A Case of Mediastinal Seminoma Presenting as Superior Vena Cava Syndrome

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Abstract

Primary mediastinal seminoma is a rare tumor usually with nonspecific symptoms such as chest pain, dyspnea and hoarseness, but superior vena cava syndrome (SVCS) is an extremely uncommon syndrome. We report a 25-year-old man who presented with superior vena cava syndrome for 1 month and a 1-week history of retrosternal chest pain. Thorax computerized tomography demonstrated a soft tissue mass in the anterior mediastinum with stippled calcifications compressing the superior vena cava and other major vessels. Transthoracic biopsy showed primary seminoma of the anterior mediastinum. Combined chemoradiotherapy was performed and the patient was considered to have a partial response to treatment.

Key words: mediastinal seminoma, anterior mediastinum, extragonadal, superior vena cava syndrome (SVCS)


Introduction

Primary mediastinal seminoma is relatively uncommon; it mainly occurs in men of the second to fourth decades (1). When present, 30% of the patients are asymptomatic, while others have nonspecific signs and symptoms (2). The occurrence of signs and symptoms of SVCS in primary mediastinal seminoma is extremely rare. SVCS occurs in only 10% of all patients (2). To our knowledge, 12 papers (3-14) in total reported mediastinal seminoma with SVCS in the English language literature from 1958 to 2010. On CT image, calcifications are infrequent while cyst formation is common (15). In the present case, we report the clinical characteristics, imaging features, especially calcifications, pathology and treatment in a 25-year-old man with primary mediastinal seminoma experiencing unusual SVCS.

Case Report

A 25-year-old man presented with signs and symptoms of SVCS for 1 month and a 1-week history of retrosternal chest pain in our hospital. Ultrasonography of his heart revealed low echogenic masses anterior to the ascending aorta. There was swelling in his face and neck. Engorgements of the jugular vein, veins of both upper extremities and the abdominal vein were observed on physical examination. Computerized tomography (CT) of the thorax demonstrated a giant anterior mediastinal mass measuring 11×8×12 cm with an irregular shape and heterogeneous density. The mass adjacent to the aortic arch pushed the right lung and bronchus and compressed the superior vena cava. Punctate calcifications and cyst formation could be observed in the CT scan (Fig. 1). There were no constitutional symptoms such as fever or weight loss but night sweats. And no enlarged cervical lymph node or supraclavicular lymph node was detected. The blood analysis, including α-fetoprotein (AFP) and β human chorionic gonadotropin (HCG), was normal. According to the signs and symptoms of SVCS and retrosternal chest pain and the findings of an anterior mediastinal mass compressing major vessels, our initial working diagnosis included lymphoma and thymoma.

Complete resection was not viable due to the adherence of the tumor to the major vessels and its extension to the

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The patient was considered to have a partial response to treatment. During the 6-month follow-up period the patient was well without evidence of disease.

**Discussion**

Primary extragonadal germ cell tumors are uncommon. These tumors usually arise in the mediastinum followed by pineal gland, retroperitoneum and sacrococcygeal area (16). Primary mediastinal germ cell tumors account for approximately 10-15% of mediastinal tumors (1, 17) and their most common histology is seminoma. Primary mediastinal seminoma usually arises in the anterior mediastinum, and it mainly occurs in men whose age ranges from 20 to 40 years (1). At the initial diagnosis, patients typically present with nonspecific symptoms such as chest pain, dyspnea, hoarseness, and SVCS caused by compression of mediastinal structures. But SVCS is uncommon in patients. Polansky et al. (2), in a review of the literature, showed that mediastinal seminoma caused SVCS in only 10% of cases. We reviewed the English language literature published from 1958 to 2010, and found that 12 papers (3-14) in total reported mediastinal seminoma with SVCS. Thus, primary mediastinal seminoma presenting as SVCS as in the current patient is rare. Occasionally, β human chorionic gonadotropin (HCG) is elevated in a very few patients (15).

The histogenesis of primary mediastinal seminoma is still uncertain. Rosado-de-Christenson et al. (15) suggested that it originates from multipotential primitive germ cells which are “misplaced” along midline structures, such as the mediastinum, during their migration from the yolk endoderm to the gonad during early embryogenesis. While others have postulated that the tumor originates from the pineal gland and the thymus (18). Ravenel et al. proposed that the primitive germ cells, which the tumor originated from, retain the ability to proliferate and differentiate into embryonic or extraembryonic tissue during embryogenesis (1).

On X-ray, there is no pathognomonic or specific imaging feature. On CT scans seminomas usually appear as a large, coarsely lobulated mass, only slightly enhanced after administration of contrast material, which compress or invade adjacent tissue. CT findings of calcification are distinctly rare, but regions of cyst formation are common in the mediastinal seminoma (1, 15). In this regard, the punctate calcifications and cyst in the present patient are in accord with previous reports. Although calcifications are common in teratoma, they can be detected in the mediastinal seminoma. When we find a mediastinal mass with calcifications, mediastinal seminoma should also be considered in addition to teratoma.

Traditional treatments of mediastinal seminoma consist of surgery and radiotherapy. Treatment should begin with surgical operation; in particular, complete resection should be performed when possible. If only partial excision can be done, the procedure should be followed by radiotherapy (19). Mediastinal seminoma is a tumor with high radiosensitivity. Thus, radiotherapy alone or associated with sur-

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**Figure 1.** Chest CT of the initial diagnosis. (A) CT scan shows a large soft tissue mass in the anterior mediastinum with stippled calcifications (arrow). The superior vena cava is compressed by the mass. (B) CT demonstrates a cyst located in the left side of the giant anterior mediastinal lump (arrow). CT: computerized tomography.
recent years, the chemotherapy has advanced in the therapeutic management of mediastinal seminoma. Liu et al. (21) evaluated the clinical characteristics and survival outcomes of fifty-five patients with primary mediastinal germ cell tumor (PMGCT). Patients who received two treatments of chemotherapy plus surgery or radiotherapy had the longest survival time of 118.3 months compared with the median overall survival time of 87.9 months (p=0.000). They recommended that chemotherapy combined with a local therapy such as surgery or radiotherapy is a reasonable treatment strategy (21). It was reported that cisplatin-based chemotherapy is curative for the vast majority of patients with mediastinal seminoma (22). Other authors have concluded that cisplatin-based chemotherapy allows long-term disease-free survival in approximately 85% of patients with primary mediastinal seminoma (23).

Surgery was impossible in the present case. Considering that single cisplatin-based chemotherapy is incapable of eliminating the residual tumor or micrometastases due to the extensive properties of germ cell tumors in the mediastinum (21), we added radiotherapy at the basic level of cisplatin-based regimens and noted a striking reduction in size of the tumor at the end of irradiation. At last, the curative effect of the present patient is remarkable, and the prognosis is favourable, which was in accord with previous reports (21, 23, 24).

In conclusion, primary mediastinal seminoma presenting as superior vena cava syndrome (SVCS) with punctate calcifications is uncommon. Primary mediastinal seminoma should be considered as a possible differential diagnosis to avoid misdiagnosis if there is a lump in the mediastinum causing signs and symptoms of SVCS, despite the fact that it is not a normal reason for SVCS. Although calcifications are commonly found in the teratoma, mediastinal seminoma should also be considered if they are found in a mediastinal mass.

The authors state that they have no Conflict of Interest (COI).

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Figure 2. Pathological analyses of the primary seminoma. Low-power view (A) reveals small round and lobules tumor cells (Hematoxylin and Eosin staining, original magnification ×100). High-power view (B) demonstrates tumor cells characterized by distinct cell borders, clear cytoplasm, and central nuclei (Hematoxylin and Eosin staining, original magnification ×400).

Figure 3. Chest CT of post-treatment. (A) CT image reveals a striking reduction in size of the tumor and the major vessels returned to normal. (B) CT demonstrates that the anterior mediastinal lump is much smaller than the previous CT, while the cyst is slightly larger than initially observed (arrow). CT: computerized tomography.
References


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