*The Journal of Laryngology and Otology*
September 1998, Vol. **112,** pp. 883–887

# Malignant schwannoma of the parapharyngeal space

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**Abstract**
We present a case of malignant schwannoma (MS) of the parapharyngeal space which completely encircled the internal carotid artery in close proximity to the skull base and required resection of the internal carotid artery along with the excision of the tumour. There have been three previous case reports in the literature of malignant schwannoma of the parapharyngeal space of which one case was associated with neurofibromatosis. Our patient did not exhibit any feature of neurofibromatosis.

**Key words: Neoplasms; Schwann cells; Carotid artery, internal; Surgery, operative**

## Case report

A 22-year-old male patient was referred from an outside hospital to King Faisal Specialist Hospital and Research Centre. According to the referral note he had presented to them initially with one-year history of hoarseness of voice and a painless swelling on the right side of his neck. At the time of examination he had a right vocal fold paralysis. Examination of the neck showed a 7 × 3 cm swelling in the upper part of the right side of the neck. Apart from this there were no positive clinical findings. After carrying out a computerized tomography (CT) scan a provisional diagnosis of schwannoma of the vagus nerve was made and excision of the lesion was carried out. This was reported as an incompletely excised malignant schwannoma. A post-operative CT scan showed residual tumour and it was at this point that the patient was referred to us for further management.

At the time of presentation to our hospital the patient complained of hoarseness and a swelling on the right side of the neck. He had no other complaints. Clinical examination showed right vocal fold and right palatal paralysis. No other cranial nerves were involved. Intraoral examination did not show any medialization of the tonsillar area. Neck examination showed a scar on the right side from the previous surgery. However apart from some fullness of the neck on the right side no definite neck mass could be palpated. The rest of the clinical examination was normal. CT scan with contrast of the neck showed a non-enhancing lesion in the post-styloid compartment of the right parapharyngeal space in close contact with the major blood vessels (Figure 1) and extending from the skull base to the carotid bifurcation. Review of the histopathology slides from the referring hospital confirmed the diagnosis of malignant schwannoma. A full metastatic workup including chest X-ray, bone scan and ultrasound of the abdomen did not reveal any distant metastasis.

After discussion with the patient a full course of radiotherapy was given. It was also explained to the patient that if the lesion did not regress with radiotherapy then a surgical excision would be required. The patient received 6600 cGy of external beam radiation in 30 fractions. Assessment at three months following radiotherapy both clinically and by CT and magnetic resonance imaging (MRI) (Figures 2 and 3) showed no regression in tumour size. A positron emission tomography (PET) scan

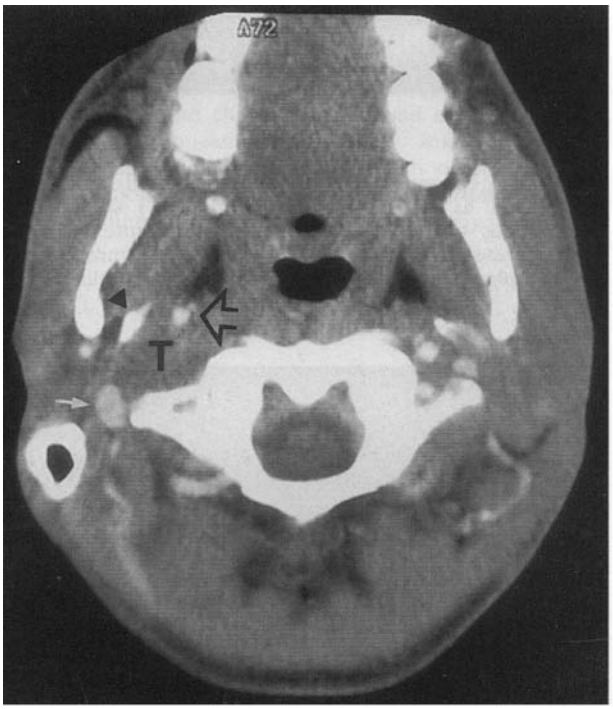


Fig. 1
Contrast enhanced CT-scan through the neck at the level of C1. A non-enhancing tumour mass (T) is found in the widened post-styloid compartment of the parapharyngeal space. The internal carotid artery (black arrow) and the internal jugular vein (white arrow) are splayed apart by the tumour.

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Accepted for publication: 30 June 1998.

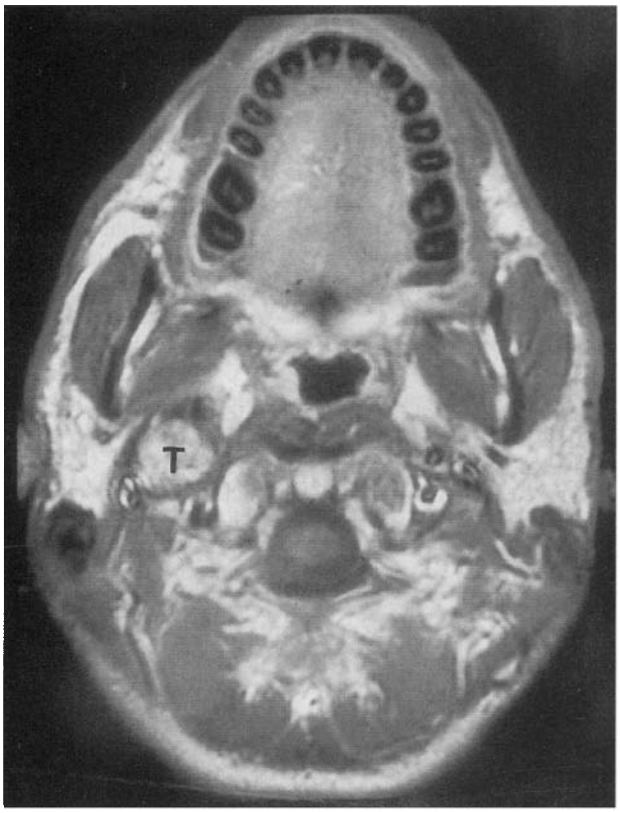


FIG. 2

Gadolinium enhanced T-1 weighted axial MRI scan at the same level as Figure 1. The tumour (T) shows non-homogenous contrast enhancement and because of better contrast resolution between the various structures of the parapharyngeal space the tumour is better outlined.

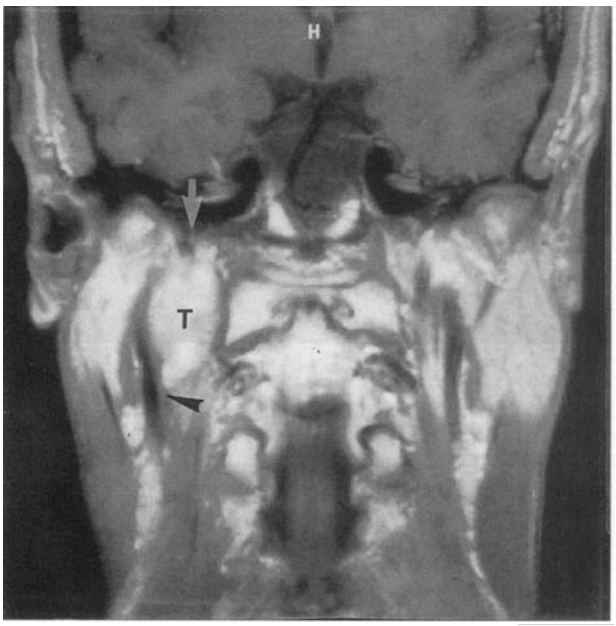


FIG. 3

Gadolinium enhanced T-1 weighted coronal MRI scan through the parapharyngeal space shows the enhancing tumour (T) in close contact with the skull base (white arrow) and also is seen causing lateral displacement and compression of the internal carotid artery (black arrow head).

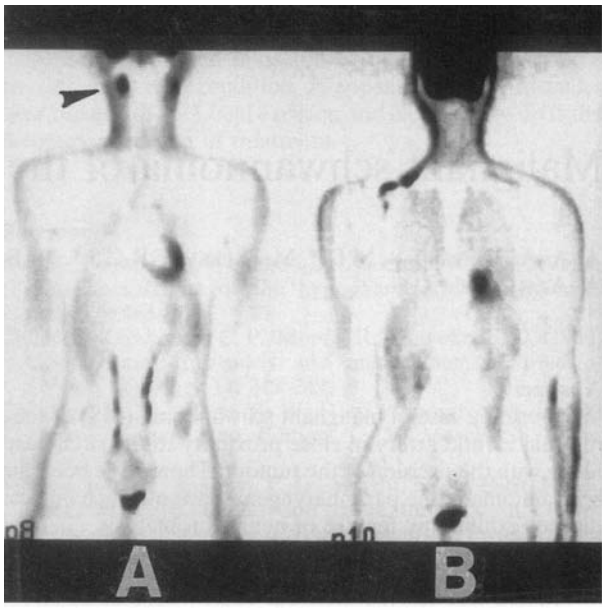


FIG. 4

**A:** FDG-PET whole body PET scan shows high uptake in the parapharyngeal tumour on the right side (arrow). Mild enhancement is seen in the left parotid gland due to sialadenitis secondary to radiation treatment. Normal uptake is seen in the myocardium and the urinary tract. **B:** Post-operative scan only shows increased uptake related to the surgery but no evidence of any residual tumour at the right skull base.

was also carried out using 2-[fluorine-18] fluoro-2-deoxy-D-glucose. It showed that there was an intense signal in the right upper parapharyngeal region due to markedly active metabolic activity consistent with a malignant neoplasm (Figure 4A).

In view of the above findings a decision was made to excise the lesion. As the lesion was malignant and was completely encircling the right internal carotid artery (ICA) very close to the base of the skull, it was decided to excise the ICA along with the tumour. Because of the proximity of the lesion to the skull base it would have been technically difficult to achieve control of the bleeding superiorly as the ICA would have to be cut flush with the skull base. Therefore after taking an informed consent a cerebral angiogram was undertaken with three objectives. Firstly to check the collateral circulation of the brain and assess that the patient had good cross circulation. Secondly on confirming a good cross circulation, performing a balloon occlusion test of the right ICA to check if the patient developed any signs of neurological deficit and thirdly following good toleration of the ICA occlusion leaving permanent balloons in the intracranial portion of the ICA to prevent any bleeding after excision. All three objectives were successfully achieved. Cerebral angiogram showed good cross circulation, balloon occlusion of the ICA for 30 minutes did not result in any neurological deficit and two permanent balloons were placed, one in the intracranial portion of the ICA and the other one in the lower part of ICA below the tumour (Figure 5). After the angiogram the patient was observed closely but he did not show signs of any neurological deficit.

Forty-eight hours following the angiogram and ICA occlusion the patient underwent excision of his residual malignant schwannoma. This was carried out via a right lateral transcervical approach to the infratemporal fossa which was facilitated by undertaking a lateral mandibular split. During the procedure the tumour was identified. It

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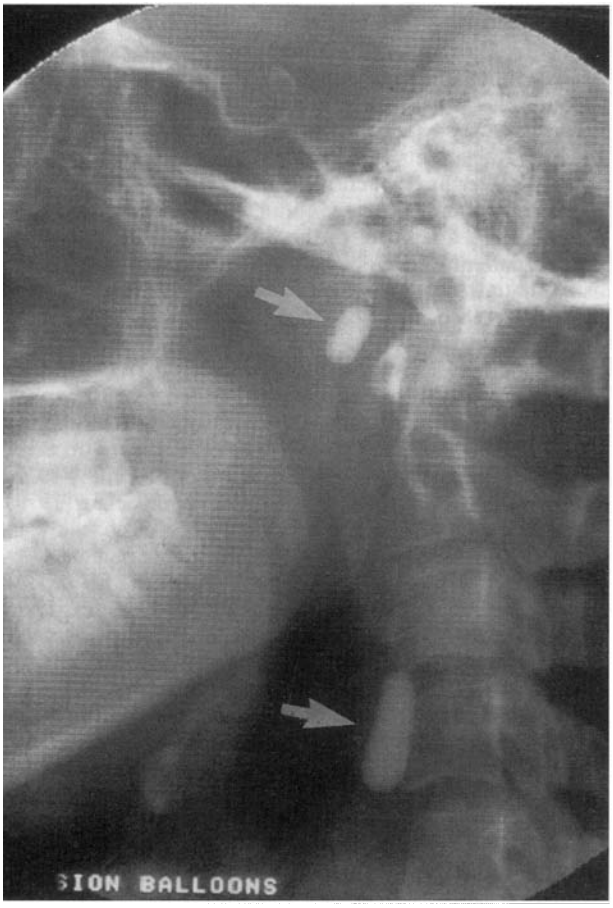


FIG. 5
Lateral view of the neck shows balloons (arrows) having been detached in the right internal carotid artery above and below the tumour.

was found that the IXth and Xth nerves had been excised during previous surgery and the tumour was adherent to the XIth and XIIth nerves. The ICA was ligated inferiorly just as it branched off the common carotid artery. The tumour was excised along with the adherent XIth and XIIth cranial nerves. The ICA was cut flush with the skull base. The balloon in the intracranial portion of the ICA could be visualized and was completely obliterating the lumen with no leakage of blood. No undue bleeding was encountered during the procedure.

Post-operative recovery was uneventful apart from slight nasal regurgitation with liquids. He had the expected right hypoglossal palsy. This, however, did not interfere with his swallowing. There were no systemic neurological deficits. There was no undue drainage from the neck and the drain was removed on the third post-operative day. The histopathological examination of the operative specimen confirmed the diagnosis of malignant schwannoma, which was completely excised. The patient was discharged eight days after surgery. At the time of discharge he did not show any sign of focal neurological deficit. The patient has been reviewed in the outpatient clinic at three, six and nine months following surgery. He does not complain of any nasal regurgitation now. Apart from the expected right-sided IXth, Xth, XIth and XIIth nerve paralysis, clinical examination does not show any other abnormality. His left vocal fold has compensated well which has resulted in his voice improvement and therefore further surgical intervention to improve his voice is not indicated. CT scan of the head and neck performed at nine months does not show any residual disease. These findings have been further confirmed by the PET scan, which shows no uptake at the right skull base (Figure 4B).

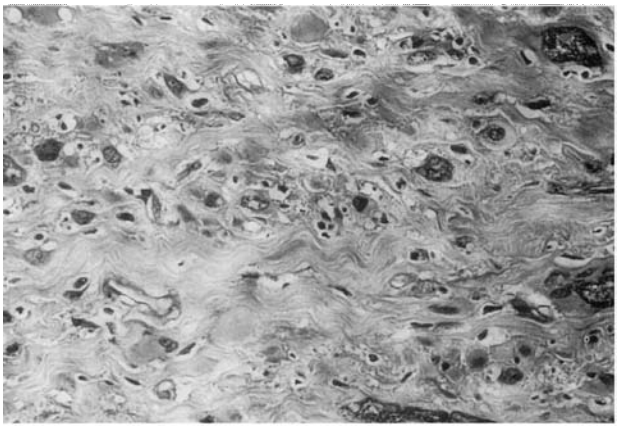

FIG. 6
Photomicrograph of the right neck node showing malignant spindle cells and giant cells infiltrating pre-existing nerve fibres. (H & E; × 250)

## Histopathological features

The tumour showed marked pleomorphism. It was in broad fascicles and clumps and showed a rare storiform pattern. Some nuclei are relatively small but multinucleated and anaplastic forms are also seen (Figure 6). In one section, there was recognizable peripheral nerve tissue. Immunohistocytochemistry was strongly positive for S-100.

## Discussion

Tumours of the parapharyngeal space are uncommon. Of the entire head and neck tumours only 0.5 per cent present in the parapharyngeal space (Allision *et al.*, 1989). Similarly malignant schwannoma is a rare tumour, as the majority of the nerve sheath tumours in the head and neck are benign (Batsakis, 1979). Nine to 14 per cent of all MS present in the head and neck (Barnes, 1985). Patients suffering from neurofibromatosis or Von Recklinghausen's disease are at risk of developing this disease. Ducatman *et al.* (1986) in a series of 120 malignant peripheral nerve sheath tumours of the whole body reported that the risk of developing this tumour is 4600 times greater in patients suffering from Von Recklinghausen's disease than in the general population. Fifty per cent of all MS are said to be associated with neurofibromatosis (NF) (Enzinger and Weiss, 1988), but there is a wide variation in the reported incidence with reports ranging from 25 to 70 per cent (Elias *et al.*, 1993). MS usually presents in adult life between 20 to 50 years of age with a roughly equal sex incidence (Enzinger and Weiss, 1988). However when it is associated with NF then the presentation is earlier and there is male predominance.

The main presenting symptom is usually a lump in the neck. As the main nerve to be involved in the head and neck is usually the vagus (Batsakis, 1979), hoarseness is a common presenting symptom. Apart from presenting as a neck lump parapharyngeal space lesions can also push the tonsil medially and present as a tonsillar swelling. It is interesting to note that out of three previously reported cases of MS, two were initially diagnosed and treated as peritonsillar abscess (Clairmont and Conley, 1977; Elias

https://doi.org/10.1017/S0022215100141970 Published online by Cambridge University Press

*et al.*, 1993). Apart from vocal fold paralysis, other lower cranial nerves may also be involved either by the tumour itself or by direct pressure.

Imaging of the parapharyngeal lesion is a vital part of the work-up in these patients as not only is it in an area which is very difficult to assess clinically, but also contains several important structures. The main objectives of the imaging are to determine the extent of the lesion and assess its resectability, delineate its relationship to the carotid artery system and the skull base or intracranial involvement. CT scan has been shown to be an excellent initial investigation. In a study of 51 patients with parapharyngeal lesions Carrau *et al.* (1990) found that the CT scan correlated 100 per cent with both the angiographic as well as surgical findings. MRI has also been shown to provide better soft tissue visualization of the neural and vascular structures while at the same time being non-invasive (Pensak *et al.*, 1994). Pre-operative angiography is performed in all cases of post-styloid lesions and in cases where malignancy is suspected (Pensak *et al.*, 1994). The typical angiographic appearance has been described as non-dilating feeding vessels, inhomogeneous blush of contrast persisting into late venous phase with no early filling of veins (Al-Ghamdi *et al.*, 1992). Functional imaging such as PET scan and Technetium-99m have not been widely used in the assessment of neural lesions. Borbely *et al.* (1992) have reported a case of cervical spinal neuromas which was positive on PET scan. PET scan has also been shown to be effective in detecting scar tissue from neoplastic tissue in an operated or radiated field (Zeitouni *et al.*, 1994). In our patient the PET scan was strongly positive and confirmed the presence of residual disease in an operated and irradiated area. To our knowledge our report is the first one showing an increased uptake on PET scan for a malignant schwannoma as well as a negative result after its complete excision. However, it should be noted that while the PET scan demonstrated the tumour, both CT and MRI were also equally effective in its delineation. Furthermore, it should be realized that the use of PET scan, as a diagnostic tool, is still in an early stage of development and indications for its widespread use in routine clinical practice are still being defined. Delayed images (two hours post-injection) of Technetium-99m scan have also been shown to be positive for a case of MS (Koch *et al.*, 1986), although in our case a similar bone scan did not prove helpful in picking up MS.

MS which is also known as malignant peripheral nerve sheath tumour and neurofibrosarcoma is not an easy tumour to diagnose. The tumour strongly resembles a fibrosarcoma (Enzinger and Weiss, 1988) and the only distinguishing feature distinguishing a MS from a fibrosarcoma is its origin from a nerve (Batsakis, 1979). This distinction may become difficult in large lesions. Electron microscopy can be helpful in diagnosis of difficult lesions and it has been shown that the presence of continuous basement membrane is a definitive sign of MS (Barnes, 1985). Immunostaining for cytokeratin and S-100 also aids in the differential diagnosis. Certain histopathological markers have been shown to have poor prognostic significance such as increased cellularity, pleomorphism and increased mitotic activity (Suit *et al.*, 1973; Greager *et al.*, 1992). Apart from this, the other poor prognostic indicators are achieving a size of more than 7 cm and association with NF (Barnes, 1985). If it is not associated with NF then the five-year survival is 47 per cent while it drops down to 23 per cent in NF patients. Reasons for the poor prognosis with NF have been suggested as increased pleomorphism, tendency towards larger tumours, poorer differentiation, a higher rate of metastasis and multifocality (Ghosh *et al.*, 1973; Ducatman *et al.*, 1986). Metastasis to regional lymph nodes is not a feature of this disease as shown by Ghosh *et al.*, 1973 in a series of 115 patients and by Das Gupta and Brasfield (1970) in 232 patients where no regional metastasis were noted. However Bailet *et al.* (1991) have reported one case of nodal metastasis in their study of 17 patients. Haematogenous metastases to lung, bone and liver have been reported and can occur in up in 33 per cent of cases (Das Gupta *et al.*, 1970). Furthermore even when local control of the disease is achieved, 44 per cent of patients develop distant metastases and it is the distant metastasis that is the cause of death in 77 per cent of the patients (Storm *et al.*, 1980).

Benign schwannomas may also undergo malignant transformation. It is interesting to note that two out of three previously reported cases of MS of the parapharyngeal space initially had a histologically confirmed benign schwannoma. Malignant transformation however occurred after two years in one case (Clairmont and Conley, 1977), and after nine years in another (Shapiro and Rickert, 1979). These reports highlight the importance of a close follow-up of those cases which are initially reported as benign.

One of the problems with rare lesions is the lack of proper controlled trials to determine the benefit of any treatment modalities. Review of the literature shows that the best outcome for the patients with MS is a combination of local resection combined with adjuvant radiotherapy and chemotherapy. Wide local excision of the tumour is the main and the commonest therapeutic option. Greager *et al.* (1992) have shown that those patients who underwent wide local excision achieved the longest mean survival (115 months compared to 74 months in patients who did not undergo wide excision). Local recurrence and multiple re-excisions are also a feature of this disease. Surgical treatment on its own is not adequate and combination with radiotherapy and chemotherapy has shown improved survival rates as shown by Storm *et al.* (1980) who achieved local control in only 55 per cent of cases who were treated by surgery alone. However, when combined with radiotherapy and chemotherapy there was only one recurrence out of six patients. In another study patients who were treated by both surgery and radiotherapy did better than those who underwent surgery alone (Basso-Ricci, 1989). However, these results should be interpreted with caution as the studies were carried out for MS at all body sites and the numbers of patients receiving radiotherapy and chemotherapy is small. It should also be noted that radiotherapy and chemotherapy are only adjunct treatment modalities and on their own have no role in the management of MS. Surgical excision is the mainstay of the treatment. Neck dissection is generally not required, as the chance of regional lymph node involvement is minimal.

MS is a rare lesion, which is difficult to diagnose histologically. The long-term prognosis for this lesion is poor and becomes even worse when associated with NF. Due to the rarity of the lesion in the head and neck it is difficult to know the best treatment modailty but the various studies, both in head and neck and at other body sites, are generally of the consensus that while wide resection is the mainstay of its treatment, the addition of radiotherapy and perhaps chemotherapy results in better control of the disease process.

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