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Parapharyngeal granular cell tumour: a unique surgical challenge

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Abstract
A granular cell tumour is a rare lesion of probable nerve sheath origin. It is typically benign but up to seven per cent may be malignant. Since its original description in the tongue in 1926, the tumour has been reported to occur at many other sites in the body. The authors report a case of a 49-year-old African woman with an oro-naso-parapharyngeal granular cell neoplasm causing mild dysphagia. The location of this tumour, which has not been reported previously, posed a unique surgical challenge. An initial attempt to remove the lesion transorally was only partially successful because it was too tough and adherent for conventional surgical dissecting instruments. Complete resection, however, was achieved with a carbon dioxide laser via the same approach. This information may be helpful in the management of other similar cases in the future.

Key words: Granular cell tumour; Parapharyngeal Space; Laser Surgery

Introduction

A granular cell tumour (GCT) is a rare lesion, which is typically benign and solitary, but may be multiple and rarely malignant.[1] The tumour is most prevalent in the 30-50 years age group and occurs predominantly in females of African descent.[2,3] A hormonal relationship between GCT and a hyper-oestrogenic state has been postulated[2,4] but the small number of cases preclude a definite association.

Since the lesion was first described by Abrikossoff[5] in the tongue in 1926, it has been documented in almost every region of the body.[3] However, the tongue remains the single most frequently reported site.[3] The authors present a new case of a submucosal parapharyngeal GCT causing dysphagia in a female adult and discuss the pathogenesis and surgical management.

Case report

A 49-year-old African woman was referred because of mild dysphagia for at least 12 months. Oral examination revealed an obvious bulge in the right lateral oro-pharyngeal wall with displacement of the tonsil medially. Palpation revealed a firm non-tender mass. There was no trismus or cranial nerve palsy. Examination of the neck was unremarkable. A computed tomography (CT) scan showed a solid non-enhancing homogenous mass (3.75 cm $\times$ 2.25 cm) lateral to the oropharynx and nasopharynx (Figure 1), with obstruction of the fossa of Rosenmüller. The lesion displaced the carotid sheath with its contents anterolaterally (Figure 2). There was neither invasion of the surrounding structures nor a clear plane of cleavage posterolaterally from the prevertebral muscles (Figures 1 and 2). The tumour did not cross the midline and no associated lymphadenopathy was seen. CT scan of the chest was normal.

Histological examination of an intra-oral biopsy of the lesion revealed features consistent with the diagnosis of a

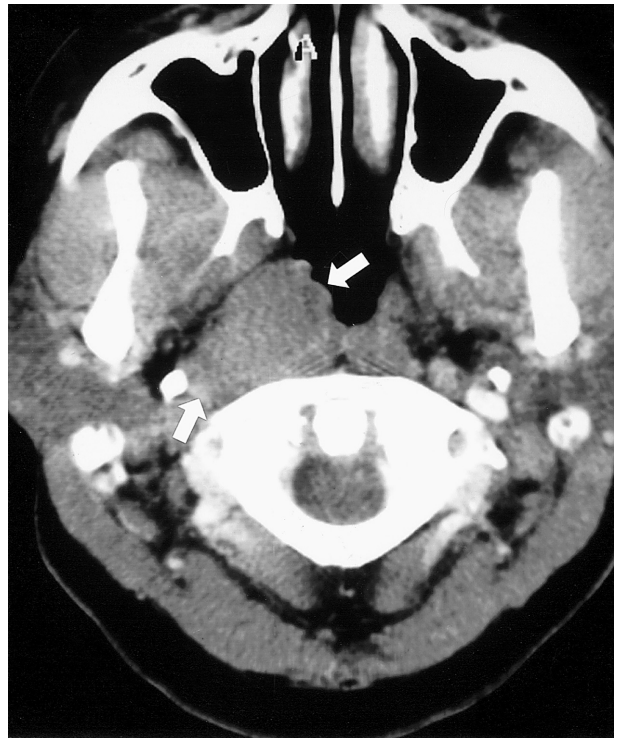

Fig. 1
Axial CT scan at the level of the nasopharynx showing the tumour (arrows) in the lateral pharyngeal region.

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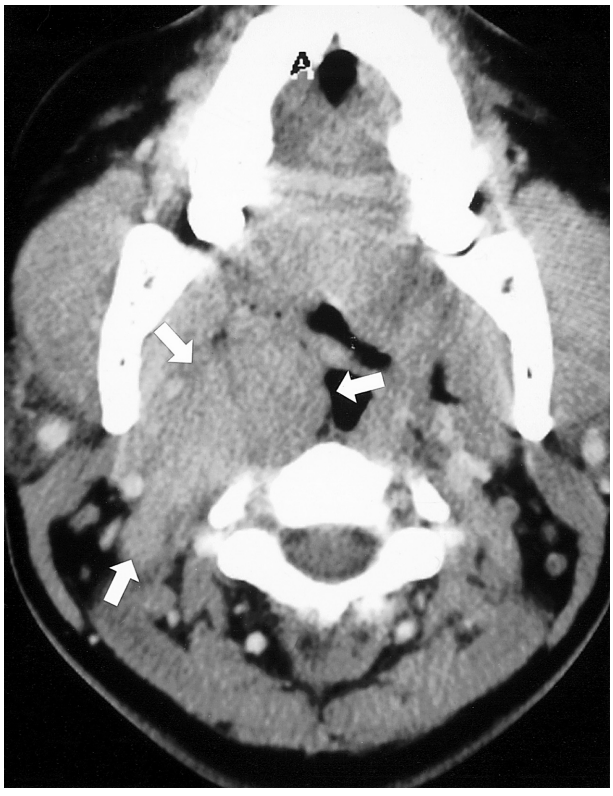

FIG. 2

Axial CT scan at the level of the oropharynx showing the tumour (arrows) displacing the contents of the carotid sheath anterolaterally.

benign GCT. These features included cells with abundant, granular, pale acidophilic cytoplasm with pyknotic nuclei showing no atypia (Figure 3). The cells displayed strongly positive immunostaining for the neural marker S-100 protein (Figure 4). An elective intra-oral surgical excision procedure was planned.

At the beginning of the operation the overlying mucosa separated easily to expose the lesion. However, the posterolateral dissection was difficult partly because of the firmness of the tumour and partly because of the extremely strong adherence of the mass to the underlying muscles as well as the transverse process of the C2 vertebra. It was necessary to divide the soft palate to gain better access to the lateral nasopharyngeal component of the tumour. After slow and careful piecemeal dissection, most of the tumour was removed. Post-operatively, the patient was noted to have a right Horner's syndrome. She was fed via a naso-gastric tube for one week, to allow some amount of healing of the lateral pharyngeal defect, before re-commencing oral intake.

A repeat CT scan performed eight weeks later showed the residual tumour. Macroscopic clearance of the residual disease was then achieved with a transoral carbon dioxide laser attached to a surgical microscope. This approach allowed precise determination of the abnormal tissue, under magnification, in an almost bloodless field. The texture of the tissue was not in any way problematical and the anatomy of the parapharyngeal region could be clearly seen. Following complete excision of the tumour, gel foam and a Whitehead varnish pack were secured in the wound and removed after two weeks. The patient was fed via a naso-gastric tube for one week before re-starting oral diet. There were no post-operative complications. Multiple biopsies taken from the tissue surrounding the tumour (after laser excision) revealed clear microscopic margins. She remains well with no evidence of any recurrence.

Discussion

This new case of a GCT beneath the lateral oro-naso-pharyngeal mucosa posed a unique management challenge. The treatment of choice of benign and malignant GCT is surgery (the role of chemotherapy or radiotherapy is unclear). The surgical approaches to lesions in the parapharyngeal spaces include a transcervical, a transoral or a mandibular splitting technique. The authors opted for the transoral approach with division of the soft palate to gain adequate access to the lateral nasopharynx. The transoral route has the distinct advantage of reaching the tumour directly with no external scar or possible keloid formation and the overlying mucosa can be resected with the tumour. However, this approach does have the potential disadvantage of possible airway complication secondary to pharyngeal swelling and less secure control of the great neck vessels.

In the first operation, conventional surgical dissection equipment (scalpel and scissors) were used, but it was difficult to resect the tumour completely because of its firm, resistant texture and its adherence to the surrounding tissue.

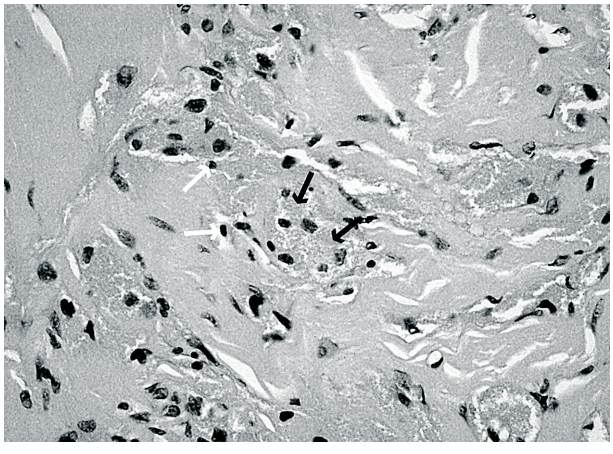

FIG. 3

Photomicrograph showing nests of cells with typical granular cytoplasms (black arrows) and densely staining nuclei (white arrows) (H & E stain; ×200).

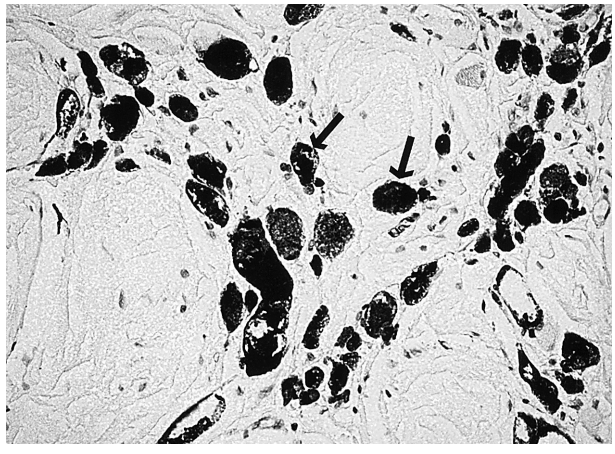

FIG. 4

Photomicrograph showing cells with positive immunostaining for the neural marker S-100 protein (black arrows) (PAP; ×200).

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It is important to have clear margins because recurrences have been reported in up to 12 per cent of cases with incomplete excision.[6] Therefore, a second operation was performed using a carbon dioxide laser to remove the residual disease transorally. It is possible that the second procedure could have been avoided if the laser was used at the inception. However, the rarity of this case resulted in some degree of uncertainty about the optimal surgical management during the pre-operative planning phase.

- **A granular cell tumour is a rare lesion of probable nerve sheath origin. Up to seven per cent may be malignant**
- **This case of an oro-nasal-parapharyngeal granular cell neoplasm shows a previously unreported location which presented a unique surgical challenge**
- **Blunt dissection via a transoral route was incomplete but the use of the carbon dioxide laser via the same approach resulted in complete removal of the lesion**

Granular cell tumours are predominantly benign lesions, but up to seven per cent may be malignant.[3] Clinically, the suspicion of malignancy arises in tumours greater than 4 cm and in lesions which show rapid recurrence or evidence of invasion into adjacent structures.[3] Pathologically, a malignant GCT tumour displays cellular and nuclear pleomorphism as well as necrosis and high mitotic activity.[7] According to some authors, a diagnosis of malignancy can only be substantiated by finding metastatic foci with histological features similar to the primary tumour.[1] In the present case, the tumour was thought to be benign because there was no nuclear atypia or histological evidence of local invasion and it was less than 4 cm.

When a GCT occurs in a submucosal location, it is common for the overlying epithelium to appear hyperplastic in 50-65 per cent of cases and this may be misinterpreted as a well-differentiated squamous cell carcinoma.[3] In the authors' patient the mucosa covering the tumour looked normal macroscopically and this was confirmed histologically. However, an awareness of the so-called 'pseudoepitheliomatous hyperplasia' is important to avoid unnecessary radical surgery and associated morbidity.

In the five cases of GCTs described by Abrikossoff[5] in 1926, all were located in the tongue and were closely associated with striated muscle. This led him to attribute the origin of the lesion to a myoblastic stem cell and to classify it as a myoblastic myoma. However, the persistent intimate relation of the tumour with peripheral nerves suggests a precursor other than skeletal muscle.[8] Immunohistochemical staining favours a Schwann cell origin because of the positive reaction for the neural marker S-100 protein and the negative reaction for myogenous markers such as actin and desmin.[9] A Schwann cell origin is also supported by the presence of myelinated nerve bundles in the majority of specimens.[3] It is conceivable that in this case the tumour arose from submucosa neural tissue and extended laterally in the parapharyngeal region.

In summary, the authors report a novel case of a 49-year-old woman with an extensive parapharyngeal granular cell neoplasm causing mild dysphagia. The tumour was resected via a transoral approach using a laser after an attempt to excise it with conventional surgical dissecting tools was only partially successful. This information may be helpful in the management of other similar cases in the future.

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