CASE REPORT

Synovial sarcoma of the transverse colon: a rare cause of intussusception

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Accepted 16 March 2018

SUMMARY

We report a third case of a primary intramural synovial sarcoma of the lower gastrointestinal tract. A 50-year-old woman presented with hematochezia, dizziness and shortness of breath. CT imaging revealed a transverse colo-colonic intussusception with a colonic mass serving as a lead point. A subtotal colectomy was performed with oncologic resection of a sausage-like mass in the mid-transverse colon. Pathological assessment showed a biphasic synovial sarcoma. The postoperative hospital course was without complications and the patient was discharged home with plans for postoperative surveillance.

BACKGROUND

Synovial sarcoma of the gastrointestinal tract is extremely rare. The management of this neoplasm is different from other colorectal neoplasms and other causes of colonic obstruction. This case emphasises the clinical significance of maintaining a wide differential diagnosis list and having a low threshold for R0 oncologic resection of masses even when colonoscopic confirmation is not obtained.

CASE PRESENTATION

A 50-year-old woman with a history of diastolic heart failure with preserved ejection fraction (ejection fraction 60%), type II diabetes mellitus, end stage chronic kidney disease on dialysis and hypertension presented with multiple episodes of bright red blood per rectum associated with mild suprapubic tenderness, dizziness and mild shortness of breath. She also reported nausea, emesis and unintentional weight loss. She denied use of tobacco, alcohol and illicit drugs. She denied a family history of gastrointestinal disease or cancer. She was haemodynamically stable on presentation.

INVESTIGATIONS

Admission laboratory values revealed a leucocytosis of 11.7 thou/mcL and chronic anaemia with a Hemoglobin of 9.3 g/dL and Hematocrit of 23.1 per cent. CT demonstrated a transverse colo-colonic intussusception with a colonic mass serving as a lead point, without signs of obstruction or perforation (figure 1). There were also two indeterminate right upper lobe pulmonary nodules measuring up to 3 mm. Colonoscopy revealed an obstructing mass in the mid-transverse colon (figure 2). Multiple colonoscopic biopsies revealed colonic ulcers without evidence of neoplasia.

DIFFERENTIAL DIAGNOSIS

Adult intussusception is rare. Five per cent of all intussusception cases are found in adults and adult intussusception accounts for only 1–5% of all intestinal obstructions.¹ It is more commonly found in the small bowel, where 50–75% of cases are caused by benign lesions, including Meckel's diverticulum and postoperative adhesions, or by benign neoplasms, including lipomas, haemangiomas, polyps and lymphoid hyperplasias.^{1–4} Thirty-three per cent of small bowel intussusceptions have malignant neoplasms serving as pathological lead points, which are most commonly metastatic disease from miliary melanomas.¹ Other malignant neoplasms, including adenocarcinoma, gastrointestinal stromal tumours (GIST), carcinoid tumours, neuroendocrine tumours and lymphomas, have also been associated with small bowel intussusception.⁵ Less commonly found in the colon, large bowel intussusception accounts for 20–25% of all intussusceptions.^{1–4} In contrast to small bowel, colonic lead point is more likely to be malignant, most commonly primary adenocarcinoma.⁴

Differential diagnosis included soft tissue sarcomas found in the abdomen, notably gastrointestinal stromal tumours, desmoid tumours and rhabdomyosarcomas, as well as more common causes of lower gastrointestinal obstruction—adenocarcinomas, carcinoid/neuroendocrine tumours, lymphoma and diverticular disease.

TREATMENT

Tissue biopsy was non-diagnostic. A decision was made to resect the mass with oncologic R0 margins. Exploratory laparotomy revealed a large sausage-like mass just distal to the mid-transverse colon with intussusception between the proximal and mid-transverse colon. The mass was well-circumscribed with no visible necrosis and no macroscopic involvement of the serosa. The splenic flexure was mobilised and because there appeared to be inflammation of the distal transverse colon and splenic flexure, a subtotal colectomy was performed with primary anastomosis between the terminal ileum and descending—sigmoid junction.

OUTCOME AND FOLLOW-UP

Pathological assessment showed an intramural biphasic synovial sarcoma measuring 9.3×6.5×5.0 cm (figure 3). There was a mitotic rate of 5/10 under high-power fields. It stained positive for cytokeratin and actin and strongly positive for

To cite: Lee YF, Cleary RK. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2018-224198

Figure 1 Transverse colo-colonic intussusception demonstrated on the axial view (left panel) and coronal view (right panel) of CT abdomen/pelvis with oral contrast.

transducer-like enhancer of split 1 (TLE-1). Stains were negative for S100, Kkit, desmin, SOX10 and Melan-A. Molecular studies were not performed. The neoplasm involved mucosa, submucosa and muscularis propria. The pathological stage was pT2b pN0 G2. The postoperative hospital course was uncomplicated. She had a slow return of bowel function and was discharged on postoperative day 6. Her postoperative follow-up at this time consists of physical exam and imaging every 3 months at the University of Michigan Sarcoma Clinic.

DISCUSSION

There are currently 11 reported cases of synovial sarcoma of the gastrointestinal tract,^{6–15} four of which were of the lower gastrointestinal tract: a jejunal neoplasm,¹² an ileal neoplasm,¹³ a colonic polyp¹⁴ and a bulky mass to the left colon.¹⁵ The jejunal mass was thought to be a metastatic lesion in the setting of a past medical history significant for a spindle cell neoplasm in the posterior neck of a young man. The ileal neoplasm was a primary synovial sarcoma in a patient with a history of left atrial leiomyosarcoma. The two colonic neoplasms were primary intramural synovial sarcomas found in a young adult man and a paediatric male patient, respectively. The patient presented here is the third reported case of synovial sarcoma found in the lower gastrointestinal tract.

Synovial sarcoma accounts for 6% of all soft tissue sarcomas.¹⁶ It can occur at any age, though there is an increased incidence in young adults in the third decade.¹⁷ They most often occur in extremities near joints (origin of the synovial description). However, they can occur at any site in the body. These neoplasms can be monophasic with spindle-shaped mesenchyme, biphasic with mesenchyme and glandular epithelium or poorly differentiated. Despite the histological heterogeneity, more than 95% of synovial sarcomas share a genetic translocation between chromosomes X and 18, t(18)(p11.2;q11.2).

Figure 2 Intramural obstructing mass visualised in the mid-transverse colon at colonoscopy.

Figure 3 Ex vivo pathology specimen of synovial sarcoma.

Five-year survival for this disease is estimated to be 66%, with better survival rates reported for children, women, non-black race, tumours located in the extremities, localised tumours and tumours <5 cm in size.¹⁸ National Comprehensive Cancer Network (NCCN) guidelines for treatment are not uniform and leave room for variation in interpretation and practice patterns depending on tumour size, grade, histology, margins and hospital type.^{19,20} Stage I disease is generally treated by surgical resection with or without postoperative radiation therapy. Management of stage II and III sarcoma varies widely—surgery alone, neoadjuvant therapy (radiation, chemotherapy or both) or postoperative adjuvant therapy (radiation, chemotherapy or both) or a combination of neoadjuvant and adjuvant therapies. For surveillance of resectable stage II disease as seen in the patient presented in this report, NCCN recommendations are postoperative imaging—CT or MRI of the chest, abdomen and pelvis every 3–6 months for 2–3 years, then every 6 months for 2 years and then annually.

Learning points

- ▶ Synovial sarcoma is a neoplasm rarely found in the gastrointestinal tract that requires consideration in the differential diagnosis.
- ▶ Non-diagnostic tissue biopsies should not preclude oncologic resection of the mass when possible.
- ▶ Although National Comprehensive Cancer Network guidelines for management of stage II and stage III synovial sarcoma allow for variations in practice patterns, all sarcomas require a multidisciplinary approach and should be presented at multidisciplinary cancer conferences.

Contributors YFL collected patient information, performed literature review and drafted and revised the paper. RKC obtained consent from the patient, performed the operation and revised the draft paper.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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