

Aggressive Osteoblastoma of Temporal Bone Causing Facial Palsy in a 9-year-old Child: A Case Report Based on 2020 WHO Classification of Bone Tumors

Osteoblastoma agressivo do osso temporal causando paralisia facial em uma criança de 9 anos: Um relato de caso baseado na classificação da OMS de 2020 de tumores ósseos

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Arq Bras Neurocir 2024;43(1):e57–e61.

Abstract

Aggressive osteoblastoma (AO) is an uncommon bone tumor that represents a borderline lesion between osteoblastoma and osteosarcoma. The vertebral column, the sacrum, the pelvis, and jaw/craniofacial bones are primarily affected. Aggressive osteoblastoma does not metastasize and is treated by surgical resection. The authors report a case of AO in a 9-year-old female patient presenting with 5th and 7th cranial nerve palsy. Prior pathological history included resection of an expansile nodule in the left temporal bone. Conventional radiological examination and computed tomography (CT) of the skull revealed an osteoblastic lesion arising in the petrous portion of the left temporal bone, measuring 5.2 cm in the largest dimension. The patient was subjected to partial surgical resection of the process. Microscopy revealed a primary neoplastic bone composed of numerous epithelioid round osteoblasts disposed in solid sheets and with mild atypia, large eosinophilic cytoplasm, and an eccentric, ovoid nucleus. The process exhibited loose stroma, low mitotic index, osteoid formation, and a few osteoclast-like multinucleated giant cells. The diagnosis of AO was thus established. After 5 months of clinical follow-up, the patient is asymptomatic, without evidence of tumoral growth on CT scans.

Keywords

- aggressive osteoblastoma
- osteoblastic tumor
- bone tumor
- pathology
- prognosis

received
March 16, 2021

DOI [https://doi.org/
10.1055/s-0042-1746195.](https://doi.org/10.1055/s-0042-1746195)
ISSN 0103-5355.

accepted
June 16, 2021
article published online
October 10, 2023

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Thieme Revinter Publicações Ltda., Rua do Matoso 170, Rio de Janeiro, RJ, CEP 20270-135, Brazil

Resumo

O osteoblastoma agressivo (AO) é um tumor ósseo incomum que representa uma lesão limítrofe entre osteoblastoma e osteossarcoma. A coluna vertebral, o sacro, a pelve e os ossos maxilares/craniofaciais são afetados principalmente. O osteoblastoma agressivo não metastatiza sendo tratado por ressecção cirúrgica. Os autores relatam um caso de OA em paciente do sexo feminino, de 9 anos, com paralisia de V e VII pares cranianos. A história patológica prévia incluiu ressecção de nódulo expansivo no osso temporal esquerdo. O exame radiológico convencional e a tomografia computadorizada (TC) de crânio revelaram lesão osteoblástica surgindo na porção petrosa do osso temporal esquerdo, medindo 5,2 cm em sua maior dimensão. O paciente foi submetido à ressecção cirúrgica parcial do processo. A microscopia revelou osso neoplásico primário composto por numerosos osteoblastos epitelioides redondos dispostos em lâminas sólidas e com leve atipia, grande citoplasma eosinofílico e núcleo ovoide excêntrico. O processo exibiu estroma frouxo, baixo índice mitótico, formação de osteóide e algumas células gigantes multinucleadas semelhantes a osteoclastos. O diagnóstico de OA foi assim estabelecido. Após 5 meses de acompanhamento clínico, o paciente encontra-se assintomático, sem evidência de crescimento tumoral na tomografia computadorizada.

Palavras-chave

- osteoblastoma agressivo
- tumor osteoblástico
- tumor ósseo
- patologia
- prognóstico

Introduction

Aggressive osteoblastomas (AOs) are very rare tumors classified as borderline lesions between osteoblastoma and osteosarcoma. Its peak age incidence is in the 2nd and 3rd decades of life.^{1–3} Overall distribution patterns are similar to those of conventional osteoblastoma, with a predilection for the axial skeleton. The vertebral column, the sacrum, proximal parts of the appendicular skeleton such as the pelvis and femur, and jaw/craniofacial bones are primarily affected.^{2–5} Aggressive osteoblastomas do not metastasize, are likely to recur (in between 20 and 30% of cases) and are characterized by the presence of epithelioid osteoblasts. The lesion is not considered a precursor to osteosarcoma.^{1,2,5–7} The present study reports a case of AO compromising the left temporal bone and causing 5th and 7th cranial nerve compression in a pediatric patient and discusses clinical and pathological findings of this rare bone tumor.

Case Report

A female patient, 9 years old, was referred to the neurosurgery service with left 5th and 7th cranial nerve palsy. On physical examination, the patient exhibited good general condition and adequate weight and height development (48.7 kg/1.45 m), without evidence of other focal neurological deficits, optic nerve edema, or alterations in other systems. Her prior pathological history included resection of an expansile nodule in the left temporal bone 2 years earlier at another institution, where the diagnosis of osteofibrous dysplasia was established. Current laboratory tests were within normal values. Conventional radiological examination and computed tomography (CT) of the skull revealed an osteoblastic lesion arising in the petrous portion of the left temporal bone, which measured ~ 5.2 cm in the largest dimension and caused compression of the 5th and 7th cranial nerves (►Fig. 1). The patient was subjected to partial surgical

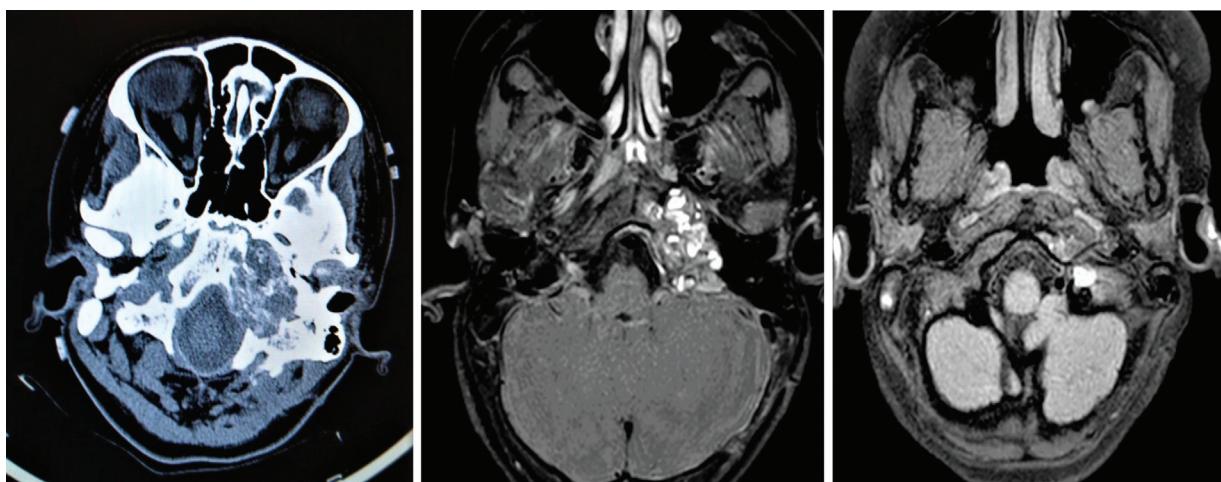


Fig. 1 Aggressive osteoblastoma: Computed tomography of the skull revealing an osteoblastic lesion (5.2 cm in the largest dimension) arising in the petrous portion of the left temporal bone.

resection of the process (~ 80% of the tumoral volume). The patient was placed in right lateral decubitus and underwent a left frontobasal craniotomy. During the surgical procedure, an expansive lesion affecting the temporal bone was identified. The process determined compression of the brainstem and the foramina at the base of the skull. The tumor was resected through the Kawase trigone in its lowest portion and lateral to the Meckel cavum. The VII cranial pair was dissected and preserved during the procedure. On gross examination, the surgical specimen was composed of several irregular, pale gray fragments of bone tissue, the largest of which measured $1.8 \times 1.2 \times 1.0$ cm. On microscopy, a primary neoplastic bone neoplasm was identified. The lesion was characterized by numerous epithelioid round osteoblasts disposed in solid sheets around irregular bone trabeculae and exhibiting mild atypia, large eosinophilic cytoplasm, and an eccentric, ovoid nucleus. The process had loose stroma, numerous small vascular channels, low mitotic index, osteoid formation, and a few osteoclast-like multinucleated giant cells. No chondroid areas were identified (**►Fig. 2**). These findings culminated in a diagnosis of AO. After 8 months of clinical follow-up, the patient is asymptomatic, without cranial nerve palsy or evidence of tumoral growth on CT scans. Previous histological slides were reviewed, and the diagnosis of AO was confirmed.

Discussion

The true incidence and age distribution of AO remain largely unknown because of the rarity of the disease.^{1,2,4,5,7} The first case series found in the literature, published in 1984 and 1996, described 15 and 36 cases, respectively.^{1,4} Clinical complaints are directly associated with compromised bone, and pain is a common symptom. Radiological findings usually comprise a circumscribed lytic defect sometimes surrounded by a rim of sclerosis.^{1,4,6,8,9} The main difference from conventional osteoblastoma is that AO is larger, usually exceeding 4 cm.^{2,5,6,9,10} The bone

contour may be expanded and have a rim of reactive bone. Eventually, the tumor crosses the joint space, thereby compromising the adjacent bone, a reflection of its aggressive biological behavior. Soft tissues may be involved if the tumor arises in small bones.^{1,4,5,8,11,12}

On gross examination, the process is an oval to round, reddish, bright, soft to hard lesion with well-defined margins.^{2,4,10,13–15} The bone contour may be markedly expanded and exhibit a thinned, disrupted cortex.^{2,4,10,13–16} The tumor stroma is characteristically rich in blood vessels. On microscopy, AO shows many similarities to conventional osteoblastoma. Aggressive osteoblastomas are composed of an irregular network of bone trabeculae distributed in a loose stroma with prominent vasculature.^{2,3,7,11,14,16,17} The most important histological finding is the presence of epithelioid osteoblasts that form solid sheets in intertrabecular spaces or rim osteoid trabeculae. Epithelioid osteoblasts are round cells with abundant eosinophilic cytoplasm, an eccentric, oval nucleus with prominent nucleoli, and some degree of atypia.^{2,3,7,11,14,16,17} Epithelioid osteoblasts are at least twice the size of normal osteoblasts and, frequently, show a large, clear cytoplasmic area with enlarged Golgi apparatus, which displaces the nucleus.^{2,3,7,11,14,16–18} Osteoid can be found around individual tumor cells or in broad zones surrounding epithelioid osteoblasts. The presence of benign osteoclast-like multinucleated giant cells and secondary aneurysmal bone cysts are common features.^{2,4,7,10,13,15,17,18} Aggressive osteoblastomas show 1 to 4 typical mitotic figures per 20 high-power fields. Necrosis is uncommon, and chondroid/cartilaginous differentiation has not been described.^{7,10,13,15,17–19}

The differential diagnosis includes osteoid osteoma, conventional osteoblastoma, and osteosarcoma.^{1,7,10,14,20,21} Osteoid osteoma and osteoblastoma measure < 4 cm in diameter and do not exhibit epithelioid osteoblasts. The main diagnostic problem regarding the entity classified as AO centers around its distinction from osteosarcoma. Classical histological findings of conventional osteosarcoma include moderate to severe cellular atypia, high mitotic index, atypical mitotic figures, prominent osteoid deposition, infiltrative/permeating growth pattern, and presence of neoplastic cartilage.^{4,9,10,17,20,22–24} Aggressive osteoblastoma also exhibits a peripheral shell of reactive bone over the soft tissue extension, which is not characteristic of osteosarcoma. Genetic studies are not useful for distinguishing between AO and osteosarcoma or for determining prognosis. There is yet no evidence that AO undergoes spontaneous transformation to osteosarcoma.^{1,4,14,21–24} Complete surgical resection, curettage, and/or partial resection is the mainstay of treatment for AO. Skull AO should be treated by wide local excision when technically feasible. Long-term follow-up is necessary to monitor recurrence.^{1,4,7,21,23,24} The **►Table 1** shows a summary of literature reports of aggressive osteoblastoma. Partial resection of skull AO have been accepted when the location of the tumor, such as the temporal bone or the base of the skull, denotes a high risk of vascular or cranial nerve damage or technical limitations.^{1,4,7,21,23,24}

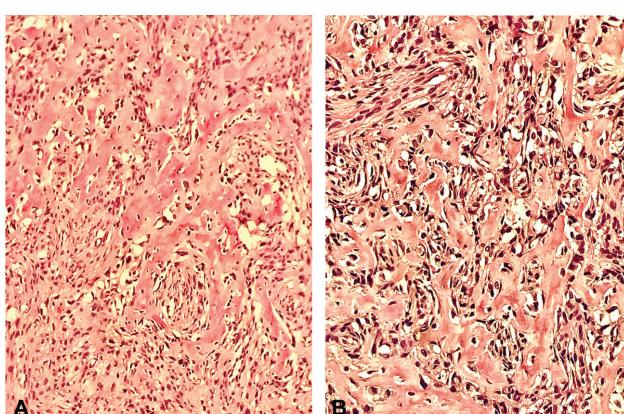


Fig. 2 Aggressive osteoblastoma of the temporal bone: (A) Epithelioid osteoblasts occupying intertrabecular spaces (hematoxylin-eosin, 200X); (B) Large epithelioid osteoblasts disposed in solid sheets and showing mild atypia. Note osteoid deposition and vascular channels in the tumor stroma (hematoxylin-eosin).

Table 1 Summary of literature reports of aggressive osteoblastoma

Reference	Gender, age (years old)	Clinical complaint	Topography	Radiologic findings	Tumor size	Clinical management	Outcome
Morris et al. ⁵	Female, 20	Pain	Left scapula	Lytic lesion	8.9 cm	Surgical resection	Disease-free at 3 years after surgery
Lu et al. ⁶	Male, 18	Local tenderness	Temporal bone	Lytic lesion	3.3 cm	Surgical resection	No signs of recurrence at 1 year after surgery
Sharma et al. ⁷	Male, 18	Progressive swelling	Right parietal bone	Lytic lesion	9.0 cm	Surgical resection	Persistent lesion at 21 months of follow-up
Salmen et al. ⁸	Male, 7	Pain	Maxilla	Lytic lesion	2.1 cm	Surgical resection	No signs of recurrence at 1 year after surgery
Al-Ibraheem et al. ⁹	Male, 25	Hemimandibular swelling	Mandible	Sclerotic lesion	2.2 cm	Surgical resection	No signs of recurrence at 1 year after surgery
Sharma et al. ¹⁰	Male, 17	Pain in left hip	Acetabulum	Lytic lesion	6.4 cm	Extended curettage	No signs of recurrence at 1 year after surgery
Sonnyal et al. ¹¹	Male, 21	Painful mass	Left femur	Sclerotic lesion	20.0 cm	Radical resection of the left femur and cryosurgery	No signs of recurrence at 32 months after surgery
Harrington et al. ¹²	Male, 25	Enlarging palatal mass	Maxilla	Sclerotic lesion	4.0 cm	Surgical resection	No signs of recurrence at 8 months after surgery
Miyayama et al. ¹³	Female, 29	Pain	Left calcaneus	Lytic lesion	3.0 cm	Surgical resection	No signs of recurrence at 10 years after surgery
Ando et al. ¹⁴	Male, 25	Neck pain	6 th and 7 th cervical vertebrae	Lytic lesion	3.5 cm	Surgical resection	No signs of recurrence at 2 years after surgery
Dixit et al. ¹⁵	Male, 20	Hearing loss and tinnitus in left ear	Left temporal bone	Lytic lesion	6.0 cm	Partial resection	Unknown
Kukwa et al. ¹⁶	Female, 12	Persistent exophthalmia	Sphenoid	Lytic lesion	5.5 cm	Surgical resection	Recurrence at 4 months after surgery
Pontual et al. ¹⁷	Male, 13	Swelling on the left side of the face	Mandible	Lytic lesion	5.7 cm	Surgical resection	No signs of recurrence at 4 years after surgery
Kashikar et al. ¹⁸	Male, 18	Swelling of the oral cavity	Mandible	Lytic lesion	1.2 cm	Extended curettage	No signs of recurrence at 6 months after surgery
Cikojević et al. ¹⁹	Female, 14	Right-sided nasal obstruction and severe headache	Right middle turbinate	Sclerotic lesion	2.2 cm	Surgical resection	No signs of recurrence at 1 year after surgery
Mohanty et al. ²⁰	Male, 23	Painful swelling	Temporal bone	Sclerotic lesion	3.0 cm	Surgical resection	No signs of recurrence at 8 months after surgery
Baker et al. ²¹	Female, 12	Right thigh pain	Right femur	Lytic lesion	5.7 cm	Surgical resection	No signs of recurrence at 9 months after surgery
Chatterjee et al. ²²	Male, 2	Swelling over dorsum of right hand	Third metacarpal shaft	Lytic lesion	3.0 cm	Surgical resection	No signs of recurrence at 2 years after surgery
Srivastava et al. ²³	Male, 24	Painful swelling	Mandible	Lytic lesion	5.3 cm	Surgical resection	No signs of recurrence at 5 months after surgery
Castro et al. ²⁴	Female, 7	Swelling	Mandible	Sclerotic and lytic lesion	5.6 cm	Extended curettage	No signs of recurrence at 6 years after surgery
Present case	Female, 9	Left 5 th and 7 th cranial nerve palsy	Left temporal bone	Osteoblastic	5.2 cm	Partial surgical resection	No signs of tumoral growth at 5 months of follow-up

Conflict of Interests

The authors have no conflict of interests to declare.

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