Osteoid Osteoma of the Atlas in a Boy: Clinical and Imaging Features–A Case Report and Review of the Literature

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Neuropediatrics

Abstract

Osteoid osteoma is a benign osteoblastic tumor, quite uncommon in the spine. We report a case of an osteoid osteoma involving the atlas in a 6-year-old boy, who presented with suboccipital pain and torticollis. Initial radiological findings were ambiguous as magnetic resonance imaging showed mainly edema of upper cervical soft tissues. The subsequent computed tomography depicted a lesion of left lamina of C1. As conservative treatment failed, the lesion was surgically resected and the patient became pain free. To our knowledge, this is the first case of osteoid osteoma involving the atlas associated with abnormal soft tissue reaction reported in literature.

Keywords
- osteoid osteoma
- cervical spine
- torticollis
- soft tissue reaction
- surgical resection

Introduction

Osteoid osteoma (OO) is a small benign tumor accompanied by abnormal pain. It can virtually arise in any bone, but it affects more frequently lower extremities. Spinal locations represent around 9% of cases.

This report is a case of a spinal OO involving the neural arch of the atlas, characterized by soft tissue reaction, unusual for a vertebral OO.

Case Description

A 6 year-old-boy was admitted to our hospital in January 2018, complaining of sub-occipital pain and torticollis. Symptoms occurred over a period of 4 weeks after herpetic lesion on his left shoulder that completely regressed with medications. There was no history of traumatic injury. On clinical examination, patient presented with torticollis and swollen cervical lymph nodes. Neither the pain nor the torticollis could be controlled with nonsteroidal anti-inflammatory drugs (NSAIDs).

On the magnetic resonance imaging (MRI) of cervical spine, the soft tissues adjacent to the left lamina of C1 showed high signal intensity on T2-weighted images consistent with massive diffuse soft tissue edema (►**Fig. 1A**) and several enlarged lymph nodes (►**Fig. 1B**); gadolinium enhancement was observed (►**Fig. 1C**). The subsequent computed tomography (CT) demonstrated a small lytic area with sclerotic central component located in the left arch of the atlas (►**Fig. 2A, B**); furthermore, the apex dentis of axis showed lytic aspects (►**Fig. 2C**).

As conservative treatment failed, surgery was undertaken with the intention of excising the lesion through a posterior approach. Through a midline incision, the paravertebral muscles were retracted and stripped off the lamina of C1 mostly on the left side. Then, the cortical shell was shaved off

received
April 1, 2020
accepted after revision
July 2, 2020

DOI https://doi.org/10.1055/s-0040-1715488.
ISSN 0174-304X.

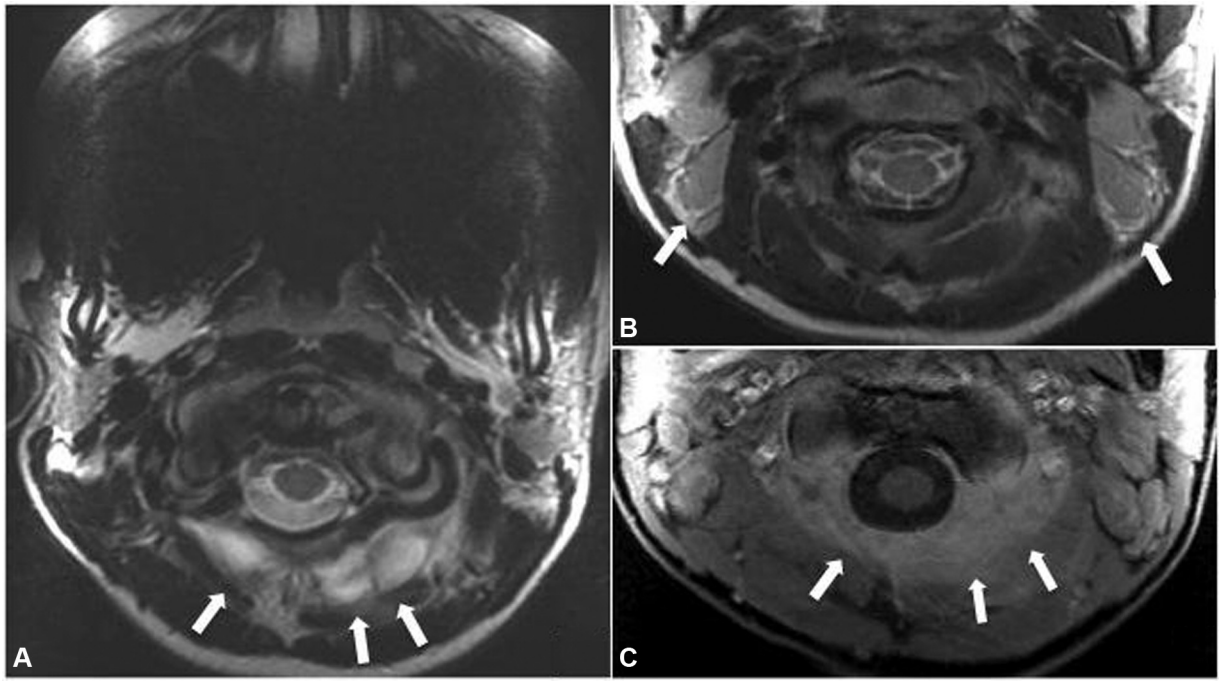

Fig. 1 Axial T2-weighted magnetic resonance imaging(**A**). There is diffuse edema of the soft tissues adjacent to the left arch of C1 (arrows). Several enlarged lymph nodes are visible (arrows) along with nuchal chains. (**B**) Axial T1-weighted magnetic resonance imaging after gadolinium injection shows enhancement of neighboring tissues (arrows) (**C**).

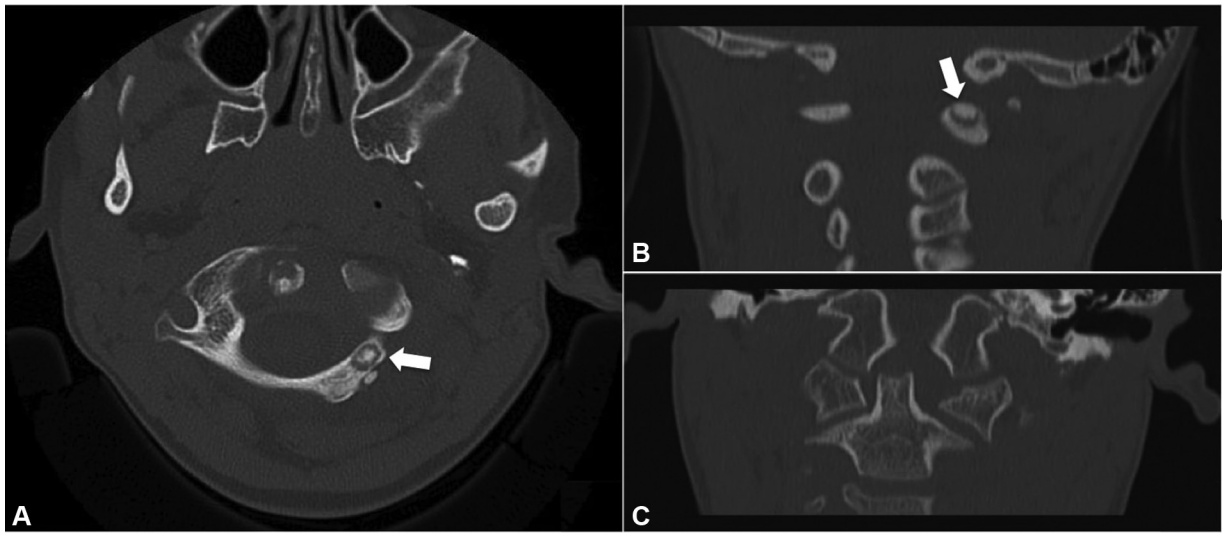

Fig. 2 (**A**) Axial computed tomographic scan shows an oval-shaped radiolucent lesion with sclerotic central component (arrow) in the left arch of C1; features are nicely revealed by the coronal reformatted image (**B**). (**C**) Coronal reformatted image at a more anterior plane shows lytic changes of the apex dentis.

using a high-speed drill and the tumor nodule could be removed. The patient had an uneventful recovery and he became pain free with full range of neck motion. The histopathology report was compatible with the diagnosis of OO (►**Fig. 3**).

CT scan performed 2 months after surgery showed complete resection of the nidus, with interruption of the posterior cortical surface along the surgical pathway (►**Fig. 4A**). The MRI performed at 6 months showed no signs of recurrence and disappearance of soft tissue edema (►**Fig. 4B**).

Discussion

OO is a benign bone-forming tumor, small in size, with a limited growth potential, accompanied by abnormal pain. It accounts for ~12% of benign bone tumors, and it affects most often patients between 7 and 25 years old, with a male-to-female ratio of 2:1 to 4:1.[1,2]

OO was first referred into the literature in 1935 by Jaffe, who identified it as a discrete pathologic entity, describing five cases of benign osteoblastic lesions. Histologically, it is composed by vascularized connective tissue with osteoblastic activity mixed

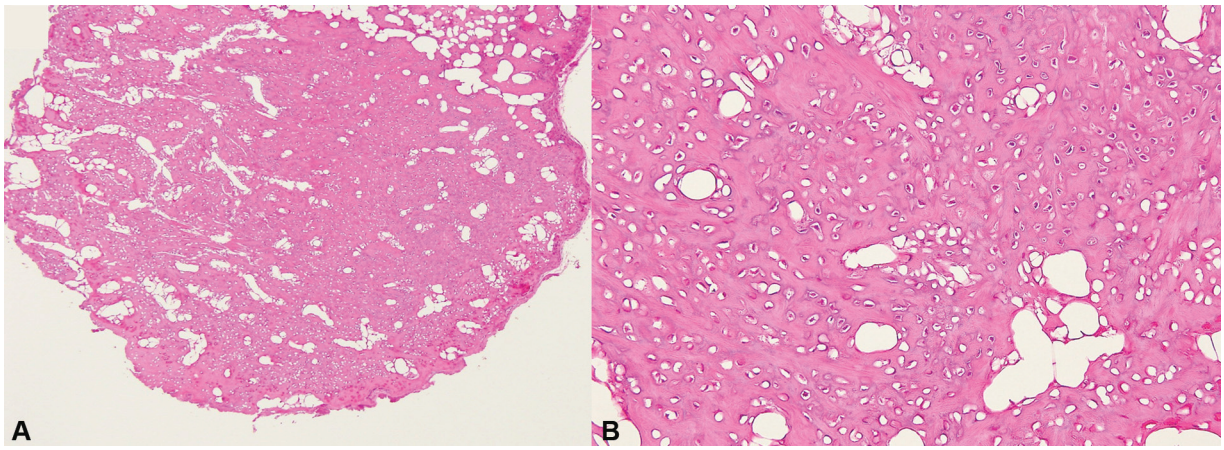

Fig. 3 Bone tissue with osteoid deposits variously anastomosed and partially surrounded by osteoblasts. ([**A**], hematoxylin and eosin [H&E] ×40) and ([**B**], [H&E] ×200).

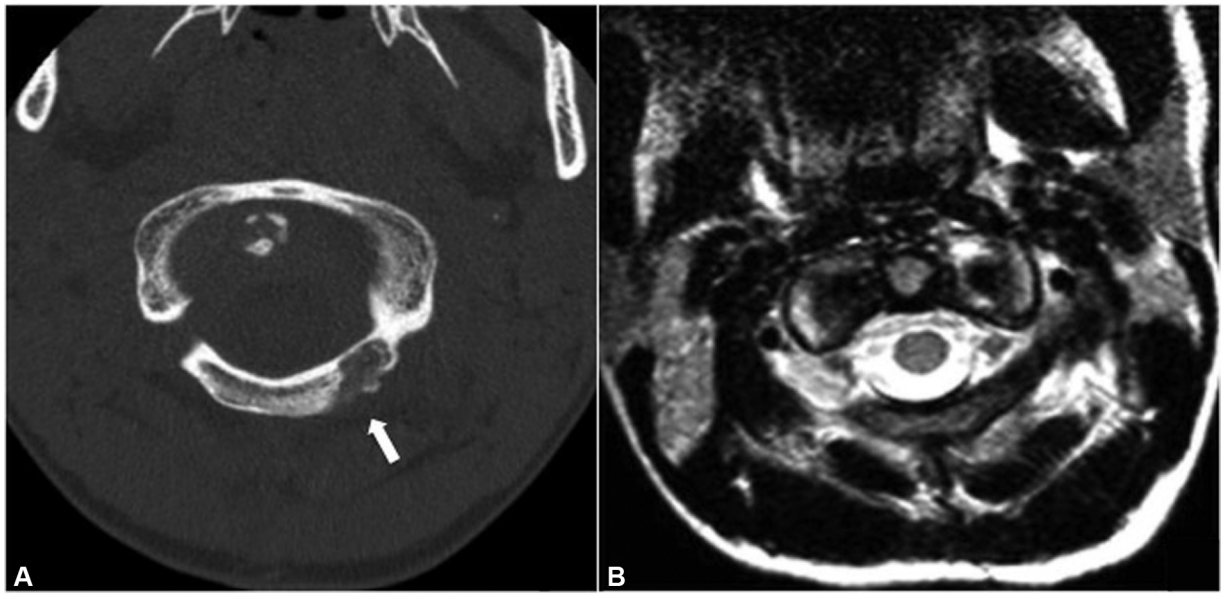

Fig. 4 (**A**) Computed tomographic images on axial plane performed 2 months after surgery confirms disappearance of the nidus (arrow). (**B**) Magnetic resonance imaging performed 6 months after surgery reveals no more edema of the soft tissues located near the left arch of C1.

with osteoid, woven bone, and few osteoclasts. These elements make the nidus that is usually surrounded by thickened hypervascular sclerotic cortical bone.[1]

OO rarely evolves to osteoblastoma; currently, size is considered the most reliable criterion to distinguish them, defining lesion smaller than 15 mm as OO.[2]

Clinically, in 70 to 75% of patients, OO manifests with nocturnal, worsening pain that can relieve with salicylates because of the presence of prostaglandins within the nidus.[3]

Depending on the affected site of the bone, OO can be classified as cortical, medullary, and subperiosteal. Regarding the proximity to the synovial tissue, they can be further categorized in extra- and intra-articular. The latter are considered as a separate entity because of their peculiar clinical and imaging aspects: pain not necessarily worsening at night and radiological severe inflammatory reaction of the adjacent tissues can make the diagnosis challenging.[4]

Despite its ability to potentially affect any bone, OO is usually located in the lower extremities.[3] Spine is affected in 9% of cases, in particular the lumbar tract (59%), followed by cervical (27%), thoracic (12%), and sacral (2%) vertebrae. The nidus is most frequently located in the neural arch and patients can suffer from radicular pain, gait disturbance, and limb atrophy.[5] According to the literature, torticollis is the main symptom for lesions located in the cervical segment, as in our case.[6,7] CT is the best modality to identify spinal OO: it can easily identify the nidus, as a well-defined area of low attenuation, with or without a variable amount of internal high density that represents the osteoid component of the nidus.[7,8] Compared with CT, MRI has limited value in demonstrating the nidus, but it is able to depict changes of the host bone and surrounding tissues.[9,10]

To date, there are only seven cases of OO involving the atlas in the previous literature, and none of them was accompanied with abnormal soft tissue reaction, as described in our case

Table 1 Summary of previously reported cases of osteoid osteoma involving C1

Reference	Year	Age/sex	Symptoms	Location	Soft tissue reaction on MRI	Treatment	Outcome
Fielding et al[13]	1977	?/M	?	?	None	En-bloc resection	?
Wedge et al[7]	1981	10/M	Torticollis	Lamina	None	Hemilaminectomy	Good
Jones[6]	1987	8/M	Torticollis	Lamina	None	En-bloc resection + radiofrequency	Good
De Praeter et al[12]	1999	21/M 22/M	Neck pain Neck pain	Lamina Lamina	None	Hemilaminectomy Hemilaminectomy	Good Good
Bruneau et al[2]	2005	25/M	Neck pain	Lateral mass	None	Drilling	Good
Amirjamshidi et al[11]	2010	14/M	Neck pain	Lateral mass	None	Drilling	Good
Yang et al[14]	2018	30/F	Neck pain	Lateral mass	None	Radio frequency ablation	Good
Present study	2018	7/M	Torticollis	Lamina	Present	Drilling	Good

Abbreviation: MRI, magnetic resonance imaging.

(►**Table 1**).[2,6,7,11–14] These features are probably due to the close relation between the nidus, located in the left lamina, and the adjacent ligaments, making this lesion seem like an intra-articular OO.

In addition, the lytic appearance of the apex dentis and the presence of swollen lymph nodes may also be related to the abnormal inflammatory reaction.

Nowadays, the appropriate therapeutic approach for OO remains controversial. The "wait-and-see" regimen using NSAIDs seems to have some success, considering the high rates of prostaglandin E2 (PGE_2) detected within the nidus, but the effectiveness of NSAIDs may be temporary. Some studies demonstrated that PGE_2 synthesis by osteoblast in culture is initially high and then rapidly decreases, since there are other proteins involved in the mechanism of pain.[15] Percutaneous minimally invasive approaches, such as radiofrequency ablation or laser thermocoagulation, constitute valuable therapeutic tools, but surgical resection is an effective instrument in selected cases, when minimally invasive strategies are not applicable.[16,17] Complete resection of the nidus is the goal of treatment for OO. The recurrence/persistence rate is statistically low for OO (4.5%), compared with osteoblastoma (9.8–-15%), usually as a result of incomplete resection.

Conclusions

The upper cervical region is a rare location for OO, but this lesion should be considered in the differential diagnosis of young patients complaining of persistent occipital pain and torticollis. CT and MRI are the preferred modalities for delineating the lesion and the associated radiological features. To our knowledge, this is the first case of OO involving the atlas accompanied with abnormal soft tissue reaction reported in literature. Conservative treatment is suggested as the initial management, but direct microsurgical resection can be advised when surgical morbidity is acceptable, especially as a radiation free-treatment in young patients.

Conflict of Interest
None.

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