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Primary Clear-Cell Sarcoma in the Mediastinum

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Key Words
Sarcoma · Mediastinal tumor · Thoracoscopy

Abstract
We report a case of primary clear-cell sarcoma (CCS) in the mediastinum. In October 2011, a 63-year-old man was admitted to our hospital for surgical resection. The tumor was completely excised by video-assisted thoracoscopic surgery. The tumor was well encapsulated and did not invade the pleura. Histological examination led to a final diagnosis of primary CCS in the mediastinum. The patient remains alive without evidence of recurrence at 15 months after surgery.

Introduction

Clear-cell sarcoma (CCS) is a rare malignant connective tissue tumor that was first described by Enzinger in 1965 [1]. CCS occurs most commonly in adolescents and young adults and the patients aged >60 years are rare. It has a high rate of local recurrence, regional lymph node metastasis, and distant metastasis [1, 2].

CCS occurs most commonly in the deep soft tissues of the extremities, with rare involvement of the head, neck, and trunk [2]. We experienced a very rare case of primary CCS in the mediastinum with successful surgical resection in an elderly patient.

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Case Report

In October 2011, a 63-year-old man presented with a mass in the right upper mediastinum on chest radiograph. Computed tomography showed a large oval mass (55 × 55 × 85 mm) with clear margins and heterogeneous enhancement in the right upper posterior mediastinum between the phrenic nerve and vertebrae (fig. 1a). The patient was admitted to our department for further investigation and surgical treatment. Magnetic resonance imaging showed areas of high signal intensity on T1-weighted imaging (fig. 1b), and 18F-fluorodeoxyglucose positron emission tomography showed abnormal uptake with a maximum standardized uptake value of 15.1. Serum tumor marker levels were within the normal ranges. Preoperatively, schwannoma was considered to be the most likely diagnosis. The tumor was excised by video-assisted thoracoscopic surgery, with negative surgical margins. The tumor was soft, well circumscribed, encapsulated, and did not invade the pleura.

Histological examination of the resected tumor with hematoxylin and eosin (HE) staining showed morphological features compatible with conventional soft tissue CCS [3]. The tumor cells were distributed in nests separated by fibrous connective tissue, showing an alveolar pattern (fig. 2a). The tumor cells had eosinophilic and clear cytoplasm and oval vesicular nuclei of varying sizes with characteristic prominent eosinophilic nucleoli. There were some melanin-producing cells (fig. 2b) and multinucleated giant cells in corymbiform arrangements.

Immunohistochemical analysis of the tumor cells was performed with a large panel of antibodies. The cells were positive for HMB-45 (fig. 2c), S-100 protein, calretinin, CD34, CD56, CD68, and CD117; and negative for Melan-A, cytokeratins AE1–AE3, cytokeratin CAM5.2, chromogranin A, desmin, caldesmon, myogenin, epithelial membrane antigens, CD57, CD99, and D2-40. The patient remains alive without evidence of recurrence at 15 months after surgery.

Discussion

CCS is a very rare soft tissue neoplasm, which occurs in the extremities in 90–95% of cases [1]. Our case was diagnosed with a primary CCS in the mediastinum. A search of the English literature revealed only one previously reported case of primary CCS in the mediastinum [4].

CCS is usually a lobular lesion with clear borders, and may be encapsulated. Immunohistochemical examination of the cells shows strong staining for S-100 protein and HMB-45 [2].

Primary CCS and metastatic malignant melanoma have similar histopathological features. Recent cytogenetic studies showed that CCS is associated with the translocation t(12;22) (q13;q12); this translocation is observed in up to 93% of patients with CCS [5], but never observed in malignant melanoma [6]. In our case, we were unable to determine whether this translocation was present, because of the lack of availability of such analysis in Japan.

The rarity of CCS in the mediastinum makes it difficult to draw conclusions regarding prognostic factors. Hocar et al. [7] reported that complete excision of the primary tumor with wide surgical margins appears to be the optimal approach to treatment, with or without adjuvant radiation therapy. CCS is an aggressive malignant tumor, and unlike most soft tissue sarcomas, it often metastasizes to regional lymph nodes [1]. In our case, we did

not have a definitive diagnosis at the time of operation, and therefore we did not perform excision of the tumor with wide margins or dissect the regional lymph nodes.

In most cases, CCS has a relentlessly progressive course and results in death because of widespread dissemination. The reported overall survival rates without local recurrence are 30% at 5 years and 16% at 10 years [7]. Some patients experience rapidly fatal progression, and late metastasis after many years of freedom from disease is also relatively common [8]. We believe that regular long-term follow-up with computed tomography, magnetic resonance imaging, and 18F-fluorodeoxyglucose positron emission tomography examination is important to identify local recurrence or distant metastasis as early as possible.

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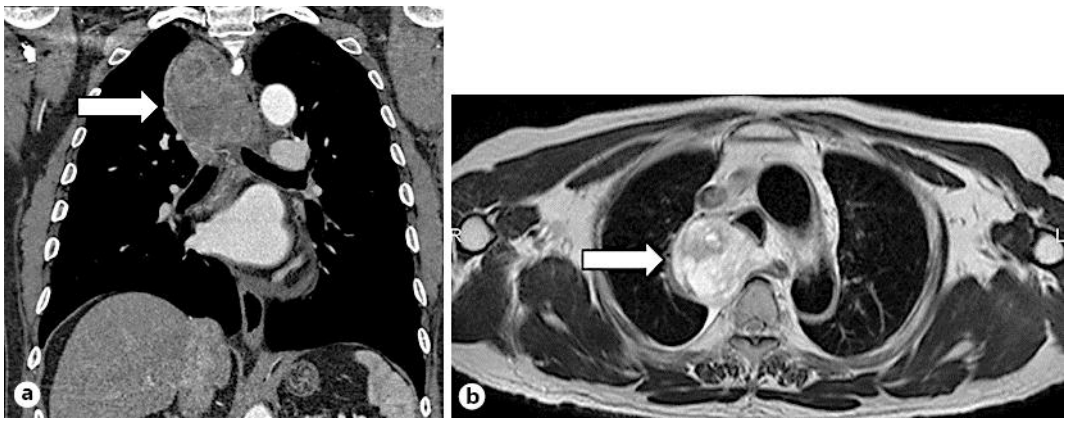

Fig. 1. Chest computed tomography (CT) and magnetic resonance (MR) (October 2011). **a** Enhanced CT image showing an oval mass (55 × 55 × 85 mm) in the right upper posterior mediastinum. The tumor mass was heterogeneous with clear borders. **b** T1-weighted MR image showing a mass with areas of high signal intensity.

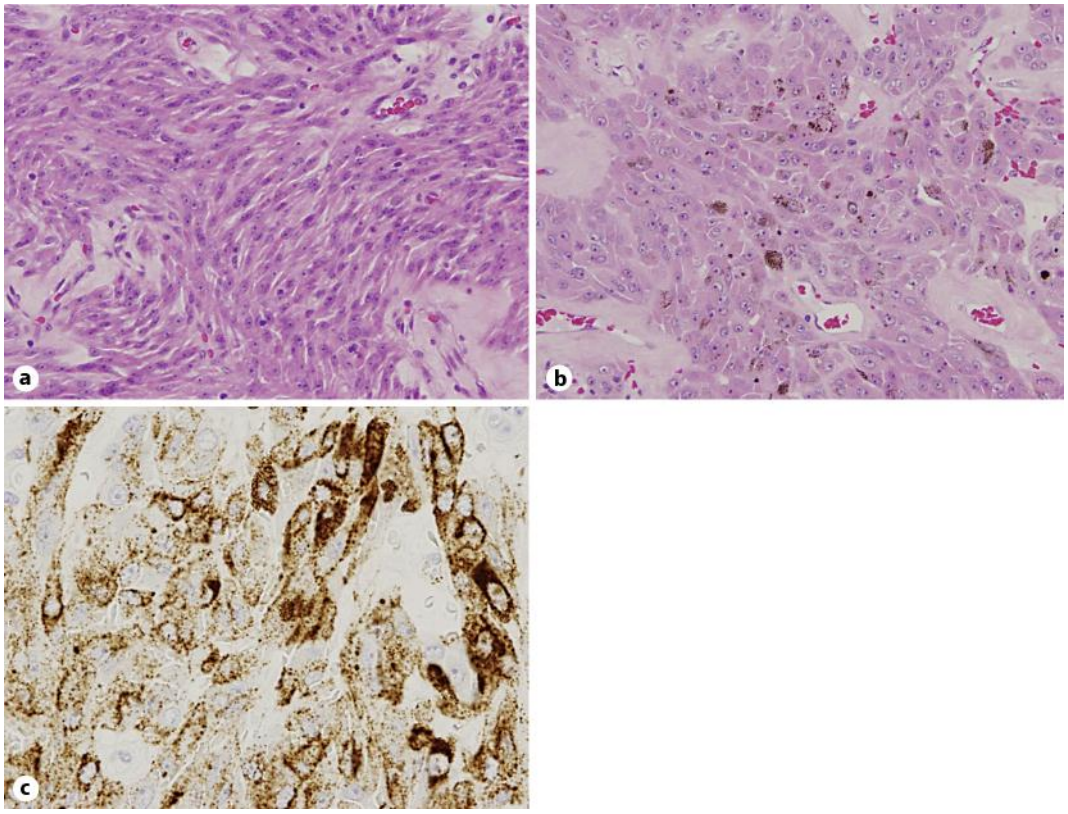

Fig. 2. Histopathological findings of the tumor. **a** The tumor cells were distributed in nests separated by fibrous connective tissue, showing an alveolar pattern (HE staining, ×20). **b** The tumor cells had eosinophilic and clear cytoplasm and oval vesicular nuclei of varying sizes, with characteristic prominent eosinophilic nucleoli. There were some melanin-producing tumor cells (HE staining, ×20). **c** The tumor cells were strongly and diffusely positive for HMB-45 protein (×20).