CLINICAL RECORDS

LEIOMYOSARCOMA OF ŒSOPHAGUS

By J. F. NEIL and E. A. HORNE (Nottingham)

A BRIEF account of a case of leiomyosarcoma of the œsophagus is presented because of the rarity of the tumour and to draw attention to the unusual but diagnostic radiological picture produced by a large pedunculated tumour in the œsophagus.

Case Report

In *April, 1949,* a male aged 54 was referred to the Aural Department from the City Chest Clinic. He had been under observation for one month, complaining of hoarseness, dyspnœa and blood-stained sputum. Fluid of a light brown colour, suggestive of altered blood, had been aspirated from his right pleural cavity.

On indirect examination of his larynx, a slight limitation of movement of the right vocal cord was seen. Further investigations could not be carried out as he failed to keep his next appointment.

In *June, 1950,* he again attended the Department complaining of dysphagia of some six weeks' duration, together with pain in the right side of the chest and the right scapular region. He could swallow fluids only. He also gave a history of syphilis and stated that he had had some treatment for it; the W.R. was positive.

Clinical examination revealed nothing new, but on radiological examination of his chest it was seen that there was a widening of the right upper mediastinum, the shadow having a smooth lateral border.

A barium swallow (Fig. 1) showed that this mediastinal shadow was apparently a dilated œsophagus, containing what was reported to be a mixture of food residue and barium above a stricture in the middle third. There was a continuous passage of barium through the stricture, the outlines of which were smooth. The radiologist, in view of the luetic history and positive W.R., suggested that the narrowing was due to syphilitic fibrosis.

Œsophagoscopy was then performed. Immediately below the crico-pharyngeus, however, the passage of the instrument was held up by blood, which seemed to come from an area of rough, bleeding mucosa: suction was of no avail and the examination was abandoned. It was decided that he had an œsophagitis and he was treated with intramuscular penicillin. Another attempt at œsophagoscopy was then made, but with the same result.

As he was losing weight rapidly, a gastrostomy was performed. He made fairly good progress until about three weeks later, when he suddenly complained of dyspnœa. On re-examination, he was found to have expiratory and inspiratory stridor with rhonchi over both lungs. His larynx still showed only a slight

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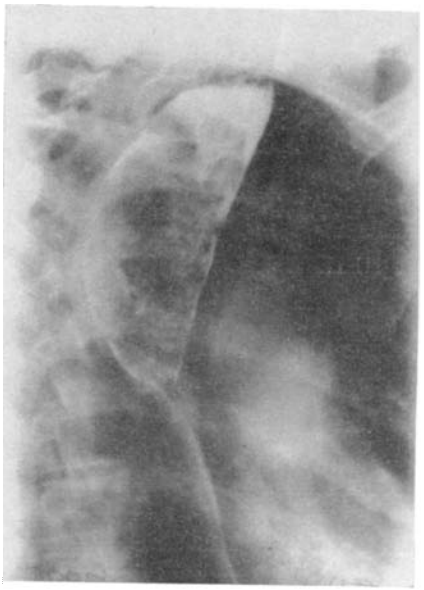


FIG. 1.

Radiograph after swallowing barium, showing the dilated œsophagus above an apparent stricture.



FIG. 2.

The œsophagus has been opened posteriorly, and the tumour drawn to one side to show its site of origin, and the smaller sessile tumours.

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diminution of movement of the right cord. Treatment with oxygen and ephedrine did not improve his condition and it was thought that he had tracheal obstruction as a result of his dilated œsophagus. A rubber catheter with sucker attached was passed down his œsophagus but produced nothing. Screening still showed the presence of the large swelling and there appeared to be some pulsation of it, which, in view of the positive W.R., raised suspicions of an aortic aneurysm. No further treatment was possible and he died the next day.

POST MORTEM

The heart, lungs and great vessels were normal.

The œsophagus was greatly dilated in its upper two-thirds. On opening it up posteriorly this was found to be due to the presence of a very large, pedunculated tumour, approximately 19 cm. long, which arose by a relatively small pedicle from the anterior wall just below the cricoid cartilage (Fig. 2). Its maximum diameter was about 6 cm. The surface was pitted, firm and greyish in colour.

In addition to this pedunculated tumour there were several sessile tumours arranged roughly in a ring near the base of the large tumour. Their average size was about 1·5 cm. in diameter. At one point the œsophageal wall had been eroded, and a tracheal ring was exposed.

The regional glands showed no evidence of invasion.

HISTOLOGY

Sections were made from the large tumour and from the secondary or multiple tumours near its origin. They all showed in general the same histological features (Figs. 3 and 4). The surface of the main tumour had lost its epithelium and was covered by very cellular granulation tissue and fibrin in which many collections of bacteria could be seen. Deep to this the tumour cells were seen to be spindle shaped, running side by side in fairly large, interlacing bundles. The nuclei in the better differentiated cells had long, parallel borders with slightly rounded ends, strongly suggestive of the nuclei of smooth muscle cells. There was, however, very considerable variation in the size, shape and staining reaction of the nuclei with moderate numbers of mitotic figures, indicating that the main tumour and the nodules were sarcomatous. Van Gieson and other connective tissue stains showed that the cells were forming only small amounts of collagen here and there in insufficient amounts to warrant a definite diagnosis of fibrosarcoma. Furthermore, no cells were found in any of the sections showing cross-striations. A diagnosis of leiomyosarcoma, probably arising in pre-existing multiple benign myomata, was therefore made.

Discussion

Nine other cases have been reported in the literature as leiomyosarcomata (Table I). They fall into two groups which are so markedly different in macroscopic appearance as to suggest that they may have a different ætiology.

Two cases were large, pedunculated tumours with no evidence of invasion at the base of the narrow pedicle, and no metastases. It seems probable that these were originally leiomyomata which grew to a large size, became pedunculated, and then underwent malignant change. The small pedunculated tumour was associated with carcinoma, and the histology was not described.

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TABLE I

CASES OF LEIOMYOSARCOMA OF ŒSOPHAGUS REPORTED IN THE LITERATURE

		Age	Sex	Type	Site
Howard	1902	51	M	Ulcerative	Lower third
Von Hacker	1908	—	—	Pedunculated	Lower third
Mallory	1935	58	M	Ulcerative	Lower third
Menne and Birge	1937	38	M	Ulcerative	Upper third
French and Garland	1941	70	M	Pedunculated	Lower third
Pennes	1942	53	M	Ulcerative	Middle third and upper part lower third
Harrington	1949	60	F	Ulcerative	Lower third
Harrington	1949	60	F	Ulcerative	Lower third
Case No. A361 University of Texas	1949	56	F	Small Pedunculated	Cardiac end, associated with carcinoma

The larger group of six cases were of the ulcerative, invasive type, and were either sessile or polypoid. Metastases to the lung were found in one case (Menne and Birge, 1937) and in all there was extension into surrounding structures. It is difficult to say whether these were primarily malignant, or secondary malignancy in a leiomyoma.

The separation of leiomyosarcomata from the other connective tissue tumours of the œsophagus, malignant and benign, is by no means easy, particularly the decision as to whether or not a leiomyoma has become malignant. The descriptions of some of the tumours reported are not entirely typical: in several cases there is no mention of the microscopical picture at all. Also, as French and Garland (1941) point out, some cases reported merely as spindle-celled sarcoma may in fact have been leiomyosarcomata. The actual incidence, therefore, is rather difficult to ascertain.

Site

The position of these tumours is usually at the lower end of the œsophagus, as smooth muscle predominates there. Menne and Birge (1937) report the only other case in the upper third. They point out that only the upper 2·5 cm. of the œsophagus is pure striated muscle: below this, smooth muscle fibres start to appear, but the possibility that smooth muscle tumours here may arise in misplaced, undifferentiated mesenchyme cannot be discounted.

Diagnosis

The signs and symptoms are those of any œsophageal neoplasm. Dysphagia is usually the first symptom, but in the case of pedunculated tumours, its onset may be delayed until the growth has reached a great size. Regurgitation of food appears later. Pain is more marked than in carcinoma and is substernal or in the back, occurring often at night. Dyspnœa and attacks of coughing may occur when the tumour is large.

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The diagnosis of the ulcerative group is fairly straightforward. A barium swallow shows narrowing and irregularity of the lumen, with filling defects: the appearance is that of a carcinoma, and final diagnosis is by biopsy.

The radiographic appearance in the large pedunculated tumours is misleading. There appears to be a smooth stricture of the œsophagus with dilatation

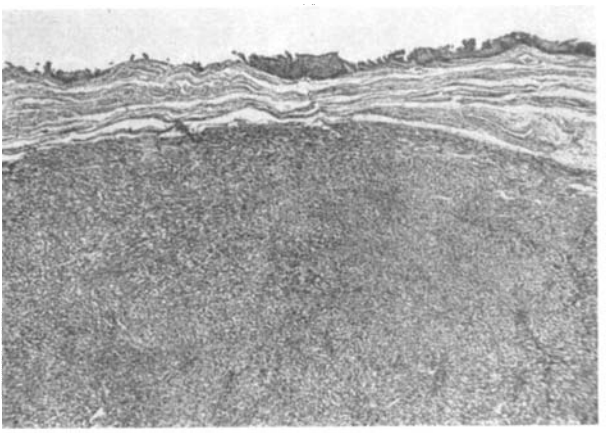


FIG. 3.

Microphotograph showing the general arrangement of cells, in large interlacing bundles. This section is from one of the smaller tumours and is covered with squamous epithelium. × 40.

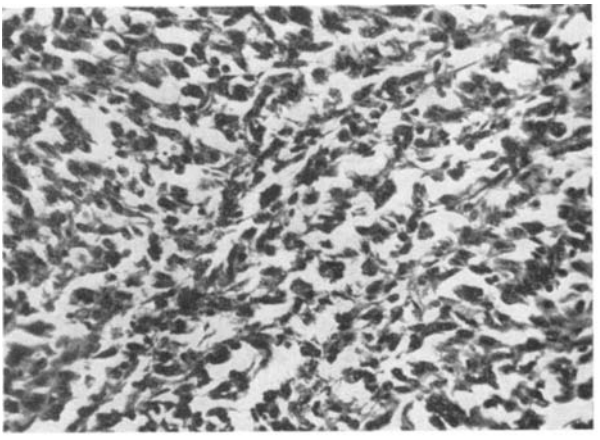


FIG. 4.

A high-power view of the same section to show the details of cell structure. × 400.

above containing food mixed with barium. Allison (1942) found it in his case of myoblastoma, describing it as a "peculiar soap-bubble appearance", and so did French and Garland (1941).

When the specimens are seen, it is easy to visualize the tumour in the radiographs, but the short history of dysphagia and the rarity of these large polyps

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makes an immediate correct diagnosis unlikely. The report on French and Garland's case suggested an examination after lavage would be necessary to confirm the diagnosis of stricture.

Œsophagoscopy is difficult when the polyp is at the upper end of the œsophagus, as the bleeding starts as soon as the crico-pharyngeal sphincter is passed. Allison was able to pass the instrument past the tumour blindly, and then visualize it while withdrawing the œsophagoscope, using suction. Even then he failed to realize that he was dealing with a pedunculated tumour.

From the point of view of operation, tumours in the lower third offer the best hope. Harrington (1949) successfully resected the growth in one of his cases and did an œsophago-gastro-anastomosis.

In the upper third, removal of a polypoid growth might be possible through a left cervical œsophagotomy, but in the present case, delivery of the growth without tearing the œsophagus would have been almost impossible.

Since this paper was written, Dr. E. A. Horne has lost his life in an accident.

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