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Case Report

Meckel's diverticulum complicated with gastro-intestinal stromal tumor: Case report

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KEYWORDS

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Abstract *Introduction:* Meckel's diverticulum is a common congenital anomaly, mostly asymptomatic. Tumors may arise rarely in these diverticulae. We claim presenting a new problem to the medical staff in Egypt.
Case presentation: We report a case of a 49 year old male patient who attended our center with pelvic mass insinuated between the bladder and the rectum. On exploration the mass was found arising at the tip of a Meckel's diverticulum, Gastro-intestinal stromal tumor (GIST) was confirmed by pathology.
Discussion: In review of recently published cases most of these tumors were presented with vague abdominal pain as in our case. Tumors were treated by resection with or without adjuvant Imatinib.
Conclusion: Surgeons and oncologists should bear in mind this rare diagnosis and know how to treat it.

Introduction

Meckel's diverticulum is the commonest congenital anomaly of the gastrointestinal tract. In spite of this, neoplasms rarely develop within Meckel's diverticulae [1]. Resection of asymptomatic Meckel's diverticulae remains a controversy. Our case raises suspicion that observational management of asymptomatic Meckel's diverticulae may leave some clinically occult diverticular neoplasms untreated. In our report we document a case of GIST arising at the tip of an accidently discovered Meckel's diverticulum in a middle aged male. To our knowledge it is the first report of such case in Egypt.

Case presentation

A 49-year-old man presented to our department complaining of vague pelvic pain and frequent micturition for about 3 months. CT revealed a well defined pelvic soft tissue mass 8×9 cm between the urinary bladder and the rectum with cystic degeneration (Fig. 1a). Complementary TRUS (Trans Rectal Ultrasonography) with biopsy shows spindle cell proliferation mostly neurogenic. No relevant finding was detected on clinical examination.

Our panel decided exploration for this retrovesical mass with provisional diagnosis of neurogenic tumor (Schwannoma or malignant peripheral nerve sheath tumor). During laparo-

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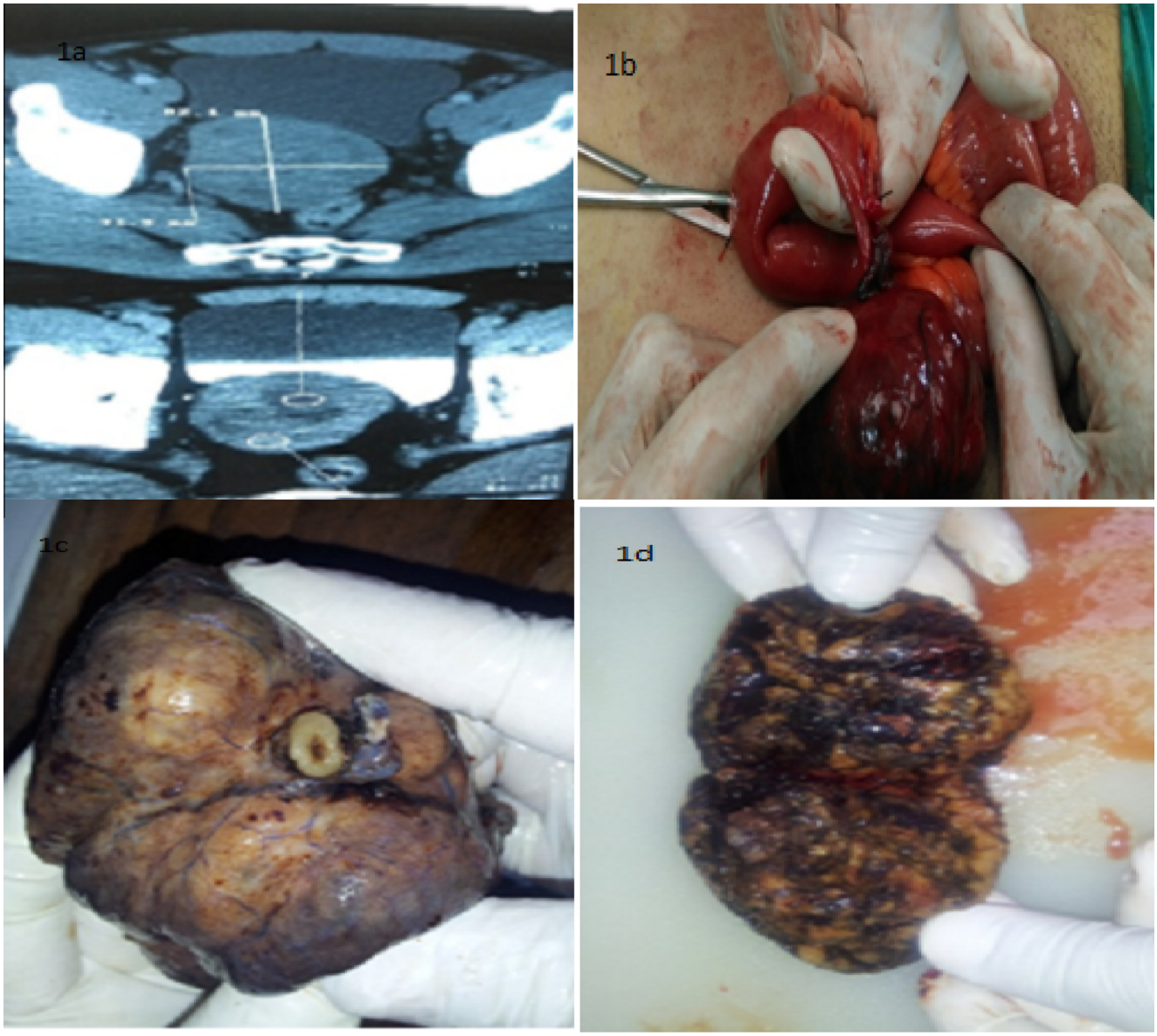

Figure 1 (a) CT showing solid mass insinuated between the full bladder and the rectum. (b) Mass on the tip of Meckel's diverticulum after ligation of its vascular pedicle. (c) Patent lumen of the diverticulum seen within the mass. (d) Cut section of the mass showing fleshy tumor.

tomy, a well defined soft tissue mass was identified at the antimesenteric ileal border 80 cm from the ileocecal valve (Fig. 1b). No other intra-abdominal lesions were found. Diverticulectomy and closure of the intestinal wall in two layers horizontally was performed as the tumor was well capsulated and away from the base of the diverticulum.

Grossly, the resected diverticulum was 1.8 cm in the maximal dimension. At the diverticular tip, a 7×6 cm oval-shaped fusiform lesion was found extending along the curvature of the diverticular wall (Fig. 1c and d). The lesion was brownish and had cystic changes.

Microscopically, a GIST arising from a Meckel's diverticulum was diagnosed by Hematoxylin and Eosin (Fig. 2a and b) and immunohistochemical stains (IHC) (Fig. 2c–e). Final diagnosis was: GIST (high risk), size >5, mitoses $>5/50$ HPF, confirmed by strong positivity for CD34, moderate CD117 positivity.

Patient was discharged from hospital after 3 days with no unforeseen events. After the removal of stitches and revision of pathology, our hospital multidisciplinary team recommended adjuvant Imatinib 400 mg daily and follow up CT every 6 months.

Discussion

Incidence of Meckel's diverticulum ranges from 0.6% to 4% [2,6]. Meckel's diverticulae are clinically occult unless complications occur. Only about 10% of Meckel's diverticulae produce symptoms, mainly due to intussusception and

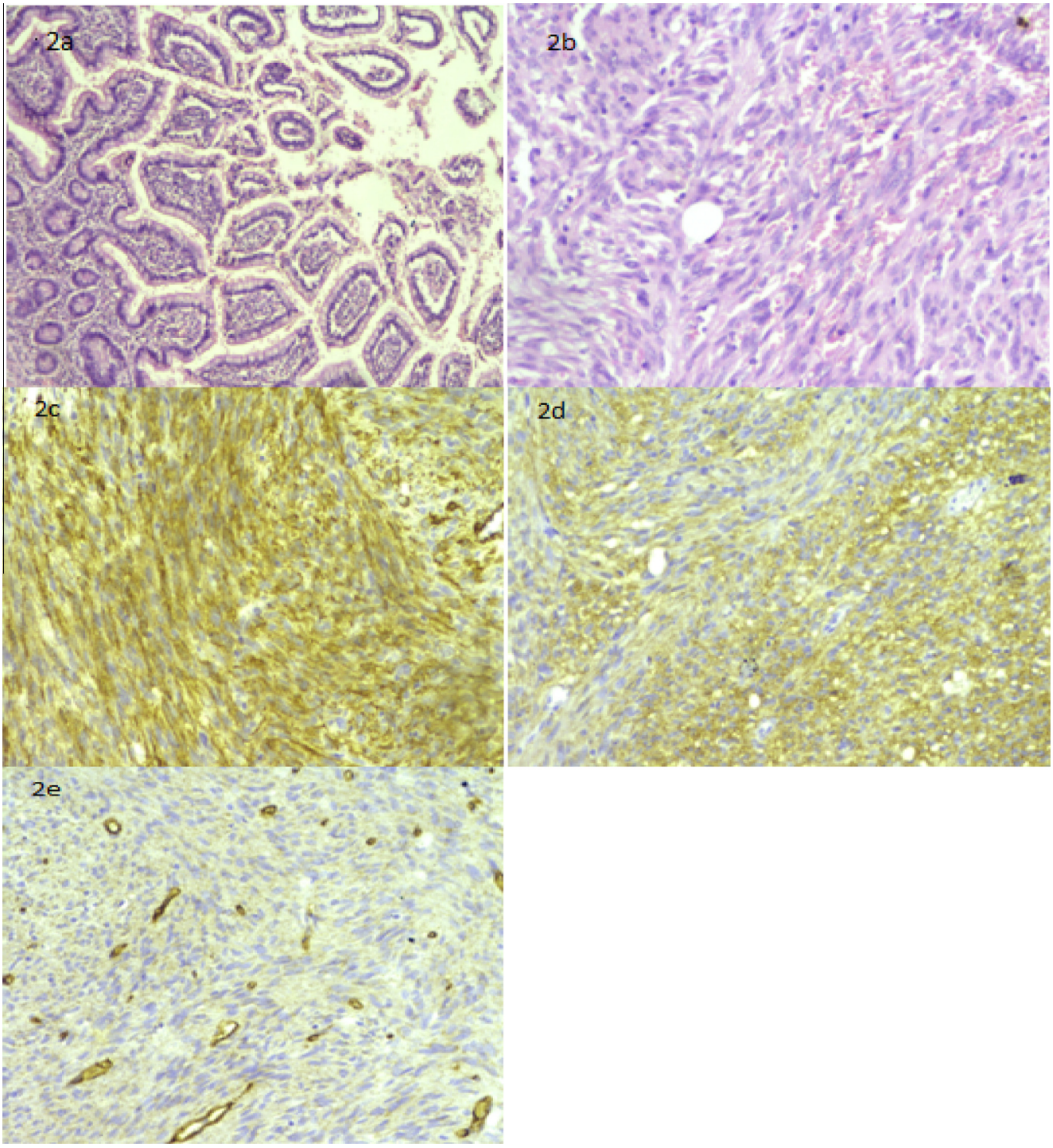

Figure 2 (a) Small intestinal mucosal lining of Meckel's diverticulum (H&E 100×). (b) Tumor formed of spindle cell proliferation arranged in long bundles. The cells exhibited plump nuclei with moderate pleomorphism. No evident abnormal mitotic activity or necrosis (H&E 200×). (c) Tumor cells showed strong cytoplasmic and membranous staining for CD34 (200×). (d) IHC for CD117 showed moderate positive cytoplasmic and membranous staining confirming the diagnosis of GIST (200×). (e) Tumor cells were negative for smooth muscle actin (SMA) (200×).

ulceration [2,4]. A neoplasm is a rare complication detected in 0.5–3.2% of Meckel's diverticulae [7–10]. Carcinoid tumors are the commonest primary diverticular tumors (about 40%), followed by leiomyosarcoma (about 22%) then adenocarcinoma (about 14%) [11]. GISTs represent only 12% of primary tumors arising in Meckel's diverticulae [7,12].

GISTs are the commonest mesenchymal tumors of the gastrointestinal tract [5,13]. The stomach is the most frequent site

(70%), followed by the small intestine (25%). About 13% of small intestinal GISTs are incidentally discovered during the management of other diseases [5,11]. A few syndromes are associated with the development of GISTs, as neurofibromatosis type 1 (Von Recklinghausen disease) and Carney's triad [5,11,13]. The clinical behavior of a GIST is related mainly to its mitotic activity and size where frequent mitoses and big size indicate higher malignant potential [5]. Under the microscope, most GISTs appear as spindle cell tumors with a storiform pattern. Epithelioid, signet ring cells, and what is called gastrointestinal autonomic nerve tumor variants may be detected in some cases. Positive CD117 and DOG1 stains are diagnostic immunohistochemical stains for GISTs [5,13].

Our patient had a Meckel's diverticulum complicated by a GIST. This rare combination has been reported in the literature [11,12,14–17]. However, our case was unique in the surgical management by just simple diverticulectomy instead of resection anastomosis.

The standard treatment for complicated Meckel's diverticulae is surgical resection; however, the management of silent Meckel's diverticulae is controversial [2]. Some authors suggested that asymptomatic Meckel's diverticulae should not be removed except if the patient is at an increased risk of developing complications. The risk factors are male gender, young age, diverticulum size >2 cm, and presence of heterotopic tissue [4,18]. This attitude toward conservation was magnified by results of other studies showing that morbidity following prophylactic diverticulectomy exceeds the lifetime complication risk of Meckel's diverticulum (9% versus 4.2%) [10]. However, since operation related complications have decreased recently, many authors accept prophylactic diverticulectomy for asymptomatic patients [16,17,19,20]. In contrast to the significant morbidity of up to 33% with complicated Meckel's, the postoperative morbidity with prophylactic diverticulectomy ranges between 0% and 6% [2].

No randomized trials addressing systemic therapy for diverticular GISTs were found but treatment can be extrapolated from treatment regimens of GISTs occurring in stomach and small intestine. Imatinib is a tyrosine kinase inhibitor which is approved for the treatment of GISTs. Imatinib may be used as adjuvant therapy in patients at a high risk of disease recurrence after lesion resection as in our case [3]. Although there is no evidence of improved survival with the use of Imatinib as a neoadjuvant therapy, it is commonly used for borderline resectable cases aiming to make surgery more feasible and/or less morbid.

Our report represents the first of such case in Egypt, we think that malignancies overlying Meckel's may be an underestimated problem due to lack of registration. Also we open another important issue for further research regarding managing such cases with simple diverticulectomy rather than segmental resection anastomosis, where in our case we could attain negative safety margin with such approach with less operative time and probably lower incidence of leak.

Conclusion

Tumors on an otherwise asymptomatic Meckel's diverticulum can occur; accurate frequency is not well established. GIST in this site is even rarer and usually misinterpreted as bladder or colorectal mass. Identifying these cases and managing with simple diverticulectomy seem curative.

Consent

Patient consent was obtained and is available for revising if needed.

Author's contributions

AF developed the idea of the paper and wrote most of the article. IH shared in writing and final formatting of the paper. SA revised the article scientifically and lingually. NM was responsible for writing, analyzing and revising pathological aspects.

Conflict of interest

The authors claim no conflict of interest.

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