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# Extraskeletal Ewing's Sarcoma Presenting With Multifocal Intrathoracic Mass Lesions Associated With Mediastinal Shift

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**Extraskeletal Ewing's sarcoma is an uncommon disease that predominantly involves the soft tissues of the trunk or the extremities. This article presents a patient with multifocal intrathoracic mass lesions involving the mediastinum and the lingula associated with mediastinal shift, eventually diagnosed as extraskeletal Ewing's sarcoma.**

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Extraskeletal Ewing's sarcoma is an uncommon disease that predominantly involves the soft tissues of the trunk or the extremities [1]. We report a patient presenting with multifocal intrathoracic mass lesions involving the mediastinum and the lingula associated with mediastinal shift, eventually diagnosed as extraskeletal Ewing's sarcoma.

A 26-year-old woman presented with shortness of breath that had progressed over the last month. On physical examination, her breath sounds were decreased on the right hemithorax as well as the lower left hemithorax. Her heart rate was 92 beats per minute and her blood pressure was 100/60 mm Hg. Laboratory investigations revealed a hemoglobin level of 9.7 g/dL, a white blood cell count of 11,000/mL, an erythrocyte sedimentation rate of 75 mm/h, and an LDH level of 2,450 U/L. Analysis of arterial blood gases revealed a pH level of 7.47, a $PCO_2$ level of 32 mm Hg, a $PO_2$ level of 64 mm Hg, and an oxygen saturation level of 93%. Chest roentgenogram demonstrated a left-sided shift of the mediastinum due to a massive lesion on the right hemithorax. Observation of the thorax by computed tomography showed a 15 × 20 cm mass lesion involving the mediastinum, occupying almost the entire right hemithorax, and displacing the diaphragm inferiorly. In addition, right-sided pleural effusion and two separate mass lesions of the left hemithorax were noted (ie, a 4 × 5 cm mass lesion involving the mediastinum and a 2 × 3 cm mass lesion involving the lingula) (Fig 1). Magnetic resonance imaging of the thorax revealed similar signal characteristics for all three mass lesions that were consistent with the presumptive diagnosis of a sarcoma (Fig 2). Fine needle aspiration cytology obtained from the right-sided mass lesion was consistent with the diagnosis of a sarcoma as well. Bone scintigraphy, computed tomography of the abdomen and bone marrow biopsy were all unremarkable. The patient underwent right thoracotomy with resection of the right-sided mass lesion. The mass lesion had originated from the mediastinum and had compressed, but had not invaded, the pulmonary parenchymal tissue. On histopathologic examination, small and round blue cells with round nuclei, small nucleoli, and scanty cytoplasms were observed that appeared to be well organized. Immunohistochemical analysis revealed positive staining for CD99, S-100 and vimentin and negative staining for cytokeratins, neurofilament, synaptophysin, and chromogranins. Based on these findings, the mass lesion was diagnosed as extraskeletal Ewing's sarcoma. Starting 1 month after resection, the patient received adjuvant chemotherapy that was composed of two cycles of ifosfamide, doxorubicin, and etoposide. On repeat computed tomography of the thorax, both left-sided mass lesions were reported to have remained stable. The decision was made to proceed with left thoracotomy with resection of both left-sided mass lesions. Based on similar findings on histopathologic examination and immunohistochemical analysis for both mass lesions, the diagnosis of extraskeletal Ewing's sarcoma was confirmed. Adjuvant radiation therapy was commenced at a dose of 50.4 Gy in 28 fractions of 1.8 Gy that was delivered on a linear accelerator using 6 Mv photons, and concurrent chemotherapy was started, which was composed of two cycles of ifosfamide and etoposide. Starting at 1 month after radiation therapy the patient was scheduled to receive further adjuvant chemotherapy that would be composed of

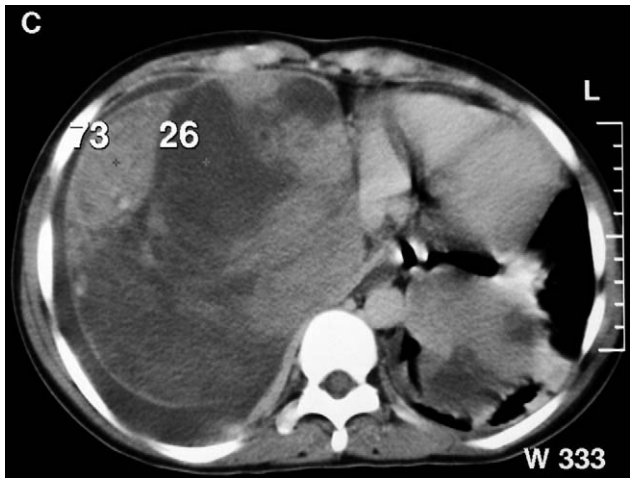


*Fig 1. Computed tomography reveals a massive lesion on the right hemithorax involving the mediastinum. A separate mass lesion involving the mediastinum is observed on the left hemithorax as well.*

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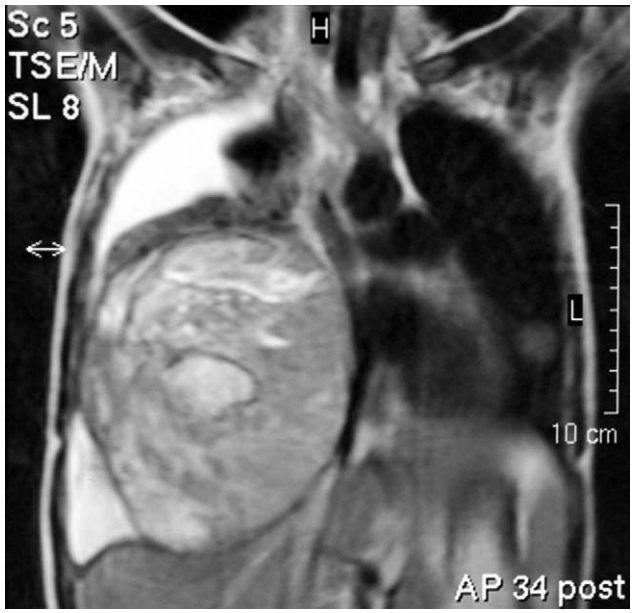


*Fig 2. On magnetic resonance imaging, the extraparenchymal mass lesion on the right hemithorax displaces the diaphragm inferiorly and the lung superiorly, and it is associated with a left-sided shift of the mediastinum. In addition, a separate mass lesion involving the lingula is observed on the left hemithorax.*

six cycles of ifosfamide, doxorubicin, and etoposide. The patient is alive with no evidence of disease at 10 months after the diagnosis.

## Comment

Ewing's sarcoma is an uncommon disease that usually arises from the skeletal bones that frequently extends to the soft tissues and most commonly affects children and young adults. Ewing's sarcoma that arises from the soft tissues and spares the skeletal bones has been identified as a much less common and distinct entity, namely an extraskeletal Ewing's sarcoma [1]. The classical histopathologic description of an extraskeletal Ewing's sarcoma is small with round blue cells that are uniform in appereance and organized in solid sheets that are divided by fibrous strands. An extraskeletal Ewing's sarcoma shares histopathologic and immunohistochemical findings with Ewing's sarcoma and thus may be confused with embryonal rhabdomyosarcoma, lymphoma, and neuroblastoma [2]. Therefore, confirmation of the diagnosis of an extraskeletal Ewing's sarcoma should rely on positive staining for CD99 on immunohistochemical analysis [3].

Extraskeletal Ewing's sarcoma rarely involves the mediastinum. In a review of 24 patients, Ahmad and colleagues [4] have reported only 1 patient presenting with extraskeletal Ewing's sarcoma involving the mediastinum. For the patient presented in this report, the mass lesion in the right hemithorax dominated the symptomatology at presentation. The observed growth pattern, namely compression without invasion of the pulmonary parenchymal tissue, limited the symptomatology until the right-sided mass lesion attained a massive size. Upon resection of the extraparenchymal right-sided mass lesion, the patient experienced immediate relief of her shortness of breath.

Although an extraskeletal Ewing's sarcoma often presents as a local disease, it should be regarded as a systemic, yet curable disease. Hence, an aggressive multimodal management strategy should be undertaken [5]. Surgery should initially be performed in an attempt to attain wide resection margins, and radiation therapy should be used as an adjunct to resection in an attempt to improve control of the obvious local disease, whereas chemotherapy should also be incorporated in an attempt to address the obscure systemic disease.

For children and young adults presenting with multifocal intrathoracic mass lesions, extraskeletal Ewing's sarcoma might be considered in the differential diagnosis. Even patients presenting with an extensive involvement of extraskeletal Ewing's sarcoma may benefit from an aggressive multimodal management strategy involving surgery, radiation therapy, and chemotherapy.

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# Very Unusual Case of Post-Traumatic Chylothorax

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**Chylothorax is a rare disease caused by both traumatic and nontraumatic events. Chylothorax can cause cardiopulmonary abnormalities and significant nutritional, metabolic, and immunologic consequences. We present an exceptional case of chylothorax due to penetrating chest trauma. The diagnosis was made by thoracentesis.**

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