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Perianal Granular Cell Tumor. Case Report and Literature Review

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ABSTRACT
The granular cell tumor (GCT) was described by Abrikossoff in 1926. Only 8% is located in the gastrointestinal tract and its perianal location is rare. According to the current literature review, this would be the thirty-second reported case. Perianal GCT is a rare and difficult to diagnose lesion. For its treatment, a complete surgical resection with free resection margins is essential, as well as an immunohistochemical examination. Subsequently, strict surveillance of patients is necessary to detect local recurrence, lymph node dissemination, and distant disease characteristic of malignant GCTs, which have a poor prognosis.

Keywords: *Granular cell tumor; Abrikossoff tumor; Perianal lesion*

INTRODUCTION

The granular cell tumor (GCT) is an infrequent lesion of the submucosa that is generally found in the oral cavity, located on the tongue in one third of the cases. About 8% of GCTs are located in the gastrointestinal tract, with the esophagus responsible for most of these cases, followed by the colon. Other sites where it can be found include the skin, breast, bile duct, respiratory and genital tracts.[1-8]

In 1926, Abrikossoff ([1]) reported the first five cases of GCT, three of which were localized on the tongue. This author postulates that the tumor originates from an embryonic muscle cell and identified the tumor as a myoblastic myoma. Subsequently, changes in the pathophysiology occurred and thus, in 1939 Leroux and Delarue postulated that GCT was not of myogenic origin but rather a non-neoplastic accumulation of granular histiocytes. In 1935, Feyrter et al. proposed the most accepted and current pathogenesis of the tumor as being of neural origin, defining it as a myoblastoma. Gullino et al., in 1949, using immunohistochemistry and electron microscopy, described the myoblast as a Schwann cell due to its positivity for S100 and the myelin-associated glycoprotein staining pattern.[2,4,9]

At the time of this bibliographic review, there are 31 cases of perianal GCT reported in the literature, which makes it an anal neoplasm with a low frequency of presentation.[3,7,6,9,10]

GCT usually manifests between the fourth and sixth decade of life and its prevalence is higher in women with a ratio of 1.5: 1, not finding differences between races.[4,6,10,11]

It generally presents as a single lesion, although multiple synchronous or metachronous lesions may also appear in 10-15% of cases.[3]

The treatment of choice in the perianal location is local excision with free margins and in patients with lymph node involvement, resection of the lymph nodes in the same surgical procedure.[3,4,6,10,12]

CASE REPORT

A 63-year-old female patient consulted for presenting a 3-month asymptomatic perianal mass, with progressive growth from its beginning.

The proctological examination revealed a perianal lesion in the left anterolateral quadrant (hours 1 to 3), rounded, approximately 4 cm in diameter, indurated, which on palpation appears not to be attached to the deep planes (Fig. 1).

A 360º transanal ultrasound is performed, showing at a low level a 37x15 mm hypoechoic mass involving the subcutaneous bundle of the external anal sphincter (Fig. 2). A colonoscopy ruled out other lesions. Complete excision of the tumor with 1 cm free margins was performed (Fig. 3).

During the postoperative period, the patient has a good progress, completely healing the wound in 4 weeks. Neither in the physical examination nor in the imaging studies was local recurrence or dissemination observed two months after excision.

In the histopathological study, the lesion was described as lined by squamous epithelium with pseudoepitheliomatous hyperplasia. No cytological atypia, mitosis or areas of necrosis were observed. The resection margins were free (Fig. 4). Immunohistochemical techniques

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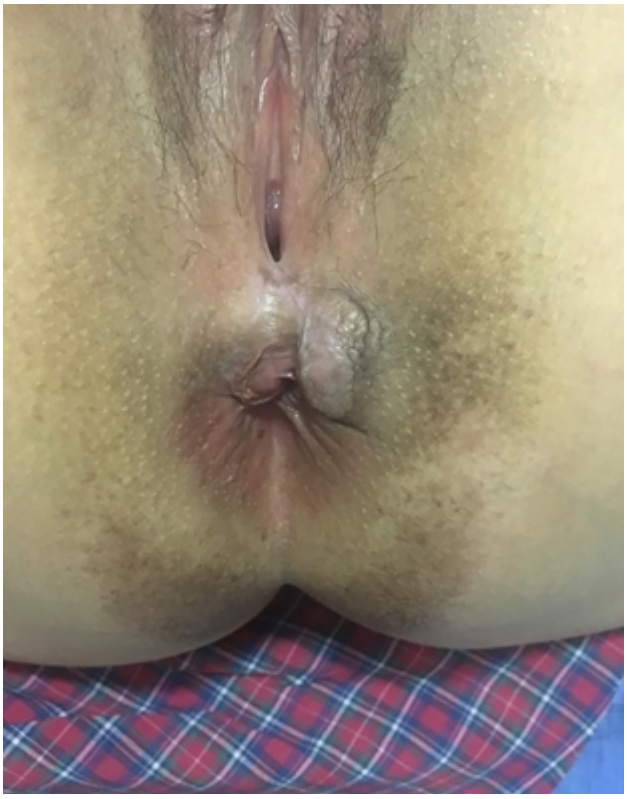

Figure 1: Perianal lesion

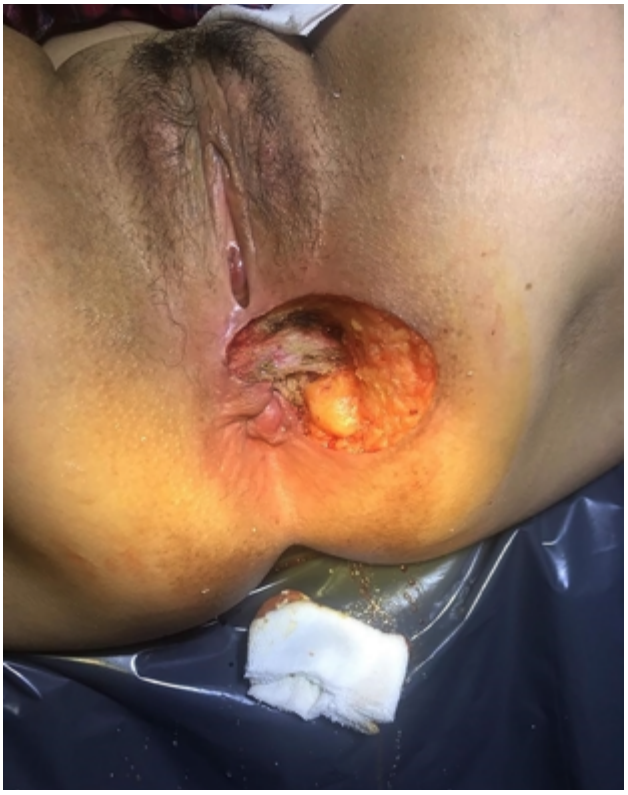

Figure 3: Complete resection

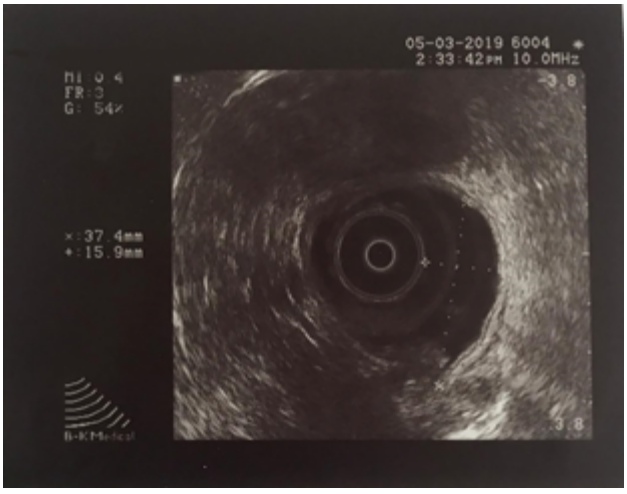

Figure 2: 360 ° ultrasound

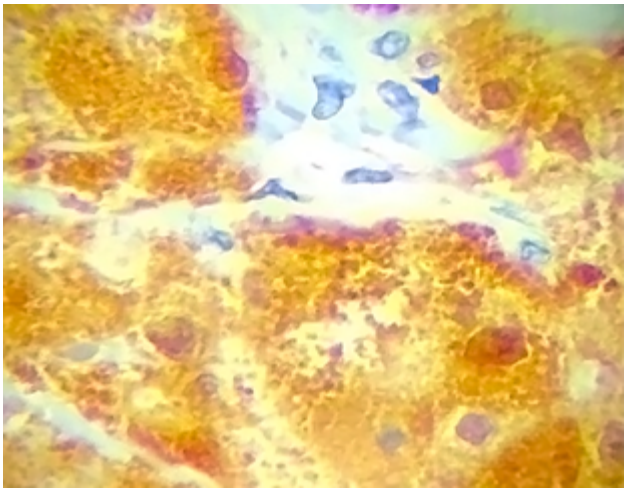

Figure 4: Histopathology

were performed: S100 positive, CD68 weak positive and Desmin negative (Fig. 5). The histopathological diagnosis was: granular cell tumor.

DISCUSSION

GCTs are generally limited to the submucosa and present as benign, asymptomatic, small, non-ulcerated, polypoid lesions, most of which are found incidentally on proctological examination. When these lesions are larger than 2 cm, they may present symptoms such as a sensation of lump, pain and bleeding. When perianal GCT is symptomatic, it can be confused with an organized abscess.[3,4-9]

In computed tomography, its characteristics are nonspecific, appearing as a heterogeneous soft tissue mass that can be difficult to assess due to the small size.[8]

From the histopathological point of view, due to the pseudoepitheliomatous hyperplasia and the overlying acanthosis the main differential diagnosis to ruled out is squamous cell carcinoma (SCC). GCT differs from SCC due to its keratin, desmin, and negative actin staining pattern specific to muscle, so the diagnosis must be made by biopsy with histological and immunohistochemical analysis.[3,10,11,13] The diagnosis can be confused with

other reactive processes or neoplastic conditions that exhibit a granular cytoplasm. However, currently available immunohistochemical staining is extremely valuable in establishing the differential diagnosis since characteristically S-100, vimentin and the lysosomal marker CD68 stain in the nucleus and cytoplasm.[3,10,11]

When a suspicious lesion is observed, surgical removal is mandatory for diagnosis and treatment which may be curative, although requires strict monitoring due to the potential for malignancy, the possibility of metastasis, and the risk of recurrence.[3,5]

Approximately 1-2% of cases are malignant.[3,5,12] Differential diagnosis between benign and malignant lesions is sometimes difficult due to their similar histological appearance. The most important predictor of malignancy is size, followed by atypical histology.[3,11,12] Benign lesions are generally smaller than 3 cm and have uniform nuclei with the absence of mitotic figures, while 60% of malignant lesions are larger than 4 cm.[4,14]

Malignant GCT can generate lung, liver, and bone metastases with typical lymphatic and hematogenous spread. Approximately 50 cases of malignant GCT have been reported in the literature, of which only 3 were reported in the anal and perianal region.[15,16]

It is not clear whether malignant lesions result from the transformation of benign lesions or begin as malignant and because of that the surgical resection with free margins is mandatory at the time of diagnosis to avoid local recurrence, and lymphatic and hematogenous spread in case of malignancy.[3] A malignant GCT may appear histologically identical to a benign one and only the appearance of metastases will lead to a subsequent diagnosis of malignancy.

The prognosis for malignant GCT is not encouraging, with an overall mortality rate of 30% to 50%. Fanburg-Smith, et al[17] reported a 2-year recurrence rate of 32 vs. 2-8% for malignant and benign tumors, respectively. This rate can increase to 20% in benign lesions with compromised margins. Therefore, it is extremely important to perform a local excision with a disease-free surgical margin.[6]

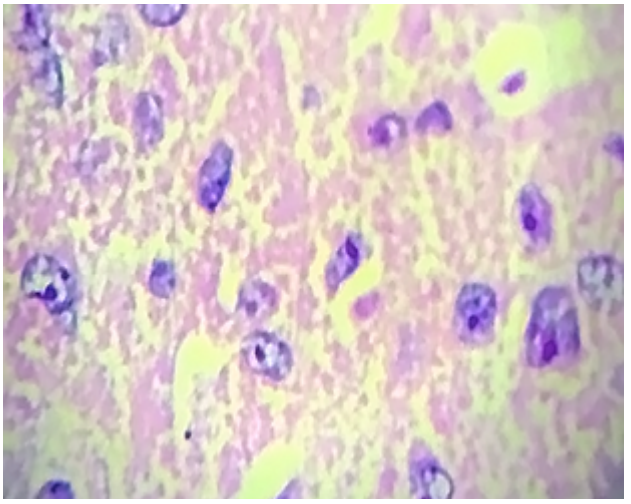

Figure 5: Immunohistochemistry

After resection and even more so in those with a malignant presentation, complementary tests should be performed to meticulously evaluate and rule out local recurrence and metastasis, since neither chemotherapy nor radiotherapy improve the prognosis.[3,6]

CONCLUSION

Perianal GCT is a rare and difficult to diagnose lesion. Because of this, we consider that it should be borne in mind when evaluating a patient with an asymptomatic or slightly symptomatic perianal submucosal lesion.

It is essential to perform a complete surgical excision with free resection margins, as well as immunohistochemistry to obtain a precise diagnosis and rule out squamous cell carcinoma, the main differential diagnosis.

After surgical treatment, close monitoring of patients is necessary because it has a high recurrence rate and there are no precise clinical, radiological or histological characteristics that differentiate a malignant from a benign GCT. During this follow-up, we must look for local recurrence, lymph node dissemination, and distant disease that characterize malignant GCT and pose a poor prognosis for the patient.

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