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Malignant triton tumour of the parapharyngeal space

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Abstract
We report the clinical course in a 25-year-old male patient of neurofibromatosis with malignant triton tumour (MTT). Triton tumour is a peripheral nerve sheath tumour with rhabdomyoblastic differentiation. This is relatively rare tumour of head and neck region and only 26 cases have been reported to date. The present case is the first reported MTT of parapharyngeal space, one of the most aggressive course among all head and neck MTTs, resulting in the death of the patient within three months following surgical resection. The aggressive nature of this tumour necessitates adjuvant therapeutic measures in addition to radical surgery in order to obtain better survival rates.

Key words: Peripheral Nerves; Tumour; Parapharyngeal Space

Introduction

Malignant triton tumour (MTT) also referred to as 'malignant peripheral nerve sheath tumour with rhabdomyoblastic differentiation', is a rare soft-tissue tumour first described by Masson in 1932.[1] This rare sarcoma variant received its pseudonym from experiments by Locatelli on the 'Triton' salamander.[2] Although malignant peripheral nerve sheath tumors (MPNST) may consist of tissues such as glandular epithelium, squamous cell, cartilage or bone, those which possess only skeletal muscle differentiation are referred to as MTT. The histogenesis of this tumour has been debated extensively. The tumour consists of two cell lines, rhabdomyoblasts and Schwann's cells. Masson postulated the induction of one cell line on the other.[1] According to Enzinger both cell lines originate from less well-differentiated neural crest cells, contributing to the formation of mesenchymal structures such as branchial cartilage, connective tissues, and muscles in the facial region.[3]

Eighty cases of MTT have been reported so far; in 26 of these cases, the tumour was localized in the head and neck. However, outcome of the patients is stated in only 23 of all head and neck MTTs.[4] This reported case is the first MTT of the parapharyngeal space, and the second case arising from the vagus, with a previous case being a vagal nerve MTT within the mediastinum. Our case has one of the most aggressive courses among all head and neck MTTs, causing death within three months following complete surgical resection. Therefore, our recent experience in treating a patient with MTT seems notable because it provided us with an opportunity to record its grim biological behaviour.

Case report

A 25-year-old white male presented with a history of dysphagia, foreign-body sensation in his throat and progressive hoarseness for six months. His medical history was entirely negative. He was a non-smoker and had not noted any weight loss. Family history revealed the death of a 27-year-old brother due to a brain tumour.

Physical examination

Physical examination revealed widespread 'café au lait' spots on the skin of the trunk and extremities. According to these findings and family history, the patient was considered to have von Recklinghausen's neurofibromatosis (VRN). Bimanual palpation of the neck revealed a firm, solid, non-compressible parapharyngeal mass on the left side. The left tonsil was found to be displaced medially in the examination of the oropharynx. Laryngoscopy revealed left-sided vocal fold weakness. The rest of the

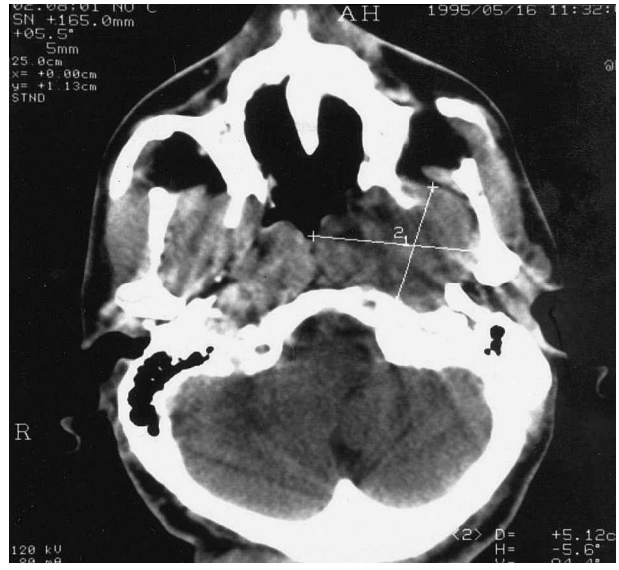

FIG. 1
Axial CT showing parapharyngeal mass.

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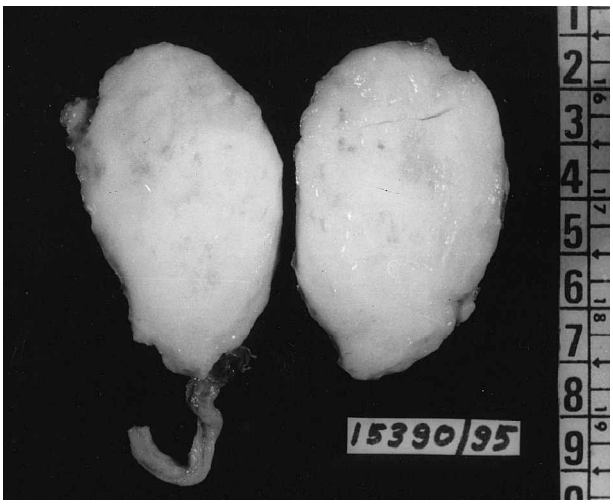

FIG. 2
Macroscopic view of the tumour: the proximal and distal edges of vagal nerve are seen in the upper and lower pole of the tumour.

physical examination was normal. Computed tomography (CT) of the head and neck demonstrated a well-circumscribed soft-tissue mass with non-homogeneous contrast enhancement in the left parapharyngeal space extending from the oropharynx to the hypopharyngeal level. The lesion measured approximately 6×4 cm (Figure 1). Imaging of the head and neck was otherwise normal.

Treatment

Transcervical horizontal incision approximately two finger breadths below the mandible was used to access the tumour. The tumour had originated from the left vagal nerve and was adjacent to the larynx. It was apparently well separated from adjacent tissues. Complete resection of the tumour with vagal nerve sacrifice was performed. The suggested adjuvant therapies were not accepted by the patient.

Pathological examination

Macroscopically, the nodular mass was well encapsulated and measured $6 \times 4 \times 3$ cm. The tumour had a white–tan cut surface and was firm in consistency (Figure 2).

Microscopically, the lesion had components of both a spindle cell sarcoma and scattered eosinophils cells. The predominant portion of the tumour consisted of pleomorphic cellular sarcomatous areas (Figure 3(a)). These areas were positive for immunoperoxidase S-100 protein stain (Figure 3(b)). Cells with round, deep eosinophilic and distinct cytoplasms were eye-catching, and they showed a positive immunostaining for desmin and myoglobin indicating the rhabdomyoblastic differentiation (Figure 3(c)). The diagnosis of MTT was made by the presence of these rhabdomyoblastic cells in a malignant nerve sheath tumour. Surgical margins of the nerve adjacent to the mass were free of tumour.

Post-operative course

At the third post-operative month, the patient developed progressive mental confusion and dysarthria. Neurological examination revealed loss of the left light and gag reflex and left peripheral facial weakness. T1-weighted magnetic resonance imaging demonstrated a dumbbell-shaped mass measuring 3 cm in the posterior cranial fossa, destructing jugular foramina, with a mass of 2.5 cm extending into the infratemporal fossa (Figure 4). Two weeks following the diagnosis of recurrence, the patient died because of uncal herniation.

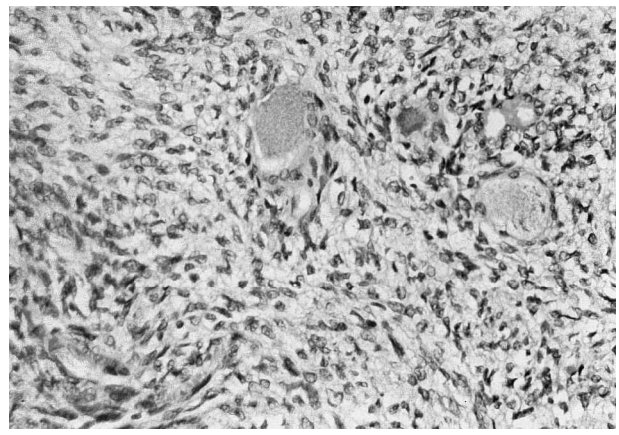

FIG. 3a
Microscopy with haematoxylin and eosin staining (×310).

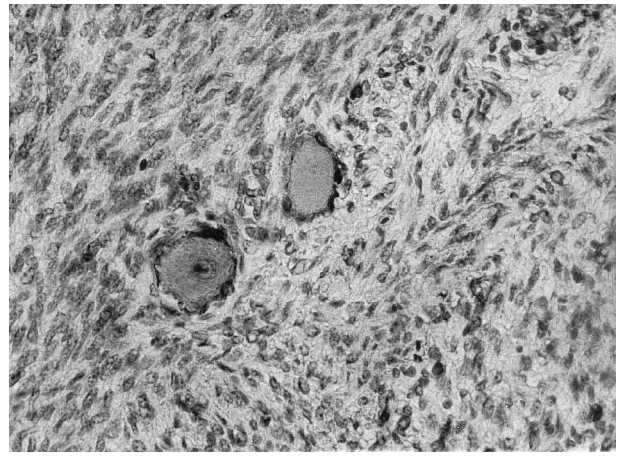

FIG. 3b
Immunohistochemical staining with S-100 protein (×310).

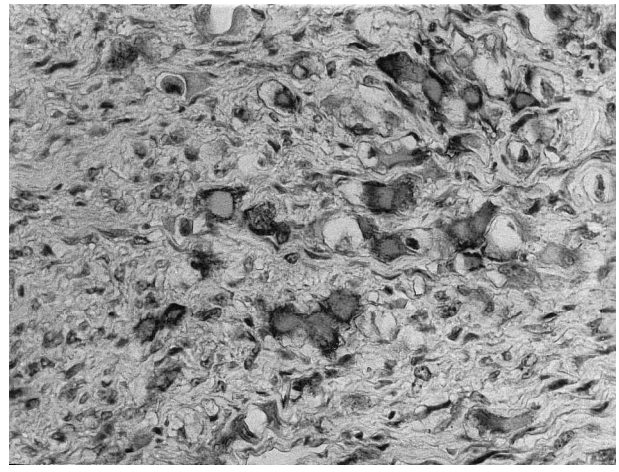

FIG. 3c
Immunohistochemical staining for desmin indicating the rhabdomyoblastic differentiation (×310).

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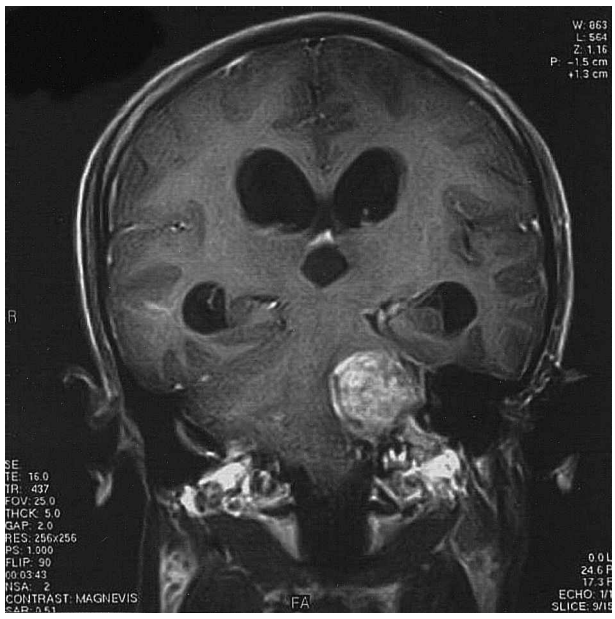

FIG. 4
T1 weighted MRI showing dumbbell-shaped mass.

Discussion

The reported case is histologically typical of MTT's arising from the vagus in a VRN case. It met the diagnostic critiera suggested by Woodruff *et al.*: 1 evidence of origin along a peripheral nerve or occurrence in patients with VRN. 2. predominance of growth characteristics of Schwann's cell type and 3. presence of true neoplastic rhabdomyoblasts.[5]

While the majority (69 per cent) of MTT cases are encountered in association with VRN, sporadic cases may also also occur.[6] Brooks has classified the MTT cases associated withVRN as group I and the sporadic cases as group II.[6] While group I cases are usually young males with a mean age of 35, group II has a female dominance with an older age average.[6] MTTs arising in the head and neck region account for nearly one-third of all MTTs (27/80), while in the remaining cases, the tumour arises from trunk and extremities equally.[4–7]

Woodruff *et al.* have defined MTT as a 'highly malignant and deadly' tumour.[5] Guccion and Enzinger have claimed that there exists no biological behavioural difference between MTT and ordinary MPNST.[8] By reviewing literature and analysing his own cases, Brooks found that the two- and five-year survival rates for MTT are 33 and 12 per cent respectively. The five-year survival rate of ordinary MPNST cases associated with VRN varies between 15 and 30 per cent.[8,9] However, in sporadic cases of MPNST, the survival for five years ranges between 23 and 75 per cent.[9–12] These data led Brooks to conclude that MTT has an equal or worse prognosis than VRN-related MPNST and a significantly worse prognosis than non-VRN-related MPNST cases.[6] The prognosis is relatively better for head and neck, and worse for buttock, trunk and retroperitoneum.[4] Survival of patients with head and neck MTT ranges between four months and 22 years.[6] The treatment protocol of choice for his highly malignant tumour is complete resection, with adjuvant radiation therapy.

Encouraging results gained by adjuvant chemotherapy have led to the option of the treatment protocol to be supplemented by adjuvant chemotherapy.[6,7] Unfortunately, despite proper treatment protocols, the local recurrence and distant metastasis rates approximate 25 and 48 per cent, respectively.[6]

The present case did not differentiate it from the appearances of a benign schwannoma. In our case, MTT had a remarkably more aggressive course than other MTTs of head and neck reported so far.[4,6] Despite adequate surgical resection with clear surgical margins, the patient died after three months with a devastating intracranial recurrence. In addition to the highly aggressive behaviour of this tumour, lack of adjuvant therapies may be one of the reasons for the rapid course of the disease in this instance.

Conclusion

MTT is a highly aggressive and rare tumour which challenges both head and neck surgeons and oncologists. The rapid course of this disease implicates that, in all cases of MTT, adjuvant therapies such as radiotherapy and chemotherapy must be considered in addition to radical surgery, in order to obtain better survival rates.

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