

## Case Report

# Granular cell tumor of the hypoglossal nerve presenting with tongue atrophy and deviation: a case report

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**Abstract:** Granular cell tumors (GCT) are rare tumors of Schwann cell origin which were more often found in subcutaneous locations than in relation to major nerves. We describe a GCT that occurred within the hypoglossal nerve located in the left submandibular area of a 32-year-old man who initially presented with a muscular atrophy and deviation of the tongue. We also review the literature related to the differential diagnoses of such a submandibular tumor, with particular emphasis on the origin of these possibilities. In this case, the lesion was observed within the hypoglossal nerve in the submandibular area. The hypoglossal nerve was transected above and below the mass in order to achieve an adequate resection. The pathology demonstrated polygonal cells with diffuse eosinophilic granular cytoplasm and positive for CD68 and S-100 which established the diagnosis of GCT. This is the first patient, to our knowledge, with a GCT arising from the hypoglossal nerve.

**Keywords:** Granular cell tumor, hypoglossal nerve

## Introduction

Granular cell tumors as rare soft tissue tumors of nervous system and originate from Schwann cell [1-4]. They tend to be found in widespread subcutaneous locations such as the oral cavity and abdominal viscera [5]. However, these tumors rarely occur along major nerves.

The hypoglossal nerve is the twelfth cranial nerve which innervates muscles of the tongue. It provides motor control of both the extrinsic muscles and intrinsic muscles of the tongue. The hypoglossal nerve passes to the submandibular region after passing deep to the posterior belly of the digastric muscle, passes lateral to the hyoglossus muscle, and inferior to the lingual nerve to reach and efferently innervate the tongue. Lesions affected the hypoglossal nerve lead to atrophy of muscles of the tongue and show deviation of the tongue.

We report the first patient, to our knowledge, with a GCT involving the hypoglossal nerve presenting with a tongue atrophy and deviation. Although GCTs are rare pathological entities, several aspects of this patient's presentation

relate to classic features of these lesions. We review these features and contrast them with other differential diagnoses of a tumor arising from the submandibular area.

## Case report

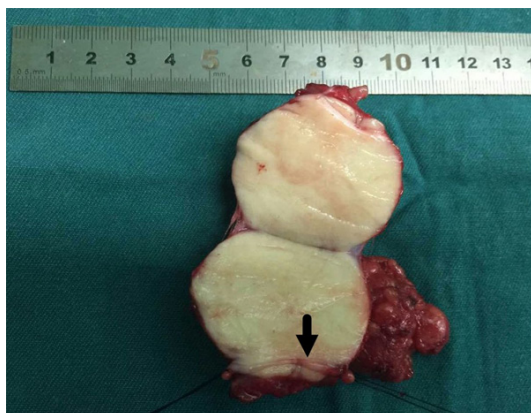
A 32-year-old man presented with tongue atrophy and deviation for one year. On examination, the patient demonstrated a 5.0 cm solid tumor in the left submandibular area and an obvious atrophy and deviation in the left tongue. No change of taste, numbness of tongue, mouth deviation or Skin perception changed in neck. An MRI demonstrated an obvious atrophy in the left tongue (**Figure 1A**) and a well-circumscribed homogenously enhancing mass of hypo signal on both T1 and T2-weighted images which lay a posteriorly to the submandibular gland (**Figure 1B, 1C**). CT angiography demonstrated the mass of intermediate intensity anteromedial to the carotid artery (**Figure 1D**). Fine needle aspiration (FNA) demonstrated polygonal cells with eosinophilic granular cytoplasm.

A 7 cm transverse incision was extended in the left submandular area, 3.0 cm below the man-

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**Figure 1.** A. Axial pre contrast T1-weighted MRI demonstrated an obvious atrophy (fatty degeneration) in the left tongue muscle (white arrow). B. Parasagittal pre contrast T1-weighted MRI demonstrated a well-circumscribed homogeneously enhancing mass of hypo signal which is posterior to the submandibular gland. C. Axial pre contrast T2-weighted MRI demonstrated a hypo signal mass without a clear capsule. D. Axial post contrast CT angiography demonstrated the isolated lesion anteromedial to the left carotid artery and jugular vein, 5 × 5 cm, non-enhanced on contrast medium injection. White arrows note the close relation of the tumor to these structures.



**Figure 2.** Macroscopic aspect on fresh tissue: The tumor was a 5.0 × 4.5 × 4.0 cm firm, whitish, non-encapsulated nodular tissue lesion and the hypoglossal nerve went through the tumor (black arrow).

dible. The submandibular gland was mobilized anteriorly from their posterior aspects. The digastric muscle anterior to the tumor with no clear boundary was divided, allowing visualization of the hypoglossal nerve, the carotid sheath, and tumor. The carotid artery and internal jugular vein were identified and preserved. The tumor was debulked and gentle traction was placed on the capsule to facilitate a total dissection. After tumor debulking, it was clear that the hypoglossal nerve went through the tumor and could not be separated. The hypoglossal nerve was sectioned above and below the tumor, facilitating tumor removal.

The tumor was a 5.0 × 4.5 × 4.0 cm firm, whitish, non-encapsulated nodular tissue lesion and the hypoglossal nerve went through the tumor (**Figure 2**). Light microscopy demonstrated diffuse eosinophilic granular cytoplasm (**Figure 3A**). No necrosis, prominent nucleoli, or increased mitotic activity were present. Immunohistochemical stains for CD68 (a marker of lysosomes), S-100 (a marker of

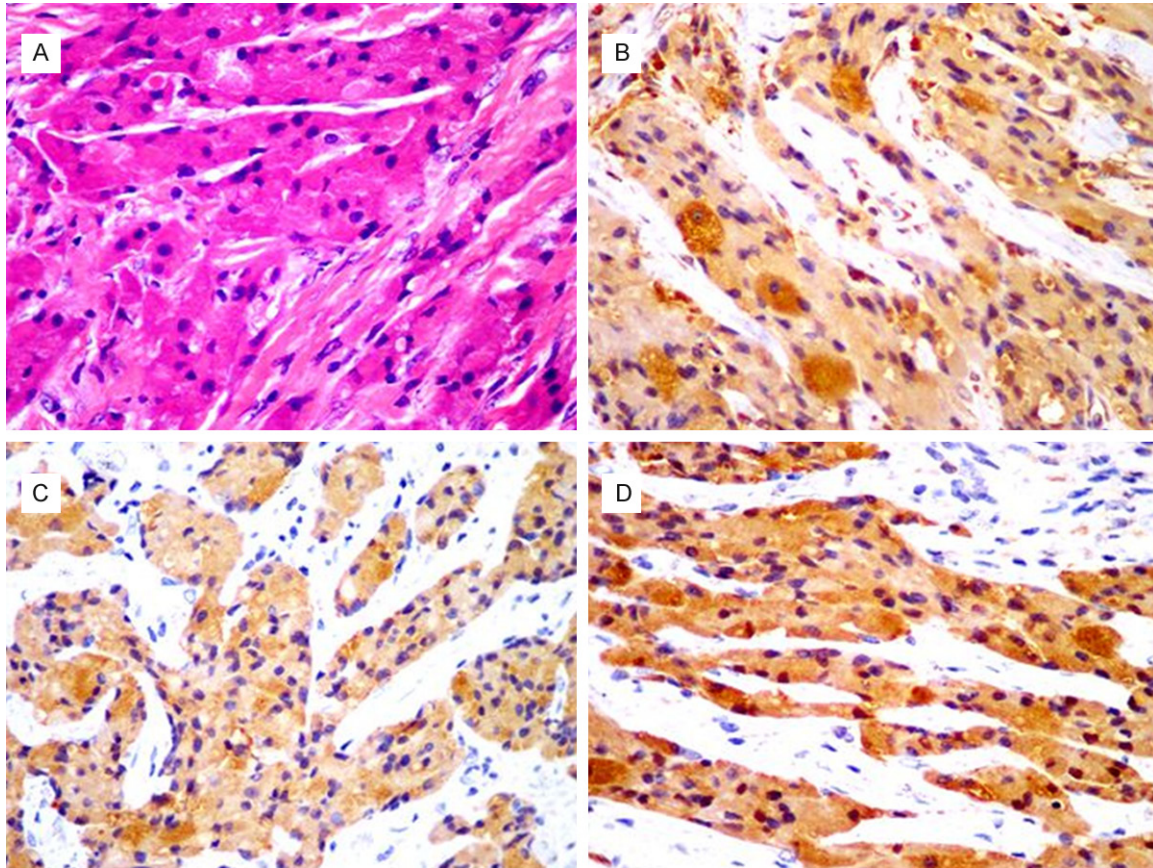
neural crest derivatives [6]) and NSE were positive (**Figure 3B-D**). These microscopic features and staining properties established the diagnosis of GCT.

Postoperatively, speech and swallowing functions were preserved and his tongue atrophy and deviation did not improve. Postoperative imaging confirmed complete resection and radiation therapy was not indicated given the benign diagnosis. A follow-up by ultrasound 3 months after surgery showed no evidence of recurrence.

### Discussion

GCTs are uncommon, usually benign neoplasms which were first reported by Abrikossoff

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**Figure 3.** A. Microscopic examination showed polygonal cells, small round nuclei and abundant granular eosinophilic cytoplasm. No necrosis, prominent nucleoli, or increased mitotic activity were present. (HE staining  $\times 40$ ). B. Immunostaining for CD68+ (SP staining  $\times 40$ ). C. Immunostaining for NSE+ (SP staining  $\times 40$ ). D. Immunostaining for S-100+ (SP staining  $\times 40$ ).

in 1926 [2]. One of its characteristics is that eosinophilic granules are contained in the cytoplasm of cells. Since the positive rates for S-100 protein are high, currently GCT was thought to originate from Schwann cells. About fifty percent of GCTs located in the submucosa or the muscle of the head-and-neck region, within which the tongue, palate and floor of the mouth are the most frequently involved organs, but the tumor seldom occurred along major nerves [7]. GCT located in the hypoglossal nerve has, to the best of our knowledge, never previously been reported.

Although usually benign, these lesions may present as solid tumors with ill-defined margins and ulcerated surface, masquerading as a malignant tumor at initial presentation. In the present case, GCT presented a submandibular mass and the atrophy of tongue. Preoperative assessment rarely enables diagnosis. In the

present case, CT suggested non-necrotic adenopathy in the left submandibular triangle. Data on the imaging aspects of GCT were rare. CT finds a solid, more or less heterogeneous tumor. On MRI, the lesion is usually in hypo signal on T1 and hyper signal on T2-weighted sequences, enhanced by contrast medium [8]. In the present case, it showed in hypo signal on both T1 and T2, and was not enhanced by contrast medium. In the present case, it showed spindle shaped cell. Clinical and radiological assessment left several diagnostic hypotheses open: hypoglossal nerve schwannoma was conceivable, although the MRI aspect did not support this; pleomorphic adenoma in the submandibular gland or metastatic malignant tumor in neck were unlikely, given FNA aspect.

Operation, with histologic and immunohistochemical analysis, provided definitive diagnosis. The histological origin of GCT has long been

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debated. A smooth-muscle origin was long supposed until electron microscopy and the presence of immunoenzymatic reactions with neurogenic markers (S100 protein and NSE) argue for a Schwann-cell origin [9]. The location in this present case strongly supports a neurogenic origin. In the present case, NSE and S100 were strongly positive.

Curative treatment in GCT is surgical. Recurrence seems to be exceptional if resection has been complete [7]. In malignant forms which were reported to be rare (2%), 2.0-3.0 cm margins should be respected, to reduce the risk of local or metastatic recurrence [10].

In summary, our case is unique because of its unusual location and rare. This hitherto unreported location supports a neurogenic origin for GCTs. Definitive diagnosis relies on pathology and immunohistochemistry, following operation. Curative treatment is surgical, with complete resection.

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### Disclosure of conflict of interest

None.

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