CASE REPORT

Synchronous oesophageal and gastric sarcomatoid carcinoma: first reported case in Australia

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SUMMARY

A 75-year-old retired teacher presents with dysphagia and weight loss for a duration of 6 months. Her gastroscopy showed two synchronous submucosal masses. A 7 cm polypoid mass was seen at the distal oesophagus, arising from a thick stalk and a 4 cm mass seen at the cardia. The biopsies showed high-grade sarcomatoid cancer. Staging CT scan and Positron Emission Tomography scan did not show any distant metastasis except a lesion in the rectum that was subsequently found to be tubulovillous adenoma on transanal excision. The patient was managed with Ivor Lewis oesophagectomy. The biopsies of resection specimen showed spindle cell/sarcomatoid carcinoma with a component of poorly differentiated neuroendocrine carcinoma in oesophageal tumour and a small component of conventional invasive squamous cell carcinoma in tumour at cardia. The patient recovered well after surgery. Since then, she has completed adjuvant chemoradiotherapy. No recurrence has been noted in 10 months follow-up.

BACKGROUND

We present the first reported case of a synchronous, oesophageal and gastric, spindle cell/sarcomatoid carcinoma in Australia. This case highlights the difficulty in making the correct diagnosis of this rare malignancy, due to the presence of both epithelial and sarcomatoid components on histology and variation in the ratio and distribution of these components in any given tumour.

Figure 1 Endoscopic appearance of the oesophageal tumour.

Figure 2 Endoscopic appearance of the tumour at the cardia.

CASE PRESENTATION

This 75-year-old woman presented with dysphagia, epigastric pain and weight loss for 6 months. She had progressive dysphagia and was only able to tolerate liquids, for 4 weeks, preceding her presentation. Epigastric pain was constant, without any precipitating or relieving factors. She lost 15 kg weight over a period of 6 months. She did not have any significant medical issues and was not on any regular medications. She was a non-smoker with no history of alcohol consumption. She was living independently at home and had stable social situation. There was no significant family history. On examination, her vitals were stable, and systemic examination was unremarkable especially no organomegaly or lymphadenopathy was found.

INVESTIGATIONS

Barium swallow showed two intraluminal masses at distal oesophagus, and she was referred for gastroscopy. Her full blood picture, renal functions and liver function tests were normal.

Her gastroscopy showed two synchronous masses. A 7 cm long, polypoid mass that arose from distal oesophagus at a 2 cm stalk. The mass was submucosal with ulceration at the proximal end. There was another 4 cm submucosal mass at the gastro-oesophageal junction extending to the cardia (figures 1 and 2 and video 1). Despite the size of the tumour, an adult gastro-scope could easily pass through oesophagus

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Video 1 Video showing the oesophageal tumour attached at a broad stalk and the cardia tumour.

without resistance. The biopsies from first gastroscopy were inconclusive in making a diagnosis, and a second gastroscopy with tunnel biopsies was done. The biopsies from the second gastroscopy revealed a high-grade sarcomatoid malignancy, with no specific line of differentiation demonstrated on morphological and immunohistochemical grounds. There was no reactivity with a wide panel of cytokeratin stains including AEAE1/AE3, CAM5.2, CK7, p63 and p40. The endoscopic biopsies did not support the diagnosis of sarcomatoid carcinoma.

CT scan of the chest, abdomen and pelvis showed few subcentimetre lymph nodes along the lesser curve of the stomach in addition to the known oesophageal and cardia masses. There was no metastasis. Subsequently, a PET scan was done that showed intense uptake at the oesophagus, proximal stomach

Figure 3 Resection specimen showing oesophagectomy and proximal gastrectomy.

Figure 4 Conventional squamous cell carcinoma in situ and underlying invasive sarcomatoid squamous cell carcinoma.

and right side of the rectum raising suspicion for a third synchronous malignancy at the rectum. Colonoscopy showed a 20 mm sessile polyp (Paris 1 s), not amenable for endoscopic resection, at the dentate line with biopsies consistent with tubulovillous adenoma.

DIFFERENTIAL DIAGNOSIS

Her case was discussed in the statewide sarcoma group multidisciplinary meeting and a diagnosis of high-grade sarcomatoid malignancy, with no specific line of differentiation, was made after considering clinical, endoscopic and histology features. The endoscopic biopsies did not support the diagnosis of sarcomatoid carcinoma.

TREATMENT

The patient underwent examination under anaesthesia and transanal excision of rectal polyp and histology of resection specimen showed traditional serrated adenoma with conventional low-grade to high-grade dysplasia. Finally, patient underwent Ivor Lewis oesophagectomy (stage 1 via laparotomy and stage 2 via thoracotomy).

The resection specimen showed two polypoid tumours, one located in the distal oesophagus measuring 90 mmx36 mm, and the other located at the gastro-oesophageal junction measuring 80 mmx60 mm (figure 3). Both tumours on histological examination were shown to be spindle cell (sarcomatoid) squamous carcinoma with a component of poorly

Figure 5 Infiltrative nests of high-grade neuroendocrine carcinoma located beneath the surface squamous epithelium.

Figure 6 Sarcomatoid squamous cell carcinoma comprising moderately pleomorphic spindle-shaped cells with a haphazard growth pattern.

differentiated neuroendocrine carcinoma of small-cell type (20%) in the distal oesophageal tumour and conventional squamous cell carcinoma (5%) in the gastro-oesophageal junction tumour (figures 4–7). Out of 15, three lymph nodes were involved by all the different tumour components (pT3+T1b N1 M0). Immunohistochemical analysis of the spindle cell (sarcomatoid) component showed patchy staining for a range of cytokeratins (AE1/3, CK8/18, CK5/6 and MNF116). The neuroendocrine component showed perinuclear dot-like staining for cytokeratins in addition to cytoplasmic expression of synaptophysin and chromogranin.

OUTCOME AND FOLLOW-UP

The patient had an uneventful postoperative period and was discharged after 13 days of surgery. She received adjuvant chemotherapy (cisplatin and fluorouracil, followed by cisplatin and etoposide for NET component) and radiotherapy. She has completed her treatment and has no recurrence of disease during 10 months of follow-up.

DISCUSSION

Oesophageal sarcomatoid carcinoma is a rare malignancy with a reported incidence of an estimated 0.3%–2% of all oesophageal cancers.^{1–3} It is more common in middle-aged males with a male-to-female ratio of 4:1. The most common site of tumour is mid-oesophagus (60%) followed by distal and proximal

Figure 7 In situ and underlying invasive conventional and sarcomatoid squamous cell carcinoma.

oesophagus.⁴ The tumours are typically polypoid in nature. The overlying mucosa is largely smooth and intact but could be ulcerated as well. The tumours are typically attached to the oesophageal mucosa with a broad stalk or pedicle.¹ Patients often present early because of its large size and obstructive symptoms.^{2,3} However, as the tumour is attached to a pedicle and grows in the oesophageal lumen, it can reach to a large size before being detected. Zhang *et al* have reported a median tumour length of 5 cm (range 2–18 cm) in a series of 71 patients.²

The hallmark of sarcomatoid carcinoma is a biphasic appearance on histology with both epithelial (carcinomatous) and stromal (sarcomatoid) components.⁵ The epithelial component is commonly squamous with variable degrees of differentiation ranging from carcinoma in situ to invasive squamous cell carcinoma. Furthermore, some studies have reported adenocarcinoma and neuroendocrine tumours in addition to squamous cell carcinoma.⁶ The sarcomatoid component is typically composed of undifferentiated spindle-shaped cells. These spindle cells are commonly arranged in a storiform or fascicular pattern, embedded in an undifferentiated matrix and have high malignant potential.^{1,2,7} The sarcomatous component has been reported to exhibit different histological forms, including spindle cell carcinoma, chondrosarcoma, osteosarcoma, malignant fibrous histiocytoma and rhabdomyosarcoma.^{2,5,8}

Moreover, the ratio and distribution of epithelial and sarcomatoid components can vary. The epithelial component is commonly found on the surface of the tumour and the spindle cell component if present in stroma and hence superficial biopsies can lead to misdiagnosis.^{2,5,6} In a series of 71 patients with sarcomatoid carcinoma, only 10 cases were diagnosed with sarcomatoid carcinoma on the initial gastroscopy and biopsies.² Immunohistochemistry and special stains help to substantiate the diagnosis of sarcomatoid carcinoma. The epithelial component is typically cytokeratin positive while the stromal component is immunoreactive to vimentin.^{7,9}

The tumour could be highly aggressive as suggested by high rate of locoregional metastasis (33%–65%), a shorter tumour doubling time (2.2–5 months) and detection of high-grade dysplasia or carcinoma in situ in the resection margins in 43%–82% patients.^{2,5,6,10,11} Depth of invasion determines the metastatic potential with a 25% risk of metastasis with lamina propria involvement and 75% with invasion to adventitia.¹ Carcinomatous, sarcomatous or mixed metastasis has been described in different case series, and it commonly involves regional lymph nodes followed by lungs and pleura.¹²

Furthermore, surgery remains the mainstay of treatment in sarcomatoid carcinoma.^{2,5} Oesophagectomy with wide resection margins and extended field lymphadenectomy could provide the best chance of cure and should be considered as the primary modality of treatment. The prognosis in sarcomatoid carcinoma is poor. In a series of 71 patients, median survival was 43 months and 5-year survival was 44.8%.²

Learning points

- ▶ Sarcomatoid carcinoma is a rare tumour and requires a high index of suspicion to make correct diagnosis.
- ▶ These tumours can be aggressive with high rate of locoregional lymph node involvement.
- ▶ Surgery and lymphadenectomy remain the mainstay of treatment.

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