subluxation

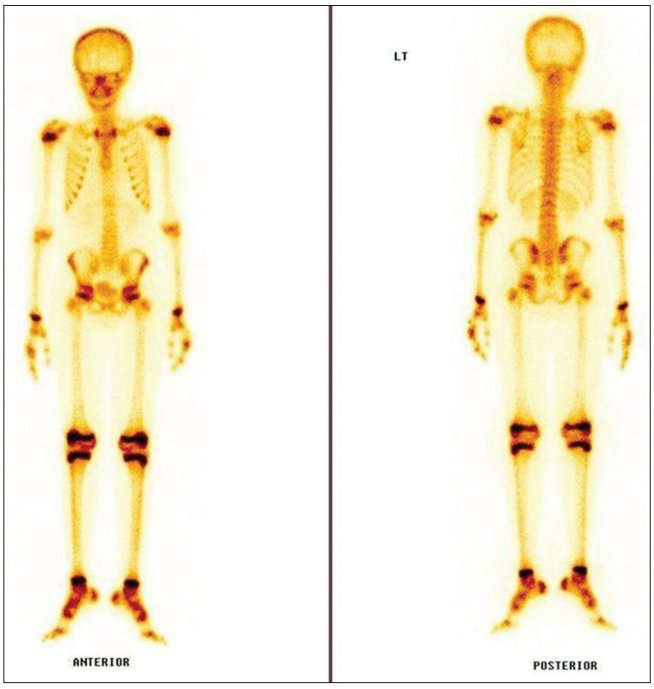

Figure 4: Post-surgery bone scan

the lack of nuclear atypia and atypical mitosis as observed histopathologically [6]. The cervical lesion is often represented as a lytic lesion involving the vertebral body with preserved disc spaces; however, rarely, it causes erosion or destruction of pedicles and posterior elements [7]. Despite the frequent reporting of vertebral involvement of EG, reports on spinal cord involvement and compression are scanty. Our patient endured neurologic deficit due to the spinal cord compression correlated with the osteolytic lesion at the axis vertebral level. Such neurological complications are reported to arise due to vertebral collapse or due to soft tissue involvement [8].

Plain radiography remains the mainstay of diagnosis in patients with EG. CT and MRI help to evaluate the vertebral lesion extension and soft tissue involvement, respectively, and assist with planning a biopsy or a surgical excision [9]. Radionuclide bone imaging is a complementary technique in the initial diagnosis of

bone lesions including EG. However, recurrences are identified more readily with fewer false-negative findings with radionuclide bone scans. The radionuclide bone scans' ability to exhibit lesional activity in addition to lesion localization is crucial for the follow-up of EG [9]. The mechanism of bone scan is based on the function of osteoblastic activity and changes in bone metabolism such as growing or healing lesions [9]. This diagnostic modality proved to be helpful in our patient in the follow-up, evaluation of response and also helped in the decision of cessation of treatment with prednisolone.

The disease management of vertebral EG has been extremely variable and controversial. Even now, there is no gold-standard treatment accepted for this condition. Considering the self-limiting nature of the lesion, which is inclined to a spontaneous resolution, different therapeutic approaches have been proposed. The goals of treatment are the preservation of spinal stability and neurological function. Cures have been reported even with conservative treatments like cervical immobilization in patients with no neurological deficits [10]. However, when the integrity of the spinal cord or of the roots is threatened, it is necessary to undertake more aggressive treatment as in our case. Our patient had progressive neurological deficit caused by cord compression which necessitated decompression. The patient endured the surgery without complications and gained complete clinical benefit early after surgery.

Steroids are recommended in patients of EG, considering the high bone regenerative potential in younger patients [4]. Post-surgery, our patient was given treatment with the oral steroid, prednisolone and he continued on the medication for 6 months, thereafter. Some authors suggest radiotherapy as a treatment choice [3], but there are also reports that warn about the risk of malignant transformation of the lesions after radiotherapy, especially in the pediatric age group [2].

CONCLUSION

The results of our case suggest that although there is no standard treatment regimen for axis EG, decompression, and surgical stabilization of the cervical spine are both essential in treating patients with neurological impairment involving cord compression. Surgical correction followed by treatment with oral steroids drastically reduces the chances of disease recurrence. Radionuclide bone scan proves to be a reliable diagnostic tool for the treatment response follow-up assessment in patients with EG.

REFERENCES

1. Willman CL. Detection of clonal histiocytes in langerhans cell histiocytosis: Biology and clinical significance. Br J Cancer Suppl. 1994;23:S29-33.
2. Yeom JS, Lee CK, Shin HY, Lee CS, Han CS, Chang H. Langerhans cell histiocytosis of the spine. Analysis of twenty-three cases. Spine. 1999;24(16):1740-9.
3. Haupt R, Minkov M, Astigarraga I, Schäfer E, Nanduri V, Jubran R, et al. Langerhans cell histiocytosis (LCH): Guidelines for diagnosis, clinical work-up, and treatment for patients till the age of 18 years. Pediatr Blood Cancer. 2013;60(2):175-84.
4. Rimondi E, Mavrogenis AF, Rossi G, Ussia G, Angelini A, Ruggieri P. CT-guided corticosteroid injection for solitary eosinophilic granuloma of the spine. Skeletal Radiol. 2011;40:757-64.
5. Ngu BB, Khanna AJ, Pak SS, McCarthy EF, Sponseller PD. Eosinophilic granuloma of the atlas presenting as torticollis in a child. Spine. 2004;29(5):98-100.
6. DiCaprio MR, Roberts TT. Diagnosis and management of langerhans cell histiocytosis. J Am Acad Orthop Surg .2014;22(10):643-52.
7. Osenbach RK, Youngblood LA, Menezes AH. Atlanto-axial instability secondary to solitary eosinophilic granuloma of C2 in a 12-year-old girl. J Spinal Disord. 1990;3(4):408-12.
8. Tutar, Suleyman. Eosinophilic granuloma of the lumbar spine in a child. Spine J. 2016;6:393.
9. Steiner M, Prayer D, Asenbaum S. Modern imaging methods for the assessment of Langerhans cell histiocytosis-associated neurodegenerative syndrome: Case report. J Child Neurol. 2005;20(3):253-7
10. Jiang L, Liu ZJ, Liu XG, Zhong WQ, Ma QJ, Wei F, et al. Langerhans cell histiocytosis of the cervical spine: A single Chinese institution experience with thirty cases. Spine. 2010;35(1):8-15.

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