Massive Chest Wall Tumor Diagnosed as Askin Tumor*  
Successful Treatment by Intensive Combined Modality Therapy in an Adult  
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A 24-year-old woman was admitted due to dyspnea on exertion. A chest roentgenogram revealed a massive tumor originating from the right anterior chest wall when pleural effusion was drained. Diagnosis of Askin tumor was made based on light microscopic findings characterized by composing with small round cells, immunocytochemical findings suggestive of neuroectodermal origin, and cytogenetic analysis demonstrating the chromosomal translocation (11; 22). After intensive combined modality therapy, including chemotherapy, irradiation, and additional surgery, she was followed up as an outpatient and has remained disease-free for 16 months after initial diagnosis. 

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Tumors arising from the chest wall are rare. Schaefer and Burton reviewed the radiographic characterization of the chest wall tumors. They classified the tumors into four groups as follows: (a) contiguous spread of adjacent neoplasms, (b) metastatic neoplasms from the other organs, (c) primary malignant tumors, such as chondrosarcoma, plasmacytoma, and fibrosarcoma, and (d) benign tumors mainly originating from cartilage. In childhood, Ewing's sarcoma is the most common chest wall tumor, whereas rhabdomyosarcoma, neuroblastoma, and Askin tumor are less common. Askin tumor was first described as a malignant tumor of the thoracopulmonary region in childhood and adolescence as a unique clinicopathologic entity. We report an adult case with a giant tumor in the chest wall, which was diagnosed as Askin tumor by extensive studies including immunocytochemical and cytogenetic examinations. Successful treatment, including an intensive combined therapy based on another report and additional surgery, is also presented.

CASE REPORT

A 24-year-old woman was admitted to our hospital on Jan 28, 1991, because of dyspnea on exertion, dry cough, and right anterior chest pain, which lasted for 1 month prior to admission and progressively worsened. She had no remarkable past history and no smoking history. Physical examination on admission showed dullness on percussion and a decrease of breath sounds in the right lung base. No other abnormalities were detected. A chest roentgenogram at the time of admission showed extensive radiopacity in the right hemithorax. Ultrasonographic examination revealed that the radiopacity was composed of a huge tumor originating from the anterior chest wall and massive pleural effusion. After evacuation of the pleural effusion, a huge tumor was observed on the chest roentgenogram (Fig 1). A computed tomography scan of the chest revealed that the mass was in contact with the right atrium and that the fifth rib of the anterior chest wall had destructive change due to the tumor. In a technetium 99m-Tc bone scintigram, the right fifth rib had high uptake of the isotope. Other studies revealed no evidence of distant metastasis. Blood tests were normal except for CA125 (460 U/ml; normal, less than 50 U/ml). Her pleural effusion showed bloody appearance and had exudative character, but the concentrations for adenosine deaminase and tumor markers were normal except for CA125 (410 U/ml).

Cytologic examination of pleural effusion revealed neoplastic cells featuring round nuclei, fine chromatin, scant cytoplasm, and a cluster formation. For further distinct diagnosis, echo-guided aspiration biopsy was performed. Histologic findings from the biopsy specimen revealed that the tumor was comprised of small round cells arranged in a diffuse pattern but not in a Homer-Wright rosette pattern, which is characteristic of neuroblastoma. Immunocytochemical studies showed that the neoplastic cells stained positively for neuron-specific enolase in the cytoplasm. In contrast, these cells were not only negative for a leukocyte common antigen, but also negative for SC11 monoclonal antibody which reacts specifically with the surface antigen of Ewing's sarcoma. They showed negative stain for periodic acid Schiff. Neurofilaments, neurosecretory granules, and rhabdomyofibrils were not observed on electron microscopy. Chromosomal analysis of the neoplastic cells in pleural effusion revealed the chromosomal translocation (11; 22 [Fig 2]). Diagnosis of Askin tumor was made based on these analyses.

Intensive combined chemotherapy (vincristine, 2 mg/m2; doxorubicin, 50 mg/m2; cyclophosphamide, 600 mg/m2; and ifosfamide, 3 g/m2) was given for 6 months, followed by reevaluation. Repeat ultrasonographic examination revealed that the effusion and tumor mass had disappeared. No recurrence had occurred over the 2 years after initial treatment.
Ewing's sarcoma and Askin tumor, we could diagnose this tumor as Askin tumor based on the fact that the neoplastic cells of the tumor were positive for neuron-specific enolase but negative for a periodic acid Schiff. This notion also was confirmed because these cells were negative for 5C11 monoclonal antibody. This is a rare adult case of Askin tumor which was diagnosed by the extensive studies indicated, suggesting that Askin tumor should be considered in the differential diagnosis of chest wall tumors, and an immunocytochemical study should be done. This is especially important when the histologic appearance of the tumor demonstrates small round cells, even if the patient is an adult.

Askin tumor was first reported to carry a poor prognosis (mean survival time: 8 months) even if many kinds of treatment were administered. Recently, Young et al reported an intensive combined modality therapy, including intensive chemotherapy, irradiation, and autologous bone marrow transplant, for sarcomas arising in the chest wall, such as Askin tumor and Ewing's sarcoma. Their results were preliminary but high CR rate and good local control with acceptable morbidity were obtained. We performed only the induction treatment, including chemotherapy and irradiation according to their protocol, but not the subsequent intensification therapy including chemotherapy, total body irradiation, and autologous bone marrow transplant because of the severe hematologic and gastrointestinal toxicities. Although Young et al noted that the extent of the surgery was not found to be of prognostic significance, we performed an additional surgery because good response to the induction treatment and because no distant metastases were observed. The present patient remains disease-free for 16 months after initial diagnosis, suggesting additional surgery in combination with intensive chemotherapy and irradiation may be considered in the future for patients with this tumor.

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