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Case Report

Granular cell tumor in the parotid gland in a pediatric patient: case report

Tumor de células granulares na glândula parótida em paciente pediátrico: relato de caso

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ABSTRACT

The granular cell tumor (TCG) is a rare and usually benign neoplasm that can be found in various sites, with preference for the head and neck region. It mainly affects adults between 40 and 60 years old, with a higher prevalence in females, and it is deeply atypical in children. This report describes a case of TCG found in the parotid gland of a pediatric patient, detailing relevant aspects for its diagnosis, including anatomopathological and immunohistochemical evaluation.

RESUMO

O tumor de células granulares (TCG) é uma neoplasia rara e geralmente benigna, que pode ser encontrada em diversos sítios, com destaque para a região de cabeça e pescoço, que é frequentemente acometida. Afeta principalmente adultos entre 40 e 60 anos, com maior prevalência no sexo feminino, sendo esse quadro extremamente atípico em crianças. Este trabalho consiste em um relato de caso de TCG encontrado na glândula parótida, observado em paciente pediátrico, descrevendo aspectos importantes do diagnóstico, incluindo avaliação anatomopatológica e imuno-histoquímica.

INTRODUCTION

Granular cell tumor (GCT) is a rare neoplasm with predominantly benign behavior. The head and neck region is the main site of involvement, with a prevalence between 45 and 65 percent, and oral lesions of the dorsum and lateral border of the tongue are the most common, accounting for 70 percent of reported cases[1].

Lesions may occur in patients from 11 months to 85 years of age, although they are uncommon in children, with peak incidence between the second and sixth decades of life, especially in women of African descent. The prevalence of GCT among all human neoplasms

is extremely low, approximately 0.019 to 0.03 percent[2,3].

Conservative surgical excision is the main treatment for GCT. However, the surgeon must be attentive to signs of infiltration into adjacent connective tissue. When the capsule is completely removed, recurrences are uncommon and the prognosis is excellent[1]. This case report was approved by the ethics committee of Hospital Erasto Gaertner – Liga Paranaense de Combate ao Câncer, under CAAE 63598422.8.0000.0098. The approval opinion number is 5.693.683.

CASE REPORT

An 11-year-old male patient, previously healthy, accompanied by his legal guardian, presented with a complaint of a preauricular swelling for four years, with progressive growth, painless and without inflammatory signs. He also reported fever and night sweats, and denied weight loss.

An ultrasound of the cervical region was performed, showing a hypoechoic nodular image extending into the subcutaneous tissue, located in the left preauricular region, with poorly defined borders, measuring approximately 1.9 × 2.9 × 1.9 cm, with a volume of 3.5 cm^3, and no Doppler vascularization. Based on these findings, a surgical approach was chosen, and the patient underwent complete tumor excision of the parotid gland (partial parotidectomy) with preservation of the facial nerve.

The surgical specimen was sent to the pathology department. Macroscopic examination identified a lesion of the left parotid measuring 2.5 cm at its greatest axis. Microscopic analysis revealed an epithelioid cellular pattern with intracytoplasmic granules, showing discrete nuclear pleomorphism, absence of mitosis, and extension to the dermis and hypodermis (**Figure 1**). The diagnostic hypothesis was Granular Cell Tumor, and complementary immunohistochemical testing was proposed to confirm the pathological entity.

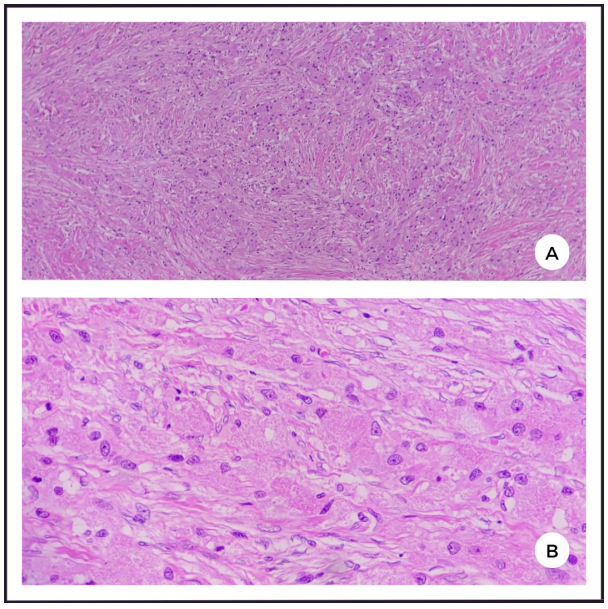

Figure 1. Histopathological appearance of Granular Cell Tumor under Optical Microscopy (OM). A. The tumor architecture shows a neoplasm composed of epithelioid cells extending into the dermis; the image demonstrates the neoplasm surrounded by connective tissue (OM, Hematoxylin and eosin, 100×). B. At higher magnification, intracytoplasmic granules are visible, with discrete nuclear pleomorphism and absence of mitosis (OM, Hematoxylin and eosin, 400×).

The antibodies vimentin, CD-68, S100 (**Figure 2**), and SOX10 were used, all showing diffuse expression in the tumor cells, and Ki-67 was positive in 0.5 percent of nuclei. With this profile of positive markers, the findings were compatible with a granular cell tumor of the parotid gland.

Postoperatively, the surgical scar showed good evolution. Subsequently, the patient developed facial paralysis with deviation of the oral commissure, for which physiotherapy sessions were indicated as treatment, along with follow-up by the oncology team.

DISCUSSION

GCT, or Abrikossoff tumor, is a benign soft tissue neoplasm that can affect various regions of the body, with malignant potential in 2 to 3 percent of cases. When first described by the pathologist Alexei Ivanovich Abrikossoff, it was believed that GCT originated from striated skeletal muscle, and from this assumption the neoplasm received other names, such as granular cell myoblastoma[3].

Currently, it is believed to originate from Schwann cells (neural cells), although the pathophysiology of its occurrence in the tongue remains uncertain; there are reports of its development after traumatic events. It is known that when damage occurs to the myelin sheath, it undergoes a process of disintegration and is phagocytosed initially by Schwann cells and later by macrophages, triggering the proliferation of granular-appearing cells, which may subsequently accumulate in organs. Although the tongue is the most affected region, more uncommon forms may involve the respiratory system, paranasal sinuses, skin, and gastrointestinal tract. In the present report, the patient presented with a GCT of the parotid gland, which is a rare finding. Moreover, the age of affected patients is generally between 20 and 60 years, whereas in this case the diagnosis occurred at age 11[4–8].

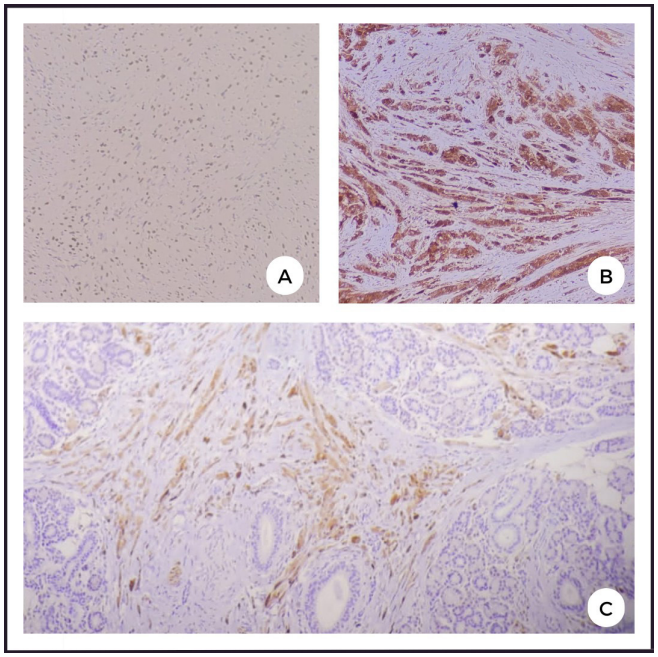

Figure 2. Immunohistochemical profile of Granular Cell Tumor. A. Positive nuclear expression for SOX10 in tumor cells (OM, Immunohistochemistry, 100×). B. Cytoplasmic positivity for CD68 in granular cells (OM, Immunohistochemistry, 400×). C. Diffuse cytoplasmic expression of S100 protein in tumor cells within the glandular tissue (OM, Immunohistochemistry, 100×).

In addition, head and neck tumors are considered atypical in children, representing fewer than 5 percent of cases. These neoplasms form a heterogeneous group generally related to congenital malformations[1]. In an extensive study presented by Stewart et al. involving 48 granular cell tumors, 19 percent were observed in patients between 0 and 20 years of age, and these findings are consistent with those of Brannon and Annand, who reported that of 61 cases, 16 percent occurred in patients aged 0 to 19 years. Furthermore, most studies show a predominance in females, unlike the case reported here[9,10].

The main clinical and pathological features of oral GCT, based on data collected in Spain and Brazil, include the presence of a single non-encapsulated nodular lesion located in the submucosa, generally asymptomatic (89 percent) and with long-term evolution. Once the definitive diagnosis of GCT is established, there is consensus that treatment should consist of complete surgical excision of the nodule with wide safety margins to prevent recurrence[11,12].

The differential diagnosis includes other entities that may present similar characteristics clinically and radiologically, such as fibromas, lipomas, neuromas, neurofibromas or schwannomas, and even squamous cell carcinoma[12].

Histologically, it is characterized by the

Table 1. Most frequent differential diagnoses in parotid lesions in the pediatric population[5].

Diagnosis	Microscopy	Immunohistochemistry	Clinical characteristics	Notes
Fibroma	Proliferation of fibroblasts in a collagenized stroma, without significant atypia or mitoses.	Vimentin +, S100 +, Smooth muscle actin −, Desmin	Firm lesion, slow growth	Rare in the parotid gland
Lipoma	Proliferation of mature adipocytes, without atypia or necrosis.	S100 positive in adipocytes, CD34 negative	Soft, mobile, and painless	Typical fatty appearance
Neuroma/ Neuro fibroma	Proliferation of spindle cells with wavy nuclei in a myxoid stroma; "hair-like" collagen may be present in neurofibroma.	Diffuse S100 positive; CD34 positive; SOX10 positive	Painless mass, slow growth	May be associated with neurofibromatosis
Schwannomas	Typically shows two patterns: Antoni A (hypercellular areas with nuclear palisading) and Antoni B (hypocellular areas). Verocay bodies may be present.	Strong and diffuse S100 and SOX10 positivity; EMA negative	Encapsulated, painless mass	Frequently arises from peripheral nerves
Squamous cell carcinoma	Islands of squamous cells with keratinization, pleomorphism, and frequent mitoses. May invade salivary parenchyma.	p40 and p63 positive; CK5/6 positive; S100 negative	Rapid growth, invasive, and may be painful	Rare in children
Hemangioma	Proliferation of vascular channels lined by endothelium, with or without lumen.	CD31 and CD34 positive	Rapid growth and reddish coloration	Most common benign lesion in children
Parotid cyst	Cuboidal or columnar epithelial lining, without atypia; may contain mucoid material.	CK7 positive; S100 and SOX10 negative	Fluctuant and painless	Generally benign
Lymphoma	Lymphoid infiltrate that may obliterate glandular architecture.	CD45 positive; specific panel depends on the subtype	Firm mass with rapid growth	May be associated with regional lymphadenopathy

proliferation of large, slightly elongated polygonal cells. The nuclei are small, with abundant eosinophilic and granular cytoplasm. These cytoplasmic granules stain intensely and diffusely with PAS (Periodic Acid–Schiff), demonstrating their glycoprotein composition[13]. The organizational pattern of the cells appears in layers, nests, or cords, with infiltration into adjacent tissues. The nuclei are usually small and central, with a subtle nucleolus and no mitoses. In 50 percent of cases, acanthosis or pseudoepitheliomatous hyperplasia involving mucosa is present[14].

Immunohistochemistry is an essential technique for tissue-based diagnoses and biomarker detection. Advances in basic chemistry, antibody design, and automation have resulted in greater sensitivity, specificity, and reproducibility[15]. In the case of GCT, diagnosis is primarily associated with positivity for the S-100 antibody. However, other markers also show immunoreactivity, such as alpha-1-antitrypsin and CD68. Nonetheless, immunoreactivity for both alpha-1-antitrypsin and CD68 in GCT may simply reflect the intracellular accumulation of phagolysosomes[16,17].

In conclusion, accurate diagnosis is achieved through anatomopathological study, which enables identification of the histological type and cellular characteristics of the lesion, complemented by immunohistochemistry and special stains. This highlights the relevance and uniqueness of the case described, emphasizing not only the importance of GCT but also the rarity of its occurrence in an extralingual location in childhood. Finally, although uncommon in the pediatric age group, granular cell tumor should be considered as a differential diagnosis in this population.

AUTHOR CONTRIBUTIONS

ACF, JSM, SHM, and JCL contributed to the conception and design of the study, data analysis, and manuscript writing. JCL performed the final revision of the text. All authors read and approved the final version of the manuscript and agree to take responsibility for its content.

CONFLICT OF INTEREST

We wish to confirm that there are no known conflicts of interest associated with this publication and that no significant financial support has influenced its results.

DECLARATION REGARDING THE USE OF GENERATIVE AI

The authors declare that they used the generative artificial intelligence tool ChatGPT to assist with language revision. The editorial board made the decision to utilize ChatGPT, an AI language model developed by OpenAI, for the translation of this manuscript from the original language, Portuguese, to English.

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