Virchows Archiv A Pathol Anat (1993) 422:423–426

Virchows Archiv A
Pathological Anatomy and Histopathology

Case report

Carcinosarcoma in Barrett's oesophagus: a case report with immunohistological examination

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Received October 2, 1992 / Received after revision February 3, 1993 / Accepted February 4, 1993

Abstract. A case of a carcinosarcoma which developed in a Barrett's oesophagus is presented. The tumour consisted of an adenocarcinoma and a spindle cell sarcoma. Immunohistological examinations demonstrated vimentin positivity in the sarcomatous portion with a negative reaction for keratin. Immunohistological and histological findings did not rule out the possibility of a double or collision tumour in this case.

Key words: Carcinosarcoma – Barrett's oesophagus – Immunohistology – Histogenesis

Introduction

Carcinosarcoma of the oesophagus is a rare tumour with both carcinomatous and sarcomatous components. The carcinomatous part is generally a squamous cell carcinoma with or without keratinization and rarely an adenocarcinoma (Du Boulay and Isaacson 1980; Ming 1973). Carcinosarcoma in Barrett's oesophagus with adenocarcinomatous component has not been described in the English literature.

The histogenesis of carcinosarcoma is obscure and controversial (Hanada et al. 1984; Iyomasa et al. 1990; Ming 1973; Ooi et al. 1986). Similar tumours have been described under the terms pseudosarcoma and polypoid carcinoma (Iyomasa et al. 1990; Osamura et al. 1978) and a precise differentiation between these tumours is not always possible. We present a case where immunohistological studies were performed, but emphasise that immunohistology has only a limited value in distinguishing between carcinoma and sarcoma (Leong 1992).

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Case report

A 76-year-old man presented to his family doctor because of increasing dysphagia. Endoscopy showed a polypoid tumour of the distal oesophagus with infiltration of the cardia. Barium swallow also demonstrated a tumour at the oesophagogastric junction, and a CT-scan revealed a circular thickening of the oesophageal wall. At subsequent oesophagoscopy, gastric mucosa was seen proximal to the tumour. Histological examination of the biopsy specimen revealed carcinosarcoma.

The patient was admitted to our hospital where a subtotal oesophagectomy with lymphadenectomy was performed. The post-operative course was uncomplicated and the patient was discharged on the 11th post-operative day. Two years post-operatively the patient is well and free of symptoms.

Pathology

The resected specimen consisted of an 8 cm long oesophagus with a 5 cm length of gastric wall. All but a 4–10 mm wide portion of the oesophagus corresponded macroscopically to a Barrett's oesophagus lined with gastric mucosa. A polypoid tumour with a diameter of 6.5 cm was seen, 2 cm distal to the oral resection margin (Fig. 1). The tumour consisted of two components, (Figs. 2, 3) an adenocarcinoma and a sarcoma which were relative sharply demarcated from each other. The sarcomatous part contained mainly spindle cells but also a few giant cells. Differentiation of the sarcomatous tissue was not seen, and numerous mitoses (over 30 in 10 HPF) were found. The adenocarcinomatous portion was located superficially with infiltration of the submucosa, while the sarcomatous part infiltrated deeper into the muscular layer. The surrounding Barrett's mucosa of intestinal type exhibited dysplastic changes. Only one lymph node containing metastatic adenocarcinoma was found among the 23 lymph nodes examined. The squamous epithelium of the oesophagus exhibited no dysplastic or neoplastic changes.

Immunohistological reactions were performed with vimentin, pancytokeratin (KL1 Dianova, Hamburg), keratin 13, keratin 18, desmin, S-100 protein alpha-1-

Fig. 1. Macrophoto of the tumour. On the right the polypoid tumour; on the left oesophagus with Barrett's mucosa. At the upper border a thin zone of normal esophageal mucosa (*arrow*). Cardia with stomach is seen in lower third of the specimen

antitrypsin and alpha-1-antichymotrypsin. Positive staining for pancytokeratin was detected in the normal epithelium and in the adenocarcinoma. Keratin 13 and 18 reacted in the squamous epithelium but not in the adenocarcinoma. The spindle cells reacted strongly reaction with vimentin (Fig. 4). No reaction with other antibodies was observed.

Discussion

Carcinosarcoma of the oesophagus is a rare malignant tumour with typical clinical and pathological features. This neoplasm can be characterized as a large, mostly exophytic, polypoid mass with infiltration limited to the oesophageal wall (Iyomasa et al. 1990; Xu et al. 1984). However, lymph node metastases can be found at all tumour stages (Iyomasa et al. 1990). In our case, a micrometastasis containing only a carcinomatous element was found. Other authors have reported that metastases contained mainly carcinoma and suggest that the sarcomatous part has a decreased capacity to metastatize to lymph nodes (Iyomasa et al. 1990).

The presence of lymph node metastases is of therapeutic significance. The tumour should be resected radically with removal of all lymph nodes (Iyomasa et al. 1990; Xu et al. 1984). Absence of spread signifies improved chance of curative resection.

It is generally agreed that carcinosarcomas of the oesophagus carry a better prognosis than carcinomas (Osamura et al. 1978) and our patient remains well two years after resection. The more favourable prognosis seems to apply for 3 year survival. After five years no differences were found in survival between the two tumours (Iyomasa et al. 1990).

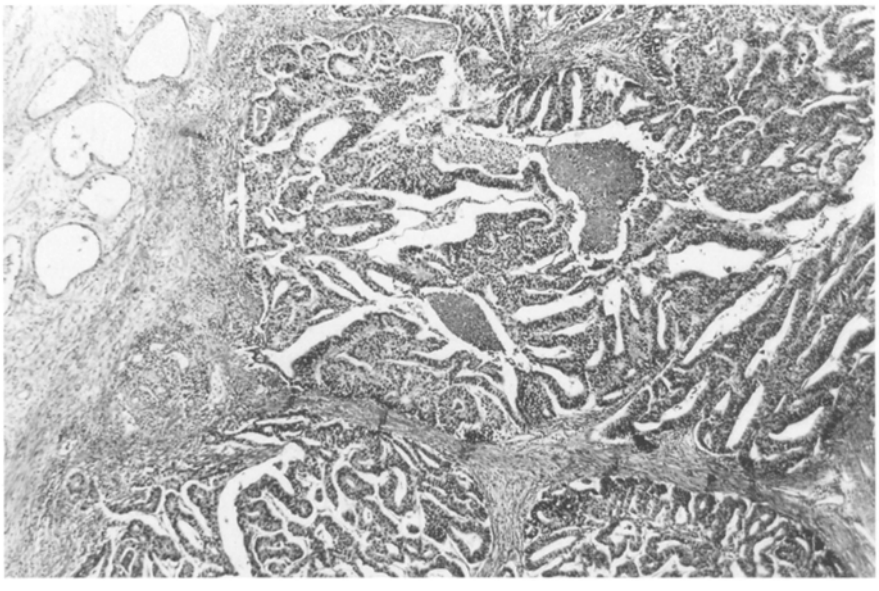

Fig. 2. The carcinomatous portion of the carcinosarcoma consisting of a moderately differentiated adenocarcinoma with invasion to the border of the muscularis propria. ×25

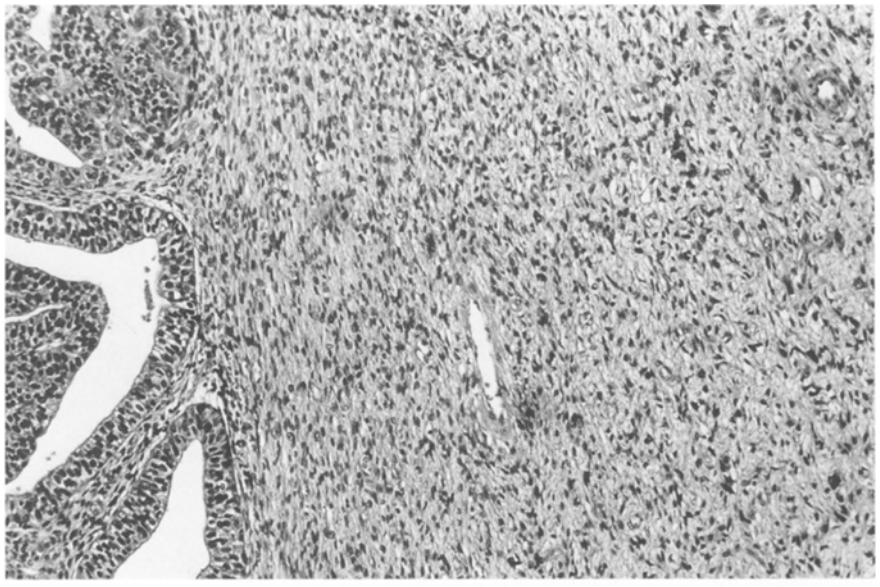

Fig. 3. Mostly sarcomatous part of the tumour with atypical spindle cells and giant cells. ×40

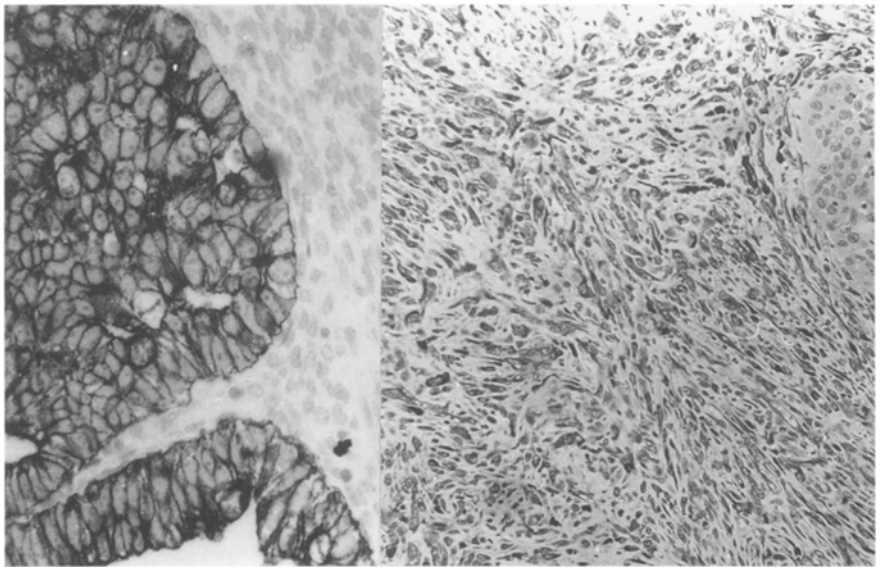

Fig. 4. Immunohistological demonstration of keratin positive adenocarcinoma on the left (×100) and vimentin positive sarcoma cells on the right (×220). In contrast there is no positivity of the carcinoma for vimentin or of the sarcoma for keratin

The histogenesis of this tumour is controversial. Tumours with specific mesenchymal (osseous, cartilagineous or rhabdomyosarcomatous) differentiation should be regarded as a true carcinosarcomas (Hanada et al. 1984; Xu et al. 1984). The classification of tumours with spindle cells is problematic. Electron microscopic and immunohistological studies suggest that the spindle cells may be of epithelial origin and represent metaplastic epithelial cells, even in the absence of keratin expression (Battifora 1976; Ellis et al. 1987; Hanada et al. 1984; Kuhajda et al. 1983; Ooi et al. 1986; Takubo et al. 1982). Other authors have reported spindle cells containing vimentin where the co-expression of vimentin and keratin was not present in the same cells (Ooi et al. 1986; Zarbo et al. 1986). These results permit no definitive conclusions about the histogenesis of the sarcomatous component. Our findings showed no keratin positive cells in the sarcomatous area. However, the spindle and giant cells reacted strongly with vimentin; nevertheless, vimentin is not a definitive marker for mesenchymal cells. Cells of different carcinomas may contain vimentin, mostly with co-expression of keratin (Leong 1992; Zarbo et al. 1986). These observations suggest that these cells probably belong to different cell lines.

Carcinosarcoma in Barrett's oesophagus has not been described in the literature. One Chinese paper, lacking illustrations, presented the case of a 58-year-old woman with a polypoid carcinosarcoma containing adenocarcinoma, squamous and basal cell carcinoma in addition to a fibrosarcoma (Liu and Zhang 1980). Other authors (Du Boulay and Isaacson 1980; Ming 1973) mention the presence of glandular structures in carcinosarcoma. In these cases, the main epithelial tumour mass consisted of squamous carcinoma. Mucoepidermoid and adenosquamous carcinoma of the oesophagus have not been reported to contain spindle cells (Bombi et al. 1991; Pas-

cal and Clearfield 1987; Smith et al. 1984). In an adenosquamous carcinoma of the gall bladder spindle cells showed the presence of prekeratin and were recognized as epithelial cells (Suster et al. 1987).

We believe that our case demonstrates unique histological features: it developed in a Barrett's oesophagus, the carcinomatous component is an adenocarcinoma without squamous or spindle cell metaplasia, and the squamous epithelium of the oesophagus showed no dysplasia or carcinoma.

The classification of these tumours is confusing and based on histological, immunohistological and electron-microscopic observations. According to the most recent WHO classification (Watanabe et al. 1990), spindle cell (squamous) carcinoma (also called pseudosarcoma and pseudosarcomatous carcinoma) and carcinosarcoma should be distinguished by the specific differentiation in the sarcomatous component. We are of the opinion, however, that tumours with spindle and/or giant cell components without immunohistological evidence of epithelial markers and in the presence of vimentin (as in our case) may be classified as carcinosarcomas. Metastases with sarcomatous components may be helpful in making a differential diagnosis (Osamura et al. 1978).

Histogenetic explanations cannot be derived from a single case. However, the relatively sharp demarcation of both neoplastic components and our immunohistological findings do not rule out the possibility of a double or collision tumour. The absence of abnormal squamous epithelium or squamous metaplasia in the adenocarcinomatous portion of the tumour seems to support this hypothesis.

Acknowledgements. We thank Dr. Wen-Chih Huang, Institute for Sinology of the University Erlangen for the translation of the Chinese paper.

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