Concomitant jejunal sarcomatoid carcinoma and gastric GIST in patient with polymyalgia rheumatica: A case report

Francesco Pata a,⁎, Manonmani Sengodan b,1, Cheuk-Bong Tang a,2, Sritharan Sangarapillai Kadirkamanathan a,4, Mike Harvey a,2, Abed Zaitoun c,3, Mahir Petkar b,4, Adriana Rotundo a,5

a Department of General Surgery, Upper Gastrointestinal Unit, Broomfield Hospital, Court Road, Chelmsford CM1 7ET, United Kingdom
b Department of Histopathology Broomfield Hospital, Court Road, Chelmsford CM1 7ET, United Kingdom
c Department of Histopathology, Nottingham University Hospital, Nottingham University Hospitals, Queen’s Medical Centre, Nottingham NG7 2UH, United Kingdom

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1. Introduction

Carcinomas involving the jejunum and ileum are rare tumours, with an estimated incidence of 0.5–0.8 per 100,000 population per year.1 Sarcomatoid carcinomas (SCA) are extremely rare, first described by Dikman and Toker,2 with only 21 cases reported in literature.

Gastrointestinal stromal tumors (GISTs) are the most frequent mesenchymal tumours of the digestive tract, more commonly found in the stomach (40–70%) and small bowel (20–40%).3 They were considered to be of smooth muscle origin and were classed as leiomyomas or leiomyosarcomas. However, immunohistochemistry studies4,5 (CD117 and CD34 expression) have suggested an origin from precursors of the Interstitial Cells of Cajal (ICC), that regulate gut motility and peristalsis. Recently, synchronous occurrence of GISTs with tumours of differing histogenesis have been reported.5

We report a case of a synchronous small bowel sarcomatoid carcinoma and gastric GIST in an 85-year-old female. To our knowledge, this is the first such presentation, and the first in a patient affected by polymyalgia rheumatica.

2. Presentation of case

An 85-year-old female was admitted in May 2010 with intestinal obstruction, following a 4-month history of dyspepsia and weight loss.

She had previously undergone a radical cystectomy and ileal conduit for transitional cell bladder carcinoma in 2000, was treated with corticosteroids for polymyalgia rheumatica, and since December 2008 was under follow-up for a gastric mass arising...
from the lesser curve of the stomach. Abdominal CT was suggestive of a GIST, but both endoscopic biopsies and Endoscopic Ultrasound/FNAB were inconclusive. The patient refused surgery. Repeat CT in March 2010, showed that the gastric mass had increased from 6.5–8.0 cm, and also revealed an additional, irregular mass in the mid abdomen attached to the wall of the adjacent small bowel. Surgery was refused again.

She was re-admitted in June 2010 with small bowel obstruction confirmed on CT scan (Fig. 1). Laparoscopy was unable to visualise the site of the obstruction and a laparotomy was performed revealing jejunal obstruction secondary to a polypoid, partly ulcerated, extra-luminal solid tumour (30 × 25 × 24 mm) in the mesentery. A large, gastric mass (101 × 66 × 22 mm) was found attached via a vascular pedicle to the lesser curve of the stomach. A partial resection of the jejunum, including the tumour and swollen lymph nodes, with end-to-end anastomosis and limited gastric resection was performed.

Histopathological examination of the obstructing mass demonstrated a sarcomatoid carcinoma, with serosal infiltration into the resected jejunal loop (Fig. 2), and five metastatic lymph nodes (pT4 N1 MX). It was a malignant biphasic neoplasm comprising of moderately differentiated adenocarcinoma admixed with a mesenchymal-like component demonstrating sarcomatoid features (Fig. 3). The adenocarcinomatous component had typical glandular and in places, a cribriform architecture whereas in other areas, it had a micropapillary pattern. Intracellular and extracellular PAS positive mucin was widely present. Focal areas of squamous differentiation with keratin pearl were also identified. The sarcomatoid element consisted of sheets of oval, round, plump and elongated spindle cells with large mitotically active nuclei. A vague myxoid appearance was also noted amongst this component. Both elements blended imperceptibly with each other. The tumour extended through the wall of the jejunum into the surrounding tissue and extensively invaded the serosa. There was also direct extension into the adjacent loop of the small bowel where it infiltrated the muscularis propria and submucosa but did not involve the mucosa. Prominent perineural and lymphovascular invasion were noted. Five adenocarcinomatous metastatic tumour nodules were present in the mesentery, presumed to be lymph node metastasis with extracapsular spread (definition as per TNM 5th edition).

A wide panel of immunohistochemical markers was performed. Both components were strongly positive for cytokeratins Cam5.2 and CK7 and were also positive for p53 and vimentin. The glandular component was positive for CD56 and Ca125, whereas focal positivity for p63 was noted in the squamous areas. The following immunostains were negative – CD117, desmin, SMA, CK20, S100, CD34, synaptophysin and chromogranin.

The gastric tumour had typical features of a GIST. The tumour was well circumscribed, arising within the muscularis propria, and was composed of bland spindle cells. 3 mitoses per 50 high power fields were seen. There was no necrosis. CD117 and CD34 were strongly positive. In view of the size and mitotic activity, the GIST was classified as a moderate risk for metastasis.

The patient was discharged on the 21st postoperative day and died on 88th post-operative day.

3. Discussion

Sarcomatoid small bowel carcinoma is a rare and aggressive variant of small bowel adenocarcinoma. It was first described by Dikman and Toker who regarded it as analogous to the blastomas found in other organs such as kidney, liver and lung and termed it “enteroblastoma”. Robey–Cafferty et al. were the first authors to give a detailed description of six cases of SCA of the small bowel, demonstrating its immunohistochemical staining patterns.
Early diagnosis of SCA of the Small Bowel is difficult due to the absence of any specific clinical features. They are usually discovered during an emergency operation for intestinal occlusion, perforation or bleeding.\(^7,8\) One case presented as superior vena cava syndrome with severe anaemia.\(^9\) Prognosis is often poor; most reported cases died within a few months of diagnosis.\(^10\) En-bloc resection with adjacent mesentry is necessary in the absence of any effective chemoradiotherapy, although effective chemotherapy and immunotherapy has been reported in cases of sarcomatoid bladder carcinoma.\(^11\) To date, to the best of our knowledge, 22 cases of SCA of the small bowel have been described in literature. This is the 23rd case and also the oldest patient with SCA of the small bowel.

GISTs are considered to have “uncertain malignant potential” and therefore all suspected GISTs of >2 cm should be resected.\(^12\) As lymph node spread and extraluminal growth are unlikely, a limited resection without lymphadenectomy is considered adequate.\(^13\) For small GISTs (<5 cm), laparoscopic resection is commonly performed\(^14\) although care must be taken to avoid tumour rupture at surgery, as this is associated with a high risk of peritoneal seeding and equivalent to incomplete resection.\(^15\) For this reason, preoperative biopsies are not indicated in resectable disease.\(^16\)

GISTs are known to co-exist with other neoplasms in frequencies varying from 4.5% to 33%.\(^17\) The most common GIST associated tumours are gastrointestinal carcinomas, lymphoma, and carcinomas of prostate, breast, kidney, lung, female genital tract and carcinoid tumours.\(^18\) Its presence in patients with epithelial malignancies is highly significant as they can easily be mistakenly evaluated as a metastatic lymph node or a metastatic deposit, thereby resulting in inappropriate treatment or tumour over staging. In our case, initial evaluation was with CT and EUS to exclude a potential metastatic lymph node, arising from a previous bladder carcinoma.

Polyvalgialy rheumatica (PR) is a clinical syndrome characterised by pain and stiffness in the neck, shoulders and hips, fatigue, weight loss and low grade fever. Although the relation between PR and giant cell arteritis is well known, its association with the occurrence of malignancy is open to debate. A few case reports detailing its possible association with breast cancer, colon cancer and non-Hodgkin lymphoma have been described.\(^19–21\) It has also been previously reported in a patient with synchronous gastric adenocarcinoma and GIST.\(^22\) The role of glucocorticoids, used in the treatment of PR, is unclear. Indeed, systemic glucocorticoid administration is known to be associated with skin cancers and non-Hodgkin lymphoma. It is interesting to note that our patient had a recurrence of PR in 2008, a few months before the appearance of GIST. In addition, there were two reported cases of SCA of the small bowel in patients with Sjogren syndrome\(^23\) and cardiac transplant,\(^24\) in which glucocorticoids are a therapeutic option. A paraneoplastic mechanism also needs to be explored and cannot be entirely excluded. The effect of PR as well as glucocorticoid usage in the development of these tumours requires further evaluation.

4. Conclusion

This is the first case report of a synchronous SCA of the small bowel, itself an extremely rare entity and a GIST in a patient with PR. SCA of the small bowel is an aggressive neoplasm with poor survival rates and surgery is the cornerstone of treatment. Given its unpredictable clinical behaviour and concomitant association with other malignancies, GISTs require adequate surgical resection with careful, long-term follow-up. In our patient, a more aggressive diagnostic approach, for example use of capsule endoscopy may have revealed sarcomatoid carcinoma at an early stage.


