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Granular Cell Tumor Occurring in the Chest Wall: A Case Report

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Granular cell tumors are uncommon soft tissue neoplasm of nerve sheath origin, which are predominately benign. Granular cells can be found at any site in the body including the tongue, skin, subcutaneous tissue, breast, gastrointestinal, and urogenital systems. However, granular cell tumors have only been rarely described in the chest wall. Here we report a case of a granular cell tumor that occurred in the chest wall of a 59-year-old woman, along with a review of the literature.

Key words: 1. Chest wall
2. Surgical operation
3. Granular cell tumor
4. Neoplasms

CASE REPORT

A 59-year-old woman presented to our hospital with a four-month history of a non-tender, hard mass at her left lateral chest wall on the 7th rib. She had no previous history of any significant illness. Physical examination revealed a 4×3 cm, movable, and mildly tender mass. It was located in the subcutaneous tissue. The overlying skin was normal. Ultrasonography showed a solid mass 3 cm in diameter, which had caused an acoustic shadowing in the posterior aspect (Fig. 1). The patient underwent a surgical excision of the tumor. The mass was carefully dissected using electrocautery as it was severely attached to the latissimus dorsi muscle. The surgical specimen was transferred to the department of pathology for accurate diagnosis. The specimen (4.0×3.5×1.7 cm) consisted of pinkish, soft tissue. On the cut section, the cut surface

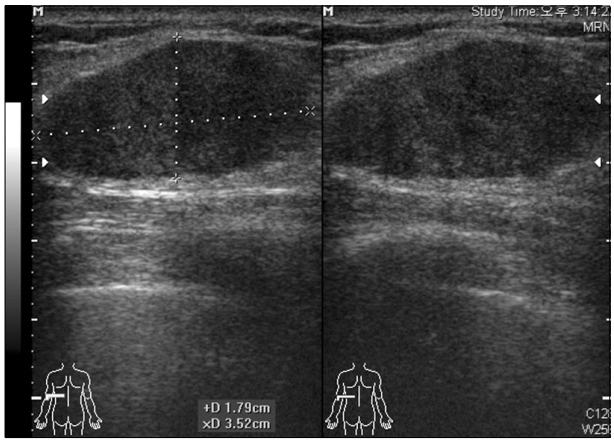

Fig. 1. Ultrasonography shows an ill-defined and markedly hypoechoic mass with posterior acoustic shadowing and a surrounding hyperechoic halo.

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showed a yellow homogeneous appearance. There was no hemorrhage or necrosis within the lesion, observed grossly (Fig. 2). The histological examination revealed polygonal cells with eosinophilic granular cytoplasm, fibrous septae between the clusters, cells with vesicular nuclei with a prominent nucleolus, eosinophilic granular cytoplasm, and eosinophilic intracytoplasmic particles surrounded by a clear halo (Fig. 3A). The immunohistochemical examination showed that S-100 protein and CD68 were positive, and cytokeratin was negative (Fig. 3B). Electron microscopic findings showed the basal membrane of the tumor cell, and autophagosome granules of varying size (Fig. 3C). These findings led us to the diagnosis of benign granular cell tumor. The patient had an unremarkable post-operative recovery and was discharged the next day. After a 3-month follow-up, the lesion has shown no recurrence.

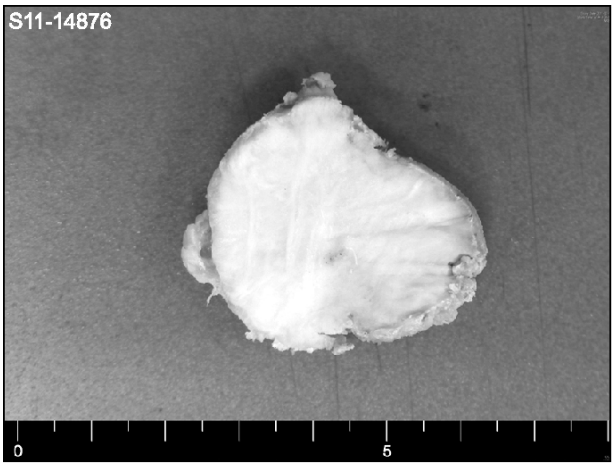

Fig. 2. Upon gross examination, the cut surface shows a yellow homogeneous appearance. There was no hemorrhage or necrosis within the lesion.

DISCUSSION

The granular cell tumor (GCT) is rare and almost benign. It usually develops as a painless mass, and most patients present with a mass as the primary symptom. GCTs were first described in the skeletal muscle of the tongue by Abrikossoff as “myoblastic myomata” in 1926 [1]. The GCT incidence is higher for women than men, with a ratio of 1.8－2.4/1. It is a rare tumor predominantly found in the skin or subcutaneous tissue of the head and neck regions. Other frequent locations are the tongue (40%), breast (15%), respiratory tract (10%), and esophagus (2%). GCTs can often be multicentric (5% to 14% of cases) [2]. In Korea, Seo et al. [3] reported a GCT in the main bronchus. GCT is very rarely found in the chest wall, and no Korean studies have reported it.

Abrikossoff [1] described this type of neoplasm as a myoblastoma, reflecting its probable origin from striated muscle cells. The histogenesis of these tumors is still controversial, but their immunohistochemical and ultrastructural features

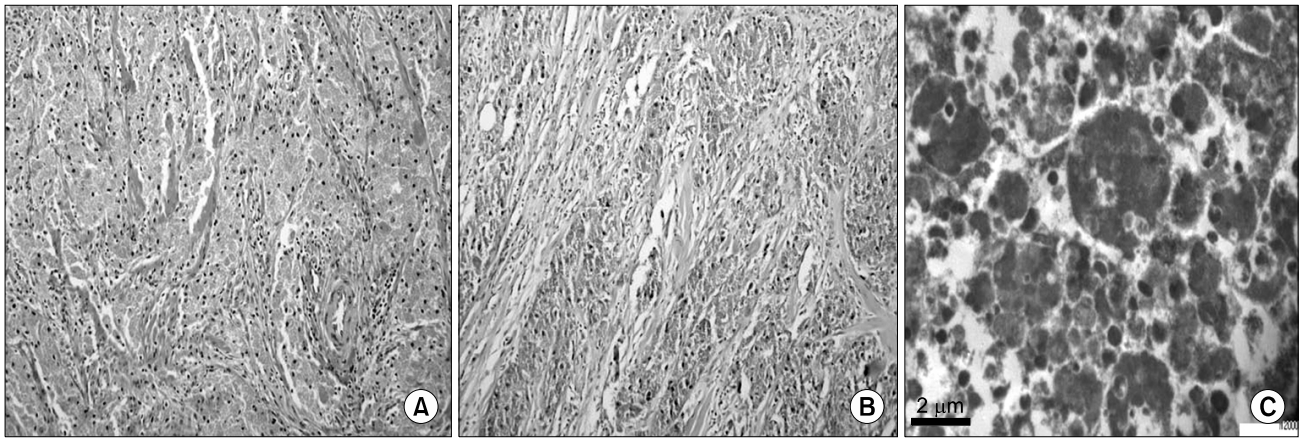

Fig. 3. (A) Upon microscopic examination, the tumor was found to be composed of large polygonal cells arranged in cords or sheets that were divided by slender fibrous tissue (H&E, ×400). (B) Upon immunohistochemical examination, the tumor was found to be positive for S-100 protein (×400). (C) Electron microscopy revealed numerous variably sized phagolysosomes in the cytoplasm.

have been accepted as neural or neuroectodermal in origin [4] because they are immunoreactive for the S-100 protein and neuron-specific enolase, and they demonstrate lysosomes on electron microscopy. They are also characterized by immunopositivity for CD68 and inhibin-alpha, but they are negative for cytokeratins [5].

Microscopically, GCTs are characterized by large polygonal tumor cells with an abundant granular cytoplasm and relatively small nuclei. The granules in the cytoplasm are periodic acid-Schiff-positive and diastase-resistant. The tumor cells may on rare occasion show a moderate degree of the nuclear atypism. The cells are arranged in nests or cords separated by fibrous connective tissue. The periphery of the tumor is not sharply defined, and this creates an appearance of infiltration that is highlighted when lymphoid tissue envelops the tumor cells.

The differential diagnosis of GCT includes rhabdomyoma, hibernoma, oncocytoma, extragastrointestinal stromal tumor (EGIST), and the reactive changes associated with trauma and injury. The histologic characteristics and reactivity toward S-100 and CD68 distinguish GCT from rhabdomyoma, which contains glycogen and hibernoma, which contains lipid droplets. Ultrastructurally, the lack of mitochondria differentiates GCT from oncocytoma. GCTs can mimic EGIST with epithelioid cells. The negative reaction to c-kit and CD34 distinguish GCT from EGIST. The inflammatory cells and areas of necrosis in the reactive change from trauma and injury are absent in GCT [6].

The distinction between a benign, atypical, and malignant GCT is challenging based upon morphology alone, as the different grades are all histologically similar. Furthermore, malignant granular cell tumors are exceedingly rare, comprising less than 1% of all granular cell tumors. The criteria proposed to distinguish between them includes: The presence of necrosis, spindle cell morphology, increased nuclear-to-cytoplasmic ratio, nuclear pleomorphism, prominent nucleoli, and increased mitotic rate (>2/10 high power fields) [7]. GCTs are classified as malignant if they display three or more of these criteria, as atypical if they display two, and as benign if they display only pleomorphism but no other criteria. The prognosis of malignant GCT is poor due to local recurrence and metastasis.

The treatment of choice for malignant GCT is radical resection with clear surgical margins [8]. Radiation and chemotherapy are not recommended due to the tumor's high degree of resistance to radiation or chemotherapy. Complete surgical resection is considered curative. Even when negative margins are not obtained, the prognosis is good. Patients with a malignant GCT are best managed with wide local excision and regional lymph node dissection [2].

We describe here a rare case of GCT that occurred at the chest wall. We expect that this report will assist in the understanding of the characteristics of GCT and in correctly diagnosing the disease.

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