CASE REPORT

Deep cervical granular cell tumor: An atypical location suggestive of neurogenic origin

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Summary

Introduction: Granular cell tumor (GCT), or Abrikosoff’s tumor, is usually benign, with predominantly head-and-neck locations. Putative Schwann-cell origin is controversial. Treatment is surgical, due to risk of malignancy.

Case report: A 41-year-old man presented with benign GCT in one of the deep cervical plexus roots, suggesting neurogenic origin.

Discussion: Surgical resection is important. Preoperative diagnosis is hindered by the ubiquity of the lesions and the poor specificity of imaging. Pathologic and immunohistochemical analysis is essential for definitive diagnosis.

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Introduction

First described by Abrikosoff in 1926 [1], granular cell tumor (GCT) is rare (0.019 to 0.03% of tumors as a whole), ubiquitous and usually benign; location is mainly (50%) in the head-and-neck region, within which it is predominantly (50%) lingual. Histologic origin was long uncertain and remains controversial [2]. The presence of S100 protein, a Schwann-cell marker, suggests a neurogenic origin. We report the case of a 41-year-old man presenting with isolated deep cervical GCT at a nerve root. To the best of our knowledge, this is the first report of such a location in the international literature, and supports the idea of a Schwann-cell origin for GCT.

Case report

Mr. D, 41-year-old, with no particular history, presented with isolated right cervical tumefaction, discovered by self-palpation, of 6 months’ evolution, without inflammation. The swelling was indurated, irregular, painless, mobile, 2 cm in diameter, and located in the Vb lymph-node level.

Fine-needle aspiration cytology was non-contributive. Cervicothoracic (CT) confirmed the presence of an isolated...
iso-to-hyper-intense lesion medially to the right sternocleidomastoid muscle and posterior to the internal jugular vein, suggestive of a Vb-level node, showing weak contrast uptake (Fig. 1). On MRI, the lesion showed in hypersignal on T1 and T2-weighted sequences, with no suppression of signal on fat-saturated images or enhancement by contrast-medium injection. Surgery found a lesion remote from the vagus nerve, posterior to the internal jugular vein, involving one of the deep cervical plexus roots, macroscopically suggestive of adenopathy rather than schwannoma. Extemporaneous examination suggested spindle-cell tumor. Benign/malignant status could not be determined.

Standard hematoxylin-eosin-safran (HES) staining revealed a tumor formed by a proliferation of round or polygonal cells with eosinophilic granular cytoplasm, poorly contoured, with small central nuclei. Some PAS-positive cytoplasmic vesicles (dilated lysosomes) could be identified. The cells expressed CD68 and PS100 markers (Fig. 2), and not neuron-specific enolase (NSE) or muscular actin. There were no signs of malignancy; the morphologic and immunohistochemical presentation as a whole was characteristic of benign GCT.

Analysis of the complete specimen, resection of which sacrificed one of the deep cervical plexus roots, confirmed the diagnosis of benign GCT and that resection was complete (Fig. 3).

Discussion

Fifty percent of GCT locations are in the head-and-neck region, within which the tongue (50%), palate and floor of the mouth are the most frequently involved organs [3]. One case of medial suprasternal cervical GCT was reported, but this was a subcutaneous lesion above the muscular and aponeurotic planes [4]. GCT located in the deep lateral cervical spaces has, to the best of our knowledge, never previously been reported.

Preoperative assessment rarely enables diagnosis. In the present case, CT suggested non-necrotic Vb-level adenopathy. There are only sparse data on the imaging aspects of GCT. CT finds a solid, more or less heterogeneous tumor. On MRI, the lesion is usually in hyposignal on T1 and hypersignal on T2-weighted sequences, enhanced by contrast medium [5]. In the present case, it showed in hypersignal on both T1 and T2, and was not enhanced by contrast medium. Several authors have highlighted the difficulty of interpreting fine-needle aspiration results, with risk of misdiagnosis [6].

Clinical and radiological assessment left several diagnostic hypotheses open: adenopathy, although the MRI aspect did not support this; X neurinoma was unlikely, given the location, which was remote from X, and the MRI aspect; neurinoma of another nerve, and especially of the deep cervical plexus, was conceivable, although the clinical aspect was non-suggestive. Surgical biopsy by cervicotomy, with histologic and immunohistochemical analysis, provided definitive diagnosis.

Given the rarity of this entity and the poor specificity of paraclinical results, diagnosis is founded on pathology following extension assessment, which usually includes cervico-thoraco-abdominal CT and MRI. Once diagnosis has been made, certain authors recommend searching

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**Figure 1** Cervicothoracic aspect: isolated lesion medial to the right sternocleidomastoid muscle, 20 × 17 mm, non-enhanced on contrast medium injection.

**Figure 2** Microscopic aspect, × 20: positive immunolabeling for tumor-cell PS100 (black arrow).

**Figure 3** Macroscopic aspect on formaldehyde-fixed tissue: poorly contoured, whitish non-encapsulated nodular tissue lesion, 3 cm diameter.
for multiple locations, found in 10% of cases [7]. As locations are ubiquitous, clinical exploration for other palpable tumefactions, as well as cervico-thoraco-abdominal CT, is recommended.

The histological origin of GCT has long been debated. The entity has had many names: Abrikosoff’s tumor, myoblastoma, myoblastic myoma, and granular cell tumor. A smooth-muscle origin was long supposed. Electron microscopy and the presence of immunoenzymatic reactions with neurogenic markers (S100 protein and NSE), however, argue for a Schwann-cell origin [8]. Muscular markers (smooth-muscle actin or desmin) are negative [2]. The location in the present case supports a neurogenic origin.

In the present case, NSE was negative but S100 protein strongly positive.

Curative treatment in GCT is surgical. Recurrence seems to be exceptional if resection has been complete [3]. In malignant forms, 2–3 cm margins should be respected, to reduce the risk of local (30%) or metastatic (50–60%) recurrence; malignancy, however, is reported to be rare (2%) [9], although cases of pulmonary lymph-node or metastasis have been reported [10].

Conclusion

We report a case of GCT in a deep cervical plexus root. This hitherto unreported location supports a neurogenic origin for such ubiquitous but rare tumors. Definitive diagnosis relies on pathology and immunohistochemistry, following surgical exploration. Curative treatment is surgical, with complete resection.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References