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Case Report

Central giant cell granuloma in a 10-year-old girl: microsurgical reconstruction with a microvascularized fibula flap

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ABSTRACT

Central giant cell granuloma (CGCG) are rare, proliferative, benign bone tumors characterized by an abundance of multinucleated giant cells. It affects children and young adults, predominantly women, in the second and third decade of life, it's commonly found in the mandible. The main treatment for giant cell tumors is surgical excision with free histologic margins; in case of mandibular involvement, the use of a vascularized composite bone flap represents the standard for a functional reconstruction. We present the case of a female patient in the first decade of life with a CGCG of aggressive type, located in the ascending branch and right mandibular angle. En bloc resection of the lesion was performed, with a 2-cm margin and mandibular reconstruction at the same surgical time with a free microvascularized flap from the right fibula. CGCG are rare and infrequent tumors during childhood. Due to its uncertain behavior, it is important to rule out other differential diagnoses prior to definitive treatment.

Keywords: CGCG, Central giant cell lesion, Mandibular diseases, Surgical approach

INTRODUCTION

Central giant cell granuloma (CGCG) are benign bone tumors characterized by the abundance of multinucleated giant cells, they represent 4 to 7% of all primary bone tumors and 1 to 7% of all benign head and neck lesions. It affects children and young adults, predominantly women, in the second and third decade of life.[1-3] CGCG affects the mandible 75% of the time, generally near the mental foramen and in the molar region.4 The main treatment for giant cell tumors is surgical excision. Different surgical approaches have been reported in medical literature depending on the size of the cyst, location, and experience of the surgeon.[1] The use of a vascularized composite bone flap represents the gold standard for functional reconstruction of the mandible.[4,5]

CASE REPORT

A 10-year-old patient referred to the reconstructive surgery department with symptoms of 2-month evolution, presenting pain in the left mandibular branch, trismus, and weight loss associated with difficulty to eat. The physical examination revealed an indurated left preauricular lesion, fixed to deep planes, non-mobile, painful, 7x5 centimeters in diameter, associated with

paresthesias in the ipsilateral mandibular branch, no regional adenopathies were palpable (Figure 1). The CGCG diagnostic test was confirmed by incisional biopsy on 07/17/20 before definitive surgery. Serum levels of calcium, parathormone, and phosphorus were measured to exclude hyperparathyroidism. In the computed axial tomography (CT) study, a solid lesion was evidenced in the left chewing space, poorly defined with an attenuation coefficient of 45 Hounsfield units, which causes bone destruction of the mandibular branch and infiltration of the masseter and medial pterygoid muscles (Figures 2 and 3). The magnetic resonance identified an irregular hyperintense tumor on T1 and T2 with heterogeneous enhancement that causes bone destruction of the ascending branch and mandibular angle (Figure 4).

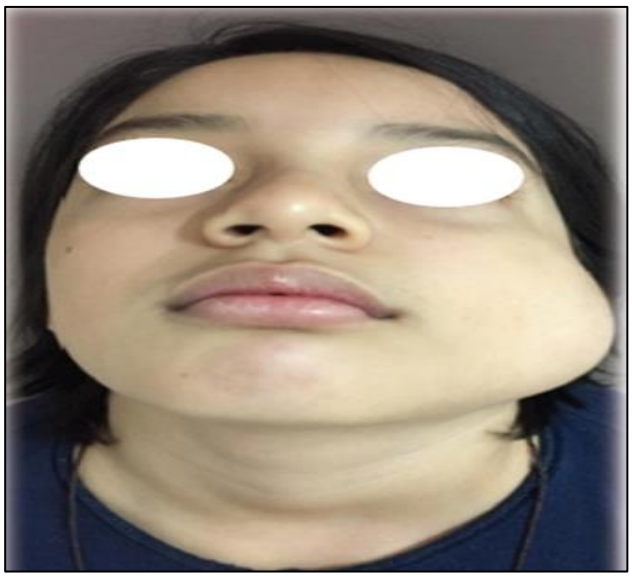

Figure 1: Pre-operative appearance.

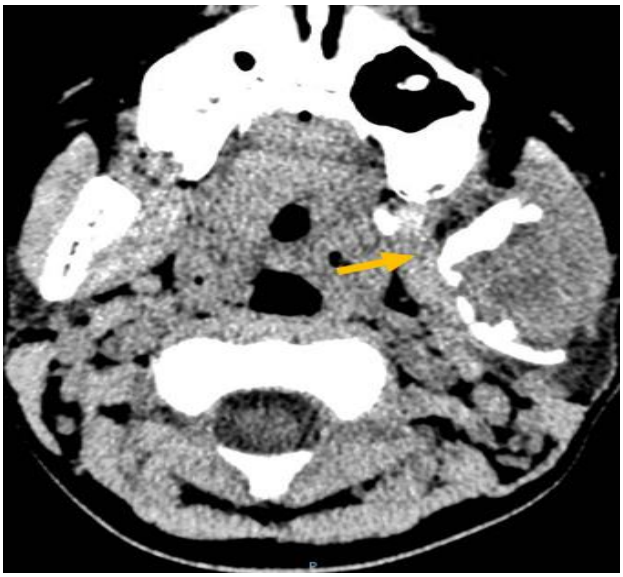

Figure 2: CT showing a solid, poorly defined lesion involving the left chewing space.

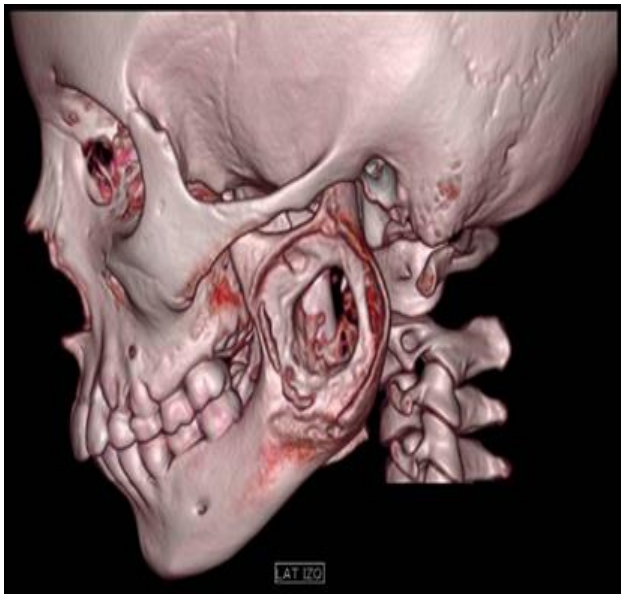

Figure 3: CT with 3D reconstruction, in which a lytic lesion involving the ascending branch and mandibular angle is observed.

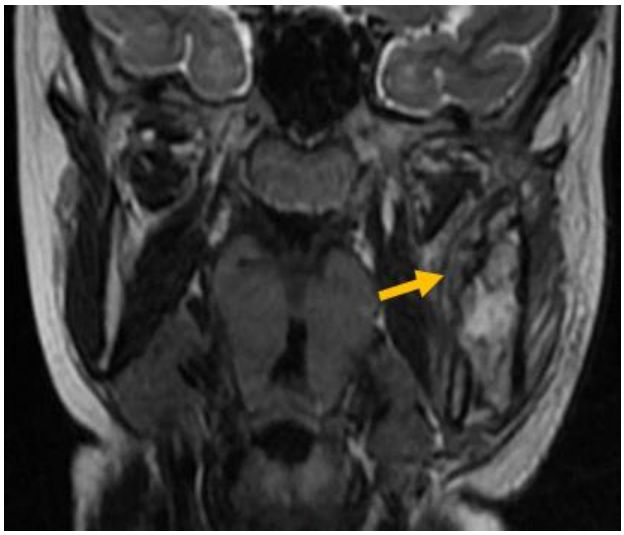

Figure 4: MR coronal view showing an irregular lesion with heterogeneous enhancement (arrow) that displaces the masseter and pterygoid muscles.

Once the diagnosis is confirmed and the extension studies have been assessed, excision and reconstruction are scheduled at the same time. A submandibular approach was performed with dissection up to the submandibular compartment with exposure of the affected portion and en bloc resection with a 2-cm margin. In the second stage, a 10-centimeter right fibular graft was obtained and osteotomized in the canine region, verifying normo-occlusion. Microvascular anastomosis is performed with an ischemia time of 30 minutes, with no evidence of thrombosis. The functional result, including chewing, swallowing and speaking were satisfactory. As of November 28, 2022, the aesthetic results were

satisfactory (Figure 5). There was no impairment of donor site growth or function.

Figure 5: Three month post-operative appearance.

DISCUSSION

CGCG can occur at any age, but is most commonly seen in the first 3 decades.[6] In a retrospective analysis of 18 patients, no cases were found in the first decade of life.[7] In our case, the patient was 10 years old at the time of diagnosis. Aggressive lesions are defined by the presence of one or more of the following signs: pain, paresthesia, root resorption, rapid growth, cortical perforation.[1] The patient started with an accelerated growth, paresthesias, trismus and cortical involvement. Aggressive lesions occur in 19 to 40% of all GCCG cases, with the non-aggressive form of presentation predominating in 60 to 80% of cases that go unnoticed for a long time, due to its growth within the trabecular bone and without pain or neurological alterations.[4] CGCG do not present defined radiological characteristics or pathognomonic radiological findings, and can be confused with other lesions of the jaws.[7] Differential diagnoses include giant cell tumor (GCT), aneurysmal bone cyst, hyperparathyroidism, and other giant cell lesions, which masquerade as osteoclast-like reactive giant cell masses.[3]

A retrospective analysis of patients with CGCG who were treated between 1991 and 2000, demonstrated that lesions confined to the molar-ascending branch region were the most common presentation at 39% (n=7).[7] This is consistent with the presentation of our case that was located at the level of the ascending branch. Regarding the size of the lesion, it was about 4.5 centimeters in its longest axis, according to Bataineh et al. His study showed that the lesions measure an average of 2.7 to 10 centimeters.[7]

The traditional treatment of CGCG is surgical resection. However, the extent of tissue removal varies from simple curettage to en bloc resection.[7] The treatment of choice is wide resection of the tumor mass. Other treatment modalities include cryotherapy, chemotherapy, and curettage with adjuvant agents.[3] Modalities other than surgery have now been tried, which may be associated with recurrence, morbidity, and severe facial mutilation. A series of seven cases describes the effective use of low and high dose denosumab in the treatment of GCT and CGCG of the jaw.[2] Within the available options for a functional reconstruction, the vascularized composite bone flap represents the reference technique for mandibular reconstruction.[4,5] Satisfactory esthetic results can be achieved using the fibula flap with virtual surgical planning, which is the ideal option for mandible reconstruction.[8] The recurrence documented in medical literature after resection of the tumor mass is 7%. In our case, the patient with an aggressive presentation is at risk of local recurrence.[1] The documented malignant transformation rate is 1-5%.[3] When fracture or rupture of the granuloma occurs, local recurrence rates are as high as 25%.[1]

CONCLUSION

CGCG are rare and infrequent tumors during childhood. Due to its uncertain behavior, it is important to rule out other differential diagnoses prior to definitive treatment, which continues to be surgical removal as well as close follow-up to document recurrences.

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