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Carcinosarcoma of the oesophagus

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The clinical and pathological details relating to a case of carcinosarcoma of the oesophagus are presented. Evidence for the true mixed nature of the lesion is demonstrated and criteria for distinguishing this from pleomorphic carcinoma are put forward. The literature relating to this rare lesion is reviewed and the problems of identifying its true pathological nature are discussed.

CASE REPORT

J. S. A., a 74-year-old man, presented with a four-week history of dysphagia, anorexia, and loss of weight. He stated that until a month previously he had been quite well apart from occasional episodes over the past two years of 'indigestion' and belching, readily relieved by medicines. The belching after meals had recently increased and was later followed by vomiting immediately after eating. For two weeks he had been unable to eat meat or other solid foods and had lived on fluids, eggs, and purée foods. He was able accurately to localize the obstruction at the level of the lower sternum. Over the period of this disability he had lost 8 lb. (3·6 kg.) in weight.

On examination the patient was thin and had lost a good deal of weight. He had a marked pectus excavatum. The pulse was irregular and the blood pressure 140/70 mm. Hg. Examination of the heart revealed occasional ectopic beats but no cardiac enlargement. There was a grade 1 systolic ejection murmur.

A barium study showed the presence of a large filling defect (Fig. 1) measuring 7×3 cm. in the mid-oesophagus with appearances suggestive of a sarcoma. The mass appeared lobulated and the oesophageal contour was virtually intact. The stomach and duodenum were normal. A chest radiograph showed moderately emphysematous lung fields. There were degenerative changes throughout the spine.

At oesophagoscopy a coarsely nodular growth, partially obstructing the lumen of the oesophagus, was seen at 28 cm. from the upper alveolar margin. The tumour had the appearance of an unripe blackberry. Biopsies were taken, and on histological examination the tumour was reported to be a leiomyosarcoma.

Though the patient was 74 years of age, thin, and frail, with a degree of pectus excavatum, he had reasonable cardiopulmonary function for his age and satisfactory ventilatory capacity. Operation with a view to resection of the tumour was therefore advised and was performed on 22 November 1966. With the patient in a semi-oblique position, an upper midline laparotomy incision was made. No secondaries were detected in the liver, and no abdominal lymph node enlargement was demonstrated.

A right thoracotomy was then performed through the bed of the resected fifth rib. A large tumour mass, distending the mid-third of the oesophagus, was present. The upper limit was at the level of the azygos arch, and the growth extended downwards for about 3 in. (7·6 cm.). The mass did not appear to transgress the muscle wall of the oesophagus.

The whole of the oesophagus from just below the thoracic inlet to the hiatal orifice was mobilized. Attention was then redirected to the laparotomy and a pyloroplasty was performed after mobilizing the

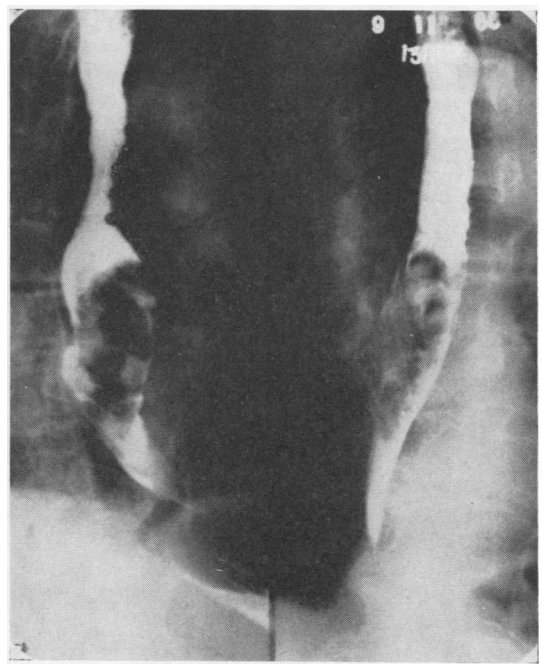

FIG. 1. *Barium study of carcinosarcoma in mid-oesophagus, showing large lobulated filling defect.*

stomach, which was then brought up into the chest through the previously dilated oesophageal hiatus, was transected obliquely to remove its fundus and upper lesser curvature, and was finally closed in layers. An oesophagogastrostomy was made just below the level of the thoracic inlet, transecting the oesophagus well above the growth and using a separate incision in the upper portion of the gastric tube. This region was anchored to the mediastinal pleura to avoid tension on the suture line. A nasogastric tube was passed through the anastomosis into the stomach. The chest was then closed in layers with two underwater tube drains, and the laparotomy incision was closed.

The patient made a smooth and uneventful recovery from the operation and on discharge had no difficulty in eating a normal diet.

When seen three months after the operation the patient reported that he had a very good appetite and was inclined to eat too much. He was eating three very good meals a day, which included a good breakfast, a meat meal at least once a day, a normal tea, and a regular late supper. In addition to this he was having morning and afternoon snacks, and his diet was supplemented by three or four pints of beer a day.

A year after operation he had gained more than half a stone (3 kg.) in weight and stated that he could eat anything. He was leading a normal life and there was no dysphagia.

PATHOLOGY

The biopsy of the tumour removed at oesophagoscopy comprised friable white fragments measuring up to 7 mm. in greatest diameter.

Histologically these consisted of bundles of pleomorphic spindle-shaped cells, generally arranged haphazardly but occasionally showing a whorled pattern (Fig. 2). The cell nuclei varied from spherical to elongate forms and the majority were hyperchromatic. A few bizarre mitotic figures were evident. The fragments were extremely vascular, with extensive areas of necrosis. There was a mild diffuse infiltration of neutrophils, plasma cells, and lymphocytes. There was no

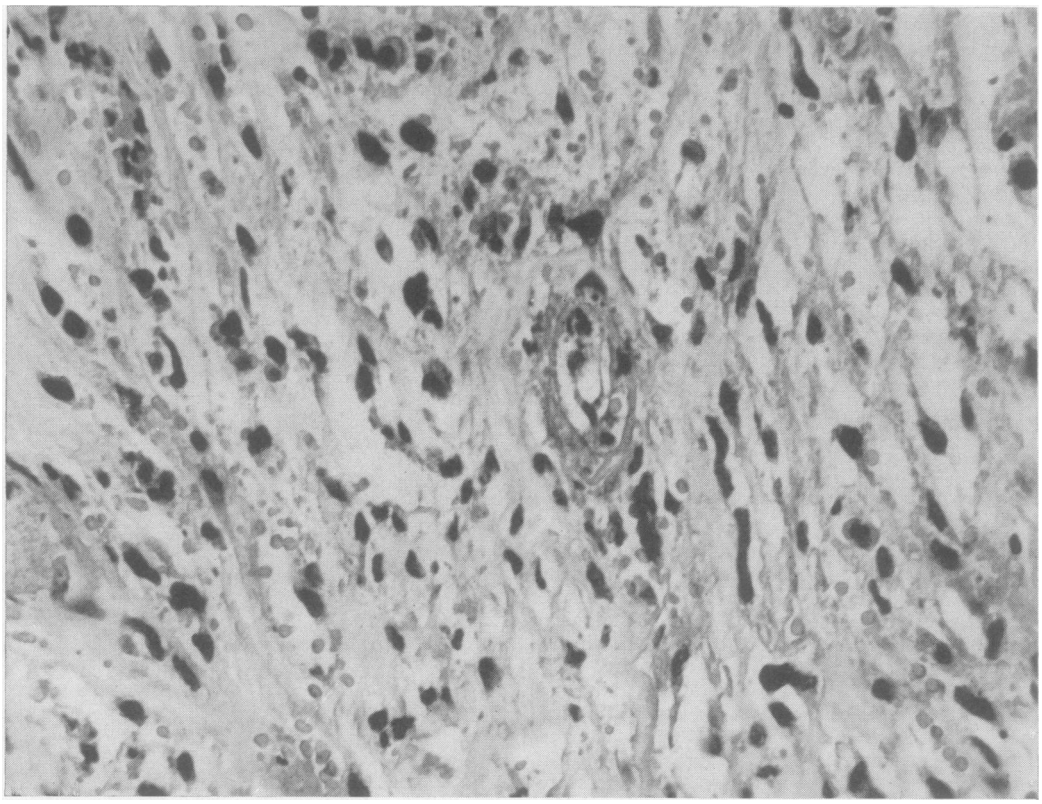

FIG. 2. *Histological section from biopsy specimen showing spindle-cell and pleomorphic sarcomatous growth. H. and E.* ×470.

FIG. 3. *The resected pedunculated specimen, bisected longitudinally.*

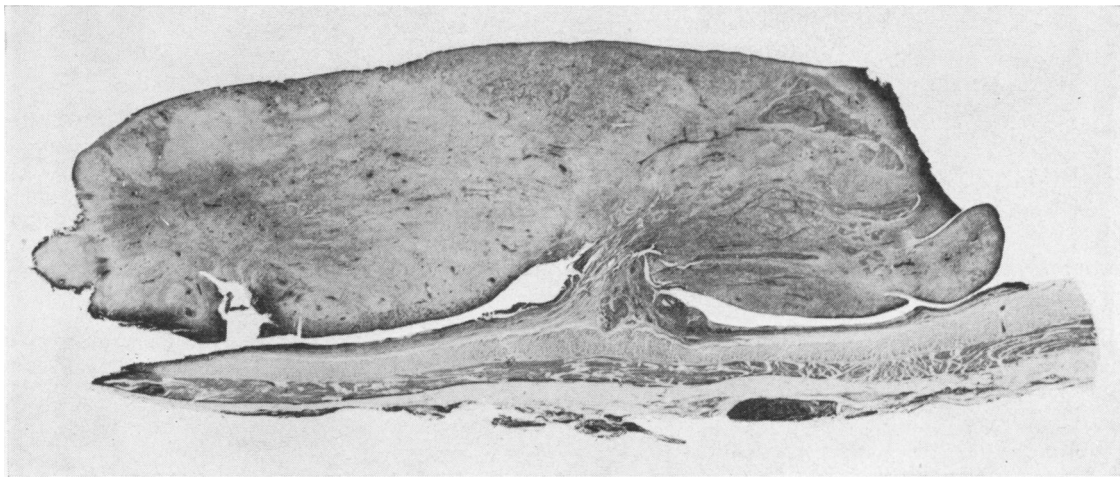

FIG. 4. *A complete section of the tumour and the adjacent oesophageal wall. H. and E.* ×2.

recognizable epithelium. A diagnosis of leiomyosarcoma of the oesophagus was made.

The resected specimen consisted of the lower oesophagus and cardia of the stomach, measuring a total of 23 cm. in length. Arising 12 cm. proximal to the oesophago-gastric junction from the posterior oesophageal wall was a polypoid tumour, measuring 6 × 1·5 × 1·5 cm. (Fig. 3). It was firm and rubbery in consistence, pale pink in colour, and was attached to the wall by a narrow stalk, 1 cm. long. There was an intact shiny membrane covering the tumour. The cut surface showed intermingled grey and pinkish areas (Fig. 4). No lymph nodes were present in the adventitia.

Microscopically, two distinct types of neoplastic tissue were present, *i.e.*, sarcomatous and carcinomatous. The bulk of the tumour consisted of interlacing bundles and diffuse sheets of spindle-shaped cells with similar morphology to those seen in the biopsy specimen (Fig. 5). They showed marked pleomorphism. The nuclei were generally fusiform with giant forms and bizarre mitotic figures. This portion showed a diffuse infiltration of lymphocytes and plasma cells and large foci of neutrophils. It was covered by an intact stratified squamous epithelium which in the region of the stalk became continuous with the oesophageal epithelium. Cytologically, this was a spindle-cell sarcoma and, in view of the tendency to whorling and the predominantly spindle-shaped cells, it was regarded as originating from smooth muscle. At the base of the stalk the sarcomatous tissue gradually merged with the oesophageal submucosa (Fig. 6).

Dispersed irregularly but focally throughout the tumour were clumps of vacuolated, pleomorphic, epithelial cells with prominent nuclear hyperchromatism (Fig. 7). Many bizarre mitotic figures were seen. A few cells had more regular polyhedral and vesicular nuclei. There was extensive tumour necrosis centrally. These clumps were regarded as representing poorly differentiated carcinoma (Fig. 8). At the base of the stalk, carcinomatous deposits were in intimate relationship with the oesophageal epithelium (Fig. 4). In the oesophageal wall in this area there was

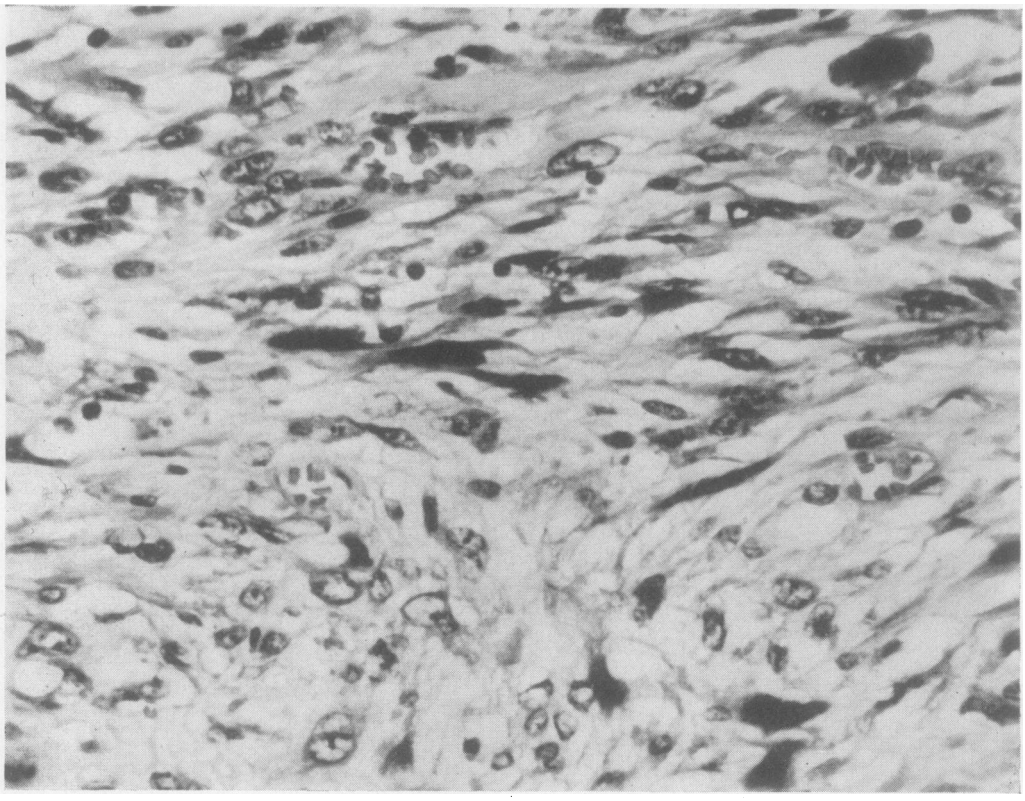

FIG. 5. *A representative field in the sarcomatous component, showing spindle-shaped cells with irregular nuclei and bizarre mitoses. H. and E.* ×470.

invasion through the muscularis mucosae into the submucosa. The circular muscle coat was intact. No carcinoma was present remote from the tumour stalk. No myofibrils were demonstrated in specially stained sections.

The definitive histological diagnosis of the resected specimen was carcinosarcoma of the oesophagus.

DISCUSSION

Carcinosarcoma of the oesophagus is a rare neoplasm comprising epithelial and smooth muscle components. Similar complex lesions have been described in many other sites, and considerable controversy has surrounded their identification.

Willis (1960) accepts a true dual tumour in the breast but denies the existence of such lesions elsewhere in the body. He maintains that anaplasia and pleomorphism in carcinomata have led histologists to false conclusions in this regard, and he cites the contribution of Saphir and Vass (1938) in his support. However, these authors, although rejecting the majority of a series of 153 cases, did accept the diagnosis in a few. Brooks (1943), like Willis, refuted the diagnosis in reviewing 110 so-called combined lesions and demonstrated transitional areas between epithelial and spindle-cell components.

Other critical authors have, however, disagreed with this extreme view and, adopting strict criteria, have established carcinosarcoma as a rare entity occurring in various sites and clearly distinguishable from pleomorphic carcinoma (Evans, 1966; Drury and Stirland, 1959; Harvey and Hamilton, 1935). The important criteria are pedunculation, sharp demarcation of the two architectural components, and relative localization of the growth.

Authoritative and convincing papers include those of Stout, Humphreys, and Rotenberg (1949) and Stout and Lattes (1957). The careful paper of Moore, Battersby, Vellios, and Loehr (1963), describing a case very similar to our own

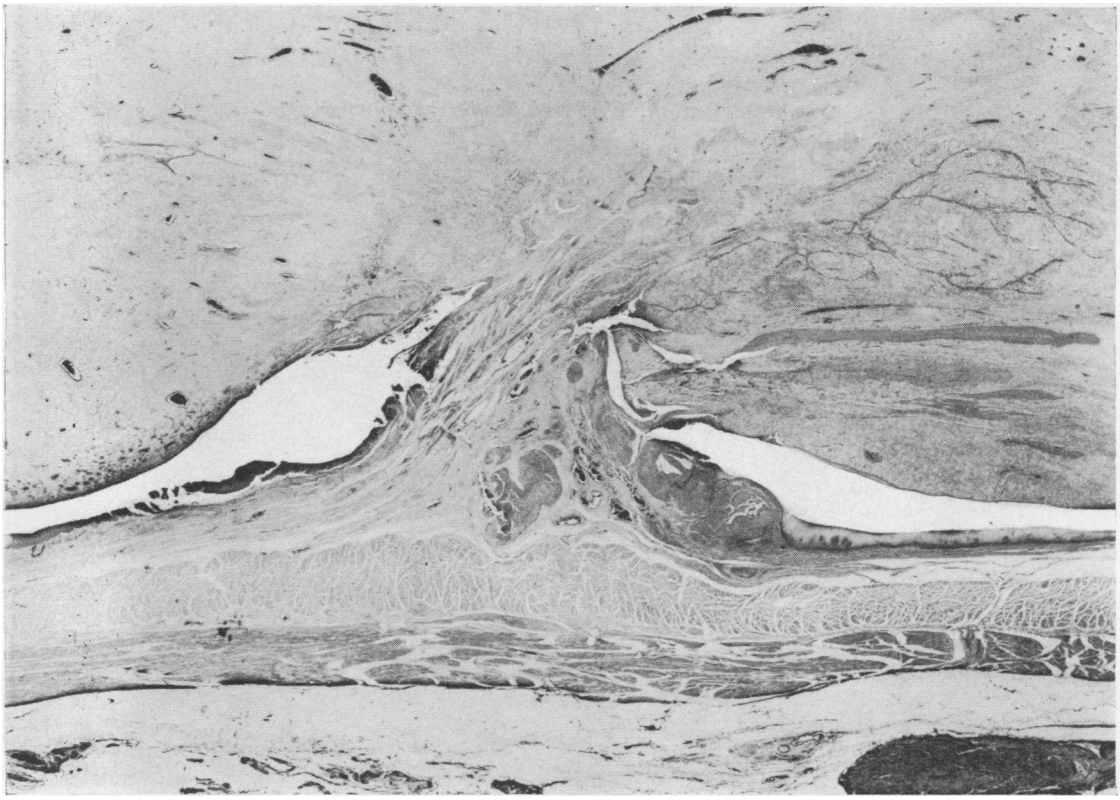

FIG. 6. *Rounded clumps of carcinoma cells at the base of the stalk. H. and E.* ×6.

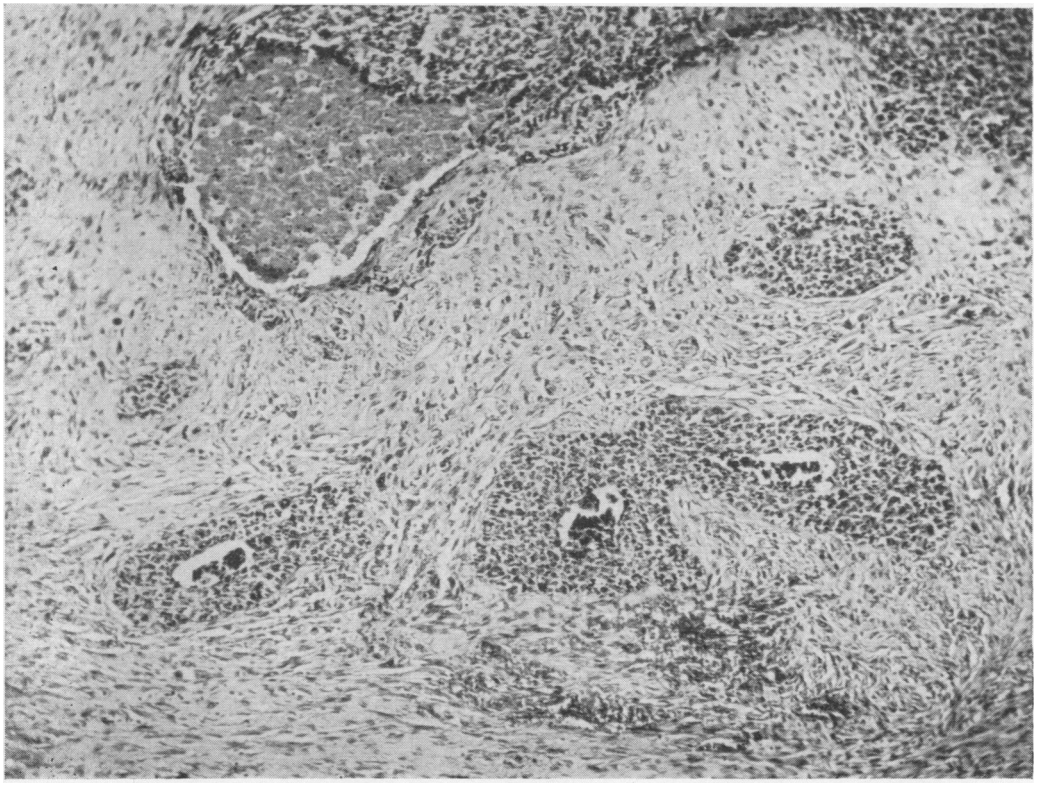

FIG. 7. *Several clumps of poorly differentiated carcinoma with central necrosis are surrounded by sarcomatous tissue. Note clear demarcation of components. H. and E.* ×88.

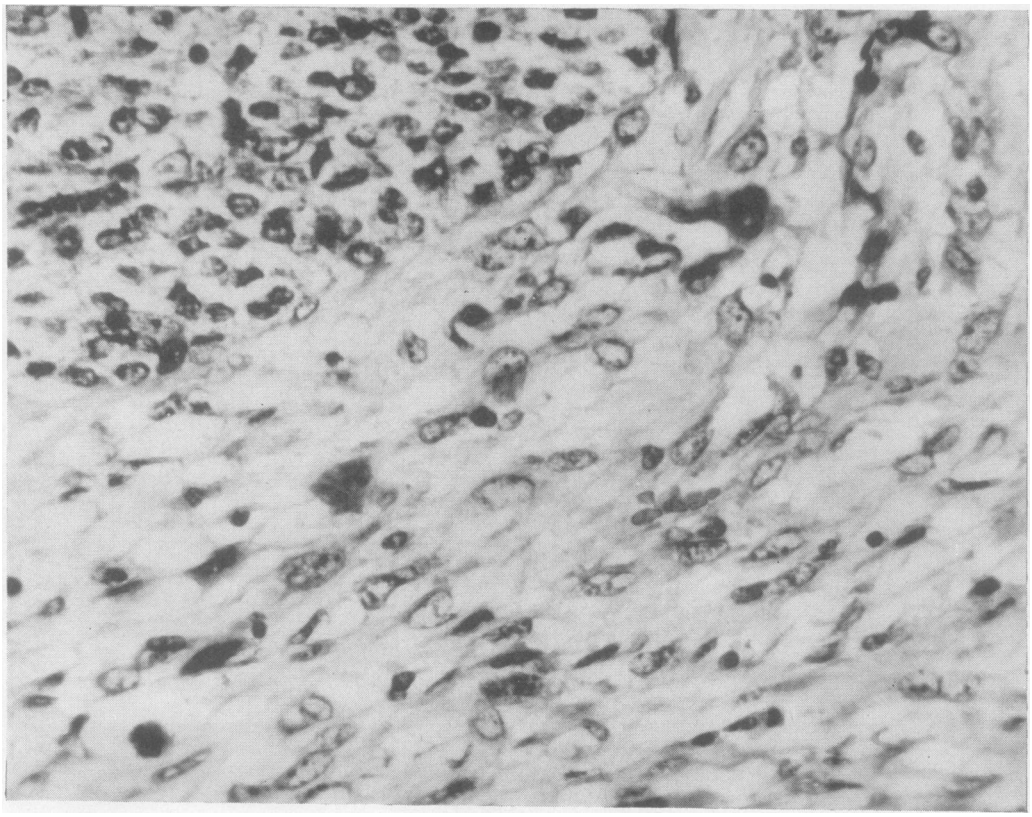

FIG. 8. *Detail of junction between poorly differentiated carcinoma cells and sarcoma.*

and reviewing the literature, lends weight to the evidence that true carcinosarcoma can be defined. Their patient was alive and well five and a half years after resection. Talbert and Cantrell (1963) gave an account of four examples of carcinosarcoma of the oesophagus and reviewed 23 cases from the literature, the majority occurring in the lower two-thirds of the oesophagus. Of these, 13 had been resected, the operative mortality being 45%. Gowing (1961) also categorizes polypoid carcinosarcoma as a clearly defined entity.

It is noteworthy that the diagnosis of carcinosarcoma does not appear ever to have been made from oesophagoscopic biopsy material. In our own case, only sarcomatous tissue was demonstrated. The finding of carcinomatous tissue in a biopsy from a pedunculated growth should suggest the diagnosis of carcinosarcoma.

The histogenesis of this curious lesion is not clear, but, of the various possibilities, the theory that one of the malignant tissues excites a neoplastic change in the other seems likely. This explanation was put forward by Harvey and Hamilton (1935). The main bulk of the lesion in our case was sarcomatous, and the carcinomatous tissue was most evident in the region of the pedicle.

In our own case the well-defined criteria of pedunculation, the sharp distinction of epithelial and muscle components, and the confinements of the lesion inside the oesophageal wall are all present.

These features differ clearly from those of anaplastic pleomorphic carcinoma, in which diffuse origin, deep invasion of the oesophageal wall, and frequent local and distant metastatic spread are regular features. In addition, the histological components of pleomorphic carcinoma intermingle haphazardly and show transitional forms between one cell type and another (Brooks, 1943).

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