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Malignant granular cell tumour of the cervical sympathetic nerve trunk

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Abstract

Granular cell tumour is a rare neoplasm that can occur in various sites. This report describes a 48-year-old female with a malignant granular cell tumour originating from the cervical sympathetic nerve trunk, who presented with hyperaesthesia of the left shoulder. The tumour had a maximum diameter of 86 mm and involved adjacent tissues directly.

Histopathologically, it showed considerable variation in the size and shape of nuclei, with occasional mitosis and an abundant granular cytoplasm. Immunohistochemically, the tumour reacted positively for S-100 protein and neuron-specific enolase, indicating its neural origin. Following total local excision of the lesion, the patient has been well without recurrence.

Key words: Granular Cell Tumour; Sympathetic Nervous System; Immunochemistry

Introduction

A granular cell tumour is a rare neoplasm. It can occur in various parts of the body, with a predilection for the skin and subcutaneous tissue. In the head and neck, it is most commonly seen in the tongue.[1] Its histogenesis has been a subject of longstanding controversy since this tumour was first described by Abrikossoff.[2] Pathological investigations, including histochemical and electron microscopic studies, have examined its histogenesis, and the majority of authors currently support a neural origin. However, only a few reports have described this tumour arising in major peripheral nerves.

Generally, a granular cell tumour is benign with a favourable prognosis; however, malignant granular cell tumours are being reported increasingly.[3,4] Malignancy is diagnosed by a combination of histopathological findings and the clinical behaviour of the tumour.[1] Despite many reports on granular cell tumours, the majority of otolaryngologists are still unfamiliar with this lesion. Moreover, there is no generally agreed upon therapeutic strategy for malignant granular cell tumours. Here, we report the first case of a granular cell tumour originating from the cervical sympathetic nerve trunk presenting with low-grade malignant potential.

Case report

A 48-year-old woman was referred to us with a complaint of hyperaesthesia of the left shoulder extending to the left neck, upper chest, and proximal arm abductors for five months. Pressing on the left neck enhanced the symptoms. Physical examination revealed a large firm mass with a relatively distinct border in the left lateral neck. It severely limited mobility. Computed tomography (CT) examination revealed a large homogeneous mass in her left lateral neck, with partial bony destruction of the cervical vertebrae

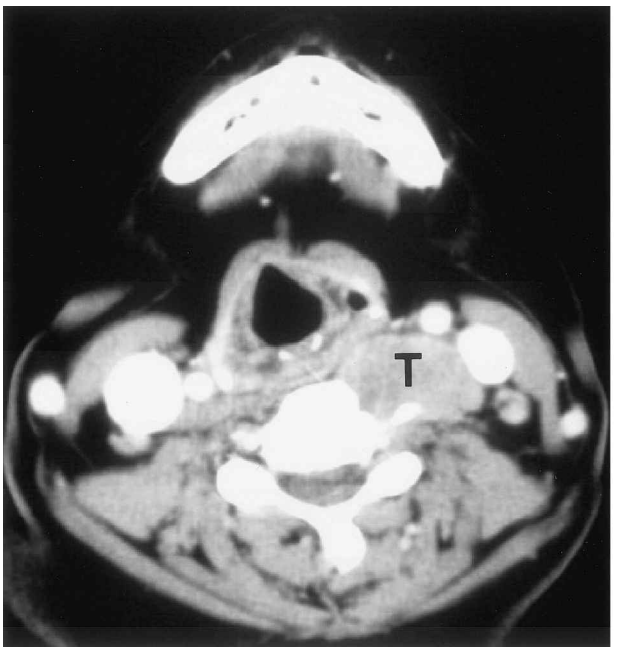

Fig. 1

Axial CT showed the tumour (T) in the paravertebral region with partial destruction of the anterolateral aspect of the cervical vertebral bone.

(Figure 1). Magnetic resonance imaging (MRI) confirmed the longitudinal extension of the tumour from C2 to C7. The tumour was isointense on T1-weighted images, with an increased gadolinium enhancement signal, and hyperintense on T2-weighted images (Figure 2). Whole body^{67} Ga

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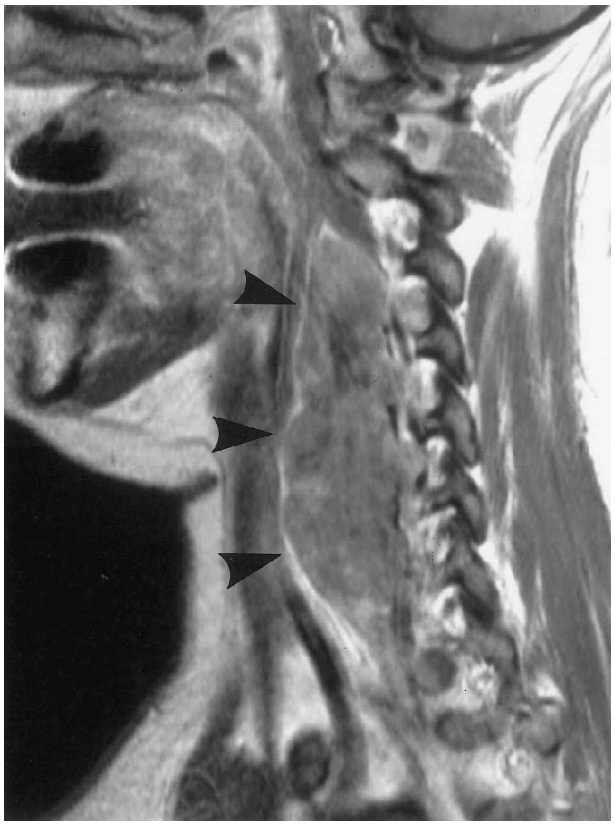

FIG. 2

Sagittal T1-weighted MRI with gadolinium enhancement demonstrated a large spindle-shaped tumour from C2 to C7 (arrow head).

scintigraphy showed no abnormal uptake. Serum neuron-specific enolase (NSE) was mildly elevated to 10.3 ng/ml (normal <9.2 ng/ml). Based on these clinical and laboratory findings, the tumour was considered to be of neurogenic origin with possible malignant features.

Total excision of the tumour was performed. The tumour was located medially beyond the common carotid artery and internal jugular vein. By retracting the common carotid artery anteriorly and the internal jugular vein posteriorly, the entire tumour was exposed (Figure 3). The cervical sympathetic nerve trunk was identified superior to the tumour. The sheath of the nerve gradually blended with the tumour capsule, demonstrating that the tumour originated from the cervical sympathetic nerve trunk. The tumour adhered to the fascia of the longus capitis muscle and the anterior longitudinal ligament. The cervical sympathetic nerve trunk was transected and the tumour, including the affected surrounding tissue, was successfully removed. The excised tumour was 86 × 31 × 32 mm in size and weighed 29.4 g. Histopathological examination revealed that tumour tissue had invaded the perimysium of the adjacent muscle. Tumour cells were predominantly round to polygonal, with ample granular eosinophilic cytoplasm and marked nuclear atypia (Figure 4). Occasional mitotic figures were seen in the nuclei. Immunohistochemically, the tumour cells stained positively for S-100 protein and NSE. Based on these findings, a granular cell tumour with low-grade malignancy was diagnosed.

Post-operatively, the patient's initial complaints disappeared and the serum NSE decreased to 6.5 ng/ml, which is in the normal range, two weeks after surgery. Her clinical course was uneventful, and there has been no local recurrence or distant metastasis in a three-year follow-up period.

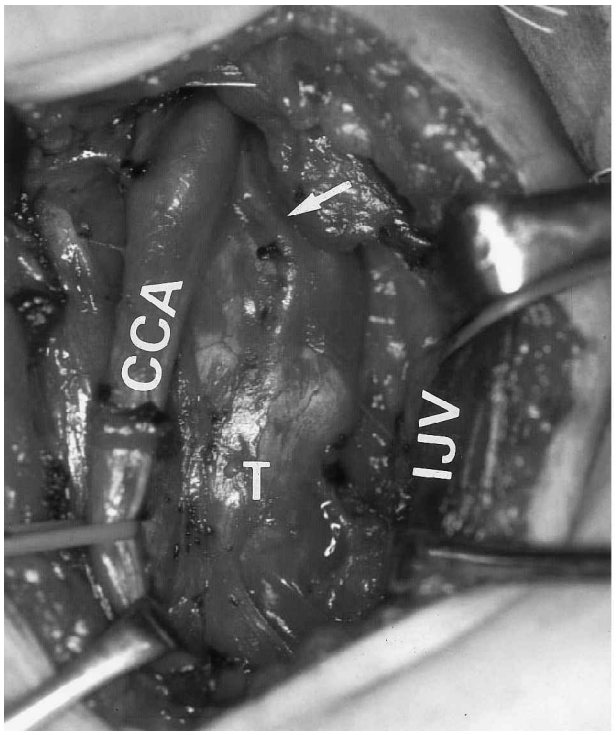

FIG. 3

Operative findings. The tumour (T) was medial to the common carotid artery (CCA) and internal jugular vein (IJV). The cervical sympathetic nerve trunk (arrow) gradually blended with the capsule of the tumour.

Discussion

Abrikossoff[2] is generally credited with describing granular cell tumour. He believed that the tumour originated from embryonic fibrils of myoblasts and called it a myoblastoma. Later, electron microscopic and histochemical studies led to the proposal of a neural theory of origin.[5] Positive immunohistochemical reactivity for S-100 protein suggests that granular cell tumours have a Schwann cell origin,[6,7] and most pathologists now agree with this idea.

Clinically, a granular cell tumour is usually a small, solitary, painless, firm nodular lesion, arising from dermal, subdermal, or submucosal tissue. This tumour may arise anywhere in the body. In the head and neck, the most common site of origin is the oral mucosa including the

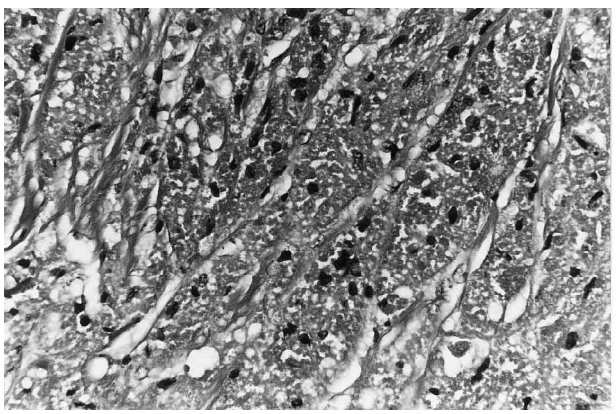

FIG. 4

Histopathological finding of the resected tumour showing polygonal cells with abundant granular cytoplasm. Nuclei were hyperchromatic and pleomorphic (H&E; ×100).

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tongue, followed by the larynx.[1,8] Some reports have demonstrated a neural origin from the trigeminal[9] and facial[10] nerves; however, there has been no published report of a granular cell tumour derived from the cervical sympathetic nerve trunk. Of clinical note, granular cell tumours are sometimes multifocal at the time of initial presentation or during follow-up.[11]

The diagnosis of granular cell tumour is made histopathologically. The tumour is characterized by nests and sheets of polygonal cells with distinct borders and abundant granular eosinophilic cytoplasm.[1] The nuclei are small and hyperchromatic. Immunohistological staining for S-100 protein and NSE is a useful tool in the different diagnosis of this tumour.[12] S-100 protein is an acidic protein that is widely distributed in the central nervous system. Outside the central nervous system, it is found in several cell types including Schwann cells, melanocytes, chondrocytes and tumours derived from these.[13] Therefore, the positive S-100 reactivity of the tumour would support a Schwann cell origin. NSE is an isoenzyme from the glycolytic pathway, and it is often found in high levels in cells of neuroectodermal origin. In this case, there was a slight increase in NSE in the serum as well as in the tumour pre-operatively, and the level decreased to the normal range post-operatively, proving that the tumour was of neural derivation.

Granular cell tumour is generally a benign neoplasm although one to two per cent may be malignant.[14] According to a review by Sonobe *et al.*,[14] 40 cases of malignant granular cell tumours were reported in the English literature for all sites in the body up to 1997. Thereafter, 12 cases including the present case were added to them by our review.[15–25] Of these, 12 (23 per cent) were in the head and neck. Malignancy in a granular cell tumour is sometimes difficult to diagnose. A combination of pathological features, including involvement of the adjacent tissue, cellular pleomorphism and mitosis of the nuclei, and clinical behaviour involving rapid growth and large size (>4 cm) attest to malignancy.[1] Metastasis may occur in the regional lymph nodes, lung, and bone.[3]

The preferred treatment for granular cell tumour is complete surgical resection. An inadequate resection margin results in subsequent recurrence, particularly if the tumour is malignant. Radiation therapy is not generally indicated, since granular cell tumours are considered radioresistant. Considering the possibility of recurrence or multifocal occurrence of this tumour, long-term follow-up is mandatory.

Conclusions

We report the first known case of a granular cell tumour with malignant potential arising from the cervical sympathetic nerve trunk. We believe that this report broadens the current awareness of granular cell tumour and the differential diagnosis of soft tissue tumours of the neck.

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