CASE REPORT

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Abrikossoff tumour mimicking cecal polyp – case report and literature review of granular cell tumours the of gastrointestinal tract

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Abstract

Granular cell tumour (GCT) or Abrikossoff tumour, is a rare submucosal neoplasm orginating from Schwann cells, and accounts for 0.03% of all submucusal tumours. Only 8% of GCTs occur in the gastrointestinal tract, mostly in the esophagus. Diagnosis of granular cell tumour is exceptionally rare. The case is presented of a 72-year-old female patient who underwent polypectomy of a colon polyp which, in the histopathology report, appeared to be a granular cell tumour. The possibility of granular cell tumour diagnosis which is extremly rare, is discussed. However, 2% of these tumours are malignant. It is very important to establish the diagnosis as early as possible. The patient was admitted to the Gastroenterology Outpatient Clinic due to a hepatic vascular lesion. She did not present any symptoms of granular cell tumour. A complete resection of the granular cell tumour fortunately confirmed it to be benign.

Key words

cancer, tumour, colon, polyp, granular cell tumour (GCT), Abrikossoff tumour, Abrikosov's tumou

INTRODUCTION

Granular cell tumours (GCTs) are rare soft tissue neoplasms that can affect virtually any organ and any age group in patients [1]. They are most commonly localized in the oral cavity, skin and subcutaneous tissue. Their occurrence in the gastrointestinal (GI) tract comprises only 8% of all GCTs [2]. They are diagnosed in patients most often between the fourth and sixth decades of life, with a female-to-male ratio of 2:1 [3]. Most lesions are benign, but 1–2% are diagnosed as malignant, which is associated with a worse prognosis [4, 5]. The case is presented of a 72-year-old female patient who underwent colonoscopy due to the high risk of colon cancer in her age group. A polypectomy of a colon polyp was performed and the histopathology report confirmed the presence of GCT.

CASE REPORT

The female patient presented to the Gastro-enterology Outpatient Clinic due to a hepatic vascular lesion found incidentally on abdominal computed tomography (CT) scan. She denied any complaints of abdominal pain, although having had a history of left mastectomy for breast cancer with no follow-up treatment. In control mammography, no changes were detected for further observation. The patient additionally suffers from chronic ischemic heart disease, hypertension, paroxysmal atrial fibrillation, diabetes mellitus, and depression. Due to years of nicotinism and diagnosed chronic obstructive pulmonary disease (COPD), she remains under the control of a pulmonology clinic. Recently, there has been an increase in persistent cough. The pulmonologist ordered a follow-up chest X-ray, which showed lesions of an unclear nature and dilatation of the hilar shadows. Due to the indeterminate nature of the lesions, a review CT scan of the chest and abdominal cavity was performed. Examination showed emphysema, atelectatic bilateral consolidation of the lung parenchyma, and multiple lesions of a calcified nodule nature, ranging from 2mm – 8mm in diameter. Within the abdominal cavity, foci of contrast enhancement with features of vascular malformation in the liver were detected. The hepatic lesion prompted the extension of further gastro-enterological diagnosis to endoscopic studies. Esophagogastroduodenoscopy revealed features of inflammation in the antrum, angle and body of the stomach which were confirmed in histopathological examination. In Warthin-Starry staining, in the mucus on the surface of the stomach, numerous bacteria corresponding to *Helicobacter pylori* infection were found. The patient was recommended to undergo *H. Pylori* eradication. During colonoscopy, a cecal polyp was removed by endoscopic submucosal dissection.

Histopathological examination of the removed polyp revealed a granular cell tumour – identified as an Abrikossoff

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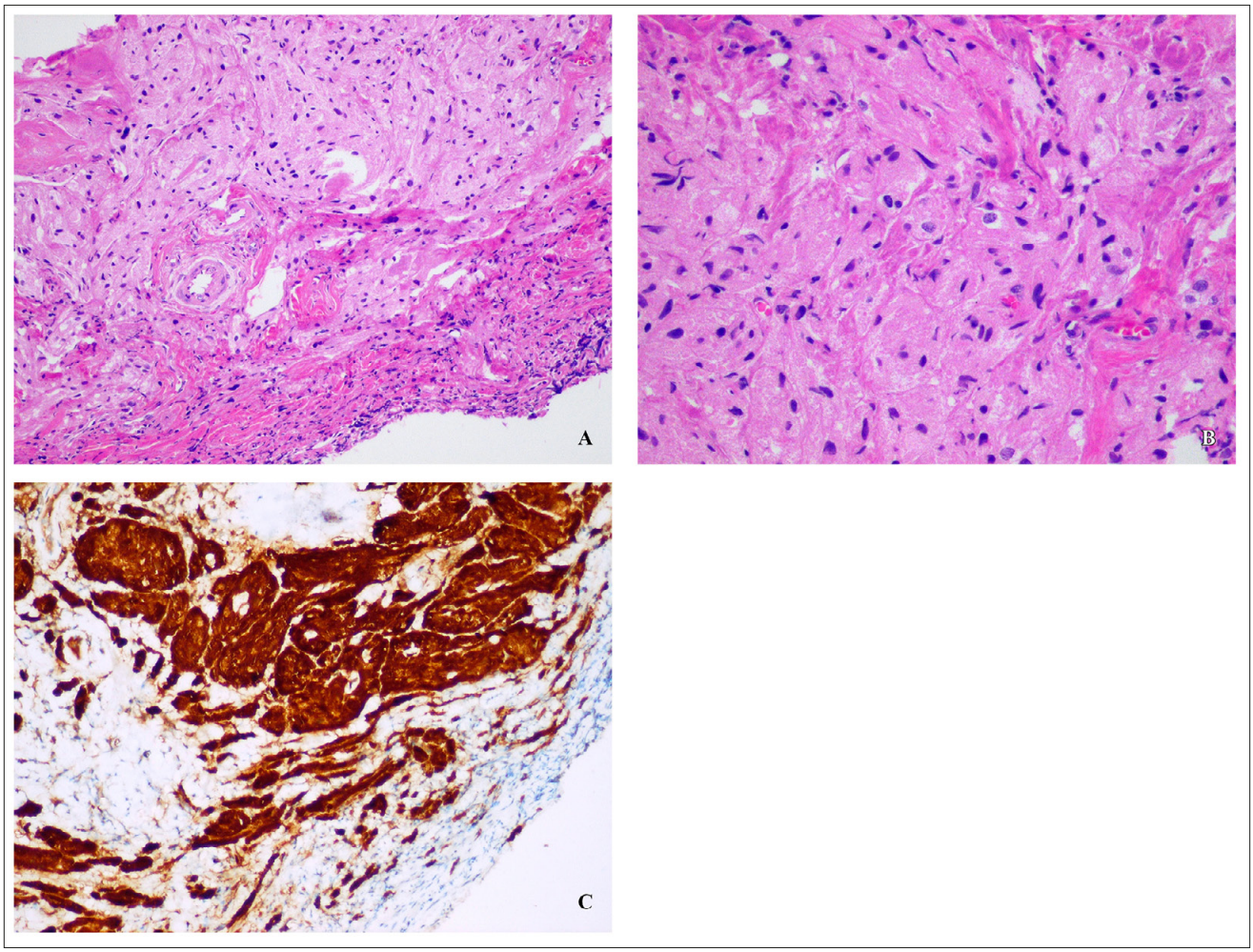

Figure 1. Microscopic appearace of caecal granular cell tumour. Sheets of large polygonal cells with aboundant granular cytoplasm and small dense nuclei with moderate atypia (A, B). Strongly positive cytoplasmic immunostaining for S-100 in neoplastic cells (C). (A, B – HE, C – IHC; objective magnification A, C – 10x, B – 20x)

tumour; however, the margins of the resected polyp were free of tumour. On immunohistochemical analysis, there was positive staining for S-100 protein expression, and negative for desmin.

The patient remains under the control of the Gastroenterology Outpatient Clinic. The patient was planned to follow-up colonoscopy in one year. Subsequent decisions regarding surveillance will be made based on colonoscopy results. At present, the patient presents no symptoms in the gastrointestinal tract.

DISCUSSION AND CONCLUSIONS

Occurence. Granular cell tumour (GCT) is a lesion of controversial origin. It was first classified as a myogenic tumour in 1926 by the Russian pathologist Abrikosoff [5]. However, it was only with the development of immunohistochemistry and electron microscopy techniques that it was properly determined to originate from nerve tissue-Schwann cells [6]. Currently, the etiology of granular cell tumours and their genetic basis is still poorly understood. However, studies show that the tumour often co-occurs with genetic syndromes, such as LEOPARD syndrome or Noonan syndrome [7]. Whole genome sequencing capabilities and a 2018 study by Pareja et al. also identified recurrent somatic mutations inactivating ATP6AP1 and ATP6AP2 in 72% of granulosa cell tumours [8]. Due to its as yet incompletely understood origin and rare occurrence, epidemiological data will begin to be limited. Knowledge of granular cell tumour originates mainly from clinical case reports [9]. GCTs can appear at any age [10], and the lesions can affect almost any organ. Their frequency is shown in Table 1 [2–3,5,7,11–13]. However, the following data remain approximate, owing to the doifferent values presented in literature reviews.

Table 1. The most common locations of granular cell tumours along with the incidence

Location	Prevalence (%)
Oral cavity [2, 5, 7, 11–12]	40%
Skin and subcutaneous tissue [2, 5, 7, 11–13]	30%
Breast [2, 5, 7, 11]	15%
Respiratory tract [2, 5, 7, 11–12]	10–15%

Only 8% of GCTs occur in the gastrointestinal tract, with the most common cases in the esophagus, large intestine and stomach [2]. Only 150 cases of GCTs in the large intestine have been reported in the literature [14]. The most common locations of GCTs of the gastrointestinal tract are shown in Table 2.

Table 2. Frequency of granular cell tumours in different locations of the gastrointestinal tract [1,5,10,14–19]

Location	Prevalence (%)
Oesophagus	65–68%
Colorectal	20–21%
Stomach	4–9%
Small intestine	~1%
Anal	~1%

Symptoms. The symptoms described below relate to locations in the gastrointestinal tract only. Most often, granular cell tumours do not produce clinical symptoms and their appearance depends on the size of the tumour. They are often detected incidentally in endoscopy procedures performed for other reasons. The situation was similar in the patient in the described case report. If present, symptoms are non-specific and depend on the location of the lesion. In the oesophagus, GCT can manifest as gastroesophageal reflux or dysphagia. In the small and large intestine, it can show bleeding or abdominal pain, and in the large intestine, it can additionally cause a change in the nature of bowel movements [1,16].

Diagnosis. The main diagnostic method for GCT is gastrointestinal endoscopy. Mostly, colonoscopy reveals GCT as a yellowish-white colour sitting polyps or nodules. This is commonly the case due to the fact that GCTs have no specific features that distinguish them from other intestinal polyps [14]. Only 7–25% of GCTs in this area occur in multiple forms [16]. Colorectal GCTs are mostly located in the ascending colon and cecum [17].

Histological findings. Histopathological and immunohistochemical examination remains the standard for the diagnosis of granular cell tumours [14]. Morphologic features include nests of polygonal, spindle-shaped cells with abundant eosinophilic, fine-grained cytoplasm [1]. A typical histologic feature of GCTs is the pustule-ovoid bodies of Milian (POBoM), which are detected under microscope as a pale halo surrounding the eosinophilic bodies [20]. They usually show positive staining for S-100, CD68, neuron-specific enolase, CD57, inhibin, TFE3, SOX10, CD56 and vimentin [15,21]. However, there is sometimes a variant that is negative for S-100, but remains positive for CD68, CD10, and sometimes neuron-specific enolase [22].

Although most GCTs are benign lesions, on the basis of histologic features, in 1998, Fanburg-Smith et al. proposed distinguishing features based on which lesions can be divided into benign, atypical and malignant. These include histological features: necrosis, spindling, vesicular nuclei with large nucleoli, and increased mitotic count [23]. Interestingly, none of them were detected in the presented case.

Some lesions are routinely removed by polypectomy followed by histopathological examination, which occurred in the presented patient,.

Treatment. Currently, there are no established guidelines for the treatment of granulosa cell tumours [1, 18–19]. However,endoscopic resection of these tumours with negative margins is recommended by most authors because GCTs can have a possible aggressive biological potential [12]. The mainstay is excision of the lesion by endoscopic mucosal resection (EMR), endoscopic submucosal dissection (ESD) or polypectomy [16, 24]. For lesions smaller than 2cm, EMR or ESD remains preferred, for lesions 2–3cm ESD and for lesions larger than 3cm, surgical excision or consideration of ESD is advised [1, 24]. Currently, radiotherapy and chemotherapy are not recommended for patients with granular cell tumour, but studies are underway on the effect of Pazopanib for metastatic lesions [25]. The prognosis of patients with complete removal of a lesion histopathologically determined to be benign is very good. In contrast, for malignant GCTs, the probability of recurrence and 5-year survival rate is 60–75% [13].

CONCLUSIONS

Although GCT is relatively rare in the colon, it should be considered in the differential diagnosis of submucosal lesions. It also remains important to recognize potentially malignant features of the neoplasm to implement appropriate monitoring and diagnostic vigilance especially in this group of patients.

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