Eosinophilic Granuloma of Atlas in a Child Treated by Surgical Resection and in Situ Fusion with 3 Years of Follow-Up: A Case Report

Mostafa Mohseni, Rouzbeh Motiei-Langroudi

Department of Neurosurgery, Shohada Tajrish Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran

INTRODUCTION

Eosinophilic granuloma (EG), a rare disorder of clonal proliferation of Langerhans cells, is a benign osseous variant of Langerhans cell histiocytosis. It involves patients of all ages, but most commonly affects children (18). It is mostly seen in the skull, long bones, pelvis and spine. When eosinophilic granuloma affects the spine, it is most often seen in the thoracic, and then lumbar and cervical vertebra (3). Moreover, if it is seen in cervical vertebra, it usually involves the subaxial vertebra in children (19), and the second vertebra in adults (3); up to date, there are only a few reports of atlas involvement by eosinophilic granuloma, either in adults or children (5,7-9,13,14,16,19,22-24).

The clinical course of eosinophilic granuloma is variable and the management of the disease involving cervical spine is challenging. A multitude of management protocols have been proposed for eosinophilic granuloma of the cervical spine, including observation, immobilization (3,6), steroid injection (21), excisional biopsy and low dose radiation (4), chemotherapy (20), and surgical excision without (12) or with fusion (14,16,17). However, no treatment modality has been shown to be superior to the others.

Here, we present a case of eosinophilic granuloma involving the atlas in a 9-year-old child, who underwent a surgical resection of the lesion, and the disease outcome after 2.5 years of follow-up is evaluated.

MATERIAL and METHODS

A 9-year-old boy presented to us with a history of occipital and posterior neck pain after he had hit the
ball with his head during a soccer game 3 weeks before. The pain did not exacerbate with mechanical motion, and did not show diurnal pattern. It was isolated to the posterior head and neck region without radiation to the upper limbs. His pain had not improved with routinely prescribed analgesics and immobilization with a cervical collar. On physical examination, no abnormality or positive finding was observed. He had unremarkable laboratory data. Cervical XR series including lateral, AP, open mouth odontoid view, and lateral flexion-extension views did not show any abnormality (Figure 1A-D). On axial thin-cut computed tomography (CT) scan with 3D reconstruction, an osteolytic lesion was seen in right atlas lateral mass and lateral lamina (Figure 2A). On Magnetic Resonance Imaging (MRI), the lesion had neither extended to paravertebral soft tissue nor compressed the thecal sac (Figure 3). Bone scan showed a zone of increased uptake in right atlas lateral mass without any other bony or systemic involvement.

The patient underwent a posterior approach to resect the lesion on April 2009. In a prone position with the head fixed in Mayfield head holder, the skin and soft tissues

Figure 1: Preoperative cervical XR series, consisting of AP (A), lateral (B), flexion (C), and open mouth odontoid views (D). Extension view is not shown here. No apparent pathology was seen in the XR.
were incised in the midline, with the muscles detached at the craniocervical junction region on the right side and the lateral mass and vertebral artery were exposed. After performing a minimal unilateral laminectomy, a soft gray low vascular lesion was found within the lateral mass and lateral part of lamina of atlas which had touched but not involved the vertebral artery. The tumor was totally resected by curettage under microscopic magnification. The vertebral artery was spared. The defect was filled with synthetic bone granules (Kasios, France) to achieve further fusion. The surgical wound was closed over a closed draining tube in anatomical layers. The patient was discharged two days after surgery without any neurological deficits or complications.

On pathologic examination, the sections revealed fragments of osseous tissue, composed of proliferated histiocytes with lobulated and grooved nuclei and sheets of Langerhans histiocytes with multinucleated giant cells, mixed with infiltrates of eosinophils. The immunohistochemic study stained positive for S100. The whole pathologic study suggested eosinophilic granuloma as the diagnosis.

On follow-up, the patient grew to be pain-free without medication early after surgery and was followed periodically every three months up to 30 months. He was completely asymptomatic at the last visit. The CT scan performed 24 months after surgery showed no evidence of tumor recurrence and the site of fusion had a good appearance with reconstruction of atlas (Figure 2B).

**DISCUSSION**

Eosinophilic granuloma is a rare disorder characterized by clonal proliferation and excess accumulation of pathologic Langerhans cells, without a known etiology.
When it involves the spine, it is most often seen in thoracic, and least often in cervical vertebra (3). Moreover, depending on the age of the patient, most cervical lesions are observed in subaxial vertebra and axis (3,19), and very rarely may be seen in the atlas.

Different treatment strategies have been proposed for eosinophilic granuloma of the cervical region. Regarding the rarity of eosinophilic granuloma in cervical region and its variable clinical course, no treatment modality has proved to be superior to the others. Moreover, a few cases of atlas eosinophilic granuloma have been reported in the literature (a total of 11 reports, including less than 20 cases), which makes the decision for the most optimal treatment even more challenging (5,7-9,13,14,16,19,22-24). The most important factors that influence the decision to choose among the multitude of treatments are the patients’ age, cervical stability, neural compression, and location of pathology (2,14). The therapeutic goals would be spinal stability, preservation of neurological function, and relief of pain (6).

Spontaneous resolution of vertebral EG without further recurrence for up to 5-8 years have been reported; therefore, some authors are prone to manage cervical EG with immobilization by external orthoses and observation, reserving surgery for those with severe or progressive neurological deficit, or those in whom a differential diagnosis is needed (1,3,6,11). However, none of these reports consisted of an atlas EG. Regarding the low rate of neurological consequences of atlas EG, this treatment modality may seem to be a rational option (9,10,16,23).

CT-guided intralesional corticosteroid injection has been proposed as a safe and effective treatment with a low complication rate for spinal EG (21); however, its feasibility in the atlas area should be taken into account, as no report up to date exists for this region. In addition, injection should be performed after confirming pathological diagnosis by biopsy, thus rendering two procedures necessary for the patient (24). This should be added to the probability of inconclusive results obtained by a biopsy, which adds to the disadvantages of this treatment modality.

Others consider low-dose radiation as a treatment choice for EG (4). However, it may have its own disadvantages, including a significant risk of malignant transformation of the lesions after radiotherapy, especially in the pediatric age group (10).

Surgery is also a well-accepted treatment for cervical and atlas EG. Ngu et al (2004) reported a case of eosinophilic granuloma in atlas lateral mass, which was successfully treated by biopsy and then, curettage. Others have also treated cervical EG with high success rate and low complications (12,14). Patients with radiographic evidence of rotary subluxation or atlantoaxial dislocation will also need stabilization and fusion (14), whether through a posterior or anterior transoral route (17).

In our case, the patient had a lesion consistency with eosinophilic granuloma in the lateral mass and lamina of atlas. The lateral mass is a well-recognized place for this lesion, but involvement of the lamina is uncommon (15). As the patient presented to us with a refractory albeit intolerable pain, we decided to choose a treatment other than sole external immobilization and observation. Regarding the potential complications of radiation, we chose surgical resection via a unilateral minimal laminectomy and curettage of the lesion, with further non-instrumented bone grafting to gain fusion. The patient endured the surgery without any complications, and gained complete clinical benefit early after surgery, which also maintained nearly 3 years thereafter. Radiologic workup shows that he is lesion free after this time period, and the bone had acceptable reconstruction.

In conclusion, the results of our case suggest that although no treatment can be stated to be the gold standard for atlas eosinophilic granuloma, surgical resection and in situ fusion with bone grafting may be an acceptable modality.

REFERENCES


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Address correspondence to: Mostafa Mohseni, Department of Neurosurgery, Shohada Tajrish Hospital, Tajrish Sq, Tehran, Iran.
Phone: +9821 227 180 27
email: mfmohseni@yahoo.com