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Malignant triton tumor of the rectum – A case report and review of the literature

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ABSTRACT

INTRODUCTION: Malignant triton tumors (MTT) are rare but highly aggressive tumors that originate from the Schwann cells. These tumors can occur in any part of the body, mostly present late and carry poor prognosis.
PRESENTATION OF CASE: We present a 24-year-old man with a rectal MTT causing non-specific abdominal pain and recurring ileus. The MRI showed a rectal mass near the urinary bladder with compression on the seminal vesical. A complete surgical resection of the tumor was performed. The immunohistological report confirmed a rectal MTT. Because of persistent ileus during the post-operative palliative chemotherapy, another tumor debulking was performed. The patient died 9 months after the diagnosis of MTT due to local recurrence under chemotherapy.
DISCUSSION AND CONCLUSION: MTTs are uncommon tumors in young age with high morbidity and mortality because of local recurrence also after complete resection.

1. Background

Malignant triton tumors (MTTs) are extremely rare subtypes of malignant peripheral nerve sheath tumors (MPNST) [1]. The reported incidence of sporadic MPNST and MTT is 0.001% [2,3]. MTT account for approximately 5% of all MPNSTs [4], and predominantly originate in the head, neck and extremities [2] with rare occurrence in the mediastinum, abdominal cavity and retroperitoneum [5]. MTT in the colorectal region is even rarely reported [6]. The presentation of MTT is always vague as the tumor originates in diverse locations. When advanced, the abdominal MTT manifests with non-specific pain and compressive symptoms [7].

Despite the availability of master-class diagnostic tools and consensus guidelines in surgical oncology for most of abdominal tumors, no standard management plan for MTT is available in literature [8–10]. Nevertheless, some recommendations have been proposed [11]. Furthermore, the prognosis of MTT is poor as reported by a 5-year survival rate of only 5 to 15% [12]. Literature has shown that the prognosis of MTT is influenced by its location (head, neck and extremities tumors carry better prognosis than the buttocks, retroperitoneal and central nervous system tumors), tumor size, degree of differentiation, tumor free resection margins, and Ki67 labeling index [13].

We present a case of an MTT of the rectum in a young adult male. To our knowledge, we are reporting first case of the rectal MTT in adults in the literature. This report highlights non-specific manifestations of the rectal MTT that invariably becomes refractory to treatment.

Written informed consent for publication was given and the case report is presented in line with the SCARE criteria [14].

2. Case presentation

A 24-year old male, without significant medical history, initially presented to a urologist with progressive lumbar, inguinal and scrotal pain since two month. Physical examination showed a distended abdomen with no evidence of scrotal tumor or inguinal lymphadenopathy. There were no clinical or laboratory abnormalities. Ultrasound showed a cystic mass dorsal to the urinary bladder, in close proximity with the rectum. For further diagnostics, a subsequent MRI was performed that confirmed the ultrasound findings. The suspected differential diagnosis included a lymphoma or sarcoma (Fig. 1). There was no infiltration of the urinary bladder or lymphatic node enlargement.

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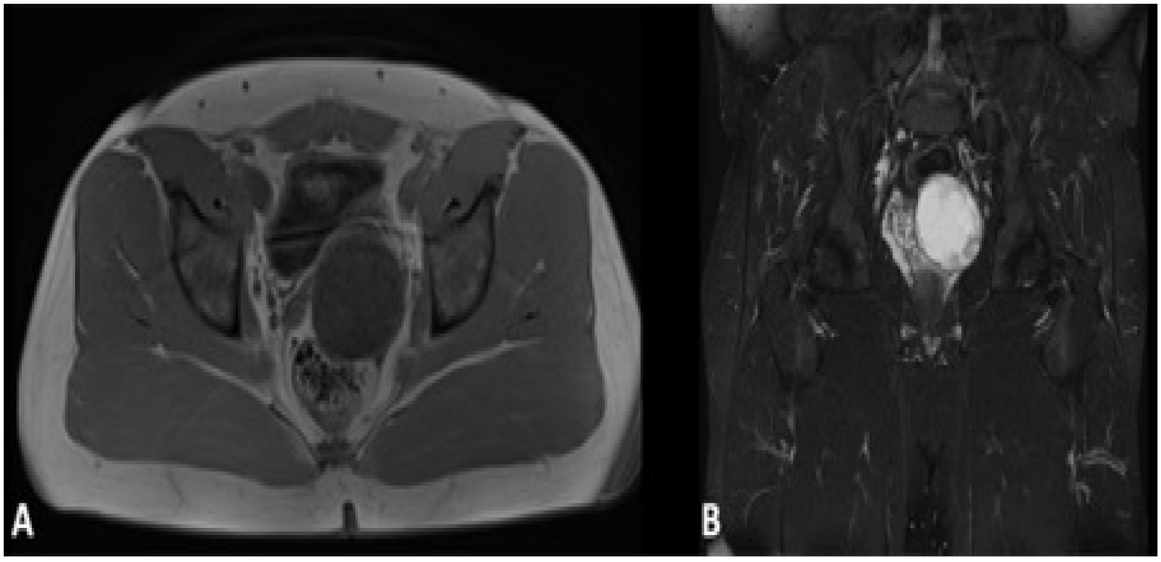

Fig. 1. Initial MRI of the pelvis. A: Axial T1, B: Coronal T2. T1 hypo intense and T2 bright/hyperintense well-defined mass arising from the left anterior rectal wall with luminal narrowing. The mass has peripheral areas of hypointense signals highlighting a more fibrous nature. There is a marked displacement of the bowel loops with a moderate amount of compression on the posterior bladder wall. No local infiltration is evident.

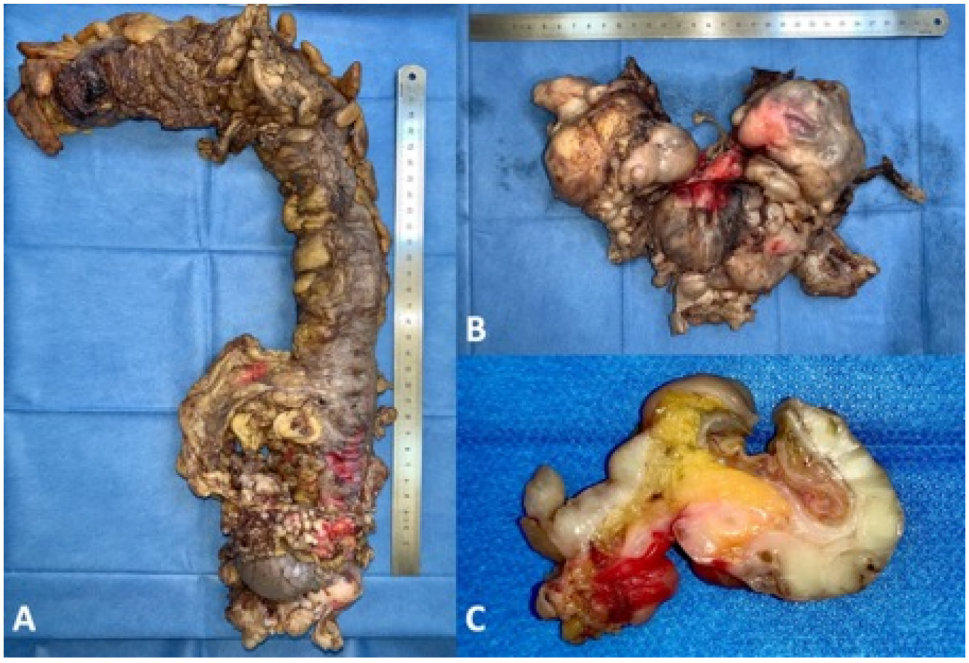

Fig. 2. Macroscopic appearance of the tumor after initial resection. A showing left colon with rectum and cystic lesion at the rectum.

With these findings, the patient then presented to the surgical clinic, where further staging examinations including CT scan were performed. There was no evidence of local or distant metastasis. A subsequent colonoscopy showed that the mass obliterated the rectal lumen without mucosal infiltration. Following the case discussion in the interdisciplinary tumor board (IDT-board), an open transabdominal lower rectal resection with Hartmann's resection was performed by a senior colorectal consultant surgeon. Macroscopically, a 9.5 cm tumor was localized on the posterior surface of the rectum without mucosal invasion. The tumor showed a capsular sheath with focal white fleshy areas of hemorrhage and necrosis (Fig. 2).

The histological examination of the resected tumor showed a heterogeneous differentiated mass with spindle, focally wavy shaped cells without a specific growth pattern. The tumor cells had abundant eosinophilic cytoplasm, irregular hyperchromatic nuclei and distinct nucleoli. The tumor mass displayed focal and central small peripheral nerve sheets. The individual tumor cells stained immunohistochemically for S100 and SOX10 and focally for NF. There was strong positivity for Desmin and MyoD1 as well as strong nuclear positivity for p53 in most of the tumor cells (Fig. 3). MDM2 was only focally and weakly positive, while no reactivity for CDK4, DOG-1, CD117 and beta-Catenin was found.

Based on the histological and immunostaining profile, a diagnosis of MPNST with rhabdomyoblastic components, also known as MTT was established.

Following surgery and further discussion in the IDT-board and with a team from the center of excellence for sarcoma, pallia-

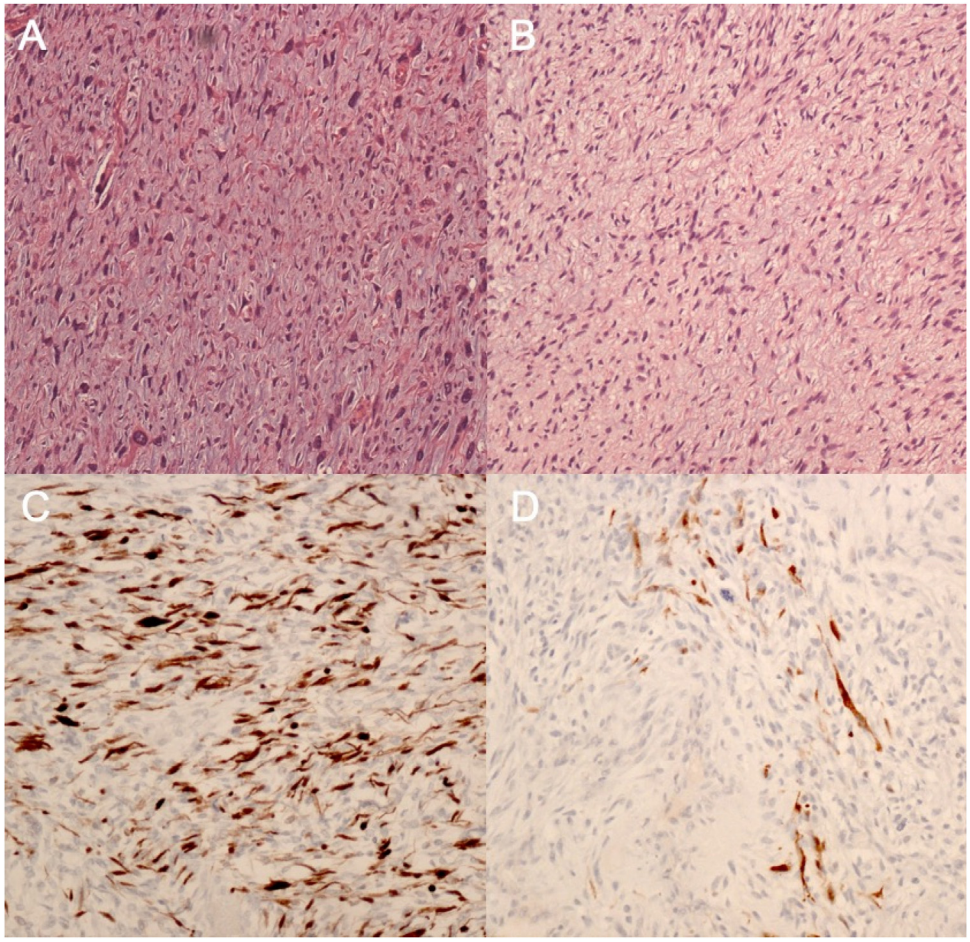

Fig. 3. Histopathological examination of the tumor. **A**: Image of hematoxylin and eosin (HE) staining with highly atypical tumor cells with abundant eosinophilic cytoplasm, irregular and hyperchromatic nuclei. **B**: Image of HE staining showing malignant spindled cells with elongated, slightly anisomorphic nuclei and wavy-shaped cytoplasm. **C**: Immunohistochemical analysis revealed positivity of the tumor cells for desmin. **D**: Immunohistochemical analysis for S100 showed individual tumor cell positivity.

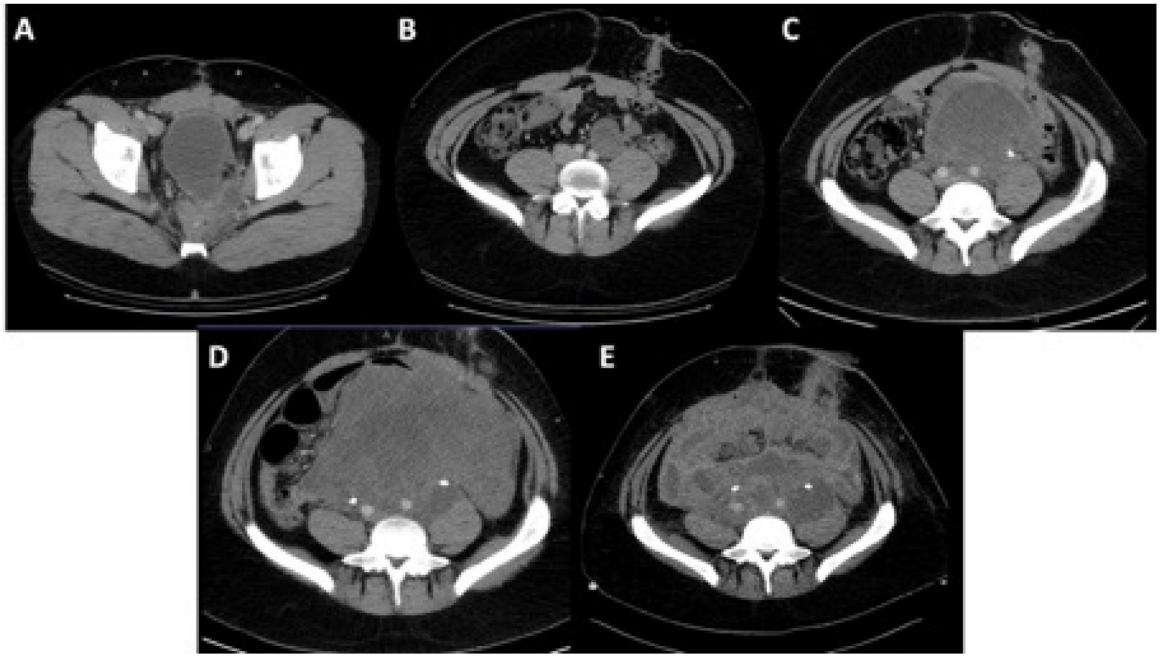

Fig. 4. CT scan of the abdomen. **A:** Primary tumor near the urinary bladder. **B–E** showing almost same level (near stoma) at different phases of treatment. **B:** after initial resection. **C:** during radiation. **D:** Ileus before second surgery. **E**: After second tumor reduction.

Table 1
Reported cases of malignant triton tumors in the gastrointestinal tract.

Organ	Age (years)	Sex	Therapy	Outcome	Literature
Rectum	24	Male	Surgery, + multimodal treatment	Died 9 month after first symptoms	Current case
Duodenum	49	Female	Surgery	Alive, 8.5 years after surgery	Asahi et al. 2018 [7]
Rectum	11 months	Male	Surgery	Unclear, alive 3 months after surgery	Ellison et al. 2005 [23]

tive chemoradiation with two cycles of Doxyrubicin and Ifosfamid was administered. This was followed by radiation and two further cycles of chemotherapy. However, during the first follow-up, CT scan of the abdomen reported a huge abdominal mass (13 × 18 cm) occluding the stoma with resultant ileus (Fig. 4). Subsequently, another tumor debulking was performed that included total colectomy and partial resection of the terminal ileum with a terminal ileostomy. Afterwards, a percutaneous nephrostomy was performed on both sides because of progressive compression on the ureters. The patient made a good initial recovery and was discharged 9 days after surgery with initiation of new palliative chemotherapy. The new chemotherapy followed a protocol of Irinotecan, Dacrabazin, Vincristine and Temozolomid. After a promising early response, the tumor grew again and led to a paraneoplastic iliac vein thrombosis and stomal occlusion. The patient died 9 months after the first diagnosis of the rectal MTT under palliative care.

3. Discussion

Until today, less than 200 cases of MTT have been reported in the literature worldwide [13,15]. Furthermore, in 1932, Masson et al. have reported the first case of MPNST, which appeared as encapsulated tumor of the peripheral nerves such as schawannomas and neuromas [16]. Our patient had MPNST with rhabdomyoblastic components, classified as MMT, which was refractory to surgical and chemotherapeutic therapies. Most patients with MTT present with vague manifestations that can hardly alert the physicians about their aggressiveness and devastating effects. As detailed, our case presented with non-specific lower abdominal pain and rectal compressive symptoms that rapidly worsened despite aggressive management and the patient eventually succumbed to death.

MTT is an aggressive tumor with a rate of distant metastases to be as high as 48% and a local recurrence rate of 43% [2,17]. MTT equally affects men and women [5], with a mean age of 31.7 years [18]. Our 24-year old patient was younger than the reported age range. There is no influence of familial predilection in MTT and the same holds true for our patient as his past history was not remarkable of any familial correlation with this tumor. According to Woodruff et al., the diagnosis of MTT should meet three criteria [17]; first, the tumor originates from a peripheral nerve, in ganglioneuroma or in a patient with neurofibromatosis type I, or in a location typical for peripheral nerve tumors; second, the tumor demonstrates growth characteristics of Schwann cells; third, rhabdomyoblasts arise within the body of the peripheral nerve tumor. Recently, these criteria have been modified and now first criterion is not considered to be essential for the diagnosis of MTT [19]. In addition to the criteria proposed by Woodruff et al., currently immunostaining is used for histopathological diagnosis of MTT. In up to 50% of all MPNSTs staining for S-100 is positive. The positivity, as seen in our case, is typically focal [20,21]. Positivity for SOX10 is also found focally. In a study by Nonaka et al., marker SOX10 showed better sensitivity and specificity than S100 for the diagnosis of MPNST [22]. In order to make a diagnosis of a MTT, the presence of rhabdomyoblasts within an otherwise ordinary MPNST is crucial. In case with equivocal results, immunostaining for Desmin and MyoD1 can complement other tests.

So far, the English literature has shown only two other cases of MTT in the gastrointestinal tract as detailed in Table 1 [7,23]. These reports have advocated that so far the best treatment for MTT is an enbloc surgical resection, which is also a mainstay of treatment as for all cases of soft tissue sarcoma. This approach has shown a correlation with lower rates of metastasis or local recurrence [24]. Because of the rarity of MTT, no standardized guidelines are available that can help physicians in dealing with such cases. Neither chemotherapy nor radiotherapy have been proven to be effective [2,25]. A repetitive tumor debulking surgery in combination with multimodal treatment is shown to be associated with longer survival [26]. Unfortunately, a complete surgical resection with disease-free margins and multimodal management could not improve outcome of our case as local recurrence developed soon after the initiation of multimodal treatment.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

This case reports the outcome of a standard treatment. Ethical Committee approval was not taken.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author's contribution

Dr. Jonas Herzberg: Conceptualization, Methodology, Investigation, Writing - Original Draft.
Dr. Gianluca Maria Corradini: Investigation, Visualization.
Dr. Cay Uwe von Seydewitz: Methodology, Supervision.
Prof. Dr. Salman Yousuf Guraya: Writing - Review & Editing, Investigation.
Prof. Dr. Tim Strate: Project administration, Supervision.
Dr. Human Honarpisheh: Writing - Review & Editing, Project administration, Supervision.

Registration of research studies

Case report not registered, as it is not first in man study.

Guarantor

Prof. Dr. med. Tim Strate

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