Carcinosarcoma of the oesophagus

Gaur D S, Kishore S, Saini S, Pathak V P

ABSTRACT

Oesophageal cancer is the third most common gastrointestinal cancer, especially in the Asian population. Only 1.3 percent of oesophageal cancers cases present as carcinosarcomas. A 43-year-old male who is a chronic smoker presented with progressive dysphagia and weight loss. On barium swallow, a large polypoidal mass was found filling the oesophageal lumen. Gross and microscopical findings along with immunohistochemistry helped in establishing its diagnosis as oesophageal carcinosarcoma. This unusual and controversial malignant tumour is biphasic in nature, composed of both carcinomatous and sarcomatous elements. This case showed a rarer feature that the epithelial element was of the adenocarcinomatous type instead of the expected squamous cell carcinomatous component.

Keywords: carcinosarcoma, oesophageal carcinosarcoma, oesophageal tumour, pseudosarcomatous carcinoma, pseudosarcoma

Singapore Med J 2008; 49(10): e283-e285

INTRODUCTION

Oesophageal cancer is the sixth most common cancer in the world, especially in the Asian population; and is the third most common gastrointestinal cancer.[1,2] While 89% of oesophageal cancers are of the squamous cell type, only 1.3% cases present as carcinosarcomas.[2] Oesophageal carcinosarcoma is a rare malignant tumour which is biphasic in nature, composed of both carcinomatous and sarcomatous elements. The multiple designations assigned to this tumour, such as carcinosarcoma, pseudosarcoma, pseudosarcomatous carcinoma and polypoid carcinoma reflect the controversy and differing views regarding its histogenesis and biology, as to whether the spindle cell component of this tumour is epithelial or mesenchymal in nature.[2]

CASE REPORT

A 43-year-old man presented with a history of progressive dysphagia for two months, increasing up to grade IV (able to ingest only liquids),[3] with significant loss of weight. The patient was a known smoker for the past ten years. He was investigated for dysphagia by barium swallow and oesophageal endoscopy, both revealing a large polypoidal growth in the lower third of the oesophagus. Ivor-Lewis oesophagogastrectomy was performed with partial oesopagogastrectomy and intrathoracic oesophagogastrostomy, handsewn by interrupted sutures using polyamide suture material (vicryl 3/0 sutures). On gross examination, the resected oesophageal segment showed a polypoidal growth, measuring 80 mm × 60 mm × 40 mm in size, including a short stalk. Cut surface showed tiny cystic areas with necrosis, with one site showing haemorrhage (Fig. 1). Consistency of the growth was firm. An entire slice of the tumour, including attachment of its stalk to the oesophageal wall, was taken for histopathological study.

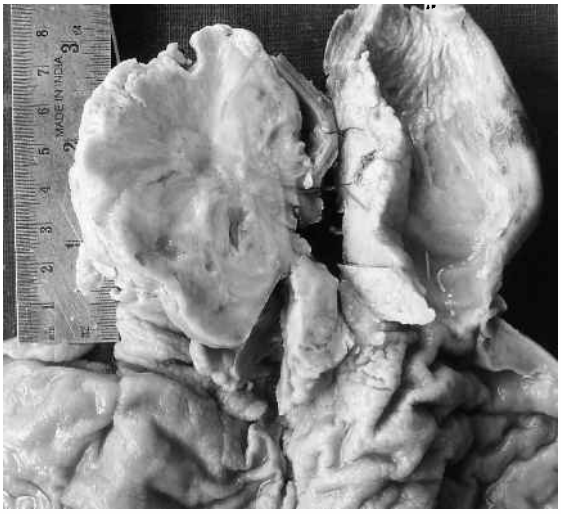

Fig. 1 Photograph shows the gross appearance of carcinosarcoma of the oesophagus situated above the gastro-oesophageal junction. Longitudinally-cut section shows areas of necrosis and the tumour stalk.

On microscopy, the neoplasm consisted of two components. The adenocarcinomatous component was composed of glands of various sizes and shapes, lined by one or more layers of pleomorphic columnar cells having hyperchromatic nuclei, many showing single nucleolus. Many mitotic figures were seen. The other component consisted of spindle-shaped cells forming groups or intersecting bundles. These had oval or elongated nuclei and many mitotic figures. The two components were lying separate in some places and were intermingled in other places. The immunohistochemistry revealed that the spindle cell component was positive for vimentin and negative for desmin (Fig. 2), while the smooth muscle cells of the oesophageal wall were negative for vimentin and positive for desmin (Fig. 3), and the adenocarcinomatous component was negative to both vimentin and desmin, but was positive for cytokeratin (Fig. 4). Diagnosis

Department of Pathology, Himalayan Institute of Medical Sciences, Dehradun 248140, Uttaranchal, India

Gaur DS, MD
Professor

Kishore S, MD
Professor and Head

Pathak VP, MD
Professor Emeritus

Department of Surgery

Saini S MS
Professor

Correspondence to:
Dr Dushyant Singh Gaur
Tel: (91) 135 247 1544,
Fax: (91) 135 247 1122
Email: dugaur@yahoo.com

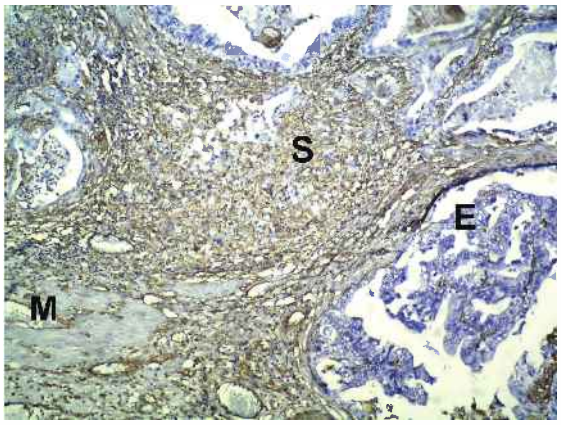

Fig. 2 Photomicrograph of carcinosarcoma shows immunomarker vimentin positivity in the sarcomatous component (S). Carcinomatous epithelium (E) and muscle fibres (M) are negative for vimentin. (Anti-vimentin [V9] with DAB chromogen, counterstain Harris' haematoxylin, × 400).

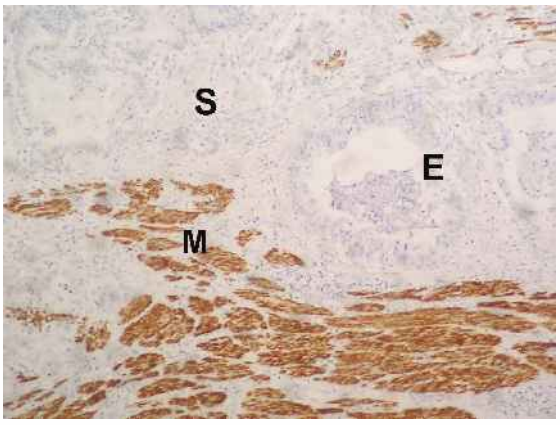

Fig. 3 Photomicrograph of the carcinosarcoma shows immunomarker desmin positivity in the muscle fibres (M). Sarcomatous component (S) and carcinomatous epithelium (E) are negative for desmin. (Anti-desmin [33] with DAB chromogen, counterstain Harris' haematoxylin, × 400).

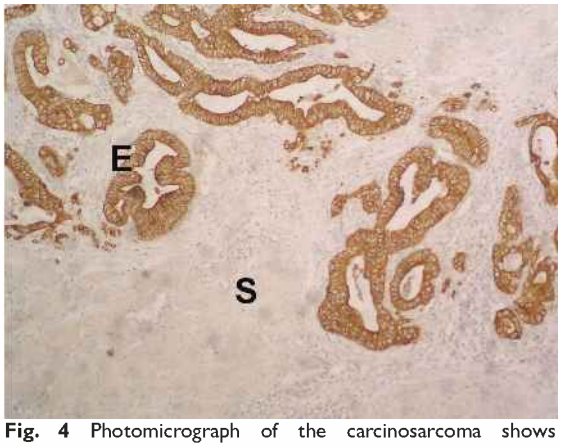

Fig. 4 Photomicrograph of the carcinosarcoma shows immunomarker cytokeratin positivity in the adenocarcinomatous epithelium (E). Sarcomatous component (S) is negative for cytokeratin. (Anti-cytokeratin [Lu-5] with DAB chromogen, counterstain Harris' haematoxylin, × 400).

of carcinosarcoma of the oesophagus was made, based on gross and microscopical features along with immunohistochemistry results.

The patient had a smooth recovery, except for prolonged chest tube drainage. He was discharged after 13 days of postoperative stay. He was scheduled for follow-up on a monthly basis. He had no complaint except for frequent gastric reflux at night, for which antacids and symptomatic treatment were given. He was prescribed a weekly dose of 5-fluorouracil 750 mg intravenously, as adjuvant therapy. The patient remained disease-free for the next six months. The chest radiograph and abdominal ultrasonography at three and six months showed no evidence of recurrence or metastasis. Later, he was lost to follow-up. On inquiry, he was reported to have died 18 months after surgery. However, the cause of death could not be ascertained.

DISCUSSION

Carcinosarcoma is an uncommon malignancy of the oesophagus that presents as a bulky intraluminal polypoid lesion of the oesophagus.[1] It predominantly affects men.[4] While the presenting symptoms and anatomic location of squamous cell carcinoma and carcinosarcoma of the oesophagus are similar, the latter often presents as a large intraluminal polypoid mass on barium swallow.[4] The more favourable prognosis associated with carcinosarcoma vs. other oesophageal neoplasia has been attributed to early onset of symptoms due to accelerated intraluminal growth, presenting as progressively increasing dysphagia, which leads to relatively early and prompt diagnosis.[4,5] Also, a lower propensity for tumour invasion has been observed, with tumour infiltration usually remaining confined to its stalk or the oesophageal wall, due to its polypoidal nature.[4,5] In the present case, the adenocarcinomatous element was seen instead of the usual squamous cell element, admixed with the sarcoma-like component, which made its presentation even more unusual and rare. There was no evidence of oesophagitis in this patient, thus excluding its origin from the metaplastic adenomatous element of Barrette's oesophagus.[6]

This patient was a chronic smoker for the past ten years. The close link between smoking and oesophageal cancers is well-established and documented.[7] However, there is no specific mention of smoking leading to carcinosarcoma in the literature. The assortment of names applied to this tumour—carcinosarcoma, pseudosarcoma, pseudosarcomatous squamous cell carcinoma, spindle cell carcinoma, metaplastic carcinoma and polypoid tumour— testifies to the differing views of its histogenesis and biology.[2,8] Histologically, both carcinomatous and sarcomatous components are seen in this tumour. The relative proportions of these components vary from case to case, though the "sarcomatous" element often predominates; while the carcinomatous element, quite often confined to the most superficial part of the mass, is prone to destruction by ulceration.[8] The sarcomatous component usually consists of haphazardly arranged or interlacing bundles of mitotically-active spindle cells.[9] Interestingly, the sarcoma-like component

appear to have a higher proliferation index, leading to production of a polypoidal exophytic growth (Fig. 1), and is more frequently aneuploid than the epithelial component; its stroma may contain conspicuous amount of collagen, or have a myxoid appearance.[7]

In the present case, only the sarcomatous component was found to be positive for vimentin, while the epithelial component lining the malignant adenocarcinomatous glands, as well as the muscle fibres of oesophageal wall were negative for it (Fig. 2). Desmin was found to be positive only in the muscle fibres and was negative in the sarcomatous and adenocarcinomatous elements (Fig. 3). Cytokeratin was positive only in the adenocarcinoma cells (Fig. 4). Thus, we concluded that this sarcomatous element does not have its origin from the smooth muscle element of the oesophageal wall. The histogenesis of the sarcomatous component is generally considered to result from metaplasia of carcinomatous cells toward mesenchymal differentiation.[10] Neoplastic epithelial cells may show dedifferentiation transforming to spindle cells, and also disdifferentiation to non-epithelial sarcoma, like chondrosarcoma and leiomyosarcoma.[11] There is no evidence of tumour in which a sarcoma appears to give rise to a carcinosarcomatous or carcinomatous subclone.[11]

Polypoid carcinomas with spindle-cell sarcomatous features have been designated either as carcinosarcoma or pseudosarcoma. The distinction between these two tumours has depended on the presence of "intermingling" of the carcinomatous and sarcomatous components in the so-called carcinosarcoma. But unlike the carcinosarcoma, the sarcomatous component in pseudosarcoma has been considered as benign.[10] Presently, studies supported by immunohistochemistry and electron microscopy suggest that carcinosarcomas are carcinomas with a varying degree of mesenchyme-like differentiation of carcinomatous cells.[7,10] However, its clinical and histopathological features are sufficiently distinctive to warrant its separation as a unique histopathologic entity.[12]

Surgical resection was chosen as the treatment of choice in the present case because the growth appeared to be exophytic, on barium swallow and oesophagoscopy.[13] Vogel et al suggested that preoperative chemoradiation in oesophageal malignancies results in significant clinical and pathological downstaging and increases survival. However, no specific observations were made regarding carcinosarcomas.[14] From the viewpoint of postoperative quality of life, reflux-related symptoms were the major problem for patients with an intrathoracic anastomosis, as seen in the present case. These symptoms cause significant insomnia and impair social and physical functions.[15] Postoperative complications might contribute to a poor prognosis in cancer patients.[16] Carcinosarcoma is associated with only a 2%–6% five-year survival rate.[17] Although the cumulative experience with this malignancy is limited, it is apparent that the presentation and clinical course of this tumour are not substantially different from the more common squamous cell carcinoma. Therefore, the diagnostic and therapeutic management should not vary from that of any malignant oesophageal lesion.[17]

REFERENCES

1. Stewart BW, Kleihues P, eds. World Cancer Report. Lyon: IARC Press, 2003.
2. Lee RG. Esophagus. In: Sternberg SS, ed. Diagnostic Surgical Pathology. 3rd ed. Philadelphia. Lippincott Williams & Wilkins, 1999: 1283-309.
3. Peters JH, Meester TR, Stein HJ. Surgical therapy for cancer of the esophagus and cardia. In: Castell DO, ed. The Esophagus. 2nd ed. New York: Little Brown, 1995: 293-335.
4. Ziauddin MF, Rodriguez HE, Quiros ED, Connolly MM, Podbielski FJ. Carcinosarcoma of the esophagus--pattern of recurrence. Dig Surg 2001; 18:216-8.
5. Madan AK, Long AE, Weldon CB, Jaffe BM. Esophageal carcinosarcoma. J Gastrointest Surg 2001; 5:414-7.
6. Dworak O, Koerfgen HP. Carcinosarcoma in Barrette's esophagus: A case report with immunohistological examination. Virchows Arch A Pathol Anat Histopathol 1993; 422:423-6.
7. Rao DN, Desai PB, Ganesh B. Epidemiological observations on cancer of the oesophagus--a review of Indian studies. Indian J Cancer 1996; 33:55-75.
8. Matsusaka T, Watanabe H, Enjoji M. Pseudosarcoma and carcinosarcoma of the esophagus. Cancer 1976; 37:1546-55.
9. Guarino M, Reale D, Micoli G, et al. Carcinosarcoma of the esophagus with rhabdomyoblastic differentiation. Histopathology 1993; 2:493-8.
10. Iwaya T, Maesawa C, Tamura G, et al. Esophageal carcinosarcoma: a genetic analysis. Gastroenterology 1997; 113:973-7.
11. Matsumoto T, Fujii H, Arakawa A, et al. Loss of heterozygosity analysis shows monoclonal evolution with frequent genetic progression and divergence in esophageal carcinosarcoma. Human Pathol 2004; 35:322-7
12. Campbell F, Bogomoletz WV, Williams GT. Tumors of the esophagus and stomach. In: Fletcher CDM, ed. Diagnostic Histopathology of Tumors. 2nd ed. London: Churchill Livingstone, 2002: 313-68.
13. Kato H, Tachimori Y, Watanabe H, et al. Superficial esophageal carcinoma: Surgical treatment and the results. Cancer 1990; 66:2319-23.
14. Vogel SB, Mendenhall WM, Sombeck MD, Marsh R, Woodward ER. Downstaging of esophageal cancer after preoperative radiation and chemotherapy. Ann Surg 1995; 221:685-95.
15. Christian E, Schmidt CE, Bestmann B, et al. Quality of life associated with surgery for esophageal cancer: differences between collar and intrathoracic anastomoses. World J Surg 2004; 28:355-60.
16. Hirai T, Yamashita Y, Mukaida H, et al. Poor prognosis in esophageal cancer patients with postoperative complications. Surg Today 1998; 28:276-9.
17. Hinderleider CD, Aguam AS, Wilder JR. Carcinosarcoma of the esophagus: a case report and review of the literature. Int Surg 1979; 64:13-9.