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An unusual cause of hemoperitoneum: case report with review of literature

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ABSTRACT

Spontaneous hemoperitoneum (SH) is a rare entity which can be life-threatening. Gastrointestinal neurofibromas are rare. Occasionally, such lesions may be the initial sign of NF1 in patients without any other clinical manifestations of the disease. The clinical presentations of isolated neurofibromatous lesions of the intestines are variable. In asymptomatic patient no treatment may be required and patient may be kept on follow up. Occasionally, they manifest with complications such as intestinal bleeding, obstruction or perforation. Surgery is the treatment of choice in symptomatic intestinal neurofibroma. We present the case of a 55 year-old male with acute abdomen due to rupture of isolated neurofibroma as one of the cause of SH. This case represents a rare presentation of isolated intestinal neurofibromatosis in a patient without systemic manifestations and highlights the need for high index of suspicion to exclude neurofibromatosis type 1 or multiple endocrine neoplasia type 2b.

1. Introduction

Spontaneous hemoperitoneum (SH) is a rare, but life-threatening condition of non traumatic etiology. It usually occurs in patients with coagulation abnormalities as a result of hepatic, splenic, vascular or gynaecologic pathology [1]. It typically presents with signs of acute intraperitoneal bleeding and occasionally abdominal compartment syndrome in severe cases. Non traumatic liver pathology such as hepatic adenomas, focal nodular hyperplasias, large hemangiomas, are considered as the most common cause of SH, when gynecologic causes are not considered. Malignant hepatic lesions, either primary or metastatic, may also rupture spontaneously. These disorders are usually associated with anticoagulation, pregnancy as triggering factors. Spleen is the second most common solid organ to give rise to SH followed by vascular causes which includes aneurysms, pseudo-aneurysms, or arterial dissection. Endoscopic technique like Angiography and embolization almost always constitute first-line therapy in the hemodynamically stable patient. Surgery should be considered in those in whom interventional techniques have failed to control the bleeding, or patient has unresponsive hemorrhagic shock [2].

Hemorrhagic shock can be rapidly fatal. The primary goal is to stop the bleeding. Resuscitation may well depend on estimated severity of hemorrhage. Patients who have moderate hypotension from moderate bleeding may well benefit from a delay in massive resuscitation in order to reach a definitive care facility. When patients are obviously in severe hemorrhagic shock, the use of intravenous crystalloids or colloids and blood products can be life saving. A hemoglobin level of 7–8 g/dl is an appropriate threshold for transfusion in critically ill patients with no risk factors for tissue hypoxia. Moreover, hemoglobin concentration should not be the only therapeutic guide in actively bleeding patients. Instead, therapy should be aimed at restoring intravascular volume and adequate hemodynamic parameters [3].

We present here with a rare case of hemoperitonium due to rupture of large pedunculated neurofibroma arising from ileum in a patient without features of associated systemic syndrome.

2. Case report

A fifty five year old male patient presented to the Casualty with features of acute abdomen and non passage of flatus and stool for one day. There was past history of intermittent pain abdomen and passage of black tarry stool. On examination, patient was hemodynamically unstable. Abdomen was moderately distended having diffuse tenderness. There was no guarding or rigidity and liver dullness was not masked. His hemogram showed haemoglobin of 7.3 g/dl. Total and differential counts were within the normal range. Blood biochemistry revealed raised blood urea (125 mg/dl) and serum creatinine (2.9 mg/dl).

Plain X-ray abdomen showed dilated small bowel loops without any free air under diaphragm. Ultrasonography of abdomen

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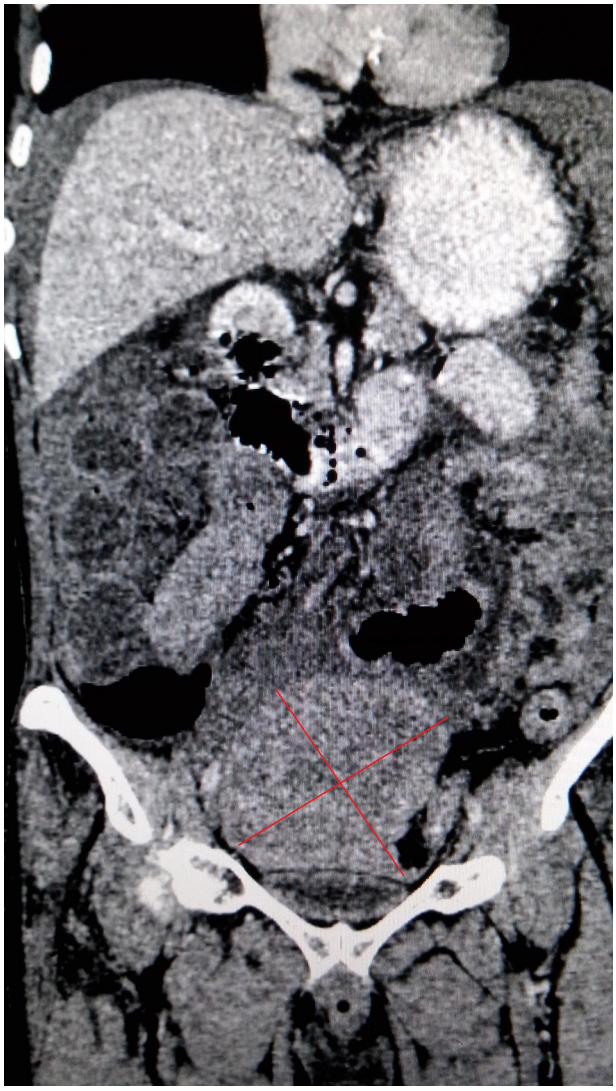

Fig. 1. Photograph of CT-scan abdomen showing diffuse echogenic collection and clot in the pelvis.

revealed gross echogenic collection with an intraperitoneal heterogeneous mass of 11 × 7 cm. Diagnostic tap was hemorrhagic in nature. Hypotension was managed with intravenous fluids and ionotropic drugs.

On resuscitation patient pulse came down to 84 beats per minutes and blood pressure of 110/70 mm Hg without ionotropic support and haemoglobin was 6.9 g/dl. Contrast enhanced tomography revealed moderately dilated fluid filled small bowel loops with pneumo-peritonium and moderate ascites with organized clot in pelvis (Fig. 1). Exploratory laparotomy was done after optimizing the patient. On laparotomy around 1.5 litres of haemorrhagic fluid and a fist sized clot was removed from peritoneal cavity. A partially ruptured 10 ×8 cm pedunculated mass arising at antimesenteric border of the ileum around 4 feet proximal to ileocaecal region was found (Fig. 2). The mass was twisted at its base by one complete turn and a sealed ileal perforation was seen at the base. As patient had low hemoglobin (Hb-6.9 g/dl) and low total protein and albumin (Total Protein/Albumin = 4.5/1.8 g/dl), hence a double barrel ileostomy was constructed after resection of the affected segment (20 cm approximate) of ileum. Postoperative recovery was uneventful. Patient was discharged on day seven. Histopathology report confirmed the mass to be a neurofibroma. Immunohistochemistry for CD-117 done which was also negative. After 6 weeks of follow-up stoma has been reversed.

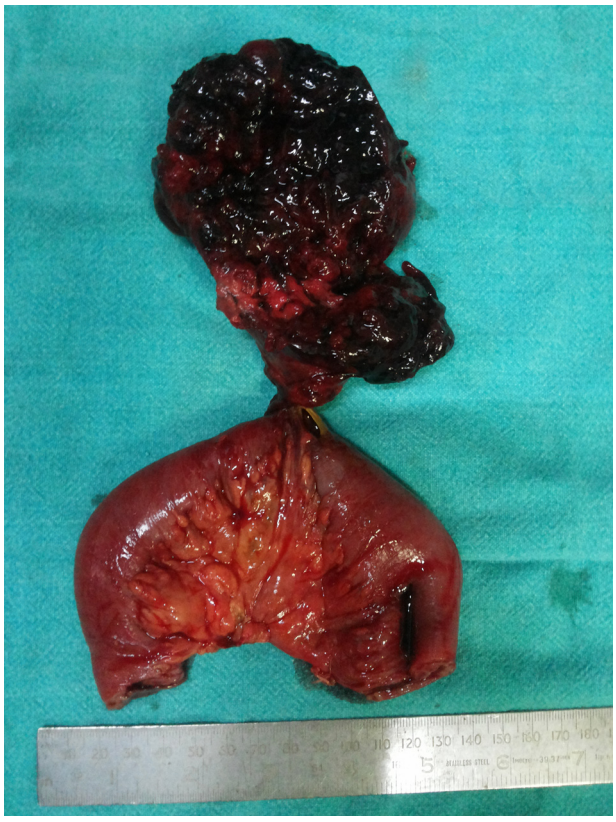

Fig. 2. Photograph of resected specimen of ileum with ruptured neurofibroma.

3. Discussion

Benign primary tumors of the small intestine are quite unusual, comprising approximately 3% of all the gastrointestinal tract neoplasms [4]. They are usually asymptomatic. Occasionally, they manifest with complications such as intestinal bleeding, obstruction or perforation.

Gastrointestinal tract neurofibromas originate from either the plexus of Meissner in the submucosa or the plexus of Auerbach in the muscularis propria or even from the serosa [5]. These lesions are often sessile and wide-based but pedunculated polyps have also been observed.

Neurofibromas are usually multiple upon presentation and are usually part of two autosomal dominant disorders with variable penetrance: Neurofibromatosis type 1 (NF1, von Recklinghausen's disease) and Neurofibromatosis type 2 (NF2, central or bilateral acoustic neurofibromatosis) [6]. Isolated neurofibromatous lesions in the gastrointestinal tract without associated systemic syndromes are a rare entity.

The first documented case of intestinal neurofibromata was reported by Albert Branca. A similar case was subsequently reported by Leriche in 1911. In one study only 18 cases of neurofibroma were found in the medical literature over a 20 year period. Of these 12 were benign and six were malignant. All of these cases, with the exception of one, were of the solitary type [7]. An isolated ileal neurofibroma has been reported by Watanuki et al. in 1995 and Al-Harakea et al. in 2013 and in both cases the patient presented with an ileocolic intussusceptions [8,9].

Gastrointestinal involvement in neurofibromatosis is uncommon and is seen in 25% patients. Gastrointestinal neurofibromas are

the most common abdominal neoplasms in NF1 but are rare in NF2. Neurofibromas have been found throughout the alimentary tract, most frequently in the jejunum followed by the stomach, ileum, duodenum and colon. Single or multiple neurofibromas occurring in the soft palate, oesophagus, stomach, gallbladder, common bile duct, small bowel and the mesentery, colon and the anal canal without associated systemic disease have been previously reported [10].

Neurofibromas and ganglioneuromas of the GIT have also been reported in multiple endocrine neoplasia (MEN 2b). However, it is rarely encountered as a separate pathologic entity. Isolated intestinal neurofibromatous proliferations may be the initial manifestation of NF1 or MEN 2b, juvenile and adenomatous colonic polyposis [11]

The clinical presentation of isolated neurofibromatous lesions of the intestines are varied and are dependent upon the focal or diffuse nature of the lesions, their location, their effect on gastrointestinal motility and their possible impingement on adjacent structures [11]. Patients may have abdominal pain, palpable masses, bleeding due to necrosis or ulceration of the mucosa, obstruction due to intussusception or extra-luminal pressure, diarrhoea and perforation. They can undergo malignant transformation in up to 5–15% of patients, especially in individuals over the age of 40.

Endoscopic biopsies are a mainstay of the diagnosis. But as neurofibromas arise deep to the mucosa and visceral neurofibromas are usually serosal, biopsies may be inconclusive. The primary therapeutic option of isolated neurofibroma of the intestines is surgical [12]. For asymptomatic and incidental findings during endoscopy, no treatment may be required and patient may be kept on follow up.

The indication for performing a primary anastomosis or an intestinal stoma has to be confirmed or negated for every individual case. Indications of intestinal anastomosis can be broadly divided into two categories: restoration of bowel continuity following resection of diseased bowel and bypass of unresectable diseased bowel. Intestinal anastomosis is contraindicated in conditions where there is high risk of anastomotic leak, such as severe sepsis, poor nutritional status like severe hypoalbuminemia, disseminated malignancy, viability of bowel in doubt and fecal contamination or frank peritonitis [13].

Isolated ileal neurofibroma with hemoperitoneum is a rare pathological entity. The clinical significance of such a diagnosis lies essentially in considering this as a possible cause of hemoperitonium as well as in the need of further follow up of these patients as the bowel involvement could be the first manifestation of neurofibromatosis type 1 or multiple endocrine neoplasia type 2b [9].

4. Conclusion

Spontaneous hemoperitoneum is a rare condition. Gastrointestinal involvement in neurofibromatosis is uncommon and majority are asymptomatic. Neurofibromas are usually multiple upon presentation and are usually associated with neurofibroma type 1 and type 2. There are very few case reports of patients with neurofibroma presenting with abdominal complications. Surgery is the treatment of choice in symptomatic intestinal tumours, although asymptomatic patients may be monitored conservatively. Follow-up of patient is mandatory to look for any syndromic association.

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Conflict of Interest

None.

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