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Parapharyngeal space tumour presenting as recurrent uvular oedema

M. J. WAREING, F.R.C.S., R. M. IRVING, F.R.C.S., D. A. MOFFAT, B.Sc., M.A., F.R.C.S. (Cambridge)

Abstract
Neoplasms of the parapharyngeal space are uncommon and usually present as an intra-oral or neck mass. They often elude early diagnosis due to their deep-seated nature. Here we report a case presenting with recurrent oedema of the uvula. The pathophysiology of this previously unreported mode of presentation is discussed.

Key words: Neck, parapharyngeal space; Neoplasm; Uvula, oedema

Case report

A 41-year-old caucasian male printworker presented via the accident service with a two-hour history of breathing difficulties and the sensation of something obstructing his throat. This had started whilst he had been working with printing dyes to which he had previously been exposed with no ill-effects. He had previously suffered an urticarial rash following exposure to packing material. There was no other relevant medical history.

On examination he was afebrile with gross uvular and accompanying soft palatal oedema accounting for his muffled speech. A diagnosis of angioneurotic oedema was made and he was treated with intravenous dexamethasone and chlorpheniramine with some resolution of the oedema. He was sent home on a reducing dose of prednisolone with arrangements for follow-up in the allergy clinic.

He represented, however, five days later with an increasingly sore throat and dull aching right preauricular pain and on examination again had an oedematous soft palate and uvula but this was accompanied by tonsillar hyperaemia and tender cervical lymphadenopathy. Flexible nasendoscopy showed a reddened postnasal space and a presumptive infective pharyngitis was treated with intravenous broad-spectrum antibiotics. Little improvement in either the palatal oedema or the facial pain occurred on this regimen.

Investigations carried out showed a leucocytosis of 14.7×10^9/l and an ESR of 2 mm/hr. Immunoglobulin and C1 esterase inhibitor assays were within the normal range. Plain temporo-mandibular joint X-rays showed some alteration of the articular surface of the mandible. In view of this and the unresolving nature of the problem a CT scan was performed which revealed an encapsulated homogenous soft tissue mass filling the right infratemporal fossa, bowing the posterior wall of the maxillary antrum forward (Fig. 1). There was no enhancement on intravenous contrast injection and the relatively avascular nature of the tumour was confirmed on carotid arteriography as no tumour blush was seen.

The tumour was resected via an anterior approach by subtotally excising the maxilla to gain access to the pterygo-palatine and infratemporal fossae. The tumour was found to be filling the superior part of the infratemporal fossa thus pushing the posterior antral wall forward (Fig. 2). The maxilla was wired back into place at the end of the procedure. Initial frozen section histological examination reported this to be a lymphoma but extensive paraffin section review showed this to be a neuroendocrine carcinoma although the site of origin was unclear.

Discussion

Tumours of the parapharyngeal space are uncommon and their deep-seated nature can allow them to attain a considerable size before becoming symptomatic. The commonest presenting feature is a mass either within the oral cavity or externally in the neck. Early presentation may occur as a result of involvement of the Vth or IXth to XIIth cranial nerves or the eustachian tube within the parapharyngeal space. This can result in facial pain or dysaesthesia, dysphagia, dysphonia or deafness (Johnson *et al.*, 1989). Uvular oedema has not previously been described as a presenting symptom but in this case may have arisen as a result of vascular and lymphatic obstruction in the parapharyngeal space.

The lymphatic drainage of the soft palate is to the deep cervical group of lymph nodes, either directly or via the retropharyngeal or parotid lymph nodes. Venous drainage is to the peritonsillar and pterygoid venous plexi (Warwick *et al.*, 1989).

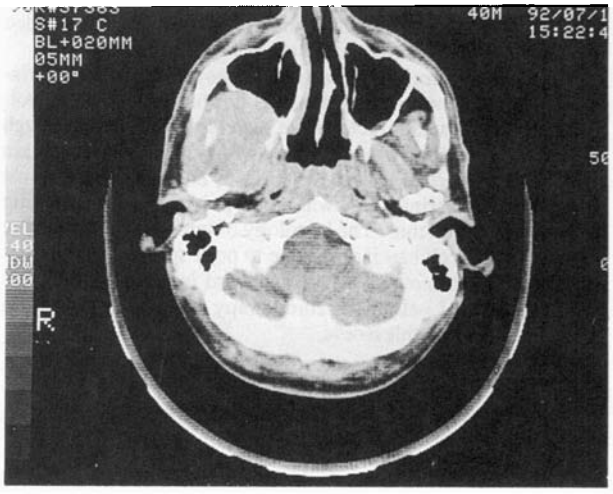

FIG. 1
Transaxial CT scan showing homogenous soft tissue mass filling the right infratemporal fossa.

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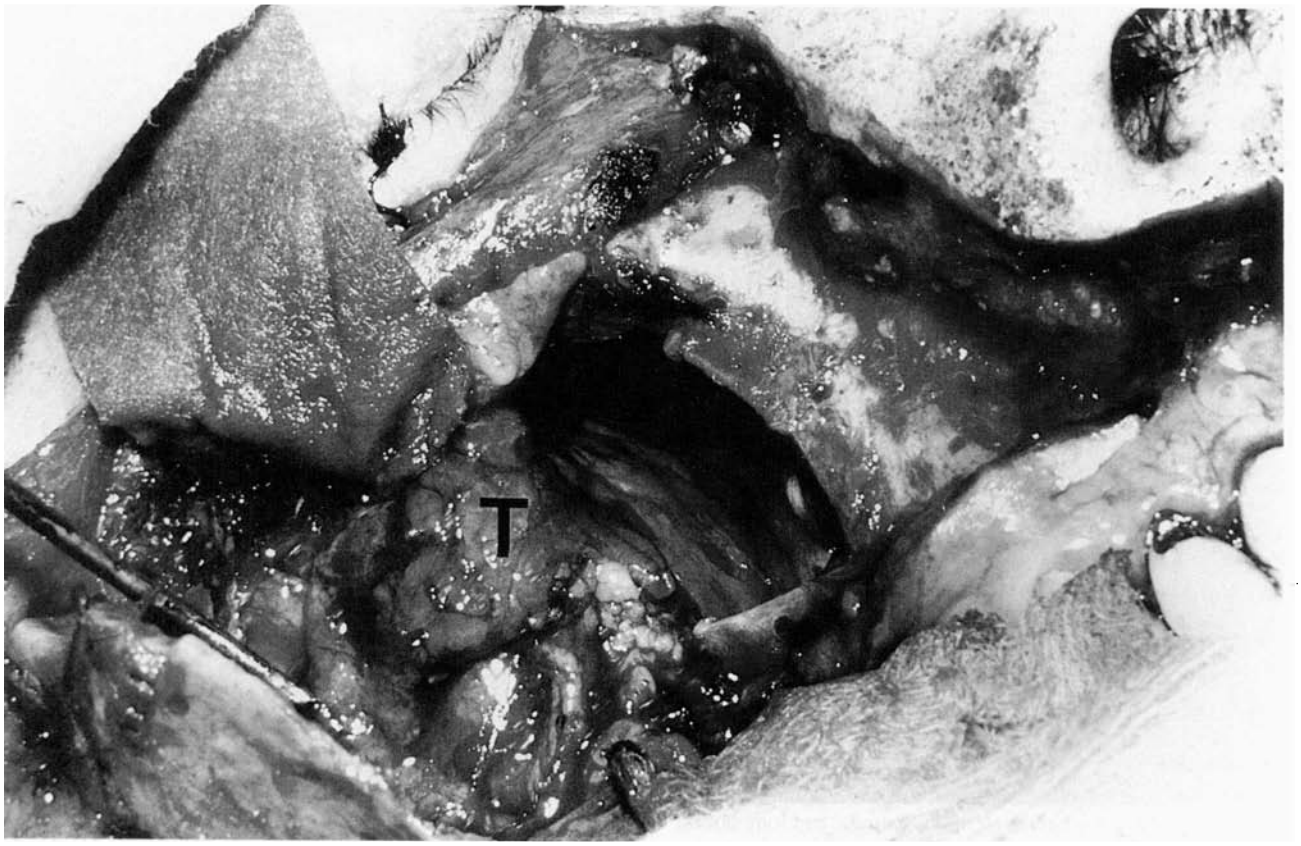

FIG. 2
Intraoperative photograph showing tumour mass (T) and forward displacement of posterior antral wall.

Improvement in the patient's condition with the administration of steroids may be explained by a reduction in peritumoural oedema alleviating lymphatic and venous obstruction. This improvement with steroids led to a mistaken diagnosis of allergic or angioneurotic oedema.

There are numerous causes of uvular oedema with it being seen most commonly in infective conditions of the pharynx. It is less commonly seen as part of angioneurotic or Quincke's oedema which is characterized by well demarcated non-pitting oedema of the skin and mucous membranes, and it should be noted that isolated uvular oedema due to an often unidentified allergen is much more frequently seen than the rare hereditary angioneurotic oedema (which shows a widespread anatomical distribution) (Merigan *et al.*, 1992). Other causes of uvular oedema are trauma including endotracheal intubation and surgery, drugs such as contrast media and hashish, certain blood disorders such as leukaemia and agranulocytosis and a miscellaneous group including aphthous ulceration and Behcet's syndrome (Hawke and Kwok, 1987; Hibbert, 1987). Uvular oedema is a rare but well recognized cause of respiratory embarrassment (Evans and Roberge, 1987), emphasizing that examination of the oral cavity and oropharynx should not be overlooked.

Conclusion

This case emphasizes the obtuse presentation of a parapharyngeal space neoplasm. It was only the development of additional symptoms that led to the correct diagnosis. We suggest that all patients with recurrent atypical uvular oedema should be investigated in order to exclude underlying malignancy.

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Address for correspondence:
Mr M. J. Wareing,
Department of Otolaryngology,
Addenbrooke's Hospital,
Hills Road,
Cambridge CB2 2QQ.