



Case Report

Granular cell tumor of rectal submucosa: case report



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ABSTRACT

This is a case report of granular cell tumor of rectal submucosa in a female, 35-years-old patient complaining of hematochezia. We describe the histological and immunohistochemical features of the lesion responsible by this clinical find. Following that, we present a discussion of the case based on the literature review, which allowed to proving the infrequency of the tumor in the rectal area and confirms the benign nature of the tumor in this case.

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Tumor de células granulares da submucosa rectal: relato de caso

RESUMO

Este artigo relata o caso de um tumor de células granulares da submucosa retal, em paciente de 35 anos, com queixa de hematoquezia. Fazemos a descrição dos achados histológicos e imuno-histoquímicos da lesão. Além disso, apresenta-se uma discussão do caso com base na revisão da literatura, que permitiu comprovar a infrequência do tumor na região retal e corroborar a benignidade do tumor no presente caso.

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Palavras chave:

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Introduction

The granular cell tumor, formerly called granular cell tumor myoblastoma or Abrikossof tumor, is a rare neoplastic process predominantly benign. It affects different regions of the body, but the lesions in the gastrointestinal tract represent only 10% of the cases.^{1–4}

The patient with this tumor is usually asymptomatic and diagnosed as an incidental finding during the investigation of other pathologies. The study with pathological assays is the gold standard for confirming the presence of this kind of tumor.^{5–7}

Although most part of the lesions are benign, a small number shows malignant behavior and about 2% of them can metastasize, especially those with atypical histology or large diameter. In these cases, it is necessary to conduct further investigations and apply more aggressive surgery resections as plan of treatment.^{8–10} We report a case of granular cell tumor, in a young female patient complaining of hematochezia for one year, at the Coloproctology Department of the University Hospital Santa Casa de São Paulo in 2013.

Case report

L.V.A., female, 35 years old, born and raised in São Paulo – Brazil, complained of an one-year history of haematochezia associated with colic abdominal pain in the lower quadrants, with no bowel habits change, during a medical attending of the Coloproctology Clinics at University Hospital Santa Casa de São Paulo. Regarding her past medical history, she had been smoking for 10 years and had a history of hemorrhoidal disease for 17 years. With regard to the family history, her mother had intestinal polyps and diverticular disease.

Physical examination

On physical examination, the patient was in pain on palpation of the right iliac fossa, with no rebound tenderness. On proctological examination, skin tags compatible with hemorrhoids were observed in the right lateral, posterior left lateral and anterior regions. The anoscopy showed hemorrhoidal protruding nipples in the right lateral, left lateral and posterior left.

Complementary exams

The upper endoscopy identified an erosive esophagitis classified as Los Angeles A, a flat and moderate erosive bulboduodenitis and an esophageal polypoid lesion. Pathologic examination revealed a mild chronic esophagitis.

The colonoscopy exam revealed two polyps in the rectum and sigmoid colon and a yellowish elevated lesion of 0.5 cm in diameter, 5 cm away from the anal edge, which was totally resected. The pathology of the early lesions showed hyperplastic polyps of colon and rectal mucosa. The latter injury, however, was a polypoid fragment coated by rectal mucosa consisting of mature neoplasm characterized by a proliferation of cells with abundant cytoplasm, micro-vacuoles,

eventually granular, and building blocks interspersed with bands of collagen fibers, as shown in Fig. 1. The immunohistochemistry revealed the expression of CD56, S100, CD34 in the normal vascular endothelium, CD68 in the normal histiocytes and a low (1%) percentage of Ki67 cell proliferation (Fig. 2).

Diagnosis

Through the proctological exam, internal hemorrhoids were diagnosed. The upper endoscopy showed mild chronic esophagitis and the colonoscopy revealed a granular cells tumor of the rectal submucosa.

Medical management

Preoperative tests were ordered to perform hemorrhoidectomy. Given the benign nature of granular cell tumor in this case, it was chosen to monitor the patient after endoscopic resection.

Discussion

The granular cell tumor was described for the first time in the oral cavity by Abrikossof, in 1926 and it is a rare neoplasm, predominantly benign.¹ It can affect any area of the body, being more frequent in the oral cavity and subcutaneous tissue.³ In the gastrointestinal tract, the organ with the highest prevalence of the tumor is the esophagus, followed by the large intestine.⁴ The incidence is higher in females (1.5:1), with wide variation in the range of age in the diagnosis, but with a peak of incidence between the fourth and sixth decades of life.²

This neoplasm has a mesenchymal origin, probably derived from Schwann cells, a hypothesis that has become strong due to the discovery of a high immunohistochemical affinity of this tumor for S-100 protein, myelin and myelin-associated glycoprotein.⁸

The granular cells have typically benign pattern, with polygonal or fusiform arranged shape in compact nests separated by collagen fibers.⁴ These cells have abundant cytoplasm, eosinophilic with PAS-positive granules and small and uniform nuclei.^{11,12} The histological and immunohistochemical descriptions found in the case are consistent with those found in the literature.

The tumor usually presents as a non-ulcerated nodule, painless, with less than 3 cm, with slow growth.^{5–7} In most of the cases, the tumor is asymptomatic, but the onset of symptoms is more common proportionally to the size or number of the neoplastic lesions.⁴ About 10% of the patients have multiple tumors.¹¹ The rate of recurrence after surgical resection is also low: around 5–10%.⁸ The finding of this injury is usually made during the investigation of other diseases through the tests such as endoscopy and colonoscopy.⁸ The tumor usually presents as a sessile polyp, yellow-grayish, with firm consistency.^{4,8} The diagnosis is made through excisional biopsy and pathological study.

The malignant potential of the tumor and its criteria are still questioned in the literature. About 2% of tumors generate metastasis.⁷ Yamada et al. suggest that the main factor for malignization is the lesion size, since 60% of metastases occur

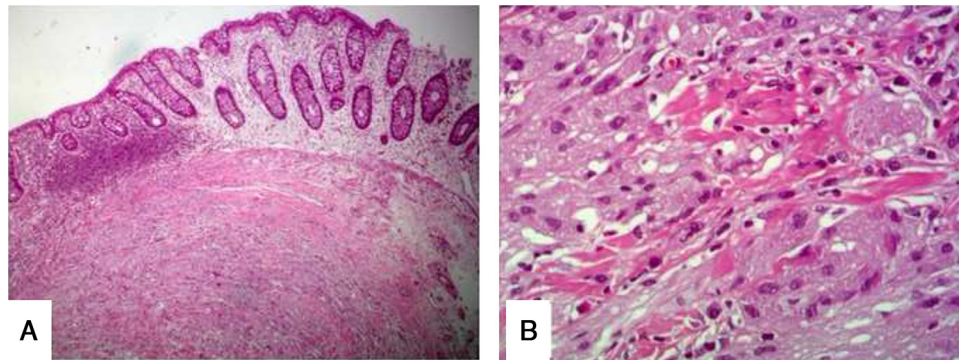


Fig. 1 – (A) Histologic overview of the lesion. (B) Detail of the histology.

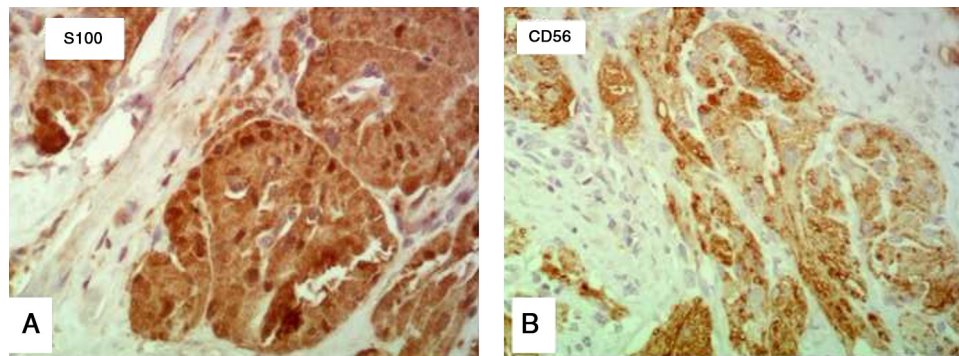


Fig. 2 – Immunohistochemistry of the lesion. (A) Immunohistochemistry for S100 marker. (B) Immunohistochemistry for CD56 marker.

in tumors larger than 4 cm in diameter.^{8,9} Fanburg-Smith et al. suggest the use of histological criteria such as cellular necrosis, pleomorphism, increased nuclear-cytoplasmic ratio, enlarged nucleoli, increased mitotic activity and cell elongation. Malignancy is suspected when 3 or more of these factors are present.¹⁰ Besides those, local recidivation or rapid growth tumor should also be considered as criteria.⁴ None of these features were present in the lesion found in the patient, suggesting benignity.

In the absence of malignancy criteria, the recommended treatment is endoscopic excision, and even the tracking of tumors smaller than 1 cm can be made without resection.^{4,13} In cases of multiple tumors, it is necessary to monitor the patient, due to the higher risk of malignancy.¹³ Due to the benign features of the tumor in our patient we chose to follow her with annual colonoscopies.

The granular cell tumor is a rare neoplasm, predominantly benign that can affect any part of the body, including the gastrointestinal tract. A small percentage of these lesions have malignant potential and generate metastasis, being important to analyze the diameter and histological features before its management. In majority of the cases, we can safely perform the endoscopic resection.

Conflicts of interest

The authors declare no conflicts of interest.

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