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Granular cell tumor of the perineum (clinical case)

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ABSTRACT *AIM: to present a clinical case of a rare granular cell tumor (Abrikosov's tumor) with perianal site.*
PATIENTS AND METHODS: a patient had a slowly progressive growth of a perianal tumor. The tumor with dimensions of 40 × 30 mm intimately adheres to the lower ampullary rectum and anterior portion of m. levator ani, no signs of malignant transformation occurred. Trepanobiopsy was performed followed by immunohistochemical study. The biopsies contain mainly fibrous tissue and clusters of cells with rounded nuclei and granular cytoplasm. Immunohistochemistry showed diffuse positive cytoplasmic reaction with antibodies to S100. The tumor was positive for CD8, vimentin and negative for GFAP. The expression of the Ki67 protein was 2%. Diagnosis: granulocellular tumor (Abrikosov's tumor), the tumor was removed by perineal access.
RESULTS: the removal of the tumor, originating from the low rectum, was performed with perineal access and with the restoration of the muscular layer of the bowel wall. No complications occurred, patient discharged 5 days after surgery.
CONCLUSION: Abrikosov's tumor is a rare benign neoplasm. Surgery is possible in coloproctological units with sufficient level of surgeons.

KEYWORDS: *granular cell tumor, Abrikosov's tumor, diagnosis and removal of the tumor, dynamic observation*

CONFLICT OF INTEREST: *the authors declare no conflict of interest*

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INTRODUCTION

Granulocellular tumor or Abrikosov's tumor (AT) is a rare benign neoplasm [1], which accounts for no more than 0.5% of all soft tissue tumors [2]. In 1925, the famous Russian pathologist — A.I. Abrikosov, based on the cases of detection of neoplasms in the striated muscles of the tongue and the similarity of tumor cells with embryonic myoblasts, suggested that it has a muscular origin as a separate nosological form [3].

According to the modern hypothesis, AT is a neuroectodermal differentiation of epithelioid cells, auxiliary cells of nervous tissue, and is regarded as a derivative of Schwann cells that form along the axons of peripheral nerve fibers [4]. On this basis, the immunohistochemical determination of the S100 protein produced by Schwann cells as a diagnostic criterion for the diagnosis of AT [5]. In women, this pathology occurs 3 times more often than in men [6]. The majority of granular cell tumors appear in the skin and subcutaneous fat — 33–44% of cases, another common localization is the tongue, palate, trachea, bladder — 23–35%, and 42.7% of patients with multiple tumors have a combined lesion of the skin and internal

organs [7]. In the literature, we have found single descriptions of AT emanating from the rectal wall [8]. The urgency of the problem is also due to the fact that the nature of the tumor development has not been definitively clarified, the diagnosis of AT is difficult, an immunohistochemical analysis is necessary to verify the diagnosis, after removal the tumor may recur, and has a risk of malignant transformation [9]. In this regard, the above observation is of clinical interest.

CLINICAL CASE

Patient F., 58 years old, was admitted to the Coloproctology Unit of the Republican Clinical Hospital of the Ministry of Health of the Republic of Tatarstan inKazan on April 4, 2023 with a preliminary diagnosis: benign rectal neoplasm.

From the history of the disease: this neoplasm appeared 5 years ago. In 2018, the patient underwent a transrectal ultrasound, the described tumor was regarded as an infiltrative perirectal inflammation in the resolution stage. Follow-up was recommended. Subsequently, during the COVID-2019 pandemic, she did not seek medical help. The tumor gradually increased in size, pain and a feeling of a foreign body joined during defecation and in a sitting position. In July 2022, she turned to a coloproctologist. With an objective examination in the perianal region along the right semicircle from 6 to 12 o'clock, a site of tissue compaction up to 4 cm in diameter was determined, with digital examination — of a tight elastic consistency, sedentary. The tumor adhered to the wall of the low rectum and slightly prolapsed into its lumen.

Anoscopy showed that the mucosa above this formation was not changed.

According to the MRI of the pelvic organs (Fig.1): in the perianal region on the right, in the projection of the muscle that raises the anus, a section of the hypointensive signal was determined by T1, T2, T2fs, of irregular shape,

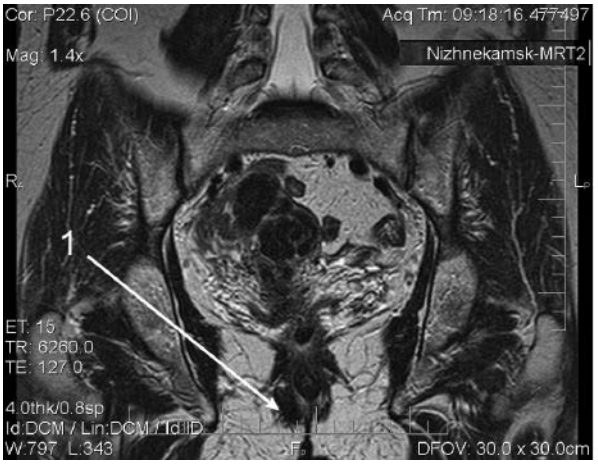

Figure 1. *The neoplasm is located in the right of low rectum and the external sphincter, close to the attachment point of the m. levator ani (arrow 1)*

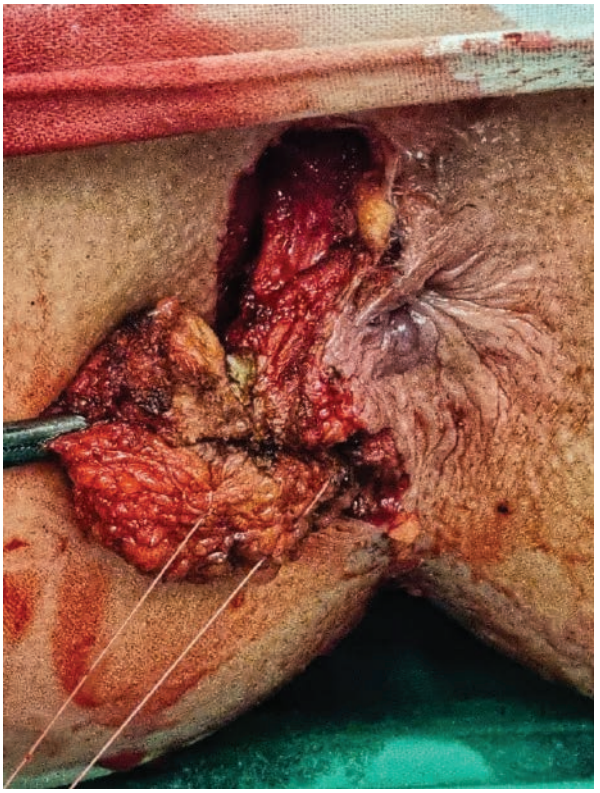

Рисунок 2. *Выделение опухоли промежностным доступом*
Figure 2. *Tumour dissection by perineal access*

with clear, uneven contours, with dimensions 29 × 11 × 22 mm, the surrounding soft tissues were without edema. Regional lymph nodes were not enlarged, their structure was preserved.

Total video colonoscopy revealed no organic pathology. According to the results of ultrasound of internal organs, overview radiography of the lungs, gastroduodenoscopy, clinical and laboratory indicators, there were no data for oncological pathology of other sites, the presence of distant metastases. To clarify the genesis of the tumor, a trepanobiopsy of the tumor was performed with subsequent cytological and immunohistochemical tests.

Cytology: there were no atypical cells, elements of inflammation, cells of flattened epithelium without features.

In the conditions of the Republican Clinical Oncological Dispensary of the Ministry of Health of the Republic of Tatarstan named after Professor Sigal M.Z., a histology of the punctate was performed: the biopsies were predominantly fibrous tissue, in the fibrous layers there were "nests" of cells with rounded or oval nuclei, abundant bright eosinophilic granular cytoplasm. Morphological picture of a granular cell tumor.

To clarify the diagnosis, an immunohistochemistry (IHC) was performed. A diffuse positive cytoplasmic reaction with antibodies to S100 was noted in tumor cells. The tumor was positive for CD8, vimentin and negative for GFAP. The expression of the Ki67 protein was 2%. Conclusion: the phenotype corresponds to a granular cell tumor.

Taking into account complaints, clinical picture, results of instrumental and laboratory tests, a decision was made on surgery.

On April 5, 2023 excision of the neoplasm of the perianal region by perineal access was performed. A semilunar pararectal incision was made under subarachnoid anesthesia from 7 to 11 o'clock. Acute mobilization of a 40 × 30 mm tumor emanating from the wall

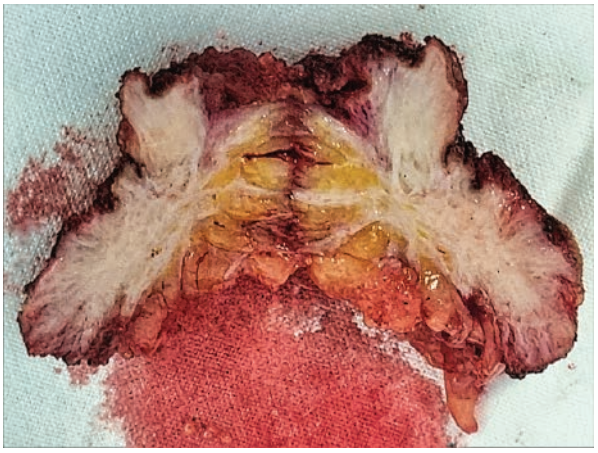

Figure 3. *Removed specimen*

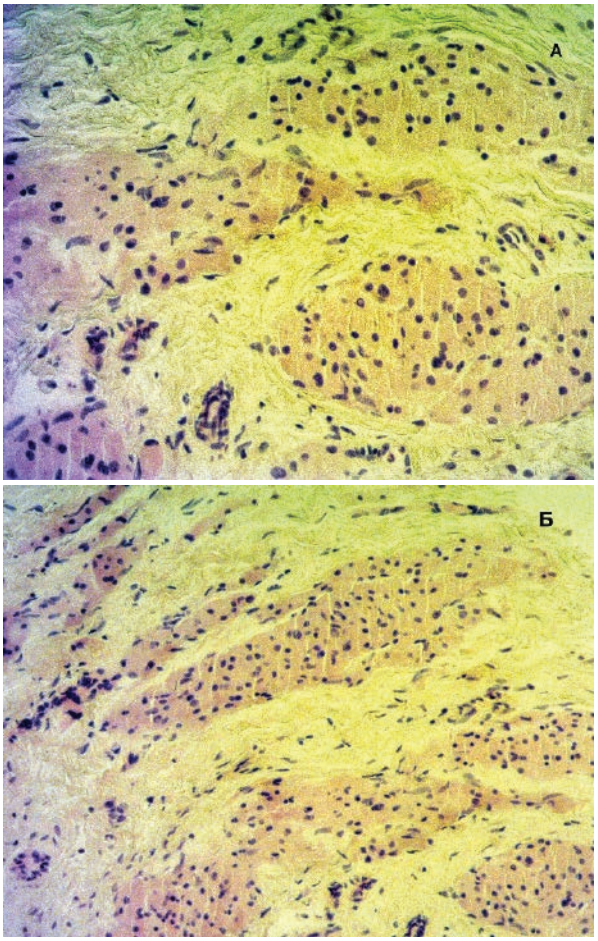

Figure 4. *There is an accumulation of large polygonal cells with centrally located, predominantly oval nuclei and visible nucleoli. Photo A. Magnification ×600, hematoxylin-eosin. Photo B. Magnification ×400, hematoxylin-eosin*

of the low rectum, intimately adjacent to the anterior portion of m. levator ani (Fig. 2), the tumor was removed. The defect of the muscle layer of the rectal wall was restored by nodular sutures with Polysorb 4–0 threads. The perineal wound was partially sutured, tamponade was with an ointment cloth. Aseptic dressing. Macropreparation (Fig. 3): tumors with dimensions of 40 × 30 mm, without an obvious capsule, with radial spread of the tumor into the adjacent adipose tissue. The section was white-gray in color with a dense consistency, with a knobby surface.

Postoperative pathohistology: clusters of large polygonal cells with centrally located, predominantly oval nuclei and visible nucleoli were determined on the slices. Clusters of tumor cells were separated by thin layers of connective tissue (Fig. 4). Rare vessels. The capsule was not detected. There were no hemorrhages and necrosis. Conclusion: granulocellular tumor.

In the laboratory of the oncological dispensary, the finished preparations from the sample of the removed tumor were revised. Microscopy revealed columns of soft tissues with groups of large cells with eosinophilic pale pink cytoplasm and a small hyperchromic nucleus. Infiltration by cells with eosinophilic pale pink cytoplasm and a small hyperchromic nucleus to the hypoderm and exit into it was visualized in the dermis. The histological picture corresponds to a granular cell tumor (Abrikosov's tumor).

The postoperative period was without features. On the 5th day, the patient was discharged from the hospital in a satisfactory condition. During the control examination on the 60th day after the surgery, the wound healed by secondary tension; held stool, gas. The patient remains under dynamic supervision.

DISCUSSION

Despite the fact that Abrikosov's tumor belongs to rare benign neoplasms, the urgency of the problem is associated with the difficulties of the initial stage of diagnosis, the recurrence of the tumor after its removal, the risk of malignant transformation and metastasis. According to literature data, tumor recurrence, detection of metastases varies from 3 to 38 months [2]. In the gastrointestinal tract, granular cell tumors are most often localized in the oral cavity and esophagus, and in the large intestine, mainly in the right colon.

AT of the perianal region and rectum can mimic fibrous post-inflammatory changes as a result of perianal infection. Vered, M. et al. (2009) suggested that granular cell tumors may be the result of a reactive lesion reflecting local metabolic or reactive changes, rather than a true neoplasm [10]. AT is often asymptomatic in the early stages of the disease. As it grows, there are complaints of pain, a feeling of discomfort in the tumor area. The progression of symptoms and the growth of neoplasm according to clinical examinations and instrumental research methods may indicate the malignant nature of the tumor. To solve this issue, it is advisable to use trepanobiopsy of the tumor with cytological, histological and immunohistochemical studies. The presence of staining in the IHC study of the soft tissue tumor marker vimentin, as well as the proteins S100, CD68, NSE, CD56 and EMA allows the diagnosis of a granular cell tumor. Evaluation of the expression of the Ki-67 marker allows, albeit indirectly, but to draw conclusions about the malignancy of the tumor with an increase in this indicator. Thus, an increase in the Ki-67 index of more than 10% is considered an unfavorable prognostic factor of tumor recurrence and metastasis. At the moment, there are no strict recommendations for surgical treatment of AT with colorectal site. It is important to note the need for a multidisciplinary approach to the treatment of patients

with AT and subsequent dynamic follow-up of patients for 3 years with control radiation and endoscopic examinations. Considering the variant of multifocal location of granular cell tumors, colonoscopy should be supplemented with esophagogastroscopy.

CONCLUSION

Diagnosis and surgical treatment of patients with rare site of granular cell tumors is possible in the conditions of the coloproctology unit of a multidisciplinary hospital, with sufficient competence of surgeons in the diagnosis and treatment of this form of tumors and good material and technical equipment of the clinic. Check up in the preoperative period, in addition to complex radiation diagnostics, should include performing an immunohistochemistry of removed neoplasm to verify the diagnosis, determine the level of mitosis and decide on treatment approach in case of tumor malignancy. After removal of the Abrikosov's tumor, patients need follow up, as well as patients with oncological disease, for 3 years with control.

AUTHORS CONTRIBUTION

Concept and design of the study: *Rais R. Shakirov, Oleg Yu. Karpukhin*
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