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A Rare Case of Ethmoido-Maxillary Osteoblastoma: Case Report and Systematic Review

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Abstract **Case Report**

Osteoblastoma (OB) is a rare benign osteogenic tumor, primarily found in the axial skeleton and long tubular bones. It accounts for <1% of all primary bone tumors, mainly affecting young adults. Maxillofacial involvement is rare, often manifesting in the mandible. Osteoblastoma's pathogenesis is unclear, with slow growth and varied clinical presentations. Radiologically, it appears as a well-defined, osteolytic lesion with sclerotic rims. The definitive diagnosis relies on histopathology, with immunohistochemistry aiding in challenging cases. Differential diagnoses include fibrous dysplasia, osteoid osteoma, osteomas, cemento-ossifying fibroma, and low-grade osteosarcoma. Surgical excision with safety margins is the standard treatment, while radiotherapy may be considered for certain cases. Prognosis is generally good, but long-term follow-up is essential due to recurrence risks, particularly in aggressive forms. Malignant transformation can occur, emphasizing the importance of careful diagnosis and management.

Keywords: Osteoblastoma, Ethmoidomaxillar bones, Surgery, Recurrence, Prognosis.

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INTRODUCTION

Osteoblastoma (OB), a rare benign osteogenic tumor, typically occurs in the axial skeleton and long tubular bones. However, its ethmoidomaxillary localization is exceptionally rare. Characterized by osteoblast proliferation, OB's clinical and radiological diversity poses diagnostic challenges. Accounting for <1% of primary bone tumors. The scarcity of maxillofacial occurrences, especially in the ethmoidomaxillary region, emphasizes the need for precise diagnostic consideration. This paper explores the histopathological, radiological, and clinical aspects of ethmoidomaxillary osteoblastomas, shedding light on their diagnostic complexities and outlining potential therapeutic approaches (Wang *et al.*, 2018) (Sanae *et al.*, 2021).

CASE REPORT

An 18-year-old patient with no pathological history presented to the clinic with a painful swelling of the left nasomaxillary area that had been evolving for 3 months, progressively increasing in size and causing exophthalmos of the left eye and unilateral nasal obstruction. Clinical examination revealed a hard, non-inflammatory, painless mass, fixed in relation to the deep plane and integral with the bone. Visual acuity was normal, with no oculomotor disturbances or diplopia. Endobuccal examination revealed an upper left vestibular filling extending from the 23 to the 25 teeth.

A facial CT scan revealed a condensing, calcium-dense, lobulated lesional process measuring 4.9x4.3x3.7 cm, responsible for a narrowing of the left nasal cavity with deviation of the nasal septum, subtotal ethmoidal filling and displacement of the internal wall and left orbital floor, responsible for grade II exophthalmos (Fig 1), suggesting an ethmoidomaxillary osteoma.

A biopsy, performed under general anesthesia, was non-conclusive, revealing inflammatory material with no sign of specificity.

The therapeutic approach was total resection of the tumour (Fig 2) with a safety margin via the vestibular approach, after a segmental maxillary osteotomy, and including the internal part of the orbital floor reconstructed with a titanium grid fixed in place with two 5-mm screws, followed by maxillary reduction and osteosynthesis with a screw plate.

The post-operative course was marked by the appearance of limited elevation of the left eye, associated with diplopia following incarceration of the inferior right muscle, which necessitated re-operation of the patient, with disincarceration of the muscle and repositioning of the titanium grid bordered by a polypropylene plate. On the other hand, the patient presented with patchy necrosis of the maxillary alveolar bone, for which he was taken back to the bloc with curettage and tooth extraction (22-23-24).

The post-operative course was simple, with resolution of ophthalmological problems and the placement of a removable dental prosthesis (Figure 3).

Histological analysis of the surgical specimen revealed osteoid tissue within edematous vascularized fibrous tissue including a large, thick-walled vessel in the center, associated with multiloculated cells, suggesting a diagnosis of benign osteoblastoma (Figure 4).

Finally, the diagnosis of an ethmoido-maxillary osteoblastoma was made on the basis of the following arguments

A follow-up CT scan (Fig 5) was performed after 10 months, showing no signs of recurrence.

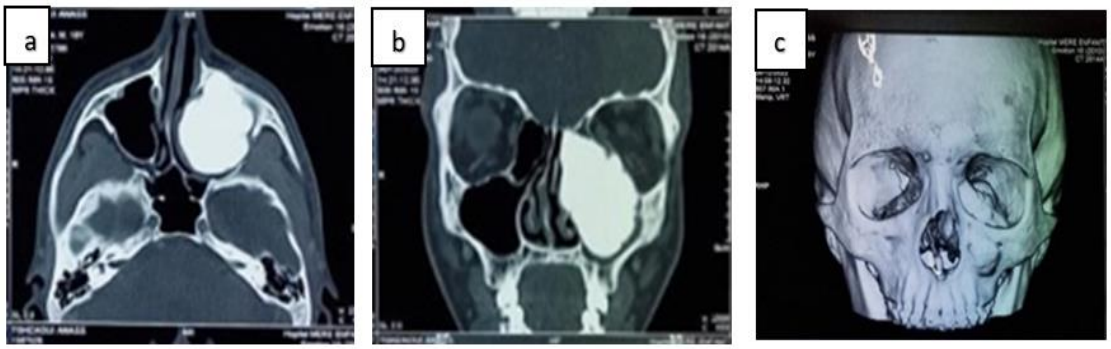

Figure 1: Left ethmoido-maxillary tumor causing narrowing of the left nasal fossa with deviation of the nasal septum (a) and subtotal ethmoidal filling and displacement of the internal wall and floor (b,c) Left orbital tumor causing grade II exophthalmos

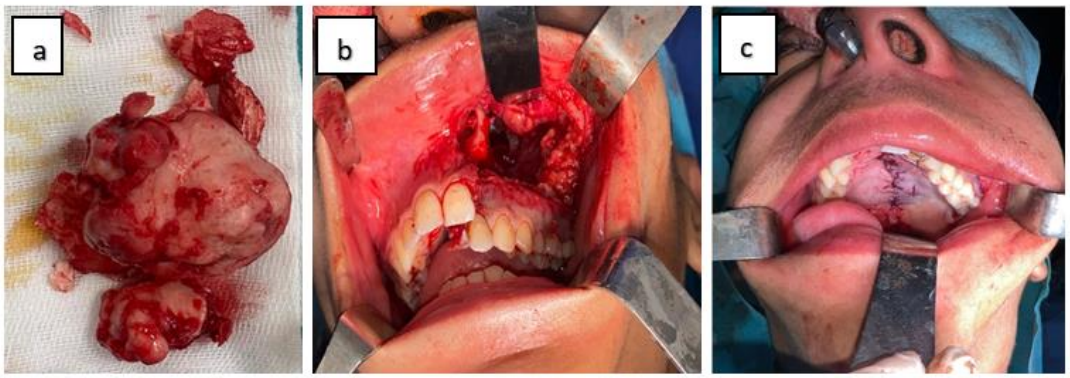

Figure 2: Total resection of the tumour (a) via the vestibular approach, after segmental maxillary osteotomy (b,c)

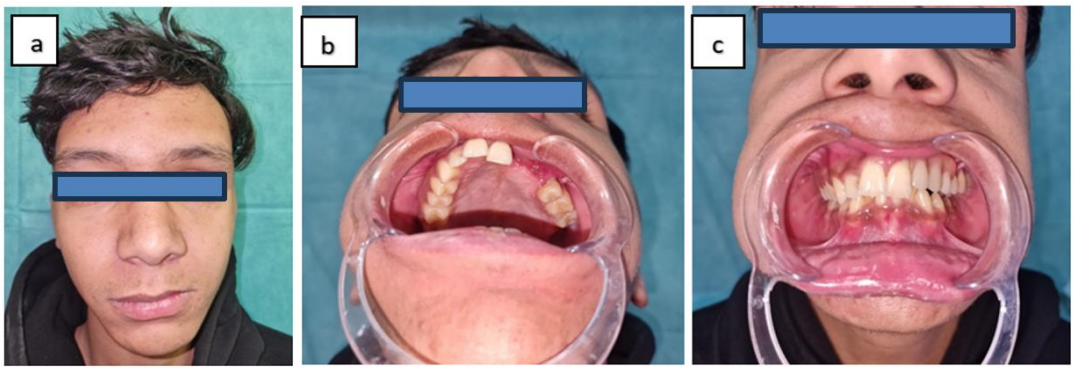

Figure 3: Slight enophthalmos in the left eye (a), good endobuccal healing (b) and rehabilitation with a removable dental prosthesis (c)

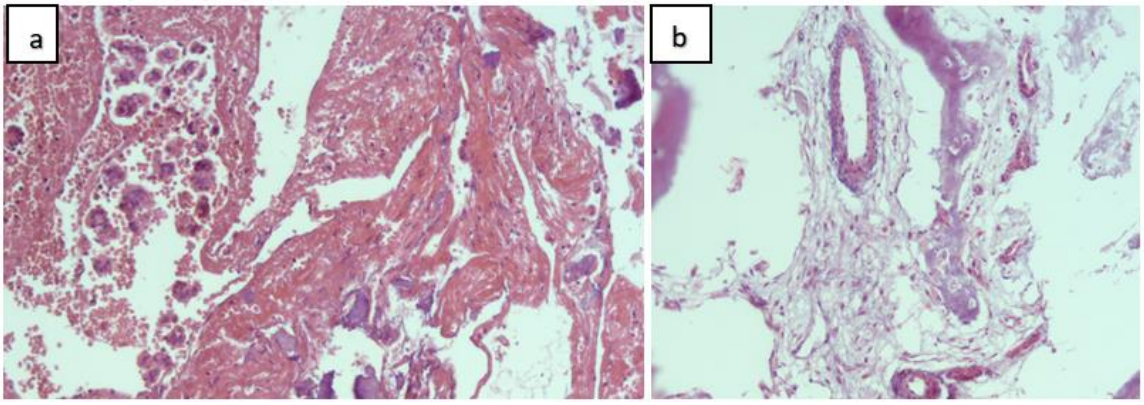

Figure 4: HEx20: (a): Osteoid tissue within vascularized edematous fibrous tissue with a large thick-walled vessel in the center (b): Bone and osteoid tissue within hemorrhagic material with multiloculated cells

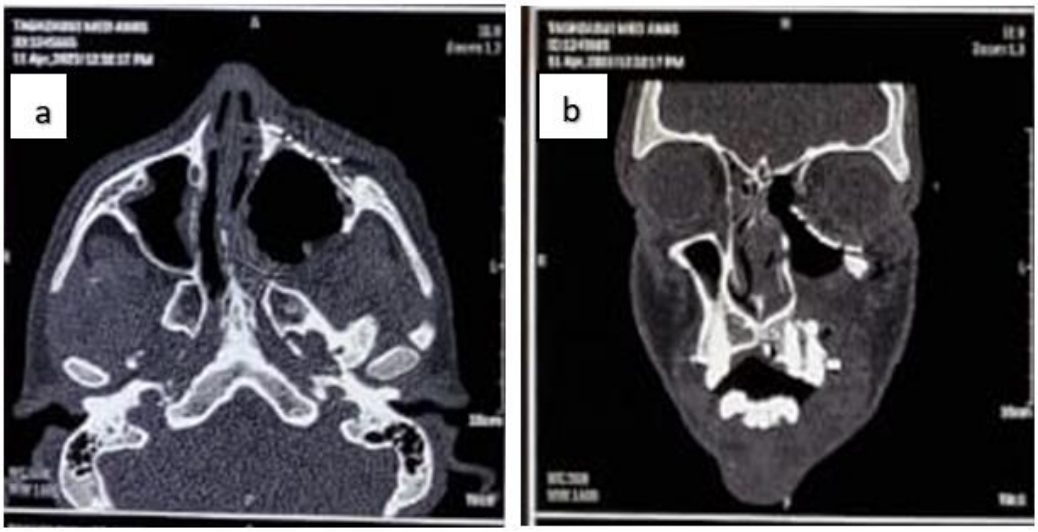

Figure 5: Control CT scan at 10 months post-operatively; (a) Post-operative residual cavity with no sign of local recurrence, (b) titanium grid in place

DISCUSSION

Osteoblastoma (OB) is a rare benign, primitive, non-metastatic osteogenic tumour. Develops mainly in the axial skeleton (spine, sacrum, pelvic bones, etc.), long tubular bones and lower extremities (Wang *et al*., 2018).

Its characterized histopathologically by a proliferation of osteoblasts forming osteoid tissue and primitive bone trabeculae set in a vascularized fibrous connective tissue stroma (Mohammadi *et al.*, 2022), which is benign and arises from non-odontogenic epithelium (Gonuguntla *et al*., 2020).

OB accounts for <1% of all primary bone tumours, and around 18% of craniofacial bone tumours (Sanae *et al*., 2021). mainly affects young adults in the 20 to 30 age bracket with a male: female ratio of 2:1 (Rana *et al*., 2016). The youngest reported case in the literature is a 7 month old female infant with an OSB of temporal bone (Miyazaki *et al*., 1987) (Mohammadi *et al*., 2022).

Its maxillofacial location is rare (10%) (Sahu *et al*., 2019) (Lim *et al*., 2021) mainly involves the mandible bone (71.4%) (Mohammadi *et al*., 2022), and more rarely the ethmoidomaxillary region. (Alvares Capelozza *et al*., 2005) (Cekic *et al*., 2016) It may be responsible for ophthalmological complications.

In the literature, 27 cases of osteoblastoma of the paranasal sinuses have been reported (Evans *et al*., 2020). Eight of these cases involved the ethmoidal sinus, and only four the frontal sinus (Kiyohara *et al*., 2013) (Cekic *et al*., 2016).

First described by Jaffe and Meyer in 1932 as an osteoblastic tumour forming osteoid tissue. Then, Dahlin and Johnson in the year 1954, suggested a name "giant osteoid osteoma," based on this histologic similarity. In 1956, Jaffe and Lichtenstein named this lesion as "Benign Osteoblastoma", which was adopted by the World Health Organization (Jones *et al*., 2006; Neville *et al*., 2023). In the other hand, the first case of mandibular osteoblastoma was reported by Bargello and Sudano in 1967 (Sahu *et al*., 2019). Dorfman and Weiss in 1984 described a subset of OB which was

characterized by locally destructive pattern, recurrence and presence of epithelioid osteoblasts and labeled them as "AO" (Rana *et al*., 2016).

Its pathogenesis is unknown, but the inflammatory nature of the lesion is suggested by the increased secretion of prostaglandins, suggesting a local response rather than a genuine tumour (Cekic *et al*., 2016).

Two types can be distinguished (Zhao *et al*., 2023) the conventional (benign one) and the osteoblastic (aggressive one) where a subtle histopathological difference can be noted between them (Panigrahi *et al*., 2016).

Generally slow-growing, sometimes can be rapid, simulating a malignant or infectious process (Sanae *et al*., 2021).

The similarity of this lesion to other lesions means that correct diagnosis based on clinical, biological, radiological and pathological evidence is essential.

No clinical specificity, symptoms have no localizing value and can be summed up as often painless swelling, headache, exophthalmos or visual disturbances (Wang *et al.*, 2018). In our case, headache and exophthalmos were the main symptoms.

Osteoblastoma has no pathognomonic radiological signs. It generally manifests as a solitary osteolytic lesion, round or oval, 2-12cm in diameter, expansive, well-defined or not, surrounded by a sclerotic rim, with or without cortication, associated with signs of ossification and expansive behavior causing remodeling of the affected bone without destruction. The degree of ossification is closely linked to the patient's age and the pathological stage of the disease. Less lytic behavior and greater calcification are generally observed in mature lesions. A ground-glass appearance may be observed depending on the extent of calcification (Cekic *et al*., 2016).

Additional angiography may be required to determine the vascular supply to the tumour and adjacent soft tissues, thus helping to plan surgery to reduce the risk of bleeding (Wang *et al*., 2018).

On MRI, osteoblastoma has a heterogeneous appearance. The sclerotic component of the lesion shows an empty signal and no contrast enhancement, while the fibrous component shows low to intermediate signal intensity on T1W and T2W images and intense enhancement after intravenous contrast administration (Sanae *et al*., 2021).

The definitive diagnosis is therefore histopathological, made on the surgical specimen. nevertheless, histological heterogeneity can be noted in osteoblastomas. Conventionally, osteoblastoma is characterized by the production of pure osteoid tissue and trabecular bone, not lamellar bone, with a variable number of osteoblasts within a fibrovascular stroma. Large osteoblasts are generally observed in conventional osteoblastomas, which contain a large nucleus with a distinct nucleolus. Because of the variable histological features, the pathologist must be experienced in distinguishing osteoblastoma from osteosarcoma (Görgün *et al*., 2016). Immunohistochemistry can sometimes contribute to an auxiliary diagnosis, relying on five antibodies, including Ki-67, PCNA, CD68, SOX2 and MDM2 in this case (Zhao *et al*., 2023).

Dorfmann and Weiss classified osteoblastomas into four groups in 1984: 1- Low-grade osteosarcomas (Osteoblastoma like osteosarcoma), 2- Pseudomalignant osteoblastomas, 3- Osteoblastomas that have undergone malignant transformation to osteosarcoma confirmed by Grace *et al*., using DNA microdensity in tissue taken four times from a lesion on the right femur of an 18-year-old man. And,4- Aggressive osteoblastoma which is named malignant osteoblastoma by Schajowicz and Lemos (Görgün *et al*., 2016).

Sometimes difficult to differentiate osteoblastoma from osteosarcoma, Lucas *et al*., have pointed out that the most reliable histological factor is the "destructive permeation" characteristic of osteosarcoma (Görgün *et al*., 2016).

Due to its craniofacial rarity, variable clinical presentations and overlapping radiographic and histopathological features with other fibro-osseous lesions, bone tumours and odontogenic tumours, the diagnosis of osteoblastoma can be difficult and requires careful consideration based on clinical, radiological and histopathological features (Lim *et al*., 2021).

Fibrous dysplasia, osteoid osteoma, osteomas, cemento-ossifying fibroma, and low-grade osteosarcoma remain the main differential diagnoses of paranasal sinus osteoblastoma (Zhao *et al*., 2023). The incidence of fibrous dysplasia is higher than that of osteoblastoma. A common feature is the ground-glass appearance on CT, which is homogeneous and diffuse in fibrous dysplasia, whereas osteoblastoma appears radiologically more nodular and coarser than fibrous dysplasia due to its mixed content of bone and fibrous tissue (Cekic *et al*., 2016).

Osteoid osteoma remains the main differential diagnosis of osteoblastoma. Their radiographic and histological features are similar, if not identical, the only difference being size. Osteoid osteoma is typically <1 cm and lesions >2 cm will be considered osteoblastoma (Söylemez *et al*., 2016; Woźniak *et al*., 2010). In addition, patients with pain that does not worsen at night and who do not respond to NSAIDs - can differentiate

osteoblastoma from osteoid osteoma, apart from the difference in size (Lim *et al*., 2021).

Osteoma, a slow-growing benign bone tumour, is the most common tumour of the paranasal sinuses. In a study by McHugh *et al*., of 45 surgically excised paranasal osteomas, 38% showed interanastomosing trabeculae of woven bone lined with hypertrophied osteoblasts and osteoclasts, are histologically indistinguishable from osteoblastomas if taken out of context, and osteomas with these features have been designated as osteomas with osteoblastoma-like features by Dorfman and Czer- niak (Evans *et al.*, 2020).

In fact, in the present case, the possibility of an ethmoidomaxillary osteoma was considered due to the radiological appearance of the tumor. It is therefore difficult, if not impossible, to differentiate osteoblastoma from osteoma on the basis of radiographic features alone.

Cemento-ossifying fibroma, a benign bone tumour of the maxilla, can be confused with osteoblastoma for several reasons: its internal appearance varies from completely radiolucent to completely radiopaque, with a tendency to have well-limited margins. Nevertheless, the absence of spindle cells in the stroma and the presence of numerous osteoblasts on histology distinguish osteblastoma from cemento-ossifying fibroma. it should also be noted that the 2 lesions may have an aggressive variant, juvenile cemento-ossifying fibroma and aggressive osteoblastoma (Lim *et al*., 2021).

It is sometimes difficult to distinguish a well-differentiated osteosarcoma from an osteoblastoma (Görgün *et al*., 2016) (Lim *et al*., 2021). But the presence of tumour progenitor cells, cytonuclear atypia and a high mitotic index point to the malignant origin of the lesion. the risk of malignant degeneration of an osteoblastoma should also be noted, and it should be considered as a distinct clinicopathological tumour entity from genuine osteosarcomas (Görgün *et al*., 2016).

The surgical approach depends on tumour location, size, extent and contiguity with noble organs. Several therapeutic modalities are possible: conservative surgical excision, excision with curettage or en-bloc resection with safety margins (Sahu *et al.*, 2019). Indeed, total resection of the tumour with safety margins is the standard treatment for osteoblastoma. However, if the lesion is adherent to important structures, curettage would be the only possible alternative to resection (Zhao *et al*., 2023).

Radiotherapy can also be a therapeutic option, and may be considered in cases of aggressive tumour behavior, incomplete resection, recurrence or non-resectable tumours. However, as radiotherapy can lead to malignant transformation of tumours, it should be used with caution (Krishnan *et al*., 2022) (Zhao *et al*., 2023)

Benign osteoblastoma has a good prognosis after surgery. Long-term follow-up is recommended because of the risk of recurrence and to assess the local post-operative condition.

The recurrence rate varies from 13.6% for benign osteoblastoma to 50% for the aggressive form (Mohammadi *et al*., 2022) (Lim *et al*., 2021).

In our review of the literature, osteoblastomas can also undergo malignant transformation, essentially in recurrent tumours (Görgün *et al*., 2016).

CONCLUSION

In summary, ethmoidomaxillary osteoblastoma is a very rare benign bone tumor requiring precise diagnosis. Surgery is treatment of choice. Despite a generally good prognosis, recurrence and malignant transformation risks necessitate long-term follow up.

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