CASE REPORT

Granular Cell Tumor Presenting as a Large Leg Mass

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Abstract

Granular cell tumor is a rare benign neoplasm most commonly appears in the head and neck region, especially in the tongue, cheek mucosa, and palate. Occurrence in limbs is even rarer. These tumors account for approximately 0.5% of all soft tissue tumors. Granular cell tumor can also affect other organs including skin, breast, and lungs. Local recurrence and metastasis is potentially higher in malignant forms with poor prognosis in respect to the benign counterparts. The average diameter of the tumor is usually about 2-3 cm. We report a granular cell tumor in the leg with an unusual size.

Key words: Head, Leg, Neck, Tongue, Tumor

Introduction

Granular cell tumor is an uncommon benign neoplasm first described in German literature by Abrikossoff in 1926 (1). These tumors are rare and account for approximately 0.5% of all soft tissue tumors (2). This tumor was initially called 'granular cell myoblastoma' because the origin was assumed to be from skeletal muscles. Various theories on the origin of granular cell tumor have subsequently been proposed including striated muscles, and histiocytes, as well as the neural origin. Nevertheless, granular cell tumors can affect any part of the body. Most granular cell tumors occur in the head and neck region, especially in the tongue, cheek mucosa, and palate (3). Granular cell tumor can also affect other organs including skin, breast, and lungs (4). The classic location of granular cell tumor, formerly known as granular cell myoblastoma, is the tongue (5). Approximately 0.5% to 2% of granular cell tumors are malignant (6, 7). In this form, local recurrence and metastasis is potentially higher with poor prognosis in respect to the benign counterparts.

Case presentation

A 36-year-old man presented with a mass in posteromedial aspect of the left leg, which was first noted approximately four years ago, however the size of the mass has been increased gradually over the past few months. The mass was not painful or tender.

Preoperative magnetic resonance imaging showed a focal, solid, well-defined, and lobulated lesion in the medial compartment of the leg with no involvement of the neurovascular bundle or no extension to the bony structures. On T1-weighted sequences of the MRI, the tumor was showing iso-intense signal compared to the surrounding muscle tissue. Perilesionl edema was absent. On T2-weighted sequences, the tumor had lower signal intensity than fat but slightly higher than muscles. The signal intensity of the tumor was heterogeneous together with a surrounding peripheral rim of high intensity signal [Figure 1].

In the operation room, the mass was approached from posteromedial aspect of the leg. We had decided to take an incisional biopsy in the first place, but as we approached, the tumor was encapsulated with no extension to periphery. Thus, we decided to perform en-block excision. We resected a lobulated and capsulated yellowish solid mass with diameters of about 11×7×5cm [Figure 2]. Light microscopic analysis showed granular cell tumor comprising of large polygonal or elongated cells with eosinophilic granules within the cytoplasm [Figure 3, 4].

We followed up the patient in the clinic for one year to detect any local recurrence or distant metastasis.

Discussion

Granular cell tumor is a rare tumor that can affect various regions of the body, but more frequently found in the head and neck region commonly involving the tongue. Few cases of intramuscular tumors are reported in the extremities (8). Abrikossoff was the first described this tumor in 1926 and named it a myoblastoma because of its origin from the tongue muscle (1). Over the following 60 years, the tumor was thought to have a neural origin because it was assumed to be from the

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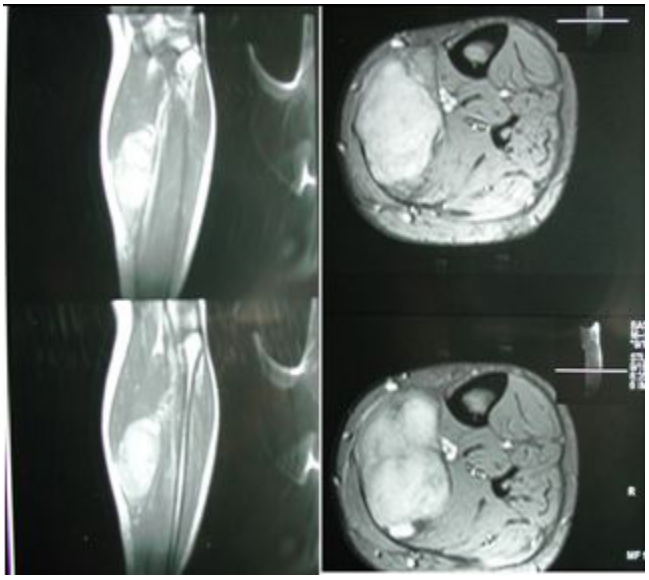

Figure 1. MRI imaging show intramuscular lesion in the leg.

radial and sciatic nerves within the extremities (9). Granular cell tumor usually presents with a painless mass occurring most commonly during the third to fifth decades of life. Women are more commonly affected with a reported ratio of 1.8–2.9:1.

The best imaging modality for the characterization of this tumor is magnetic resonance imaging (MRI) (9). Clinical findings for prediction of malignancy includes large tumor size (>5 cm), older age, female gender, oval or round shape, deep location (intramuscular), occurrence in the lower extremities, rapid recent growth after an extended period, and local recurrence (7, 10). On MRI, findings in support of the benign tumor include a superficial lesion with iso-intense or brighter than muscle signal on T1-weighted sequences. Furthermore on T2-weighted sequences, the center signal should be

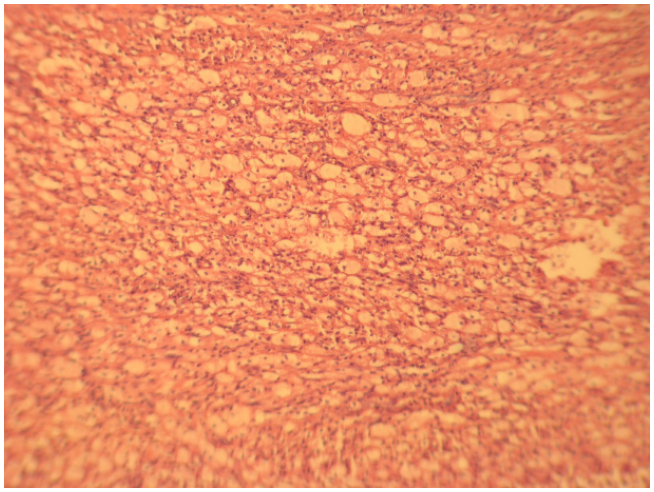

Figure 3. Pathologic slide of tumor with low magnification.

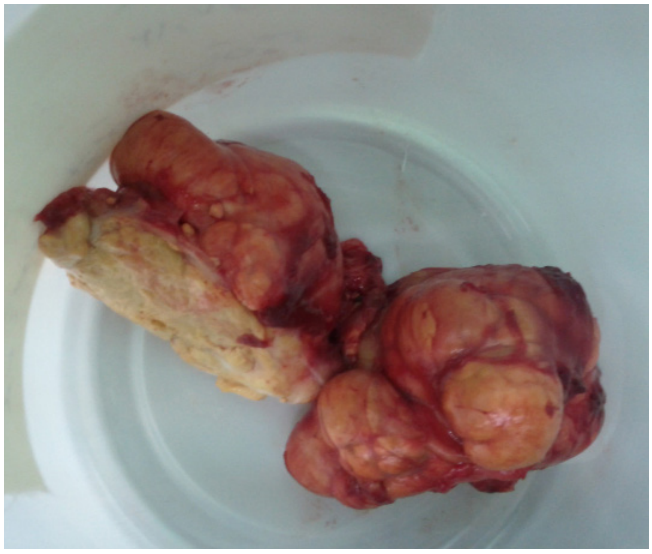

Figure 2. Macroscopic appearance of tumor that bissect from longest diameter.

iso-intense to muscle or suppressed fat with high signal in the periphery of the lesion. However, any invasion to the adjacent structures demonstrates a malignant granular cell with signal intensity characteristics similar to other aggressive neoplasms.

Fanburg-Smith et al reported histologic criteria for malignant granular cell tumors by analyzing the clinical and histologic data of 73 cases of granular cell tumors. According to their report, they explained six features including necrosis, spindling of cells, vesicular nuclei with prominent nucleoli, increased mitotic activity (two mitoses per 10 high-powered fields), high nucleocytoplasmic ratio, and pleomorphism. Having three or more out of six should be considered malignant, which may result in death in 40% of cases because of the

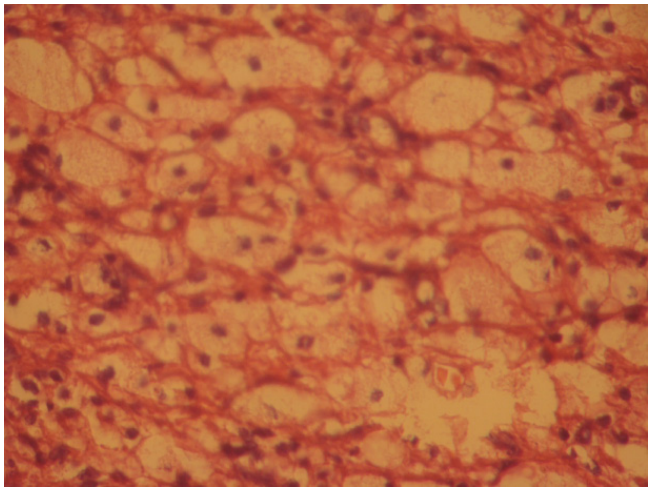

Figure 4. Pathologic slide of tumor (medium magnification) consisted of large polygonal or elongated cells with eosinophilic granules in cytoplasm.

high chance of local recurrence and metastasis. Tumors having one to two criteria should be considered atypical. Tumors with none of those criteria should be considered benign having an excellent outcome with no metastasis or local recurrence after adequate resection (10).

Surgical excision with a safe and clean margin is the treatment of choice for this tumor although this is not always possible because of lacking a surrounding capsule or proximity to structures such as nerves or vessels.

We reported a case of granular cell tumor in the lower extremity, which was interesting in terms of rarity in this location (intramuscular), and large size (the maximum reported size was 5cm while the tumor in our report was 11cm), however it was well encapsulated. The patient underwent wide resection of the tumor, which was benign with no recurrence so far.

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