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Desmoplastic ameloblastoma of the jaw: An infrequent paediatric case report

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Abstract

Ameloblastoma is a neoplastic process of the jaw that originates from the odontogenic epithelium. Its early diagnosis is of particular interest because of the need for resection with a wide margin of safety, which reduces the likelihood of local recurrence. The neoplasia is difficult to differentiate from other common lesions in pediatric patients, such as osteomyelitis, dental cysts, and fibrous lesions. We report a 14-year-old female patient of Asian descent presented with a palpable painful mass in the left jaw region. Doppler ultrasonography revealed a cystic image with poorly defined boundaries. Symptomatic treatment is indicated in cases of suspected osteomyelitis. The persistence of the symptoms led to performed biopsy, diagnosing ameloblastoma. Subsequently, the injury was completely resected, and the margins were preserved. Finally, mandibular reconstruction was performed. Demoplastic ameloblastoma is uncommon in paediatric age and if not suspected, the diagnosis can be delayed with the risk of insufficient margins.

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Keywords: Ameloblastoma; Desmoplastic subtype; Jaw; Type IV collagen; Osteoclast-like cells; Bone persistence.

Introduction

Ameloblastoma is a benign neoplasm of the jaw that originates in the odontogenic epithelium at any stage of development of the enamel organ, its remnants, or in the epithelium of the dental cyst [1]. It is the second most common tumor of this origin, after odontoma [2]. Ameloblastoma is locally aggressive, with an initial overlapping growth followed by rapid expansion, and early diagnosis is of great interest due to the need for a wide resection margin, reducing the patient's relapses and morbidity. The peak incidence is in the fourth and fifth decades of life, in an age range of 8-92 years, with equal distribution by sex. The demoplastic histological pattern of the neoplasm simulate an "osteofibrous" lesion by X-ray imaging. Different histological patterns can coexist in the same tumor, called hybrid lesions [3]. In this respect, biopsy provides a precise diagnosis of the neoplasm, because an important overlap can be observed between the different morphological variants of ameloblastoma and osteomyelitis, dental cyst, and fibrosis pathology. We read the Declaration of Helsinki and followed the guidelines for this report.

Clinic case

A 14-year-old female patient of Asian descent, with no personal background of interest, presented at our institution with a palpable painful mass in the left jaw region of 30 days of evolution. When evaluated by the Dental Service, facial asymmetry and odontogenic focus were found, and Doppler ultrasound revealed a voluminous cystic image with poorly defined boundaries and fine internal septations. The dimensions were 60×36×31

mm (Figure 1A). Symptomatic and antibiotic treatments were indicated by suspected osteomyelitis.

The increased pain prompted further consultation and hospitalization. A panoramic radiograph was performed, which showed a multilocular radiolucent image generating a separation between the roots of the second premolar and the first and second molars. Computed axial tomography of the facial mass showed an expansive osteolytic lesion compromising the horizontal and ascending left branches of the lower jaw (Figure 1B).

Biopsy of the cystic wall and jawbone was performed, histological findings revealed large sections of trabecular bone tissue that exhibited marked osteoblastic activity and medullary spaces with fibrosis. A few areas showed epithelial cell strands in different layers, one of basaloid appearance, another with cubic to cylindrical cells and hyperchromatic nucleus, above, cells with stared cytoplasm and loose stroma that were arranged in a reticular form (Figure 2A,B) and a third layer of squamous differentiated cells. Immunohistochemically, the neoplasm showed a 10% nuclear expression for Ki67, diffuse cytoplasmic immunomarquage for cytokeratin (AE1-AE3) in the epithelial component, focal positivity for S100 and desmin in fusiform interstitial cells, positivity for type IV collagen associated with interstitial osteoid material deposits, and negativity for caspase-3 and p53. With a diagnosis of ameloblastoma, complete resection of the injury and the widest possible conservative margins were performed. A partial left mandibulectomy was performed from the canine to the mandibular branch with a size of 7×4x2,3 cm. Macroscopic examination of the outer surface extending to the bone body showed a solid tumor formation of brown color, focally friable and rounded. Inside it corresponded to a brown-shaped area, measuring 3.3x1.3 cm (Figure 3A,3B). The margins were 0.5 cm from the mucous resection edge, 2.6 cm from the bone edge corresponding to the mandibular branch, and 1 cm from the bone resection border corresponding to the right jaw body. Microscopic examination of the specimen revealed another histological pattern: follicular, acantomatous, reticular, and a predominance of large sectors with cords and cell nests trapped and compressed by a dense fibrosclerotic stroma, identifying the neoplasm as a demoplastic ameloblastoma (Figure 3C). Mandibular reconstruction was performed with right rib self-adhesion (6th and 7th ribs) and placement of a titanium plate. The patient is currently under follow-up and has shown favourable development.

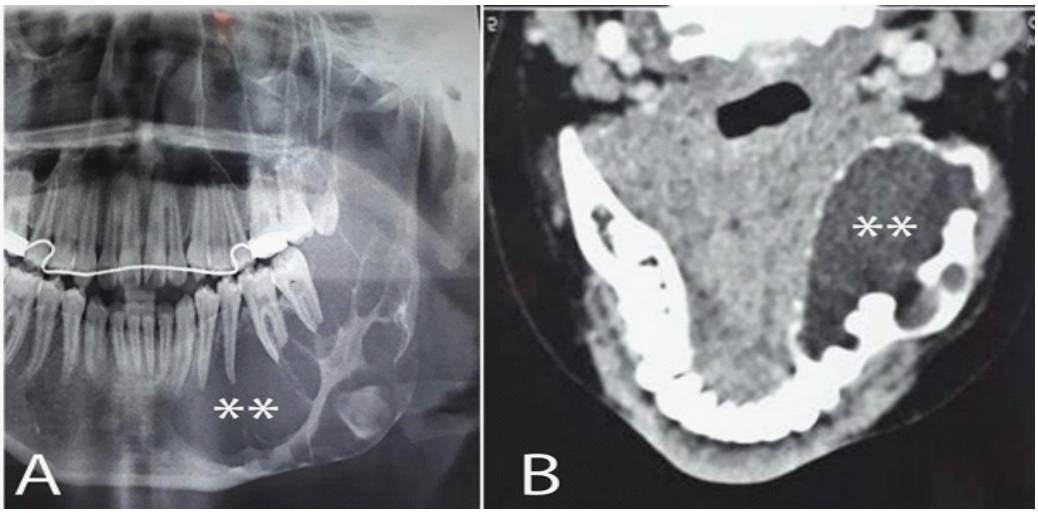

Figure 1: **(A)** The Doppler ultrasound revealed a voluminous cystic image with poorly defined limits, fine internal septa, and particulate content adjacent to the bone surface, that do not demonstrate emission of Doppler signal. **(B)** The computed axial tomography of the facial massif showed an expansive osteolytic lesion compromising the left horizontal and ascending rami of the lower maxilla.

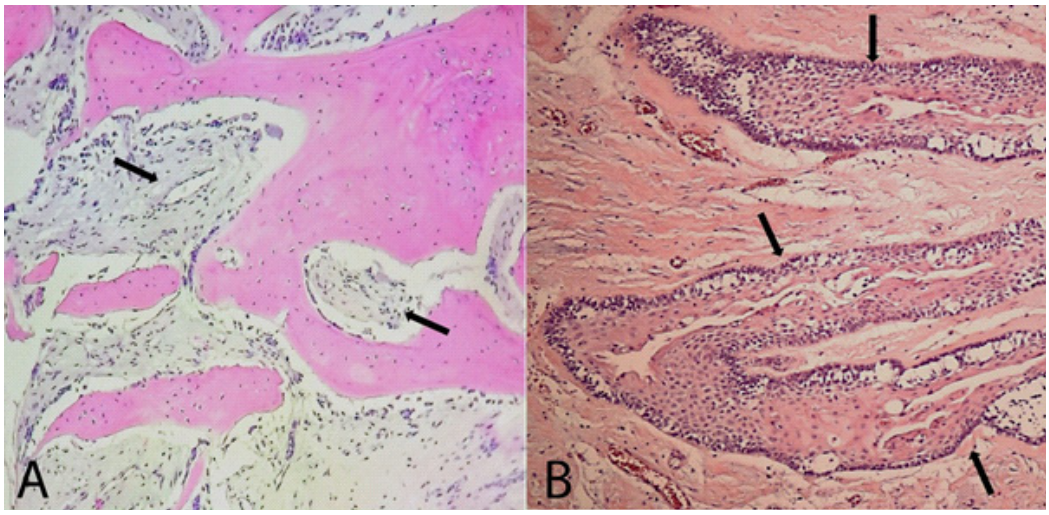

Figure 2: **(A)** The primary biopsy that gave the diagnosis of ameloblastoma showed large sections of trabecular bone tissue that exhibited marked osteoblastic activity and medullary spaces with fibrosis (Hematoxilin & Eosin, 200X). **(B)** In a few areas, epithelium with squamous differentiation were observed (Hematoxilin & Eosin, 200X).

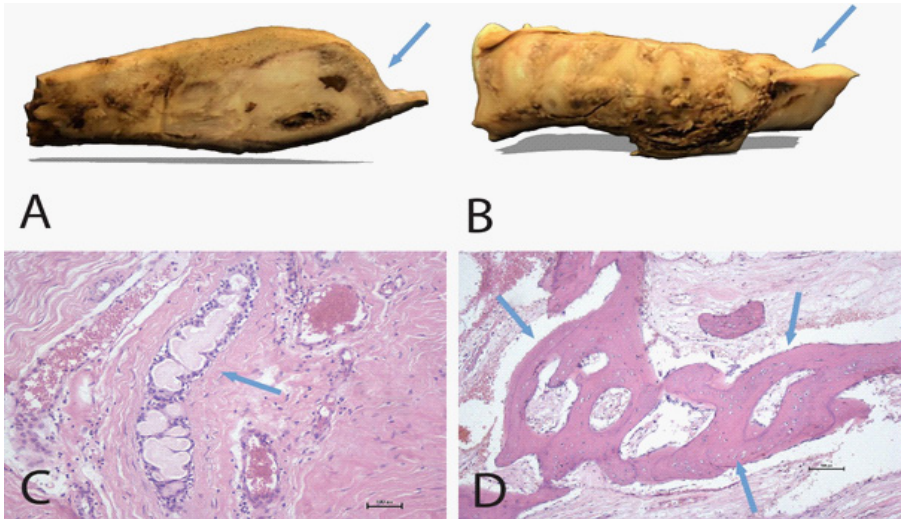

Figure 3: **(A,B)** A partial left mandibulectomy was performed from the canine to the mandibular ramus with a size of 7x4x2.3 cm that included 5 teeth, gingiva, and associated mucosa. In the macroscopic examination, a solid brownish tumour formation, focally friable, rounded with clear limits that measured 2x1.5 cm, was identified on the external surface extending to the bone body. **(C)** The microscopic study of the specimen identified another histological pattern such as follicular, acanthomatous, reticular and the predominance of wide sectors with cords and cellular nests trapped and compressed by a dense fibrosclerous stroma, identifying the neoplasm as desmoplastic Ameloblastoma (Hematoxilin & Eosin, 400X). **(D)** The increased synthesis of type IV collagen favours the formation of metaplastic bone trabeculae, grooved by active and hyperplastic osteoblasts (Hematoxilin & Eosin, 200X).

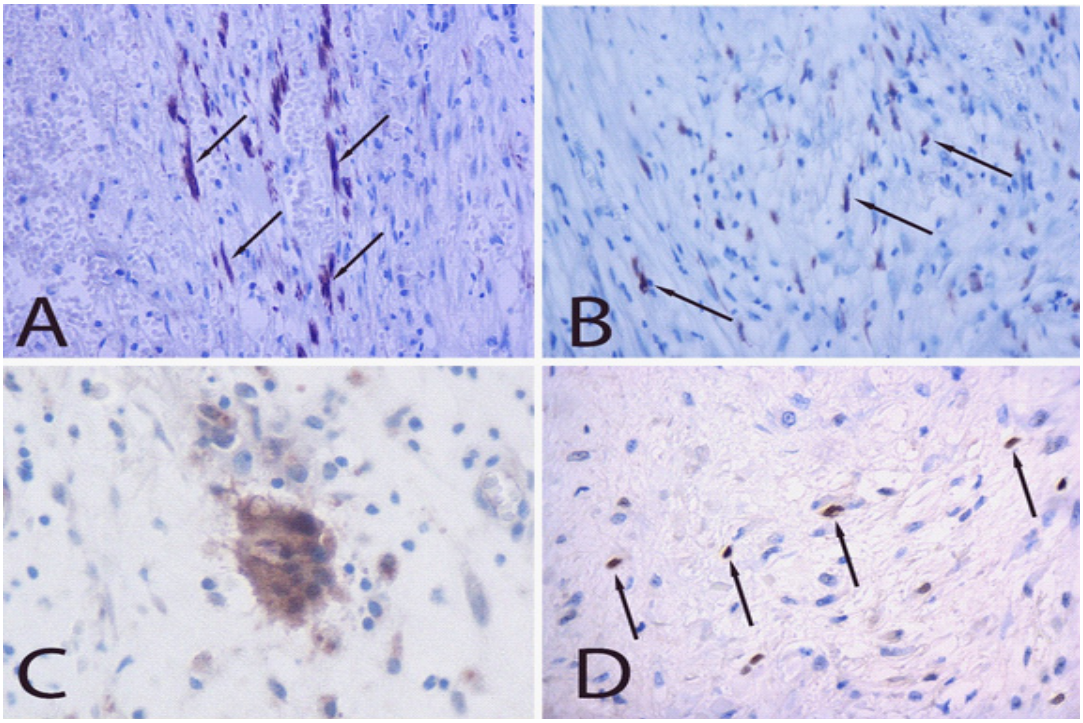

Figure 4: **(A,B)** Focal immunolabeling for desmin and S100 protein was observed in fusiform interstitial cells (immunohistochemistry, 200X). **(C)** Osteoclast-like cells expressing caspase-3 in the cytoplasm (immunohistochemistry, 400X). (D) Low population of neoplastic cells expressing nuclear Ki67 (immunohistochemistry, 200X).

Discussion

The incidence of Demoplastic Ameloblastoma (DA) varies between 0.9% and 14% [2,4,5]. The first detailed report was published by Eversole in 1984 [6], and previously Takigawa et al. in 1981 and Uji et al. in 1983 [2]. In 2017, the World Health Organization (WHO) reclassified ameloblastoma into three forms: unicystic, solid or multicystic, and peripheral or extracystic. DA is a variant of ameloblastoma with specific clinical, radiological, and histological characteristics [7], with an average presentation of 43 years, without gender predilection [8]. The tumor size is variable, between 1.0 and 8.5 cm in diameter, and dental displacement is a finding observed in 92% of cases [2]. The pediatric cases are infrequent, difficulty the diagnosis, assuming it to be as osteomyelitis. Histologically, it is characterized by an extensive, densely collagenized and demoplastic stroma with variable odontogenic epithelium organized in islands and cords, alternating with richly vascularized areas and low cellularity (Figure 3C). It has been suggested that the new synthesis of extracellular matrix proteins involved in the support, adhesion, proliferation, migration, and differentiation of tumor cells. Immunohistochemical studies have shown an interstitial increase in type IV collagen expression by the active synthesis of proteins favouring the formation of metaplastic bone trabeculae (Figure 3D), sucked by hyperplastic osteoblasts [9]. In our case

we observe focal immunomarquage for desmin and S100 protein (Figure 4A,4B) in cells phenotypically compatible with myofibroblasts, in proximity to osteoid deposits expressing type IV collagen. The presence of osteoclast-like cells expressing cytoplasmic caspase-3 suggest a phenomenon of bone persistence. (Figure 4C). This possibility was previously described by another group [10]. The phenomenon of transdifferentiation was proposed by our group [11], contributing to desmoplastic changes. The low population of neoplastic cells expressing nuclear Ki67 suggest a low duplication rate of this neoplasm (Figure 4D). On the other hand, a "hybrid lesion" of DA, showing microscopic features of desmoplastic and classical ameloblastoma was described [3,12]. Differential diagnosis includes fibrous dysplasia, chronic osteomyelitis and ossificant fibroids [6]. The treatment is complete resection with an adequate margin of unaffected tissue, with long-term follow-up due to the recurrence possibility [13].

Declarations

Consent to participate, patients' consent form, ethical approval and/or institutional review board (irb) approval: It is not applied in our Institution for this type of work.

Data sharing is not applicable to this article as no data sets were generated or analysed during the current study.

References

1. Bologna-Molina R, Mosqueda-Taylor A, de Almeida-Oslei P, Toral-Rizo V, Martínez-Mata G. Peripheral desmoplastic ameloblastoma: Histopathological and immunohistochemical profile of a case. Med Oral Patol Oral Cir Bucal. 2010;15(6): e846-9. doi: 10.4317/medoral.15. e846.

2. Sharma A, Ingole S, Deshpande M, Meshram D. Retrospective analysis of Desmoplastic Ameloblastoma: Clinical review. Med Oral Patol Oral Cir Bucal. 2021; 26(2): e246-e255. doi: 10.4317/medoral.24152.

3. Philipsen H, Reichart PY, Takata T. (2001). Desmoplastic ameloblastoma (including "hybrid" lesion of ameloblastoma). Biological profile based on 100 cases from the literature and own files. Oral Oncology. 2001; 37 (5): 455-460. doi: 10.1016 / s1368-8375 (00) 00111-1.

4. Majumdar S, Uppala D, Kotina S, Veera SK, Boddepalli R. Desmoplastic ameloblastoma. Int J Appl Basic Med Res. 2014; 4(Suppl 1): S53-5. doi: 10.4103/2229-516X.140743.

5. Rajendra Santosh AB, Ogle OE. Odontogenic Tumors. Dent Clin North Am. 2020; 64(1): 121-138. doi: 10.1016/j.cden.2019.08.008.

6. Eversole LR, Leider A, Hansen L. Ameloblastoma with pronounced desmoplasia. J Oral Maxillofac Surg. 1984; 42: 735-40. doi: 10.1016/j.joms.2021.12.007.

7. Alves PM, Pereira KMA, Vasconcelos MG, Souza LB, Queiroz LMG, et al. Desmoplasic ameloblastoma in maxilla- report of case and review of the literature. Int J Morphol. 2008: 26(2): 263-268.

8. Atalay B, Soluk M, Brkić A, Emes Y, Cetin O, et al. Desmoplastic ameloblastoma. J Craniofac Surg. 2009; 20(6): 2256-9. doi: 10.1097/SCS.0b013e3181bfc537.

9. Savithri V, Janardhanan M, Suresh R, Kumar RV. Desmoplastic ameloblastoma with osteoplasia: Review of literature with a case report. J Oral Maxillofac Pathol. 2013; 17(2): 298-301. doi: 10.4103/0973-029X.119784.

10. Yan L, Lu L, Hu F, Shetti D, Wei K. Piceatannol attenuates RANKL-induced osteoclast differentiation and bone resorption by suppressing MAPK, NF-κB and AKT signalling pathways and promotes Caspase3-mediated apoptosis of mature osteoclasts. R Soc Open Sci. 2019; 6(6): 190360. doi: 10.1098/rsos.190360.

11. Cao G, Ottaviano G, Fusaro A, Mendez J, Navacchia D. Desmoplastic fibroma of bone: A morphological and immunohistochemical characterization. J Clin Images Med Case Rep. 2021; 2(6): 1399. doi: 10.52768/2766-7820/1399.

12. Effiom OA, Odukoya O. Desmoplastic ameloblastoma: analysis of 17 Nigerian cases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2011; 111(1): e27-31. doi: 10.1016/j.tripleo.2010.06.021.

13. Keszler A, Paparella ML, Dominguez F. Desmoplastic and non-desmoplastic ameloblastoma: A comparative clinicopathological analysis. Oral Dis. 1996: 2: 228-23.