Pharyngeal leiomyosarcoma

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Malignant neoplasms of smooth muscle origin are rare. In a review of 200 cases of soft tissue sarcoma, Hare and Cerny (1963) found only 13 (6·5 per cent) leiomyosarcomas; the uterus, broad ligament and gastro-intestinal tract being the areas most often involved, and the stomach being the main site in the gastro-intestinal tract.

In the pharynx, only two cases of leiomyosarcoma have been reported, those of Fuller *et al.*, (1966), and Mankodi *et al.*, (1970).

The patient of Fuller *et al.*, presented with a 'lump in the throat', following negative investigation for a pyrexia of unknown origin. The tumour was a smooth spherical mass on the posterior pharyngeal wall, and, after diagnosis by frozen section, was removed via lateral pharyngotomy.

The leiomyosarcoma was spherical, 3 cm. in diameter, and arose from a pedicle. It consisted primarily of spindle cells, and there was a uniform pattern of mainly straight and parallel reticulin fibres throughout.

The patient of Mankodi *et al.* presented with hoarseness and dysphagia; the tumour was again a solitary lesion, 4 × 3 cm. arising from the posterior hypopharyngeal wall. The diagnosis of leiomyosarcoma was made on the operative specimen, removed by laryngo-pharyngo-cervical oesophagectomy.

Microscopy again showed spindle-shaped cells, accompanied by straight reticulin fibres arranged in interlacing bundles. Myofibrils could be demonstrated in the cytoplasm. The tumour was covered by a mucosal epithelium showing marked hyperplasia.

It is the purpose of this paper to present a third case, to compare the presenting features and management of these three cases, and to outline the pathology.

Case report

The patient was a 65-year-old female with a one month's history of excessive nasality of speech, nasal regurgitation and dysphagia for solids.

Clinical examination revealed a spherical mass in the hypopharyngeal region. It was on an obvious pedicle, was excessively mobile, and appeared midline. A lateral soft tissue X-ray of the neck showed the mass apparently attached to the posterior pharyngeal wall, adjacent to cervical vertebrae 3 and 4. A barium swallow (Fig. 1) confirmed the plain film and further outlined the mass. Angiography was not indicated since the mass was midline.

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The patient was placed in the tonsillectomy position, and using a Boyle-Davis gag with general anaesthesia, a more detailed inspection was possible. The mass arose from a small pedicle on the posterior pharyngeal wall in the midline. It was slightly larger than a golf ball, spherical, freely mobile, of firm consistency and had an apparently intact mucosal surface; that is, similar in appearance and site to the two leiomyosarcomas previously described.

The presumptive diagnosis was that of a fibroma or leiomyoma, that is, of a benign lesion. In part, the reason for this, besides its clinical features, was that a tumour of such magnitude, if a carcinoma, would almost certainly have had nodal metastases whereas there were in fact no palpable nodes. It was thus removed per-orally as an excision biopsy by clamping and then ligating the pedicle, removing the tumour and applying diathermy to the base. Postoperative recovery was uneventful.

The histology indicated leiomyosarcoma. The aim therefore was to remove any remnant of the tumour by a lateral pharyngotomy. However, although



FIG. 1.
Barium swallow; showing tumour outlined.

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only one week later, it was difficult to ascertain precisely the extent of the sarcomatous involvement of the pharynx due to the rapid healing of the area involved. Because of this, a per-oral removal with excision diathermy was chosen in preference to the more desirable lateral pharyngotomy approach.

The patient remains well and there is no evidence, one year later, of any recurrence.

The specimen was a smooth-surfaced oval mass of tissue, 30 × 30 × 15 mm. (Fig. 2) with, at one point, an approximately circular hollow or pit, 5 mm. across and 5 mm. deep, having the appearance almost of a hilum (Fig. 3). This was the original site of attachment, the 'hilum' being in fact a completely retracted short pedicle.

Sections showed a mass of neoplastic tissue with a well-vascularized and oedematous pedicle containing scanty glands of salivary type and a few skeletal muscle fibres. The surface was wholly ulcerated and no longer bore epithelium.

In general the pattern was that of a highly cellular, markedly polymorphic but essentially spindle-celled neoplasm: (Figs. 4, 5) the differential diagnosis appearing to comprise leiomyosarcoma, fibrosarcoma, rhabdomyosarcoma and spindle-cell carcinoma. In analysis of this selection, the following staining procedures were used: a reticulin method, phosphotungstic acid haematoxylin and van Gieson.

Reticulin staining showed an essentially pericellular pattern throughout and was regarded as excluding the diagnosis of carcinoma virtually completely.

Phosphotungstic acid haematoxylin was used in a search for cytoplasmic

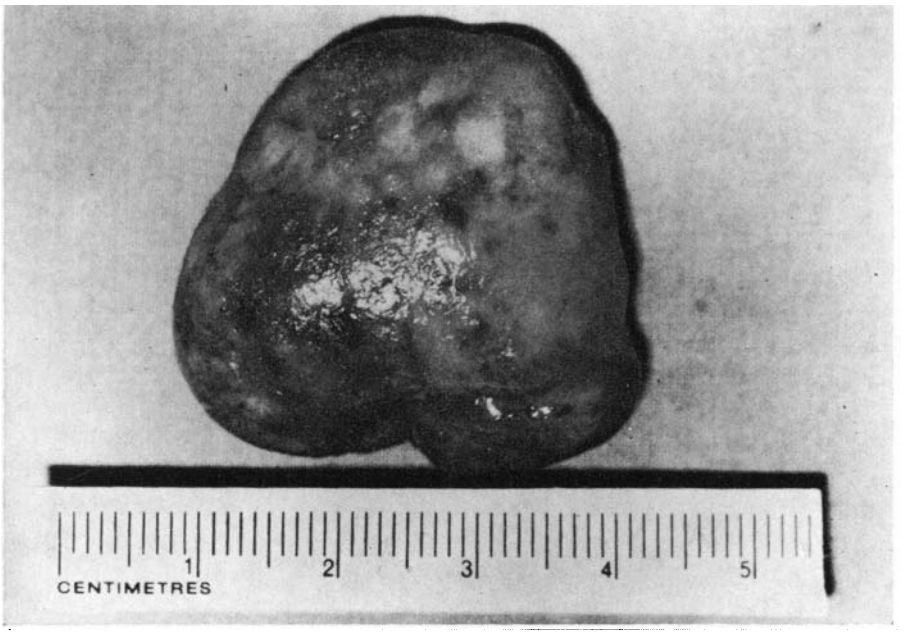


FIG. 2.
Specimen; showing the apparent smooth surface.

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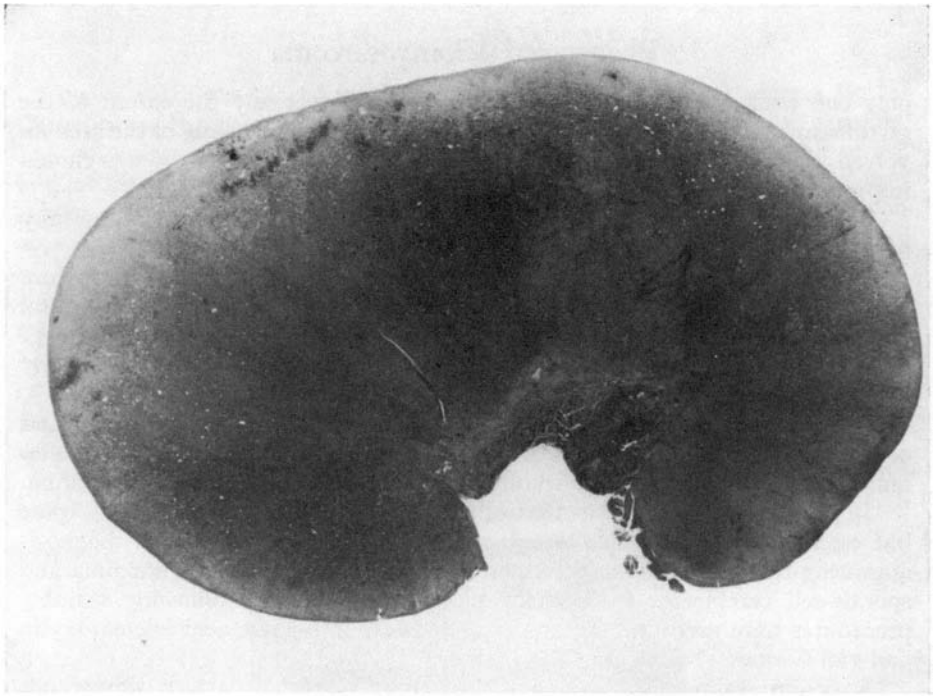


FIG. 3.
A section through the middle of the mass. The 'hilum' is the deeply retracted pedicle. H. & E. × 6.

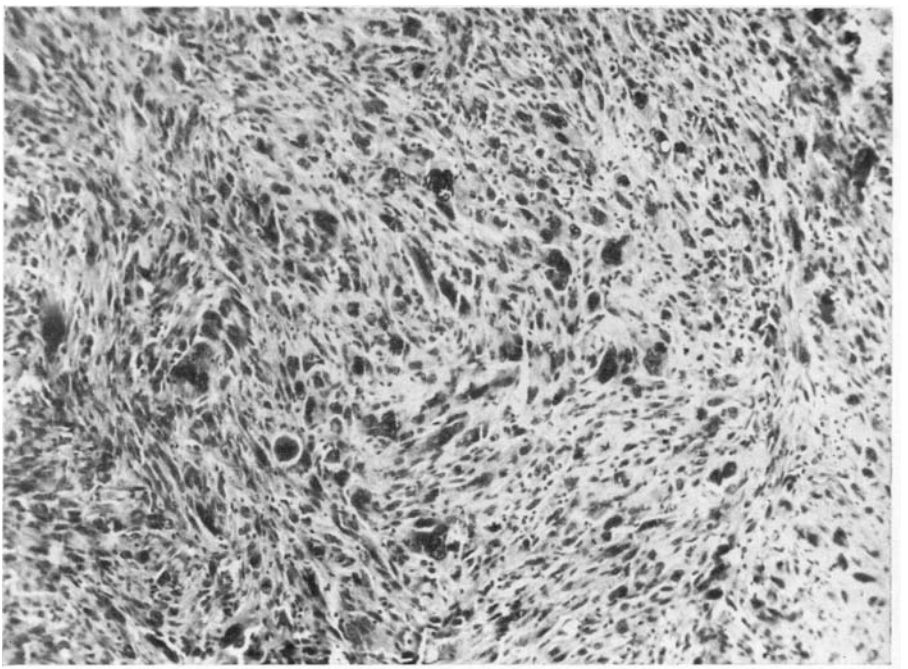


FIG. 4.
An area showing marked cellular polymorphism. This was the predominant pattern. H. & E. × 140.

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striations amongst the abundant large polymorphic cells. No striations were found after prolonged search; positive evidence of rhabdomyosarcoma was therefore lacking.

The van Gieson method showed for the most part an intimate mixture of red-staining collagenous fibres and yellow or neutral-staining non-collagenous spindle cells. No firm conclusion could be reached on the nature of the many neutral-staining cells but in many areas where the cells were forming relatively well differentiated fascicles the staining strongly suggested a smooth muscular nature. Also, not infrequently, the bizarre giant cells gave a yellowish reaction strongly suggesting that they too were essentially muscular.

The preferred diagnosis was therefore *polymorphic leiomyosarcoma*.

Tissue obtained from the second and definitive per-oral removal comprised two specimens. Firstly, a single piece of tissue, 6 mm. across, from the site of excision of the main mass. It showed only inflammatory changes and no residual neoplasm in the many sections. Secondly, two pieces of tissue, 8 mm. and 6 mm. across, also from the posterior wall of the pharynx.

Sections from the latter showed, as expected, much inflammatory coagulum, cell debris and granulation tissue, but also, quite unexpectedly, areas of aberrancy in the superficial epithelium satisfying all the morphological criteria of carcinoma *in situ* (Fig. 6). There was no identifiable remnant of the earlier-removed spindle-celled neoplasm.

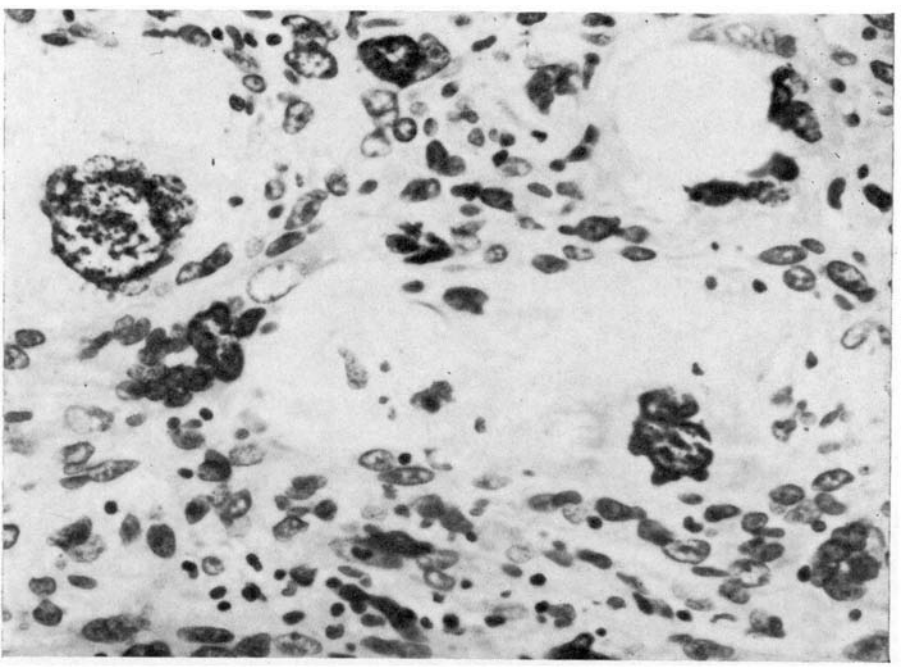


FIG. 5.
This shows that the polymorphism is produced by a combination of irregularity in nuclear size, the presence of many nuclei in some single cells, and (the two largest masses of chromatin) highly aberrant mitotic figures.
H. & E. × 560.

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The problems raised by this unusual conjunction of histological patterns are clear: (a) is the area of carcinoma *in situ* quite certainly so? (b) is the first-removed mass not in fact a sarcoma but a carcinoma? (c) if it is not a carcinoma but indeed a sarcoma, is there any explanation for their co-existence other than pure chance?

No certain answer can be given to these questions but the problem can at any rate be assessed. On purely histological evidence, the staining behaviour of the main mass is almost totally unacceptable as indicating carcinoma. Next, nowhere is there any tissue intermediate in appearance between the apparent carcinoma *in situ* and the highly polymorphic malignant neoplasm, and this adds further doubt. Either, then, the conjunction must be accepted as a curious coincidence or the diagnosis of carcinoma *in situ* must be looked at with increasing suspicion. The tissue in question was not irradiated but it was cauterized, and in this lies at least a possible explanation. The apparent carcinoma *in situ* could just conceivably be a highly aberrant reparative reaction or an unusual hyperplastic regenerative overgrowth.

Discussion

A leiomyosarcoma of the upper air and food passage is a rarity. Ash, Beck and Wilkes (1964) have no record of an unequivocal leiomyosarcoma of the pharynx.

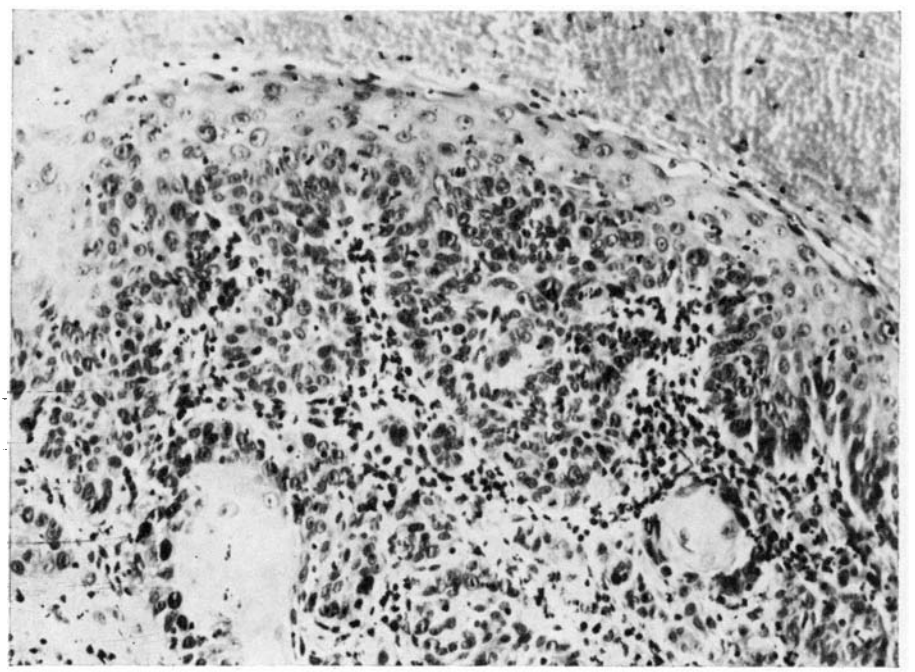


FIG. 6.
From the second biopsy. There is much irregular overgrowth of the pharyngeal epithelium with a degree of nuclear polymorphism and mitotic activity highly suggestive of carcinoma *in situ*.
H. & E. × 280.

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The feature in common to this and the two earlier-reported cases is that the lesion was a firm almost spherical mass with a smooth surface and relatively mobile pedicle, these last two characteristics suggesting a benign nature. The presenting symptoms varied widely from hoarseness and dysphagia in Mankodi's case; pyrexia of unknown origin, weight loss, and the feeling of 'something in the throat' in Fuller's case; to that of hypernasality and nasal regurgitation in our own.

A hypopharyngeal polyp may be asymptomatic. When symptoms are produced, they are essentially those of a space occupying lesion in a hollow structure. Depending on location there may be obstruction causing nasal regurgitation, dysphagia and/or stridor; pressure on and displacement of surrounding structures causing hoarseness, overspill, hypernasality, the 'feeling of a lump'; or systemic effects such as pyrexia, weight loss and fatigue, secondary to rapid growth with necrosis and infection.

Sohn and Feuerstein (1967) describe a fibromatous polyp of the hypopharynx completely unsuspected and asymptomatic, that projected suddenly from the mouth during the course of electro-convulsive therapy. These authors discuss the clinical silence of such a hypopharyngeal polyp and attribute it to the distensibility of the upper oesophagus. In these three cases of leiomyosarcoma the tumour was already of considerable size when diagnosed.

The differential diagnosis of a hypopharyngeal polyp must include lesions arising in the retropharyngeal or parapharyngeal space. These *extrinsic pharyngeal lesions*, such as neoplasms of the deep lobe of the parotid, chemodectoma, branchial cysts, neurilemmoma of the vagus or sympathetic trunk, chronic retropharyngeal abscess may give the appearance of a sessile polyp. *Intrinsic pharyngeal lesions* must include neoplasms both benign and malignant in the diagnosis, this case demonstrating the difficulty of making a clinical diagnosis of a benign tumour.

Clinical assessment without an anaesthetic is unrewarding, as all that can be seen is a 'large swelling'. Radiography only confirms this, although angiography will help in excluding the extrinsic lesions. There were no palpable nodes in the three reported cases of leiomyosarcoma. Direct examination under general anaesthesia is essential for accurate assessment of the site of origin of the tumour, its appearance, consistency, vascularity, relationship to surrounding structures, and of its encroachment or fixation, if any, to the prevertebral region or the parapharyngeal structures. The use of a Boyle-Davis gag is recommended as it allows the base of the tongue to be retracted from the posterior pharyngeal wall.

The three reported lesions have each been removed by a different approach; lateral pharyngotomy, radical pharyngectomy and per-orally. The latter is by far the simplest and quickest, and radical surgery is to be deprecated.

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The lateral pharyngotomy approach allows better assessment of the pedicle length, of its attachment, and of any encroachment beyond the pharyngeal wall. It therefore allows for more complete removal and safer control of haemorrhage. A frozen section diagnosis prior to the removal via a lateral pharyngotomy is advisable, but if these facilities are not available then a routine biopsy report should be obtained prior to removal by this approach. It is advantageous with a pedunculated tumour to do an excision biopsy per-orally with clamping and then ligation of the pedicle.

The healing of the area of excision after removal of the polyp was so rapid and complete in our patient, that the planned lateral pharyngotomy was contra-indicated.

Radiation therapy should not categorically be deemed useless in dealing with these tumours; it may be of benefit in the pre-operative management of very large leiomyosarcomas—as reported in the oesophagus—as well as in palliation (Wolfel, 1963). The question of the use of radiotherapy, however, did not arise at any time in this third case report.

A wait and see policy was adopted, despite the second report of carcinoma *in situ*. It appears, so far at any rate, that this policy has been correct.

Summary

A third case of pharyngeal leiomyosarcoma is presented. The clinical features of polypoidal pharyngeal lesions are discussed, together with a résumé of an adopted mode of therapy. The histopathological diagnosis is also discussed.

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