A Rare Case of Mediastinal Granular Cell Tumor

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The granular cell tumor (GCT) occurs extremely rarely in the mediastinum. Few mediastinal GCT cases have been reported in Japan or other countries. Here, we report a case of a 24-year-old man with superior mediastinal GCT. The mass was located just above the aortic arch. It was firm, oval in shape, and well encapsulated. The tumor was removed completely with video-assisted thoracoscopic surgery, but we had to resect the vagus nerve, which was already included in the tumor, along with the tumor. After the operation, the patient recovered without any specific complications except for a mild degree of hoarseness.

Key words: 1. Granular cell tumor 2. Mediastinum

CASE REPORT

A healthy, 24 year-old male was admitted for an evaluation of a mediastinal mass that was incidentally identified on a chest X-ray. On the chest X-ray, a round mediastinal mass measuring 3.5 cm in diameter, was found above the aortic arch (Fig. 1). A computed tomography (CT) scan was performed to reveal a large, oval mass measuring 3.1×2.1 cm, in the aortic arch region (Fig. 2). Based on the CT data, we suspected thymoma and lymphoma. For a pathologic confirmation, we performed a CT-guided biopsy, which confirmed the granular cell tumor (GCT) located at the superior mediastinum. In the microscopic findings, the tumor cells were polygonal or ovoid, with abundant eosinophilic and granular cytoplasm. The nuclei were small, or hyperchromatic, eccentrically located (Fig. 3A). Immunohistochemistry staining with CD56 and S100 was positive, which was consistent with the diagnosis of GCT (Fig. 3B).

Because of the growth tendency of GCT, we decided to perform surgery to remove the mass. Video-assisted thoracoscopic surgery was performed successfully, but we had to resect the vagus nerve along with the tumor, as the mass encircled the nerve (Fig. 3). After the operation, the patient had no specific complications except hoarseness attributable to injury of the laryngeal nerve. When we followed up after 2 years, he showed no evidence of recurrence but his voice still had mild hoarseness.

DISCUSSION

Previously, GCT was considered to be a type of myoblastoma because it resembles a skeletal muscle cell. Immunohistochemistrical and electron microscopy studies revealed the similarity between GCT and Schwannian cells [1]. However, there are some differences in the ultrastructural features. Therefore, the histogenesis of GCT is still unclear.
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Fig. 1. Preoperative chest X-ray and computed tomography (CT) images. (A) A 3.5-cm round shape mass was located above the aortic arch. Preoperative chest X-ray. (B) 3×2 cm large, oval shaped mass located on the left paratracheal region. Axial CT image (white arrow).

Fig. 2. Intraoperative image and gross finding image. (A) Intraoperative image: the encapsulated round shape tumor was located above aortic arch. (B) Gross image: 3.5×2.6×1.7 cm.

Fig. 3. Microscopic images. (A) Granular cell tumor (GCT) with nerve fiber. Round or polygonal GCT cells were scattered between hyalined collagen bands (H&E, ×200). (B) Immunohistochemistry staining with CD56 (×400). (C) Immunohistochemistry staining with S-100 (×400).

[2]. GCT can appear anywhere, but it is frequently observed in the head, neck, breast, and skin regions. It has been reported that GCT is more common in women, black people, and people in their 40s to 60s [1].

The GCT in the lung or mediastinum has rarely been reported worldwide. Machida et al. [2] reported seven posterior mediastinal GCT cases. Three of them were accompanied with nerve involvement [2]. In another three reports, all three superior mediastinal GCTs involved the sympathetic nerve [3-5]. Half of the mediastinal GCTs reported thus far have been asymptomatic, while the others have shown symptoms including cough, chest pain, and wheezing [2].
The most common mediastinal tumors are thymoma, lymphoma, and neurogenic tumors. It is difficult to diagnose mediastinal GCTs on the basis of radiologic findings as there are no specific radiologic findings for mediastinal GCT and of the extremely low incidence of mediastinal GCT. Chest CT is generally useful in supporting the diagnosis of a mediastinal tumor. To diagnose mediastinal GCT, magnetic resonance imaging (MRI) is more helpful than CT because the MRI of GCT corresponds well with the different histological features as compared to that of a neurogenic tumor. On the T2-weighted images of GCT, the high-intensity regions on the periphery correspond to myxoid degeneration and the central high intensity corresponds to cystic, hemorrhagic, and necrotic features. Further, a gadolinium diethylene triamine penta-acetic acid-enhanced T1-weighted image shows contrast enhancement on the tumor surface [2].

Most cases of GCT are benign; therefore, they rarely develop a local recurrence. However, in incomplete resection patients, GCT can be locally aggressive and may recur [1,2]. Lack et al. [6] reported that 20.8% of incomplete tumor excision patients developed local recurrence. Therefore, a pathologically confirmed GCT should be completely resected, and the patient should be monitored regularly.

Generally, all cases of mediastinal tumors don’t have to be confirmed pathologically before operation even though it is suspected to be malignant. Further, the possibility of GCT should be kept in mind while diagnosing and managing a mediastinal mass.

All cases of mediastinal tumor do not confirm the pathology except suspected malignant tumors. In our case, the radiologist recommended a biopsy because of the possibility of lymphoma. Therefore, we performed a CT-guided biopsy before the surgical resection.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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