Case Report

# Isolated enchondroma of the atlas


## ABSTRACT

Periosteal enchondromas located in the spine are rare. We reported an even more infrequent occurrence of a periosteal enchondroma in the cervical spine of a 19-year-old girl. During the operation, a giant (>8 cm × 5.5 cm × 5 cm) ossified periosteal enchondroma with involvement of posterior structures and muscles of the axis was resected. The pathological examination revealed that the tumor consisted of enchondroid tissue with typical chondrocytes, confirming the diagnosis of periosteal enchondroma. Early identification of the initial lesion should be coupled with total surgical resection, as a definitive treatment, to prevent malignant transformation. Enchondromas grow in an expanding manner which makes easy total resection.



**Keywords:** Atlas, isolated enchondroma, vertebra


## INTRODUCTION

Enchondroma is one of the rare tumors of the spine.[1] The symptoms are most commonly pain and swelling at the site of the lesion; however, compression of the cord can cause myelopathy.[2-4] Curettage is the traditional treatment of enchondromas; however, block excision of the tumor with margins of the normal bone is the best treatment which we could achieve in our case. Here, we presented a 19-year-old girl with enchondroma of the atlas without any neurological symptom.

## CASE REPORT

A 19-year-old girl presented with a history of progressive onset of swelling at her neck for the past 5 years without any neurological symptom. There was no history of trauma. The patient was evaluated, and the clinical examination revealed all the features of no cervical myelopathy. Computed tomography (CT) scan showed a partially ossified lesion (8 cm × 5.5 cm × 5 cm) arising from the spinous process of cervical C1 [Figure 1a and b]. Magnetic resonance imaging, in addition to confirming the CT findings, also confirmed that there was no compression of the spinal cord by this lesion [Figure 2a and b]. The patient was operated upon through a posterior midline approach. A well-defined bony lesion was found to be arising from the spinous process of the atlas. The lesion was excised completely.

Histopathological examination of the specimen showed outer perichondrium, cartilage cap, and underlying stalk. Endochondral ossification was also seen at the base which is a normal feature and should not to be interpreted as a malignancy invading the stalk. The patient was followed up for 12 months, and there was no pathological swelling on her neck.

## DISCUSSION

Enchondroma, a common benign cartilaginous tumor involving the acral skeleton, is extremely rare in the vertebral column and even more rare in a vertebral body of the cervical spine.[5,6] Enchondroma is accounting for 5% of all bone tumors, affecting the short tubular bones of hands and feet


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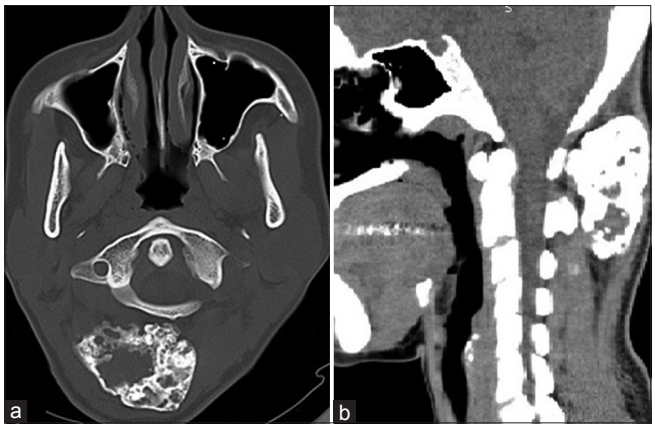

Figure 1: (a and b) Axial and sagittal sections of computed tomography showing enchondroma of the atlas

in over 50% of cases.[7] Spinal involvement is seen in only 2% of cases, and enchondroma is almost unknown in the skull.[7] The most common tumors present in the upper cervical region are neurofibroma and meningioma. Enchondromas have been described as occurring in the second to eighth decades of life, with most presenting during the second to fourth decades.[7] There is no sex or racial predilection in these tumors.

Enchondromas are often asymptomatic because of their slow growth but may have varied presentation. There are many ways of classifying benign chondrogenic tumors, and the widely accepted classification is the one proposed by Lichtenstein.[8] There are four histological types in this classification which are as follows: osteochondromas, enchondromas, chondroblastoma, and chondromyxoid fibroma.[9] Each type is distinct in its frequency and location of occurrence. Although there may be clinical similarities, the X-ray presentation, histopathology, and propensity to undergo malignancy vary a lot according to the subtype. Enchondromas are often asymptomatic because of their slow growth.[9]

An enchondroma may occur as an individual tumor or as several tumors together. The conditions that involve multiple lesions including the following: Ollier's disease (enchondromatosis) – when multiple sites in the body develop the tumors and Maffucci syndrome – a combination of multiple tumors and angiomas. Solitary enchondromas are most often seen in the age group of 20–40 years. Oliver's disease is usually detected at 0–10 years.

In our case, tumor growth was so slow, and expanded peripheral structures for 5 years and caused no neurological symptom. However, this slow growth may give a clue about the benign nature of the lesion; but so large mass(8 cm × 5.5 cm × 5 cm) be should be resected and investigated for malignancy. However, surgical morbidity may be substantial,

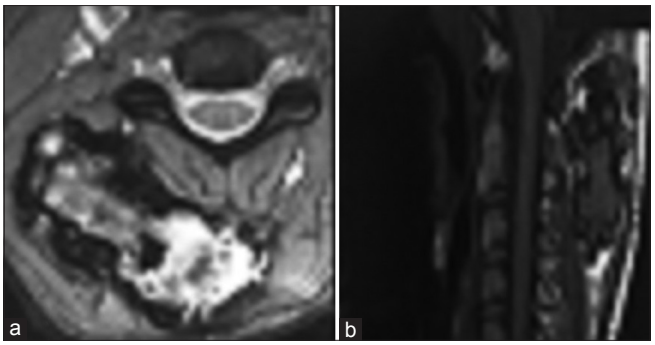

Figure 2: (a and b) Axial and sagittal sections of magnetic resonance imaging showing enchondroma of the atlas

given the propensity for chordomas to abut or surround neural, vascular, and visceral structures. Thus, early recognition is essential.

We think that early identification of the initial lesion should be coupled with total surgical resection, as a definitive treatment, to prevent malignant transformation. Enchondromas grow in an expanding manner which makes easy total resection. We should aim to total resection. Although enchondroma of the cervical spine is an uncommon diagnosis in the spectrum of benign tumors of the spine, it should be considered in the list of differential diagnoses when an expansile lytic lesion involving the spine is encountered.

## 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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