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Primary liposarcoma of the sigmoid presenting as colonic intussusception - A case report

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ABSTRACT

Liposarcomas are malignant soft tissue neoplasms usually located in the retroperitoneum, head, neck and extremities. Although secondary invasion of the gastrointestinal tract by retroperitoneal sarcomas is frequent, primary involvement is uncommon. We report a young patient with Crohn's disease in remission presenting with intussusception of the colon due to a primary liposarcoma of the sigmoid. Emergency resection confirmed the diagnosis. The patient remains in remission after five years. This represents the youngest diagnosis of liposarcoma to date and the first occurring in the sigmoid. We also highlight the concomitant diagnosis of Crohn´s disease.

Key words: Liposarcoma. Colon intussusception. Crohn's disease. Soft tissue sarcoma.

INTRODUCTION

Soft tissue sarcomas are uncommon malignancies in the adult (less than < 1%) and about 15% are located in the retroperitoneum (1). Liposarcoma is the most common variant accounting for up to 20% of all soft tissue sarcomas and over 50% of retroperitoneal sarcomas (1,2). They are usually found in the extremities, retroperitoneum and, less often, in the head and neck area. Involvement of the gastrointestinal tract occurs frequently through extension of retroperitoneal tumours, but primary involvement of the colon is uncommon with very few reports on this matter (2-11) (Table I). Due to their rarity, the mode of presentation, progression and prognosis are not well known.

In this paper, we report the first case of primary liposarcoma of the sigmoid colon presenting with colonic intussusception in a patient with a previous diagnosis of Crohn's disease. We discuss clinical, endoscopic and histological findings and review the existent literature.

CASE REPORT

We report the case of a 32-year-old female with ileal Crohn´s disease diagnosed at 27 and medicated since then with azathioprine (100 mg/day), in clinical remission for 2 years. She was admitted to our Emergency Department with acute lower left quadrant abdominal pain associated with a prolapsed rectal mass. She was diagnosed with hemorrhoid prolapse and discharged. Five days later, she returned to the Emergency Department with persistence of symptoms, now associated with rectal passage of blood and mucus, tenesmus and constipation. Physical examination revealed a slightly tender abdomen without palpable masses; rectal examination showed an ovoid ulcerated mass that prolapsed through the anus. Laboratory tests were relevant for mild leukocytosis and elevation in C-reactive-protein (CRP) (Table II). Colonoscopy revealed an intraluminal yellowish ovoid lesion in the sigmoid colon. The surface of the lesion was ulcerated and prevented further progression of the endoscope (Fig. 1). Twenty-four hours later, the patient developed severe abdominal pain and distention. Emergency abdominal CT (Fig. 2) revealed findings suggesting intussusception of the sigmoid colon due to an intraluminal mass consisting predominantly of fat. An urgent

exploratory laparotomy confirmed a sigmoid-rectal intussusception caused by a short-stalked lesion located in the sigmoid. Hartman's procedure was performed. Macroscopically, the tumor measured approximately 3.5 x 3 x 4 cm, had a rubbery consistence and the surface was ulcerated. The cut surface revealed an encapsulated, yellowish-pink, solid tumor (Fig. 3 A and B). Anatomopathologic examination revealed a well-differentiated liposarcoma with ulcerated areas growing from the submucosal layer and invading the *muscularis* and subserosa. The mucosa of the resected colon revealed no histological evidence of active Crohn's disease (Fig. 4 A-D). Azathioprine was suspended. One year later, intestinal reconstruction was successfully performed. At 5-years follow up the patient remains in clinical remission and with no evidence of disease recurrence.

DISCUSSION

Liposarcoma is a malignancy of fat cells arising frequently from the deep-seated stroma rather than the submucosal or subcutaneous fat. The most recent World Health Organization classification recognizes 5 categories of liposarcomas: 1) well differentiated or atypical lipomatous tumour, which includes the adipocytic, sclerosing, and inflammatory subtypes; 2) dedifferentiated; 3) myxoid; 4) mixed type; and 5) pleomorphic. The first case of primary colonic liposarcoma was reported by Wood and Morgenstern in 1989 (3). Since then 9 other cases have been published (2,4-11). Patients were predominantly females (7/10) between the ages of 41 and 79. The most common locations were the ascending colon (5/10), the descending colon (3/10) and the cecum (2/10). Our patient was 32 years old, the youngest case reported to date, and presented with intussusception of the sigmoid, a location not previously reported. Most patients presented with abdominal pain, weight loss, altered bowel habits, haematochezia and palpable mass. Only 2 cases reported intussusception as a manifestation of colonic liposarcomas (7,8). Intestinal intussusception, although relatively common in children, accounts for only 1 to 5% cases of bowel obstruction in adults and over 75% of cases are related to malignancy, namely colonic adenocarcinomas (12). Tumour size ranged from 3.5 to 15 cm, a feature that is not uncommon among liposarcomas in general (1,2). The histological subtypes included

well differentiated (5/10), myxoid (3/10), pleomorphic (3/10). Curiously, Choi et al. described a mixed liposarcoma with histological characteristics of both well differentiated and myxoid types (10). It is unknown if this particular histologic subtype has any influence in prognosis. Histological subtypes of liposarcomas have been demonstrated to correlate with clinical behaviour and prognosis. The well-differentiated type and most myxoid types are considered as low grade malignancies with 5-year survival of 100% and 88% respectively (13). Although they rarely metastasize, repeated local recurrences may cause the tumour to evolve into a higher grade of sarcoma or to dedifferentiate, in which case metastisation is possible (14). Pleomorphic and poorly differentiated types have a poor prognosis with 5-year survival rates between 0 and 20% (13). Other factors associated with poor prognosis have been identified including age > 45 years, presence of round cells, necrotic areas, tumor depth size, more than 20 mitosis per 10 high power field and disseminated disease (13,14). Two patients died during follow-up (2.5 and 4 years following diagnosis). However, we can only speculate over survival since most reports have short follow-up (mean 20.5 months). No standardized guidelines have been established for the treatment of colonic liposarcomas but, similar to other liposarcomas, surgical resection with wide excision is considered to be the treatment of choice. The exact role of radiotherapy and chemotherapy in the treatment of colonic liposarcoma is yet to be defined. Radiation therapy may be a valuable adjunct to surgery, especially in those of the myxoid variant and as shown to affect survival rates in soft tissue sarcomas (15). A retrospective analysis suggested that myxoid liposarcoma is relatively chemosensitive in comparison to other liposarcomas, in particular dedifferentiated and well-differentiated tumours (15). Our patient also had a previous diagnosis of Crohn's disease restricted to the ileum. To our knowledge there is no known relationship between these 2 entities. Our patient had a normal colonoscopy 3 years before. We can only speculate over the possible role of azathioprine in the development of the neoplasia. In conclusion, primary colonic liposarcoma is an extremely rare entity, ours being the 11th one reported, the 3rd presenting as colonic intussusception and the 1st presenting in the sigmoid. To date there is no known relationship between this type of tumour and Crohn's disease, so our case may represent an incidental finding. The

optimal treatment strategies are still to be established, although surgery is considered as the mainstay of curative treatment.

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Table I. Reported cases of primary liposarcoma of the colon

Year	*Age (yr)/sex*	*Location*	*Size (cm)*	*Histology*	*Survival*
1989 (3)	62/F	Caecum	12	Myxoid	Dead at 4 yr
1994 (4)	54/F	Ascending colon	6	Pleomorphic	N/A
2000 (8)	65/F	Caecum	5	Well-differentiated	Alive at 6 mt
2004 (5)	53/F	Descending colon	7.5	Well-differentiated	Alive at 2 yr
2006 (6)	46/M	Ascending colon	12	Myxoid	Alive at 12 mt
2006 (7)	66/F	Descending colon	4.5	Well-differentiated	Alive at 10 mt
2006 (2)	56/F	Ascending colon	3.5	Well-differentiated	N/A
2008 (9)	69/M	Descending colon	7	Pleomorphic	Alive at 10 mt
2009 (11)	79/F	Ascending colon	5	Pleomorphic	Dead at 2.5 yr
2010 (10)	41/M	Ascending colon	15	Well-differentiated and myxoid	Alive at 2 yr
Present case	32/F	Sigmoid	3.5	Well-differentiated	Alive at 5 yr

M: Male; F: Female; yr: Year; mt: Month; N/A: not available.

Table II. Laboratory evaluation at admission to our institution

Item	*Value*	*Normal values*
Hemoglobin (g/dL)	12.4	12.0-16.0
Leucocytes (x 10^9/L)	16.3	4.0-10.0
Neutrophils (x 10^9/L)	14.0	1.5-7.5
Platelets (x 10^9/L)	430	150-400
Urea (mg/dl)	14.0	15.0-39.0
Creatinine (mg/dl)	0.7	0.6-1.3
CRP (mg/dl)	1.2	< 0.8
LDH (UI/ml)	250	65-387

There is mild leukocytosis and elevated CRP. CRP: C-reactive-protein.

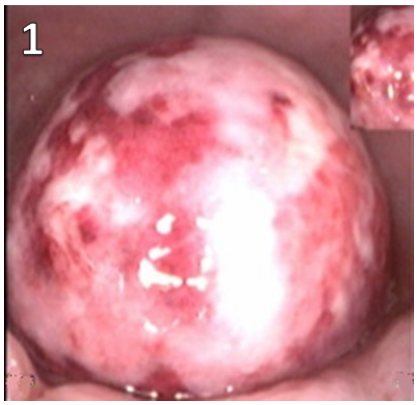


Fig. 1. Colonoscopy showing an intraluminal, ulcerated in the sigmoid colon.

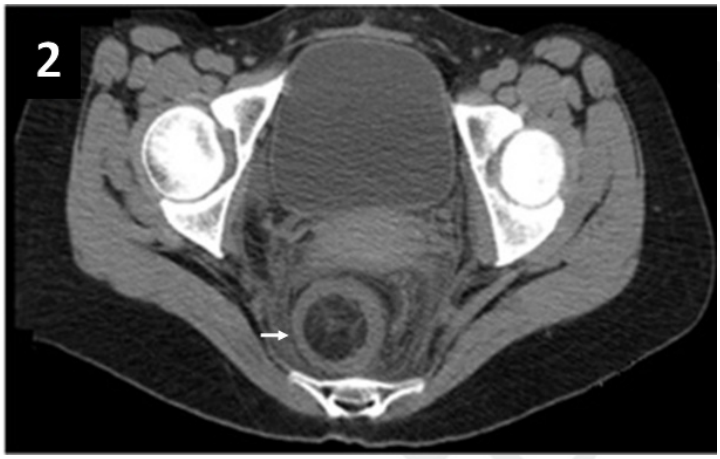
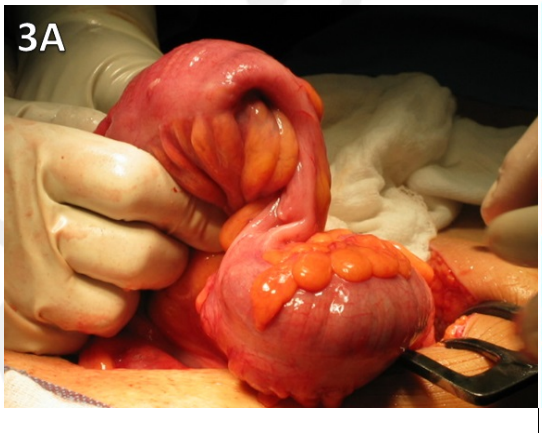


Fig. 2. Contrast-enhanced CT showing sigmoid-colonic intussusception by an intraluminal round mass with dominant fatty density (arrow).



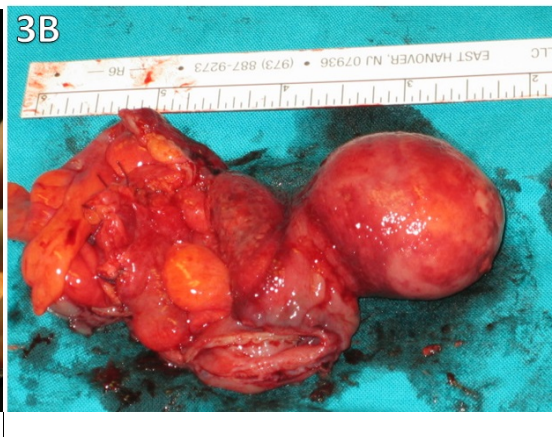


Fig. 3. Intraoperative findings. A. Telescoping of the sigmoid into the descending colon. B. The resection specimen showing a luminal short-stalked mass with an ulcerated surface and rubbery consistence measuring 3.5 x 3 x 4 cm.

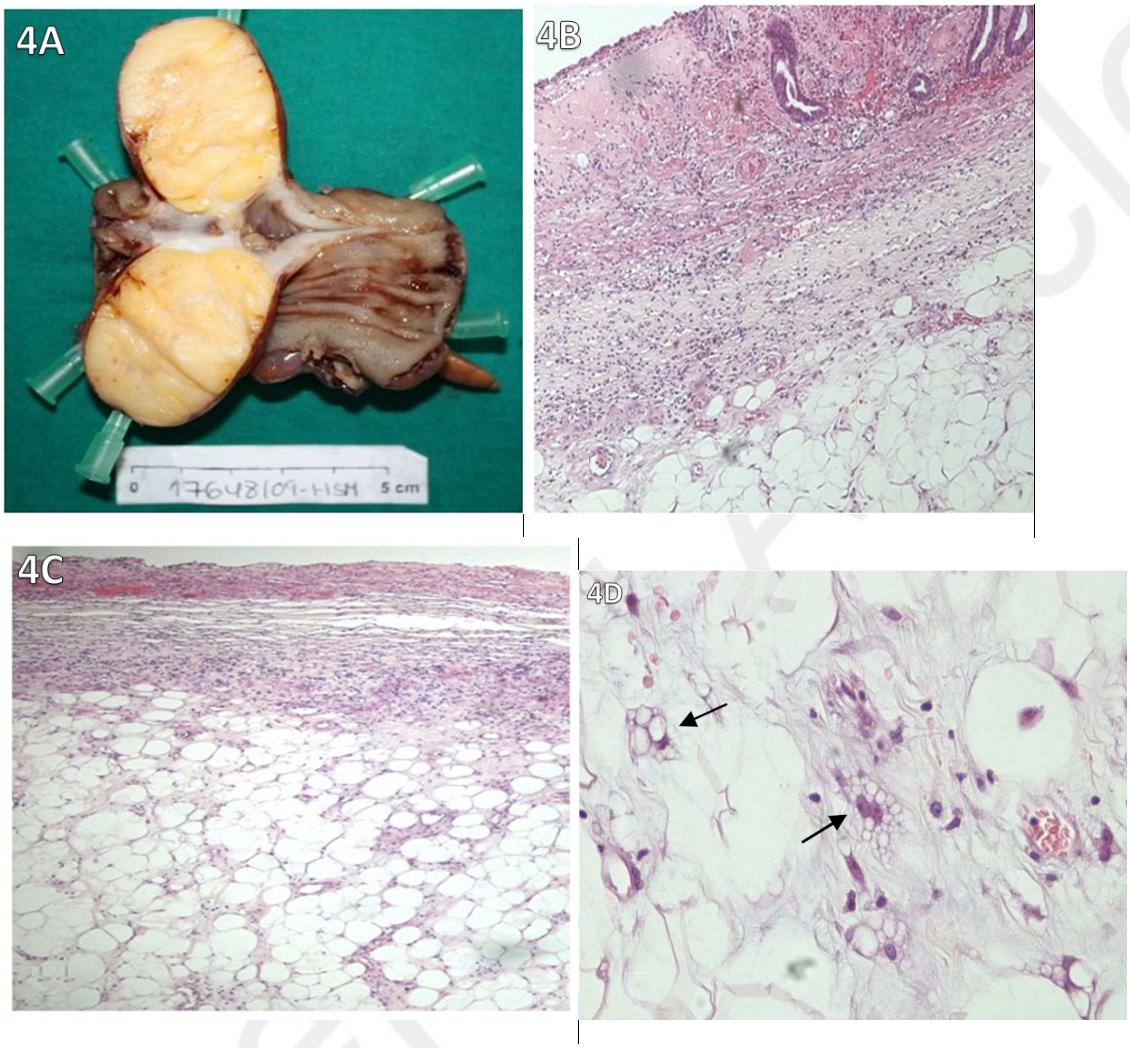


Fig. 4. Macroscopic and microscopic pathologic findings. A. Well circumscribed, lobulated, yellow tumor, in section. B. Microscopic aspects of ulcerated surface. C. Proliferation of mature lipocyte-like cells with marked variation in size in the submucosa. D. Multivacuolated lipoblasts (arrows).