Sclerosing rhabdomyosarcoma (SRMS) is a newly recognized and rare variant of rhabdomyosarcoma. This soft tissue tumor has not yet been reported as a thoracic lesion. We report a case of a 26-year-old woman who presented with a large chest wall tumor. The tumor originated from the right anterior chest wall and protruded into the intra- and extrapleural cavity. A transcutaneous needle biopsy revealed spindle cells in an abundant hyalinized and fibrous stroma. Although the tumor was considered as a malignant soft-tissue neoplasm, a definitive diagnosis could not be established. A wide excision of the chest wall including the second, third and fourth rib and a part of sternum was performed. Histologically, cytoplasmic cross-striations were found in a portion of the tumor cells. The tumor cells were positive for muscle markers, and the tumor was diagnosed as rhabdomyosarcoma consistent with a sclerosing type of rhabdomyosarcoma. Eighteen months after the complete resection, the patient has pleural disseminations but is alive and undergoing chemotherapy. This case highlights the histologic features of a rare form of rhabdomyosarcoma, and emphasizes the importance of awareness of its existence and the utility of skeletal muscle markers in distinguishing sclerosing rhabdomyosarcoma from its mimics.

Keywords: sclerosing rhabdomyosarcoma, chest wall, thoracic surgery

Introduction

Although rhabdomyosarcoma is the most common malignant soft-tissue tumor of childhood and adolescence, this tumor originating from chest wall is comparatively rare. Sclerosing rhabdomyosarcoma (SRMS) is a rare variant of rhabdomyosarcoma that was first described by Mentzel and Katenkamp in 2000. This variant is characterized by spindle-shaped cells in prominent sclerosing hyaline stroma. Because of its densely hyalinized collagenous matrix and its occasional expression of a pseudovascular pattern of growth, this soft tissue tumor may be confused with sclerosing epithelioid fibrosarcoma, angiosarcoma, osteosarcoma and some other types of sarcoma. Although only 41 cases have been reported in the literature, the clinicopathologic characteristics and treatment procedure of this rare entity have not been clearly defined. We report a case of SRMS that originated from the right anterior chest wall and discuss its clinical, histologic, and immunohistochemical features.

Case Report

A 26-year-old woman visited our hospital with a right
Sclerosing Rhabdomyosarcoma of a Chest Wall in an Adult

breast mass and a dry cough. Her family and personal medical histories were unremarkable. A chest radiograph demonstrated a large mass occupying the majority of her right lung field and a pleural effusion. A chest computed tomography showed a 13.0 × 11.0 cm mass that originated from the right anterior chest wall and protruded into the intra- and extrapleural cavity. A compressive atelectasis of the right lung by the tumor and a pleural effusion were found (Fig. 1). A transcutaneous needle biopsy revealed atypical spindle cells with scant cytoplasm and hyperchromatic nuclei in an abundant hyalinized and fibrous stroma. The neoplastic cells stained positively for vimentin and bcl-2 but not for pancytokeratin, epithelial membrane antigen, CD 34 or S-100. No antibodies against muscle markers were used because the abundant hyalinized stroma was inconsistent with a typical rhabdomyosarcoma, and the rhabdomyoblasts generally observed in rhabdomyosarcoma were not identified. Although a definitive diagnosis could not be established, the tumor was considered to be a malignant soft-tissue neoplasm. We decided to perform surgery for resection to obtain a definitive diagnosis.

We performed the surgical resection through a right anterior thoracotomy in the fourth intercostal space in addition to a median sternotomy with an inverted L-shaped skin incision. The tumor originated from the second intercostal muscle and spread from the second rib to the fourth rib and to the right edge of the sternum. We performed a wide resection of the chest wall that included these parts. The chest wall defect was reconstructed with polypropylene mesh. Histologically, atypical spindle-shaped tumor cells with enlarged fusiform nuclei and the abundant collagenous stroma showing prominent hyaline sclerosis were observed. Strap rhabdomyoblast cells were not identified. The tumor cells were arranged in diverse growth patterns, including fascicular, small nests forming microalveolar architecture and pseudovascular structures (Fig. 2A and 2B). Mitotic activity was increased up to 35 mitotic figures per 50 high power fields. Cytoplasmic cross-striations were found in a portion of the tumor cells (Fig. 2C). This finding suggested the diagnosis of rhabdomyosarcoma. The tumor cells stained positively for desmin, myogenin, myoglobin and MyoD1 (Fig. 2D–2F). The tumor was diagnosed as rhabdomyosarcoma consistent with a sclerosing subtype. After surgery, we allowed time for the postoperative wound infection that developed in the subcutaneous tissue around the lower end of the median wound to heal and for her appetite to recover. During this time, no additional treatment was given, and pleural disseminations developed five months after surgery. Eighteen months after the complete resection, the patient still has metastases but is alive and undergoing chemotherapy with vincristine, actinomycin-D and cyclophosphamide (VAC therapy).

Discussion

Based on the morphologic features and molecular analysis, the current World Health Organization classification categorizes rhabdomyosarcoma into three main subtypes: embryonal, alveolar and pleomorphic. SRMS is a rare variant of rhabdomyosarcoma that was first reported in 2000 by Mentzel and Katenkemp.1) Although some cytogenetic studies have suggested that there is a link to the embryonal rhabdomyosarcoma subtype, it remains unclear whether SRMS is a variant of alveolar
or embryonal rhabdomyosarcoma or whether it represents a distinct subtype. To the best of our knowledge, only 41 cases have been reported in the literature. Like other rhabdomyosarcoma subtypes, most SRMSs arise from the extremities (53%) and head and neck regions (37%). SRMS in the chest wall has never been described in the literature. Unlike other rhabdomyosarcoma subtypes that occur mainly in children and adolescents, patients of all ages with SRMS have been reported. Histologically, SRMS is characterized by atypical spindle-shaped cells and the production of an abundant hyalinized matrix. The tumor cells occasionally exhibit peculiar microalveolar and pseudovascular patterns. Such findings are not part of the criteria for the standard rhabdomyosarcoma subtypes. SRMS is therefore easily mistaken for sclerosing epithelioid fibrosarcoma, angiosarcoma or osteosarcoma upon the initial evaluation. Immunohistochemical staining for the muscle markers: desmin, myogenin and MyoD1 ultimately leads to a diagnosis of SRMS. SRMS should be included in the differential diagnosis, and immunohistochemical staining for muscle markers should be performed when a soft-tissue mass is composed of spindle-shaped tumor cells with an abundant hyalinized stroma.

The number of patients with this type of rhabdomyosarcoma is small, and the follow-up results are short or inconclusive. Among the cases with available follow-up reported, 9 of 32 patients (28%) experienced distant metastases, 6 patients (18%) developed local recurrence, and 5 patients (15%) died of the disease. There is no consensus on the treatment of SRMS. Four patients were treated by surgery alone. Of those 4 cases, 3 cases developed metastatic disease. In our case, the adjuvant chemotherapy could not be performed due to the postoperative wound infection and loss of appetite. Therefore, the pleural disseminations developed after surgery. As in conventional rhabdomyosarcomas, a multimodal approach that includes surgery and combined chemotherapy and irradiation may be necessary for most cases of SRMS. Patients with chest wall tumors are frequently asymptomatic, and tumors are consequently found after growing large as in our case. If further studies can demonstrate the effectiveness of chemotherapy and irradiation, preoperative therapy would be more helpful for such large tumors, thus ensuring negative surgical margins and reducing the size of the chest wall defects. As more cases are recognized and reported, we hope to be able to better define the clinicopathologic characteristics of this rare entity.

Fig. 2 Histology of the tumor. (A) The tumor was composed of atypical spindle cells that were arranged in fascicular, microalveolar and pseudovascular structures with an abundant hyalinized and collagenous stroma (hematoxylin and eosin, ×10). (B) At a high power view, the tumor demonstrated a uniform population of spindle cells with scant cytoplasm and hyperchromatic nuclei (hematoxylin and eosin, ×20). (C) The cytoplasmic cross-striation was found in a portion of the tumor cells. (D, E, F) Immunohistochemical staining: tumor cells were positive for desmin (D), myogenin (E) and MyoD1 (F).
Disclosure Statement

The authors have no conflicts of interest to declare. No external funding was obtained for the work presented here.

References