

Case Report

An Unusually Large Granular Cell Tumor of the Pharynx: A Case Report and Literature Review

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Received 6 August 2008; Accepted 9 September 2008; Available online 1 October 2008

Abstract: We report a granular cell tumor of the pharynx in a 53 year-old woman who presented with a large retropharyngeal mass. Surgical excision revealed a 5.5 cm tan rubbery unencapsulated but circumscribed mass. Histologically, the tumor is composed of diffusely arranged oval and spindle cells with abundant eosinophilic granular cytoplasm and mildly pleomorphic nuclei without necrosis or mitoses. Immunostains show the tumor cells to be positive for S-100, vimentin, non-specific esterase and focally positive for inhibin. In addition to its unusual location, this tumor is extremely large while most granular cell tumors are small (<2 cm). This case represents a unique example of a large granular cell tumor at a rare location: the pharynx.

Key Words: Granular cell tumor, pharynx

Introduction

Granular cell tumor is a relatively uncommon neoplasm occurring in all ages and throughout the body. The incidence peaks in the third decade and the tumor occurs more frequently in women (2:1) and in African Americans (5:1) [1].

Approximately half of the cases occur in the head and neck region, with one third of these occurring in the tongue. Granular cell tumor of the larynx is a well recognized but uncommon lesion, accounting for approximately 3-10% of the cases. Granular cell tumor of the pharynx, however, is a very rare lesion.

Granular cell tumor was first described by Abrikossoff in 1926 [2]. Since then, more than 300 cases have been reported [3]. It often occurs in the skin and subcutaneous tissue. About 50% of the tumor occur in the head and neck area, of which one third occur in the tongue. Less common sites include the lip, buccal mucosa, maxilla, palate, trachea, bronchi, gastrointestinal tract, breast, urogenital tract, neurohypophysis, and orbit [3]. Involvement of the larynx is uncommon [4]. Involvement of the pharynx is extremely

rare. Only two cases were reported in the English literature in the last 50 years after extensive literature search [5].

Case Report

A 53 year old female patient presented with a history of difficulty swallowing, which first started in the early 1990's and progressed since then. In June, 2004, when the patient underwent a hernia repair surgery, the anesthesiologist was unable to pass the endotracheal tube. Physical examination revealed a muffled voice. No stridor or shortness of breath was found. CT and MRI (**Figure 1**) showed a retropharyngeal mildly enhanced soft tissue mass (5.5 cm). The impression of the CT imaging was lymphoma with differential diagnoses including mucoepidermoid carcinoma, adenocystic carcinoma, pleomorphic adenoma and benign lymph node. The patient was subsequently treated with surgical resection of the mass.

Incisional biopsy was sent for intra-operative frozen diagnosis and diagnosed as spindle cell neoplasia. An excision of the mass was performed after frozen section diagnosis. Gross examination showed a tan rubbery

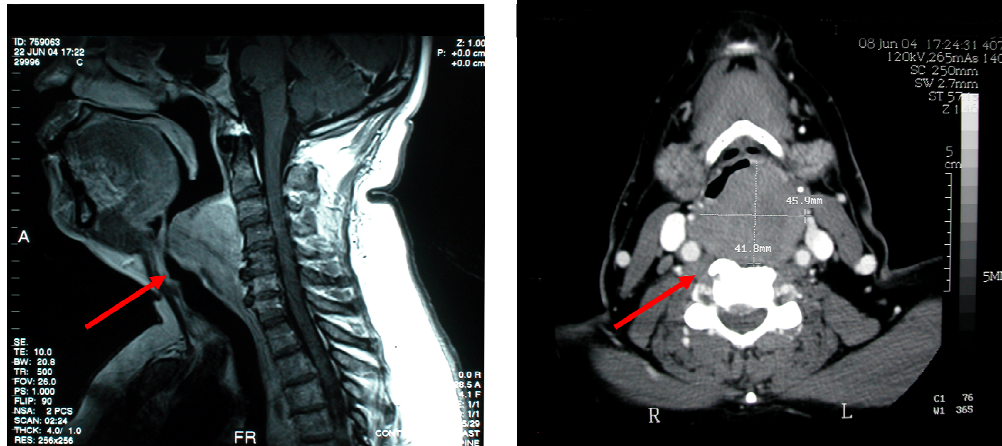


Figure 1 MRI (left) and CT (right) showed a large mildly enhancing retropharyngeal soft tissue tumor (red arrows).

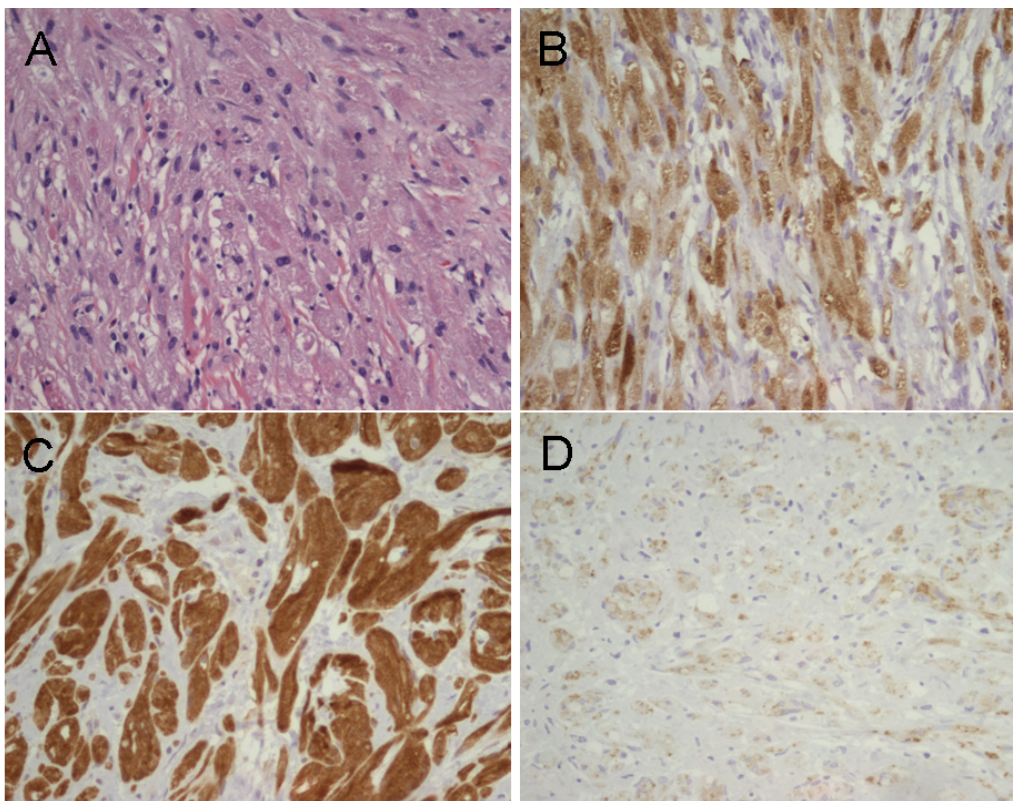


Figure 2 Histological appearance and immunostains of the granular cell tumor. **A.** Medium power view (200x) of the tumor shows a mixture of epithelioid and spindle-shaped cells with abundant granular cytoplasm. No mitosis was identified. The tumor cells show diffuse nuclear and cytoplasmic immunostaining for S-100 (**B**) and NSE (**C**) with only focal and weak cytoplasmic immunostaining for inhibin (**D**).

mass with homogenous texture. The haematoxylin & eosin (HE) stain showed a fairly demarcated nodular lesion. The cells varied from epithelioid to spindle-shaped with

abundant granular cytoplasm. The tumor cells showed slight variability, but no bizarre cells or mitosis was identified. No necrosis was identified (**Figure 2**).

A battery of immunostains demonstrated that the tumor cells are diffusely positive for S-100, vimentin and nonspecific esterase (NSE), and focally positive for inhibin. They are negative for CD34, smooth muscle actin, cytokeratin and desmin (Figure 2). Follow-up of the patient has not revealed any sign of recurrence 4 years post surgical resection.

Discussion

Granular cell tumor usually presents as a single slow-growing benign small nodule. More than two thirds are less than 2cm [3, 6]; however concurrent multiple nodules [7, 8] and malignant counterparts have been reported [9]. Granular cell tumor can induce pseudoepithelial hyperplasia, which can mimic squamous cell carcinoma [10]. Granular cell tumors usually either present as asymptomatic small nodules or show nonspecific symptoms such as hoarseness, cough, pain, dyspepsia or dysphasia with bigger size [3, 11]. Malignant granular cell tumors show high nuclear/cytoplasm ratio, pleomorphism, high mitotic activity and necrosis [11, 12].

The typical histological appearance of benign granular cell tumor is large epithelioid or spindle-shaped cells with syncytial abundant eosinophilic granular cytoplasm. The tumor cells are positive for S-100 and NSE, which are suggestive of its Schwann cell origin [8, 13].

Small tumors (less than 1 cm) can be closely monitored and large benign tumors are usually endoscopically or surgically resected [3, 11, 14]. Recurrence of granular cell tumor after adequate resection has been reported [11].

In summary, our case is unique because of its large size and rare, unusual location. However, it has the characteristic histological appearance and immunohistochemical profiles as other atypical small granular cell tumors.

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