Clinical Case Report

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The outcome of eosinophilic granuloma involving unilateral atlantoaxial joint

A case report and literature review

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Abstract

Rationale: Solitary eosinophilic granuloma (EG), the most benign, common form of Langerhans cell histiocytosis, has a self-limiting process and is associated with a good prognosis. Immobilization is recommended as the first treatment strategy for solitary EG, although the treatment protocols are still controversial. Radiotherapy and surgery are secondary treatment choices. Lesions of the upper cervical spine react differently to treatment because of their specific anatomical and motor features.

Patient concerns: We discuss the case of a 29-year-old man with axis EG who underwent immobilization, radiotherapy, and finally surgery.

Diagnosis: Eosinophilic granuloma (EG).

Interventions: An initial conservative protocol, including immobilization with a collar and radiotherapy, effectively relieved his neck pain, whereas torticollis secondary to atlantoaxial subluxation was not improved. Therefore, he underwent tumor resection through the anterior approach and spinal reconstruction, fixation, and fusion through the posterior approach.

Outcomes: The deformity was well corrected and follow-up was satisfactory.

Lessons: Upon review of the literature, we found that EG lesions affecting the axis have more risk factors for instability or deformity, and they have particular anatomical and motor characteristics; thus, they require more consideration and attention in terms of treatment, prognosis, and follow-up.

Abbreviations: CT = computed tomography, EG = eosinophilic granuloma, LCH = Langerhans cell histiocytosis.

Keywords: adult, atlantoaxial rotatory subluxation, axis, eosinophilic granuloma, spinal instability, surgical intervention

1. Introduction

Langerhans cell histiocytosis (LCH) represents a series of diseases caused by the abnormal proliferation and tissue accumulation of dendritic cells with features similar to epidermal Langerhans cells in various organs. Nevertheless, it has a low disease incidence of 1:1,500,000. As one of the most benign tumors, eosinophilic granuloma (EG) usually involves the skeletal system, and it is often a solitary osseous lesion, accounting for 60% to 80% of individuals with LCH.[1,2] The common lesion sites include the skull, femur, mandible, pelvis, and spine, in that order. Spinal involvement accounts for 6.5% to 25% of all skeletal cases of LCH.[3,4]

The classic radiographic characteristics enable the basic assessment of osseous lesions.[4] Computed tomography is best for determining the characteristics and extent of the bone lesion, and magnetic resonance imaging is ideal for detecting an abnormal signal intensity, paravertebral soft-tissue mass, and spinal cord compromise. Moreover, some studies have reviewed the signal intensity on T1-weighted and T2-weighed images to make a more accurate diagnosis.[5,6] In addition, for more detailed radiological features, a definitive diagnosis of LCH should always be based on histological and immunohistochemical examinations of lesional tissue, which is characterized by abundant eosinophilic cytoplasm and positive CD1a and/or CD207 (Langerin) staining of lesional cells.[1]

Solitary EG has the potential to spontaneously remodel itself through a self-limiting process, and its treatment protocols have not been approved until now. In recent years, more researchers have recommended conservative treatments, such as immobilization, bed rest, nonsteroidal anti-inflammatory drugs, and close follow-up, for patients with a mildly isolated spinal lesion. The atlantoaxial spine has a complex anatomical structure and biomechanical characteristics. Here, we describe the case of a 29-year-old man with atlantoaxial rotatory subluxation secondary

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YS and WG have contributed equally to this work.

The patient has consented to any related data for publication in this article.

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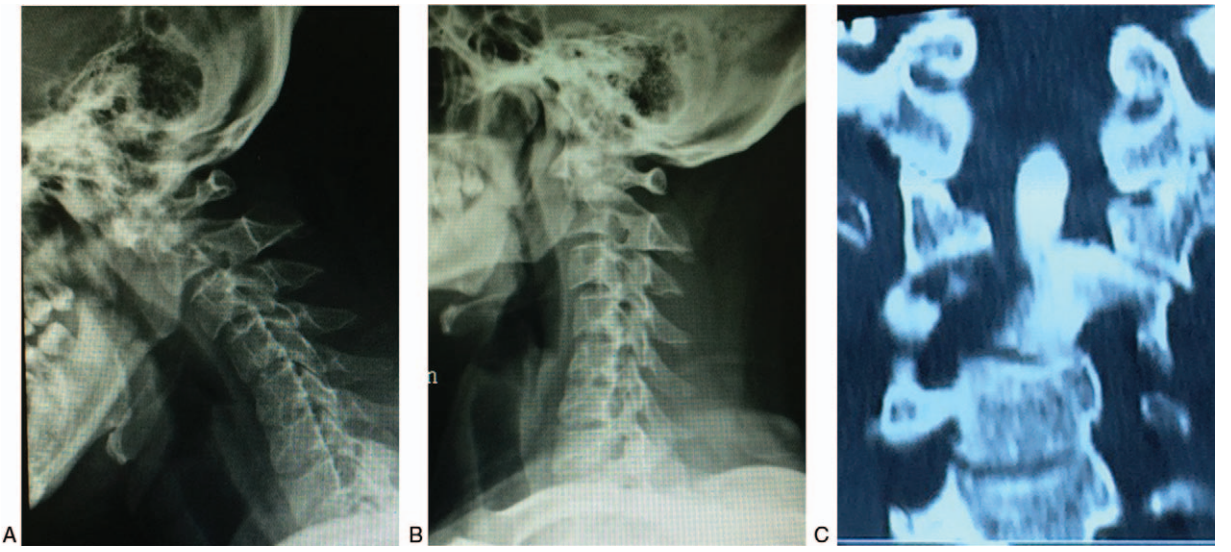

Figure 1. Lateral flexion–extension radiographs (A and B) and coronal reconstruction computed tomography scans of the upper cervical spine (C) at the first clinic visit. These plain films show no apparent odontoid lateral mass asymmetry or atlantoaxial instability.

to axial EG, and we review the literature regarding treatment of EG in the upper cervical spine.

2. Case report

A 29-year-old man was evaluated because of a 1.5 month history of severe neck pain and a mobility limitation that was unresponsive to physiotherapy and nonsteroidal anti-inflammatory drugs. The neurological examination showed no abnormality, and his medical history was unremarkable. Radiographs of the cervical spine showed a marked osteolytic lesion at the axis vertebrae with no apparent atlantoaxial instability (Fig. 1A and B). The computed tomography (CT) scan confirmed an osteolytic lesion in the right part of the C2 body (Fig. 1C). The bone scan and positron emission CT scan confirmed hypermetabolic foci at the C2 level and the isolated lesion. A needle biopsy was performed through the transoropharyngeal approach. The histological examination showed LCH lesions stained with hematoxylin–eosin (Fig. 2A and B). In addition, LCH was diagnosed based on the immunohistochemical staining results: Langerin (+), CD68 (+), S-100 (+), and CD1a (+) (Fig. 2C–F). After the oncologist was consulted, we treated him with a cervical collar and radiotherapy (14 Gy/7 F). His neck pain decreased.

One month later, he returned with torticollis, a mobility limitation, and slight neck pain. Radiographs and the upper cervical CT scan showed collapse of the right atlantoaxial joint and atlantoaxial rotatory subluxation (Fig. 3A–C). To treat the torticollis deformity, he underwent posterior atlantoaxial fusion

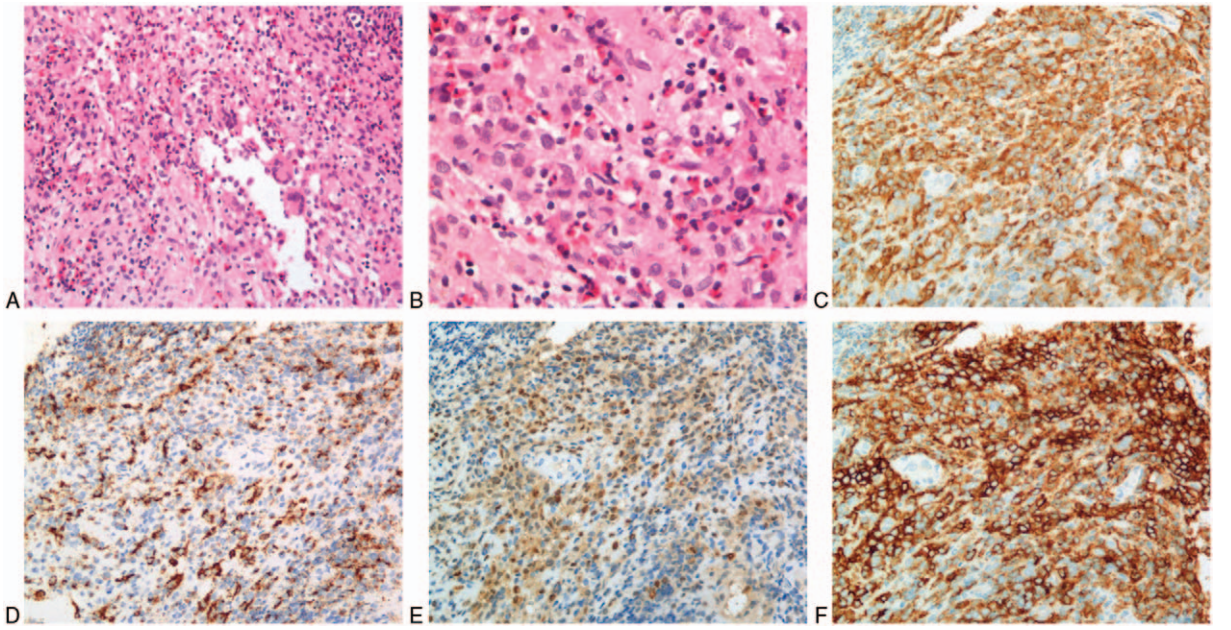

Figure 2. Hematoxylin–eosin staining indicates a proliferation of large cells, accompanied by variable number of eosinophils, lymphocytes (A and B). The diagnosis of Langerhans cell histiocytosis was supported by demonstration of Langerin (C), CD68 (D), S-100 (E), and CD1a (F) immunoreactivity in the lesion cells.

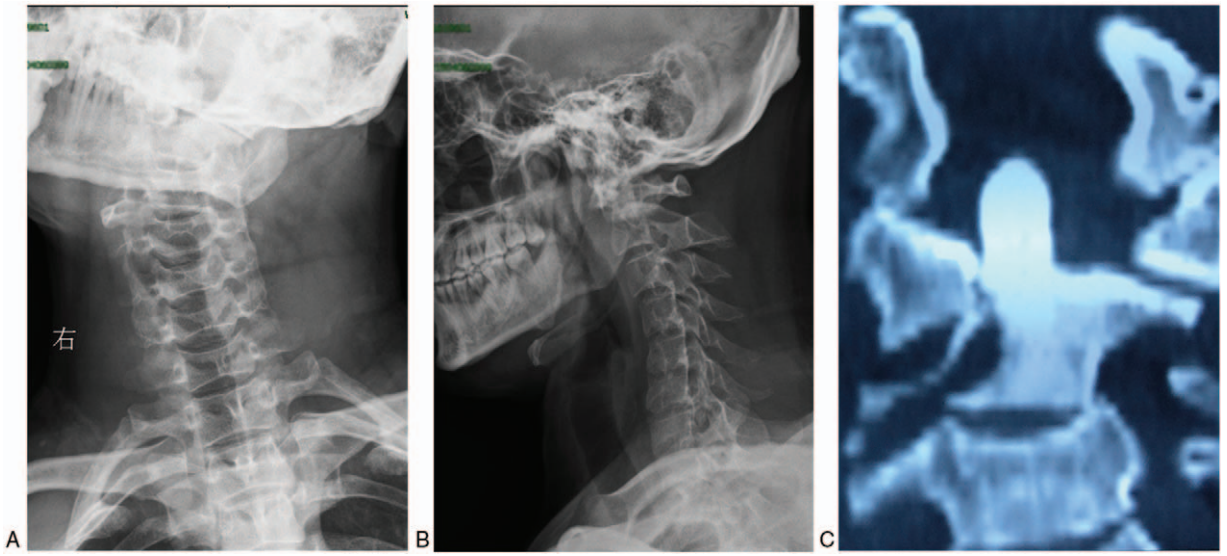

Figure 3. Anteroposterior lateral radiographs (A and B) and coronal reconstruction computed tomography scans of the upper cervical spine (C) after conservative therapy for 1 month. Stiff torticollis occurred secondary to collapse of the right atlantoaxial joint and apparent lateral displacement of the odontoid process.

with pedicle screw fixation and autogenous iliac transplantation, and anterior lesion resection with a titanium mesh filled with allograft bone (Fig. 4). At the 3-, 6-, and 12-month follow-ups, the torticollis deformity was satisfactorily corrected, and no migration or breakage of implants was observed (Fig. 5A–C). No obvious recurrence was seen at the last follow-up. The study was approved by the Ethics Committee of Tongji Medical College, Huazhong University of Science and Technology.

3. Discussion

The occipitocervical junction is a complex anatomic location that includes the craniocervical and atlantoaxial junctions, which contain the occipital bone, atlas vertebrae, axis vertebrae, and important complex ligamentous structures. Furthermore, it is the most mobile area in the spinal column, and it can move in the sagittal, coronal, and axial planes. Thus, any lesion affecting these anatomical structures most likely results in obvious restricted cervical motion, torticollis, or instability.

Previous studies have demonstrated that EG affected almost all of the vertebral bodies at the lesion segment,[5] even at the C2 level.[2,4,7–10] In our patient, the right half of the C2 body was damaged by EG. Without a normal vertebral body, a lateral mass in the C1 vertebra is most commonly involved in cases of EG.[3,10–12] Unlike the subaxial cervical spine, the atlantooccipital and atlantoaxial joints are adjacent to the C1 lateral mass

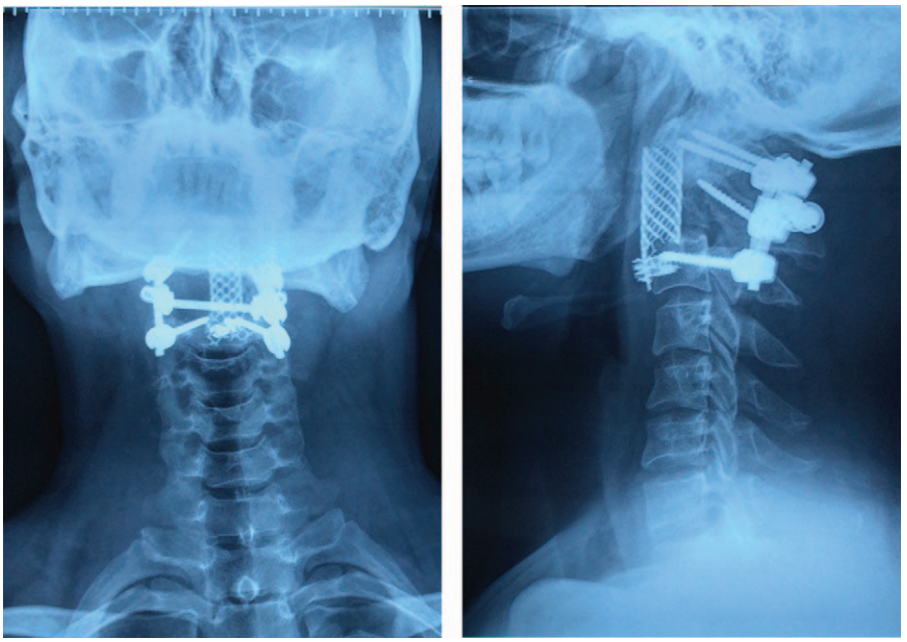

Figure 4. Radiographic images of the cervical spine postoperatively. Tumor resection through the anterior approach and spinal reconstruction, and fixation and fusion of the C1–C3 levels though the posterior approach.

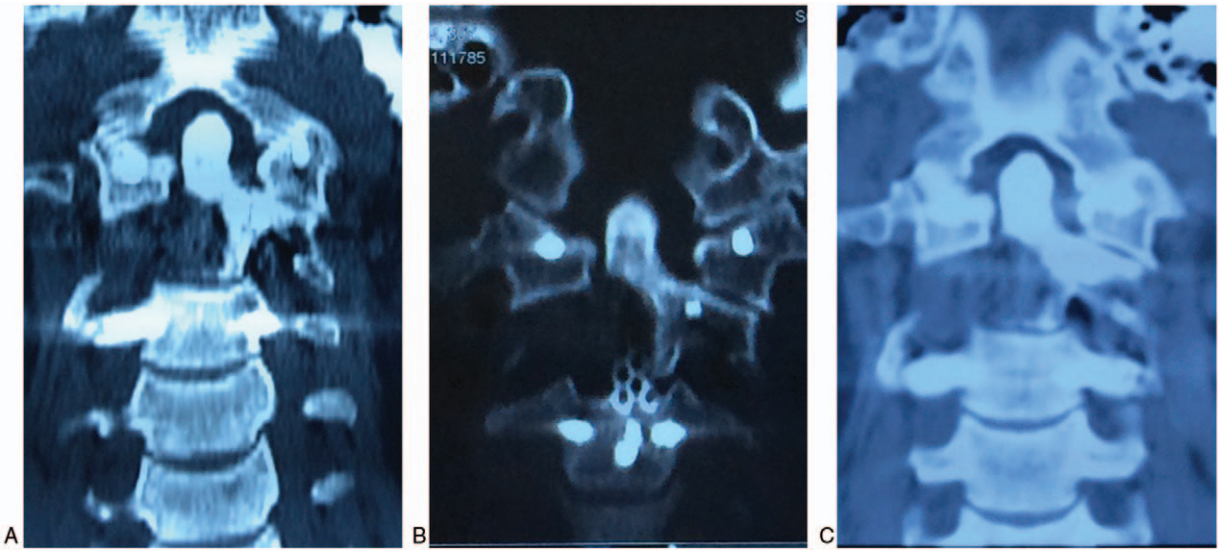

Figure 5. Follow-up computed tomography scan. Reconstruction of the upper cervical spine is satisfactory at the 3-, 6-, and 12-month follow-ups (A–C, respectively). No significant tumor recurrence was found during the 1-year follow-up.

and C2 vertebral body, and the stability of this region is maintained with the help of ligaments. The atlantooccipital joint contributes to the 23° to 24.5° flexion/extension, and the atlantoaxial joint provides 25° to 30° rotation and 10.1° to 22.4° flexion/extension.[13] Furthermore, anterior–posterior and cranial–caudal translations exist with motion of the atlantooccipital and atlantoaxial joints.[14] Thus, the C2 lesion is more likely to affect the stability of the atlantoaxial joint and lead to cervical deformity, and atlantoaxial subluxation or instability has already been observed on presentation in patients with EG.[2,7–10] Some of these patients have a history of immobilization with a collar, but the effect of this treatment was unsatisfactory. Neck pain with or without a torticollis deformity was the most common complaint when the C1 vertebral body was affected.[3,10–12] Atlantoaxial subluxation or instability is rarely observed.

However, various conservative measures have been recommended, such as simple observation, bracing, nonsteroidal anti-inflammatory drugs, or casting with or without initial bed rest. There is still no approved treatment guideline until now. Our patient presented without obvious instability and subluxation. Although the collar and radiotherapy effectively relieved our patient's neck pain, atlantoaxial subluxation did not improve. Similarly, Jiang et al[10] also reported 3 adult patients in whom the C2 vertebral body was involved; they underwent no treatment or conservative treatment, and they developed fixed atlantoaxial dislocation. Huang et al[15] reported 5 of 6 adult patients in whom the C2 vertebral body was affected, and they underwent lesion resection and spinal reconstruction after failure of conservative treatment. In Ha et al's case,[7] conservative treatment, including rigid immobilization with a Minerva jacket and low-dose systemic chemotherapy, achieved a satisfactory prognosis in a 6-year-old boy with an EG lesion at the C2 vertebral body. The lesion that affected the C1 vertebral body showed a good response to immobilization with a collar. This is partially because of the mild initial presentation at the C1 level compared to that at the C2 level. Thus, on the basis of limited reports, we propose that adequate strength must be ensured when immobilization is used for a lesion affecting the occipitocervical junction. A rigid collar is probably sufficient for a lesion at the C1 level, whereas a stronger instrument, such as Halo-vest immobilization, is needed for a lesion at the C2 level in patients with secondary deformity and instability.

Another issue worthy of attention is that the epiphysis in adults has ossified and stopped growing, as reported by many researchers.[2,10,15] Thus, the vertebrae cannot remodel itself, especially if they are affected by a large lesion, as in our patient. Once malunion occurs at the lesion site, the surgical difficulty and risk increase. In one patient described by Jiang et al[10] skeletal traction and surgical intervention were ineffective for the fused C1/C2 lateral mass and fixed C1 anterior dislocation. Currently, an advanced surgical technique is sufficient for resecting the lesion and reconstructing spinal alignment, even in the upper cervical spine. However, the surgical protocol is still preserved for specific cases with marked instability, deformity, neurological deficit, or suspected malignancy. On the basis of our patient and literature review, we assume that these risk factors are more likely to induce spinal instability and deformity, which include C2 body damage, an asymmetrical lesion, facet joint involvement, and adult stage and large osteolytic lesions (the criterion may be recommended as 2 cm).[1] Thus, a more radical protocol such as immediate surgery is proposed for these high-risk patients.

In conclusion, more motor and biomechanical characteristics should be considered when immobilization is recommended for an EG lesion involving the occipitocervical junction, especially in adults and in those with asymmetric osteolytic damage. The close follow-up and strict compliance are more essential in patients with these risk factors significant. Surgical intervention can effectively treat a solitary lesion and maintain spinal alignment, which is one option for existing instability or deformity.

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