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CASE REPORT

# Pseudomalignant osteoblastoma of the odontoid process

**Pedro Fernandes[1] · Joaquim Soares do Brito[1] · Adília Costa[2] · Jacinto Monteiro[1]**

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## Abstract

**Introduction** The anterior elements of the spine, particularly the odontoid processes, are a rare location for osteoblastomas. Pseudomalignant osteoblastomas are themselves rare histologic types and are also extremely rare in this location. Most osteoblastomas are Enneking stage 2 lesions; less frequently, they can be more aggressive with extra-capsular extension (Enneking stage 3). En bloc resection is recommended for aggressive lesions, but the literature is less clear regarding the approach to stage 2 tumors, particularly those with pseudomalignant histologic features.

**Case report** A 6-year-old male child presented with a type III pathologic fracture of the odontoid. The fracture healed but upon 6-month follow-up CT scanning, an expansile lesion was detected. Surgical biopsy revealed an osteoblastoma which was treated with intralesional excision. Meanwhile, the excised specimen showed histological features of a pseudomalignant osteoblastoma. Despite this diagnosis, no further treatment was undertaken. At a 10-year follow-up, the patient was free from pain and had full range of motion of the cervical spine; no recurrence was detected.

**Conclusion** This unique case of odontoid osteoblastoma illustrates that malignant behavior may not be predicted only by the presence of pseudomalignant features on histology.

**Keywords** Osteoblastoma · Pseudomalignant · Odontoid process

## Introduction

Osteoblastoma is a rare and benign osteoid-producing primary bone tumor which occurs mainly in long bones [1–4]. Nonetheless, osteoblastomas in the spine represent nearly 35% of all osteoblastomas and the cervical spinal is most frequently affected (25–39%) [5–7]. Involvement of the anterior spinal elements is unusual and is normally secondary to anterior extension of the tumor [8]. The involvement of the odontoid process is particularly rare and has only been reported in the literature on a few occasions [9–11].

Since the first descriptions of osteoblastoma, the issue of distinguishing it from osteoid osteoma has been continually raised [1, 2, 12, 13]. It is commonly accepted that osteoblastoma differs from osteoid osteoma in its ability to grow larger than 2 cm in diameter [7, 14]. Other major issues to address with osteoblastomas are the broad spectrum of clinical behaviors presented by these usually benign tumors [15]. Therefore, while some osteoblastomas are difficult to differentiate from osteoid osteoma, others could present with pseudomalignant features and must be differentiated from osteogenic sarcoma [16, 17].

Taking into consideration the difficulty in obtaining a proper classification, and knowing that en bloc resection is usually the best option for aggressive osteoblastomas, the authors herein present a rare Enneking stage 2 pseudomalignant osteoblastoma of the anterior spine odontoid process that at 10-year follow-up had an excellent outcome following intralesional resection.

## Case report

A six-year-old male child was admitted to the emergency department with severe neck pain after a minor fall. He was unable to support his head independently but a full neurological examination was normal. Although the first computer tomography (CT) was erroneously reported as negative for

✉ Joaquim Soares do Brito
joaquimsoaresdobrito@gmail.com

[1] Orthopaedics Department, Centro Hospitalar Lisboa Norte, University Hospital of Santa Maria, Avenida Professor Egas Moniz, 1649-035 Lisbon, Portugal

[2] Pathology Department, Centro Hospitalar Lisboa Norte, University Hospital of Santa Maria, Lisbon, Portugal

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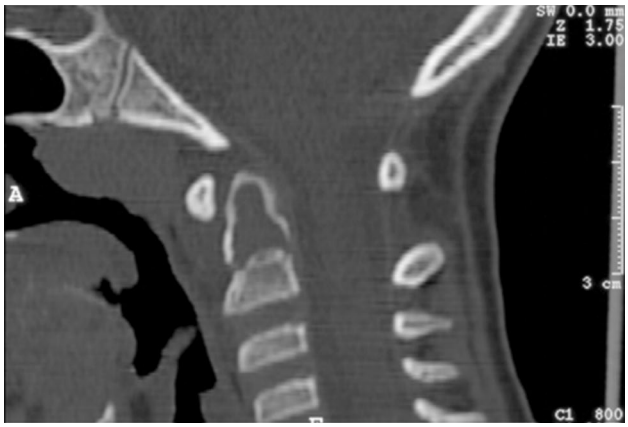


**Fig. 1** Pathologic type III odontoid fracture fracture—CT scan sagittal view

any acute pathology, the fact that the patient had to hold his head up with both hands to allow any movement led us to re-evaluate the examination; central osteolysis in the odontoid process was then validated. The odontoid margins were well defined with a sclerotic rim and a small pathologic fracture (Fig. 1). Magnetic resonance imaging (MRI) scanning was consistent with a benign condition and was suggestive of an eosinophilic granuloma with high-intensity bony and soft tissue signal considered part of the acute fracture.

Conservative treatment with a hard collar was begun with immediate improvement in the patient's symptoms. 6 months later (after missing a 3 months appointment), radiographs showed an unexpected expansive lesion confirmed on CT scan having a scalloped edge with undefined margins (Fig. 2). Almost all the vertebral body and both pedicles, more on the right than the left, were now involved. An MRI continued to show a high-intensity bony and soft tissue signal in almost all the ondontoid and the body of C2 body, despite fracture healing. A scintigram revealed very high osteoblastic activity not supporting an eosinophilic granuloma diagnosis. Open biopsy through an anterior approach was performed and the histopathology showed the usual characteristics for an osteoblastoma. Due to the location of the tumor, an intralesional extended curettage and high-speed burring of any approachable transitional area was planned. Surgery was performed through a high antero-lateral approach with removal of the anterior part of the C2 body and odontoid under the C1 arch (which was left intact). Tumor tissue was removed and the gap was filled with a mixture of iliac bone autograft and synthetic bone paste. A halo-vest was applied and worn for 8 weeks.

To our surprise, the histopathology of the removed tumor revealed unusual and worrisome features like those described in aggressive osteoblastoma and in pseudomalignant osteoblastoma (Fig. 3). Due to the CT and MRI findings and the absence of mitotic figures, it was decided in

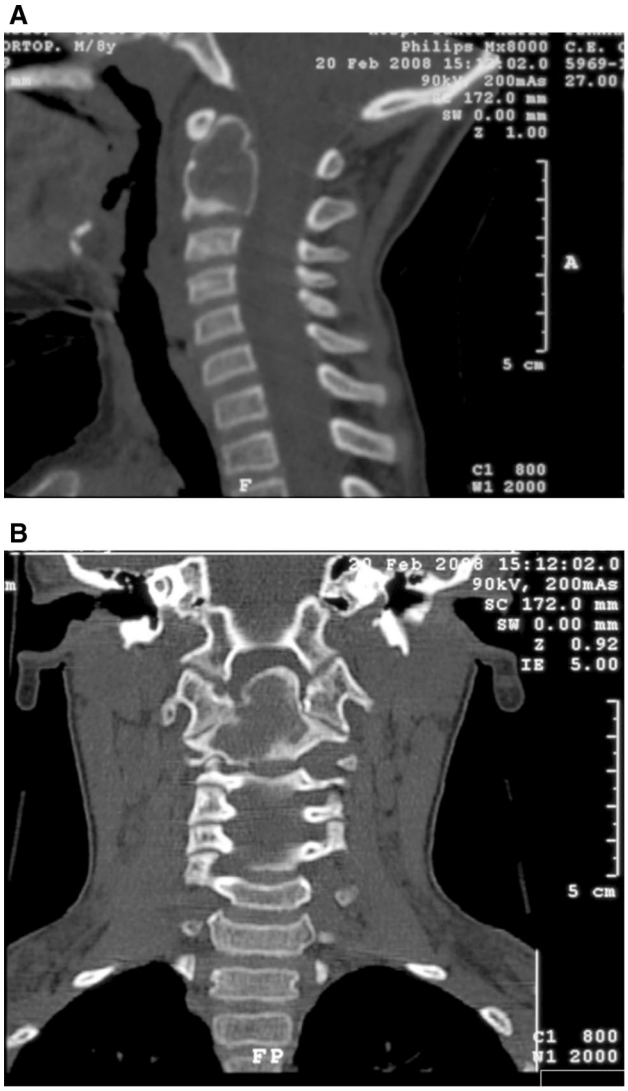


**Fig. 2** Healed fracture but continuous expansible lesion—CT scan sagittal (**a**) and coronal (**b**) views

our multidisciplinary conference to follow closely for local recurrence without any further treatment. At a subsequent long-term follow-up (10 years), the patient presented with full and painless cervical spine range of motion with no local recurrence (Fig. 4).

## Discussion

Osteoid-producing primary bone tumors are rare lesions with the most prevalent being osteoid osteomas and osteoblastomas [18]. Early on, Lichtenstein called today's osteoblastoma an "osteogenic fibroma of bone" [19]. Then in 1954, Dahlin and Johnson reported 11 unusual tumors, all of which appeared to originate within bone. They chose to call these tumors "giant osteoid osteoma" for their histologic

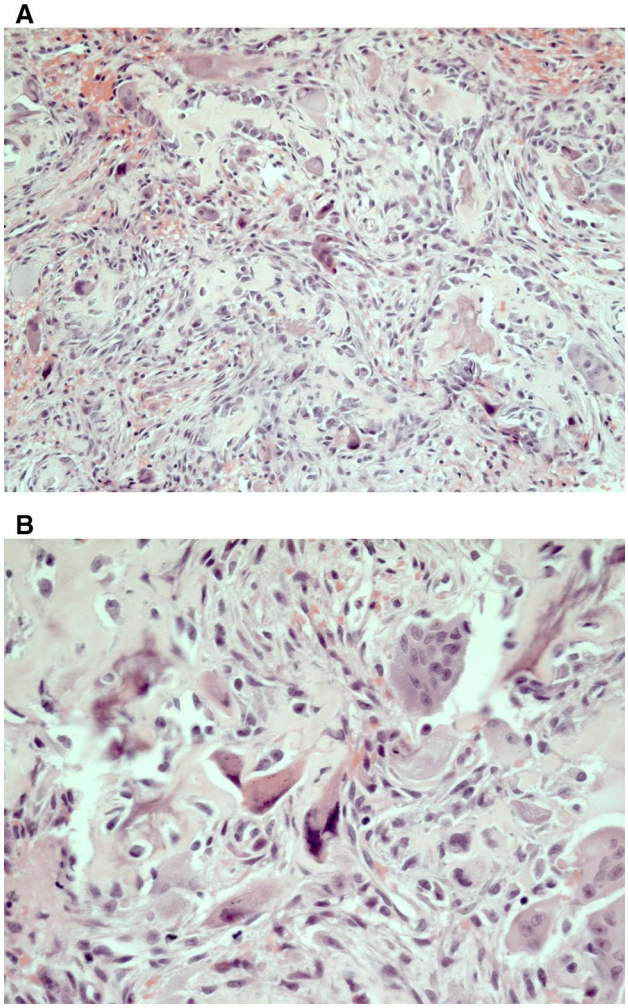


**Fig. 3** The tumor has enlarged epithelioid osteoblasts with abundant cytoplasm and scattered large cells with bizarre and hyperchromatic nuclei. Some osteoclasts are also seen. No mitoses were clearly found and there was no necrosis (H&E, **a** ×200; **b** ×400)

similarity to osteoid osteoma [12]. The current and generally accepted name, osteoblastoma, was arrived at from different and independent publications by both Lichtenstein and Jaffe [1, 20].

Osteoblastomas are rarer than osteoid osteomas, with a reported incidence between 10 and 25% of all primary osseous spine tumors [18–21]. Classification is based on radiographic characteristics of the tumor, where well-demarcated borders are indicative of latent lesions and indistinct borders result from permeation into host bone and indicate a more aggressive entity [22, 23]. Most osteoblastomas are Enneking stage 2 lesions; more aggressive tumors are less frequent [5].

The radiographic findings in classic osteoblastoma include a well-circumscribed, osteolytic-osteosclerotic lesion with an expansile, scalloped or lobulated appearance. Sometimes there is erosion of the cortex, and a common finding is the presence of a sclerotic rim representing a reaction from the bone and periosteum [7, 14, 24]. Some osteoblastoma cases may mimic malignancy, particularly when there is expansion, destruction of the bone cortex and new periosteal bone formation [25]. Lucas et al. in their series on osteoblastomas noted radiologic findings suggestive of malignancy in about 12% of cases [26]; in another osteoblastoma series, Rocca et al. found radiologic features for aggressive Enneking stage 3 osteoblastomas in 16% of all cases [27]. Osteoblastoma local aggressiveness has been well documented by Marsh et al. in 1975, when he reported on 25 new cases of osteoblastoma, reviewing also 172 previously reported cases. In two of the new osteoblastoma cases, Marsh described a clinically aggressive presentation requiring several surgical procedures, with some difficulty in establishing a proper histological diagnosis. One particular case ended in the patient's death 34 months after the initial surgical procedure, allegedly due to continued local tumor expansion [15]. On the other hand, in 2012 Boriani et al. published a review of 51 surgically treated patients with osteoblastoma of the spine, and classified the tumors with the validated Enneking staging system. They concluded that 80% of the reviewed cases (41 of 51) would have been better classified as aggressive Enneking stage 3, due to features such as erosion of the cortex, and invasion of the vertebral canal and/or the surrounding soft tissues [29]. The aggressive nature of these tumors was also well illustrated in a study by Raskas et al., in which 56.6% of osteoblastomas invaded the epidural space, requiring dissection of the dura mater. However, these features occurred in none of 159 osteoid osteomas [7].

Of all imaging techniques, CT scanning is the most useful since it can identify the lesion, the degree of sclerosis, and the extent of bony involvement [29]. If the radiologic findings suggest an aggressive lesion, biopsy should be done, including the periphery of the tumor or the cortical bone surrounding the tumor [16]. MRI has a limited role in primary osseous tumors, since it poorly visualizes bone marrow. Additionally, visualization of the margin between osseous and soft tissues in MRI is less defined, which can result in an inaccurate diagnosis of an aggressive or malignant lesion [30, 31]. Nonetheless, MRI is important to evaluate soft tissue involvement and the relationship with neurovascular structures [32, 33].

Differentiation between osteoblastoma and osteosarcoma poses great challenges [27, 28]. Osteoblastomas with aggressive radiographic and histologic characteristics have been titled by several authors as aggressive osteoblastomas [26, 34]. Other authors use the term pseudomalignant osteoblastoma or even malignant osteoblastoma to define osteoblastomas with clinical, radiologic and histologic features of aggressiveness. To our knowledge, the term pseudomalignant was first applied to osteoblastoma by Joseph

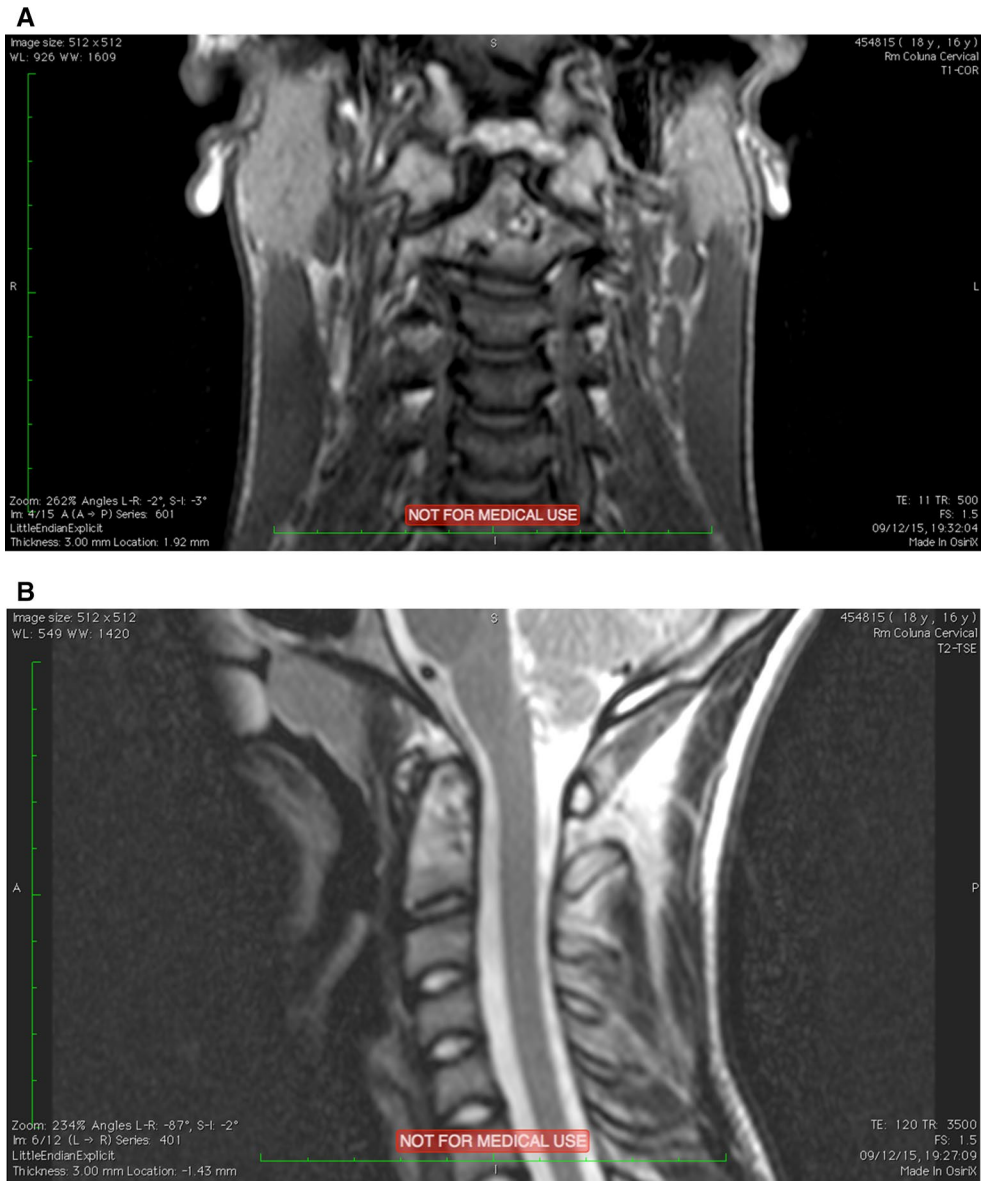


**Fig. 4** 10 years post-operative—MRI T1 (**a**) and T2 (**b**)

Mirra in 1976 [17]. In his paper, Mirra presented a case report wherein the clinical, radiologic and histologic findings supported the diagnosis of a proximal fibular pseudomalignant osteoblastoma; however, at the time he could not completely exclude the diagnosis of osteogenic sarcoma. In the same year, Schajowicz and Lemos reported an eight-case series describing osteoblastoma features that should be considered for a malignant osteoblastoma diagnosis [35]. Later, Cheung et al. better described the diagnostic criteria for pseudomalignant osteoblastoma [24]. Sometimes it is difficult to differentiate osteoblastoma from osteogenic sarcoma; however, osteoblastoma lacks the necessary criteria to be considered a sarcoma [17, 35]. Terminology presently used to describe these tumors is still controversial as it is difficult to predict their behaviors and the expected prognosis [21, 36].

Aggressive osteoblastoma, as traditionally described by Dorfman, is nonetheless considered to be a benign lesion with distinctive histologic features, which include the presence of large epithelioid osteoblasts, stromal mitosis, areas of increased cellularity and a disordered osteoid matrix with absence of atypical mitosis [34]. Despite these histologic features—which are likely associated with an aggressive behavior—in the absence of local recurrence, the osteoblastoma is usually classified as epithelioid [37]. However, this variant is expected to have a higher probability for recurrence, although without the potential to produce metastases [34].

The distinguishing histological features of pseudomalignant osteoblastoma as described by Mirra include the presence of large, sometimes multilobulated nuclei with dark chromatin (degenerative atypia) in the absence of mitotic figures [17]. This diagnosis should be established with great caution and be reserved for tumors that show atypical clinical behavior [37]. Moreover, degenerative atypia may be difficult to distinguish from true anaplasia and may be related to ischemia [17]. In 1997, Cheung et al. developed diagnostic criteria for pseudomalignant osteobastoma; areas of classical osteoblastoma coupled with atypical osteoblasts and an absence of mitoses. These constituted key points to allow the diagnosis of pseudomalignant osteoblastoma [24]. On the other hand, Schajowicz and Lemos considered the presence of immature bone tumor (similar to that observed in conventional osteosarcoma) to be the main feature to designate an osteoblastoma as malignant [35].

In our clinical case above, the first histopathologic study obtained through an open biopsy showed the usual characteristics for a classic osteoblastoma, and with this diagnosis we performed a successful, extensive, intralesional resection. However, the histopathology of the removed tumor revealed large epithelioid osteoblasts, degenerative atypia and disordered osteoid matrix, with an absence of mitotic figures. Despite these worrisome features, which are usually seen in aggressive and pseudomalignant osteoblastomas, no further intervention was instituted. We also hypothesize that the degenerative atypia seen in our case could have been related to changes induced by the previous biopsy [17]. In fact, Boriani et al. showed a recurrence rate 10 times higher in patients after intralesional excision previously submitted to open biopsy. The authors also concluded that even en bloc resection was scarcely effective in "non intact" patients [29].

To our knowledge, this is the first pseudomalignant osteoblastoma of the odontoid process reported in the literature. This unique case illustrates that aggressive tumor behavior may not be predicted simply by the presence of histologic epithelioid and pseudomalignant features. An excellent clinical result was achieved 10 years following an extensive intralesional resection. The clinical course of this particular case does not disprove the need for en bloc excision in aggressive or pseudomalignant osteoblastomas, but rather highlights the need for further investigation to better understand this clinical entity, especially so we may better address this tumor in certain areas where en bloc resection can be challenging.

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## Compliance with ethical standards

**Conflict of interest** None of the authors has any potential conflict of interest.

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