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Abstract

Granular cell tumor is a rare tumor that can occur at any location. The most frequent one is the buccal mucosa especially on the tongue, dermis and subcutaneous tissue. Few cases were reported to occur in the perianal area. Although it is an almost always-benign entity, it can be associated to an alteration of the patient's quality of life. Although a malignant variant of this tumor exists, it barely presents 2% of all cases. The basic treatment is surgical excision. In our work, we report a case of perianal granular cell tumor in a 52 years old female evolving during a period of one year. In our work, we also discuss the most important epidemiological, clinical, pathological, therapeutic and prognostic features of this rare entity.

Keywords

Granular cell tumor; Gastro-intestine; Buccal Mucosa; Prognosis

Introduction

The granular cell tumor is a pathological entity which was first described by Abrikossof in 1926. Many sites have been described. The most common is the oral mucosa, especially on the tongue, dermis and subcutaneous tissue. The granular cell tumor is rarely located in the perianal region. Granular cell tumors often occur between the 4th and 6th decade with a male prevalence of up to 68%. Although there is a malignant variant of this tumor, it represents barely 2% of all cases. The basic treatment is surgical excision with local recurrence being the only possible complication when the resection is incomplete. In our work, we report the case of a granular cell tumor of the perianal region which has been clinically mistaken for squamous cell carcinoma. 6 months after surgery, the patient had a good course, with no signs of local recurrence. We also discuss the most important features of this entity.

Case Report

Our patient is a 52-year-old woman, married, without clinical or surgical morbidity. No relevant family neoplastic history was found. She complained for about a year of perianal pain, without diarrhea, abnormal discharge, rectal bleeding or fever. The patient has already received anti-hemorrhoids treatment for more than 2 months without any improvement. She was then admitted to our hospital, where a rectal examination revealed a right perianal nodule without local inflammatory changes. No hemorrhoids or other lesions were identified. General

the patient's examination was normal. We performed a contrast-enhanced pelvic MRI, which identified a lesion of the hamstring which appeared isointense on the T1- weighted images and hypo-intense on the T2-weighted images (Figure 1). The lesion showed progressive and delayed improvement on T1-weighted images with fat saturation with contrast. He was in close contact with the levator ani and gluteus maximus mascules. Other investigations included colonoscopy, laboratory tests and a chest x-ray and were found to be normal. Complete gross surgical excision and the surrounding subcutaneous tissue under local anesthesia were performed. The local anesthetic used was Marcaine 25%, with a safety margin of 0.5 cm around the mass. Pathological examination of the resection sample, a 5.5x4.5x2cm subcutaneous tissue, revealed a diffuse proliferation of large cells. These cells have a polygonal oxyphilic granular cytoplasm and a regular, often pycnotic, hyperchromatic nucleus. The tumor stroma was essentially fibrous with no inflammatory infiltrate. No mitotic activity or necrosis was observed. The proliferation locally invades the skeletal muscle (Figures 2 and 3). The cells have a PAS + cytoplasm under histochemical study (Figure 4). In an immunohistochemical study, the tumor cells were positive for the proteins CD68 and S100 (Figure 5). 6 months after surgery, the patient had a good course, with no signs of local recurrence.

obtained on resection specimen and rarely on biopsy specimens. Granular cell tumors occur in the gastrointestinal tract in only 5 to 19% of all cases. The largest series of

Discussion

Granular cell tumour is a tumour that occurs especially on the buccal

granular cell tumours was reported by Lebranchu et al. He reported 263 cases, showing man predominance at about 68%, and among all cases, only one patient had a perianal location, whereas the others frequent locations were skin (38%), esophagus (19%) and tongue (10%). A high incidence rate in African descent patients were found in up to 29% of cases in a study done by Lack et al about 110 patients [4]. Most interestingly, at least in our case, the largest number of perianal location for this tumor was reported in a series of 74 cases of gastrointestinal granular cell tumors. This study has found only 48 patients to have a perianal location. The median patient's age was of 39 years old for the gastrointestinal location. In a 2016 review of literature made by Araújo et al. among 48 cases of recto-anal localized granular cell tumor, 40 had an anal/ perianal location. Another case of peri-anal granular cell tumor was recently reported by Emily F Kelly et al. When this tumor is located on the perianal area, it manifests most often in form of a painful nodule as in the case reported in our work, although the identification of a nodule was possible only one year after the beginning of symptoms. Other reported symptoms include rectal bleeding and also incidental discovery of the tumor when its symptoms are hidden by those of other perianal pathologies such as hemorrhoids and perianal fissures. On histopathological level, the granular cell tumor is non-capsulated and typically made of large granular eosinophilic cells [3]. The tumor cells are known to express the S-100 protein, neuron specific enolase (ENA) and CD-68, thus justifying a both neural and macrophage/histiocytic origin for this tumor, although it was initially thought to have a muscular origin. The link to histiocytes/macrophage is more supported by morphological and structural features. Nevertheless, the histogenesis of granular cell tumors is not completely elucidated.

Conclusion

In our work, we report a case of perianal granular cell tumor in a 52 years old female evolving during a period of one year. We also discuss the most important epidemiological, clinical, pathological, therapeutic and prognostic features of this rare entity.

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Mucosa (Particularly on the tongue), the skin and subcutaneous tissue. Nevertheless, it can occur on any location. In up to 8,5%, these tumours can occur in a multifocal fashion. The diagnosis is most

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