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Retro- and para-pharyngeal ganglioneuroma

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Abstract
A patient with slight dysphagia and dyspnoea on exertion was referred to us. We diagnosed a large retro- and para-pharyngeal ganglioneuroma. The mass was surgically removed *in toto*. The case is presented and the features are discussed.

Key words: Head and neck neoplasms; Ganglioneuroma

Introduction

Ganglioneuromas of the retro- and para-pharyngeal space are a rare entity compared to other benign neural tumours. These unusual benign tumours arise from the sympathetic chain, the most common locations being mediastinal, retroperitoneal and adrenal (Brandenburg, 1972; Enzinger and Weiss, 1988; De Heyn, 1990; Joshi *et al.*, 1992).

Ganglioneuromas result from the maturation of neuroblastoma cells into ganglion cells and may develop spontaneously or after chemotherapy or radiation of neuroblastoma (Hayes *et al.*, 1989). The age distribution shows a maximum between 10 and 29 years (Enzinger and Weiss, 1988; Hayes *et al.*, 1989; De Heyn, 1990). Cases of malignant transformation have been reported (Chandrasoma *et al.*, 1986; Enzinger and Weiss, 1988).

Case report

A 57-year-old male patient was referred with moderate dysphagia and dyspnoea on exertion. Clinical examination showed a slight bulge of the posterior wall of the hypopharynx. The mobility of the left vocal fold was reduced. The patient was in good general condition and routine laboratory values were normal. Ultrasonography showed a well circumscribed, homogenous, hypoechogenic mass pre-vertebrally without echolucent components. During swallowing studies a widening of the pre-vertebral soft tissue shadow with a ventral displacement of the larynx and the trachea of up to 4.5 cm was revealed. CT scan and magnetic resonance imaging (MRI) showed a retro- and parapharyngeal mass with a maximum width of 9 cm, a maximum anteroposterior diameter of 4.5 cm and a maximum craniocaudal extension of 19 cm (Figure 1). The expansion extended cranially up to the level of the soft palate and the atlas, the jugulum and laterally on both sides to the carotid neurovascular bundle, displacing the left one. There was a ventral displacement of pharynx, larynx, oesophagus and trachea. On CT, the lesion showed a density unequivocally neither in the cystic nor in the soft tissue range (35 HU) with minimal enhancement of contrast media (44 HU). On MRI the lesion showed intermediate signal intensity on T1-weighted sequences, a

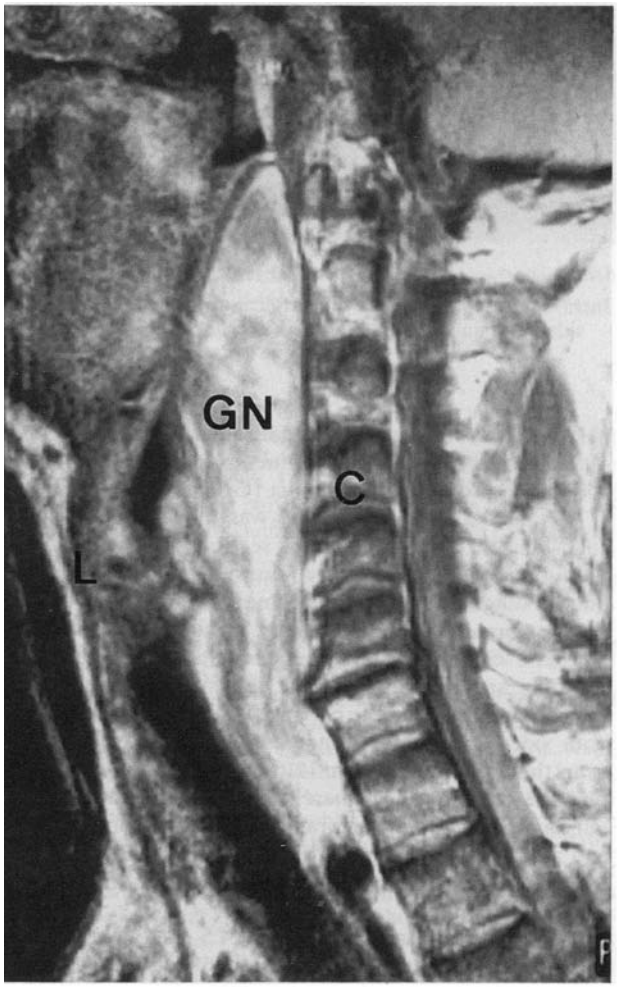

FIG. 1
MRI sagittal section of the neck. GN: ganglioneuroma; C: cervical vertebral column; L: larynx.

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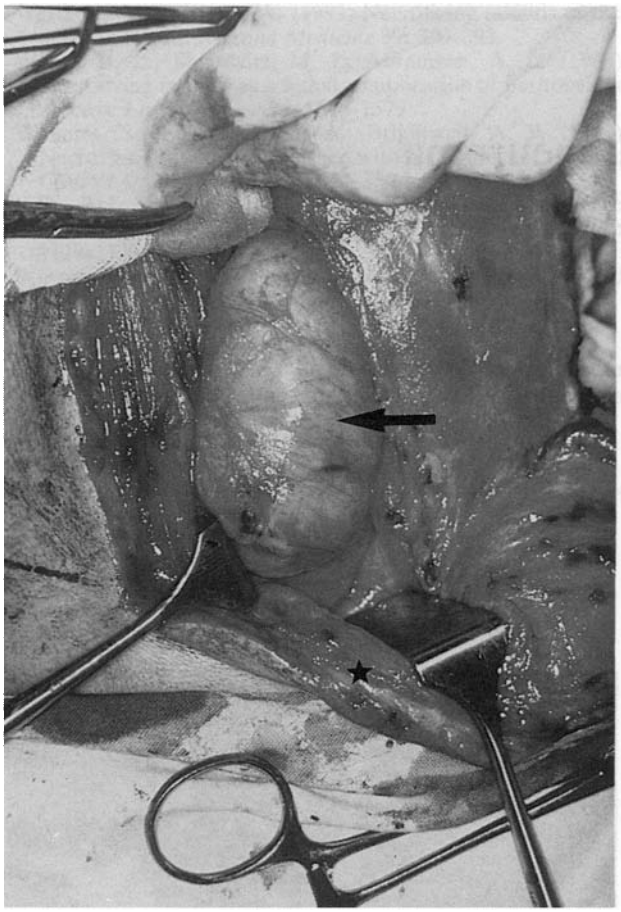

FIG. 2
Intra-operative picture of the surgical approach from the left side. ←: ganglioneuroma; *: M. sternocleidomastoideus.

heterogenous hyperdensity/enhancement on T2-weighted sequences and a non-homogeneous enhancement after administration of contrast media (GdDTPA).

A transoral incisional biopsy was taken under general anaesthesia. The immediate section for microscopic examination under surgery showed a benign neural tumour diagnosed as neurofibroma. The surgical approach was performed without delay by a modified (U-shaped) Kocher's collar incision. The encapsulated tumour was carefully approached from the left side (Figure 2) and easily separated from the perivascular and neural sheath of both sides, anteriorly from the pharynx and posteriorly from the vertebral column. In the caudal third on the left side the tumour was adherent to a small neurovascular bundle which contained a branch of the thyrocervical trunk and a branch of the sympathethic trunk (Figure 3). Three enlarged jugulodiagastric lymphatic nodes on the left side were also removed.

The tumour measured 19 × 9 × 4.5 cm (Figure 4) and was soft. Its pale cut surface resembled fish meat.

Histopathology of the resected specimen revealed a tumour composed of clusters of ganglion cells contained within a background of Schwann cell bundles, the typical picture of a ganglioneuroma (Figure 5). The lymphatic nodes showed a non-specific lymphadenitis.

The post-operative period was uneventful apart from some oedema of the larynx on the fourth post-operative day, treated by parenteral application of corticosteroids. The patient was discharged home in good condition on the eighth post-operative day.

On follow-up after three months the patient was asymptomatic. An MRI scan showed only post-operative scarring, there was no sign of recurrence. The larynx showed normal mobility of both vocal folds without oedema.

Discussion

Ganglioneuromas are fully differentiated tumours with no immature elements, composed of ganglion cells, Schwann cells accompanying the neuritic processes, and fibrous tissue. The presence of any immature elements would negate the diagnosis of ganglioneuroma (Enzinger and Weiss, 1988; Joshi *et al.*, 1992; Shotton *et al.*, 1992).

Neurogenic tumours comprise between 32 per cent (Work and Hybles, 1974) and 40.5 per cent (Shoss *et al.*, 1985) of parapharyngeal space lesions. Salivary gland tumours are more common (48 per cent (Work and Hybles, 1974)), (38.1 per cent (Shoss *et al.*, 1985)). Seventy to 80 per cent of the parapharyngeal space tumours are benign lesions (Work and Hybles, 1974; Shoss *et al.*, 1985). Our patient, aged 57 years, was outside the usual maximum of age distribution (10–29 years) (Enzinger and Weiss, 1988; De Heyn, 1990).

Ganglioneuromas of the parapharyngeal space are uncommon and rarely reported. They were not reported

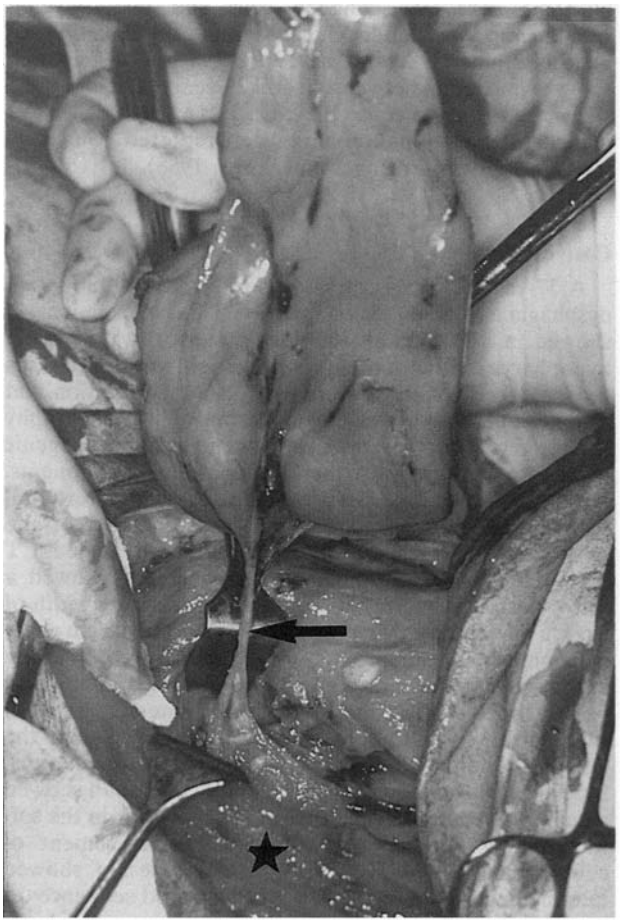

FIG. 3
Mobilized tumour adherent to a small neurovascular bundle. ←: neurovascular bundle; *: M. sternocleidomastoideus.

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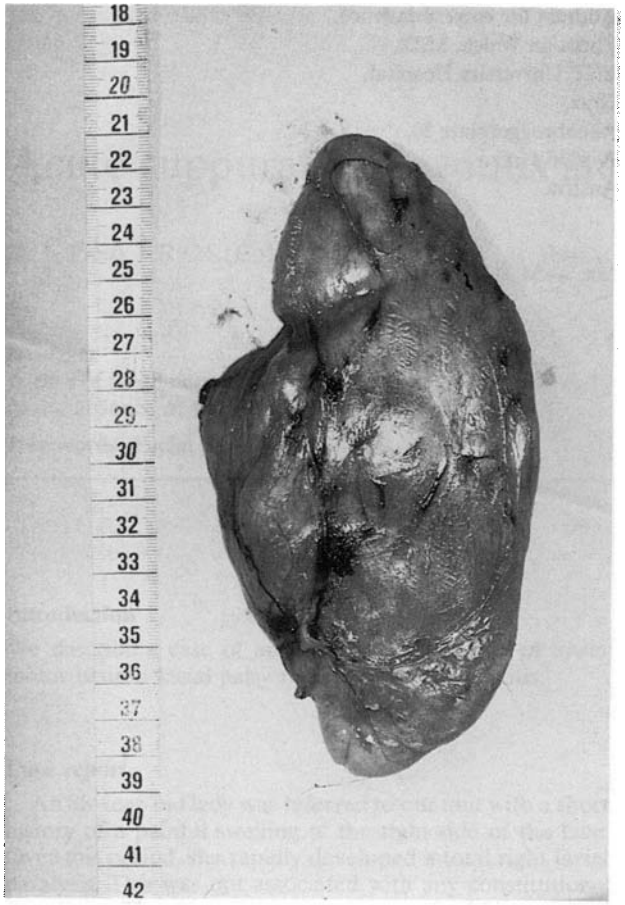

FIG. 4
Resected ganglioneuroma sized 19 × 9 × 4.5 cm.

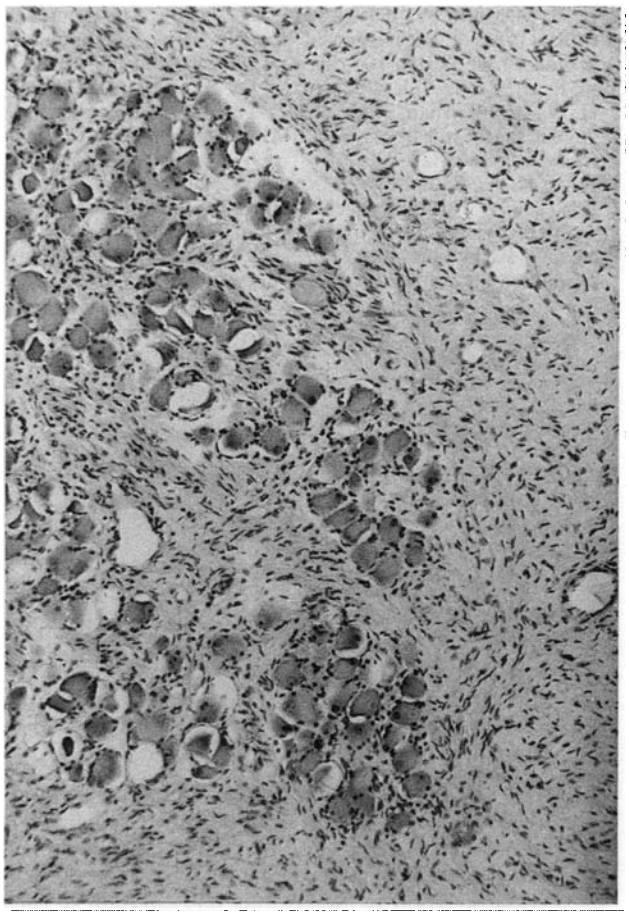

FIG. 5
Typical histological picture of the ganglioneuroma with clusters of ganglion cells (H & E; × 160).

by Som *et al.* (1984; 1987) in two comprehensive studies. When they do occur, displacement of the medial wall of the parapharyngeal space is pathognomonic (Maran *et al.*, 1984) as found in our patient.

MRI is an established technique which is becoming increasingly important in the diagnosis, treatment planning and follow-up of ENT tumours. The soft tissue contrast demonstrated by MRI is superior to that obtained with CT scanning (Langnickel and Held, 1992).

Intraoral incision biopsies may be contra-indicated in the diagnosis of parapharyngeal lesions as significant haemorrhage may be encountered. However, the decision to proceed to complete removal had already been made and the position of the carotid sheath identified by pre-operative scanning. Under these circumstances it is an acceptable technique.

Although it has been reported that a mandibulotomy is required if the tumour extends to the skull base or if its vertical diameter exceeds 8 cm (Tandon *et al.*, 1992) or that resection may even be impossible (Hayes *et al.*, 1989), in our case surgical excision was performed *in toto* without mandibulotomy.

Cases of malignant transformation of ganglioneuroma have been reported (Enzinger and Weiss, 1988), nevertheless the prognosis of our patient is very good because the tumour was totally removed.

Totally surgical resection and follow-up are the best way to treat a ganglioneuroma.

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