CASE REPORT

A case of extraskeletal Ewing sarcoma originating from the visceral pleura.

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Abstract

Extra skeletal Ewing Sarcoma (EES) is a rare entity which predominantly occurs in adolescents and young adults. It usually arises from the soft tissues of the trunk or the extremities. We present a case of EES arising from the left visceral pleura in a 21 year old female patient who presented to the emergency room of our institution with fever, productive cough and sternal pain radiating to the back for the last 3 days. Chest radiograph was firstly performed, followed by chest CT examination. Finally open lung biopsy revealed a small round cell malignancy. The mass was resected and the histological examination revealed Extra skeletal Ewing Sarcoma (EES) of the visceral pleura without involvement of the adjacent lung. Secondary multiple nodules at the lateral wall of the pleura were also noticed and so postoperative multiagent chemotherapy was performed. EES should be considered in the differential diagnosis of any patient, especially adolescents or young adults, with a soft tissue mass of the trunk or the extremities. Hippokratia 2011; 15 (4): 363-365

Key words: extraskeletal Ewing Sarcoma; visceral pleura; soft tissue tumor

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Ewing Sarcoma, first described by Ewing in 1921, is regarded as an uncommon malignancy but it is the second most common primary bone malignancy in childhood. Extra skeletal Ewing (EES) Sarcoma is a very rare disease. It was recognised as a distinct entity in 1969 by Tefft at al. who treated 5 patients with paravertebral soft tissue masses similar to Ewing Sarcoma but without affecting the adjacent bony structures. EES usually arises from the soft tissues of the trunk and the extremities but it has also been reported at other more unusual locations[1-8]. We present a case of EES of the visceral pleura in a 21 year old female patient.

Case Report

A 21 year old female patient with no history of prior medical problems, presented to our hospital with fever up to 38˚. Her blood pressure was 120/70mmHg, her pulses 120 and SpO2 was 97%. The left hemithorax was silent during the auscultation. The chest radiograph demonstrated opacification of the left hemithorax with mediastinal shift to the right (figure 1). WBCs were 12.600, CRP 25.3, ESR 40 and the tumor markers CA 125: 247.5 and CA 15-3: 11.1. Thoracic CT examination performed after contrast administration revealed a large pleural effusion and a heterogeneous, enhancing mass located at the left pleural cavity, displacing, compressing and causing atelectasis to the adjacent lung (figure 2a, b). The mass was extending to the left paravertebral space and was infiltrating the paravertebral fat tissue. There where no signs of bone destruction or calcification. Multiple enhancing nodules were noticed at the lateral wall of the pleura (fig. 3a, b). The patient underwent thoracoscopy and biopsy. The histological examination revealed Extra skeletal Ewing Sarcoma of the visceral pleura without involvement of the adjacent lung.

Discussion

Ewing Sarcoma was first described by J. Ewing in 1921 and since then it is regarded as the second most common malignant bone tumor in childhood. It usually arises from the bone and invades the adjacent soft tissues in 90% of cases. Extra skeletal Ewing Sarcoma (EES) is recognized as a distinct entity since 1969, when Tefft et

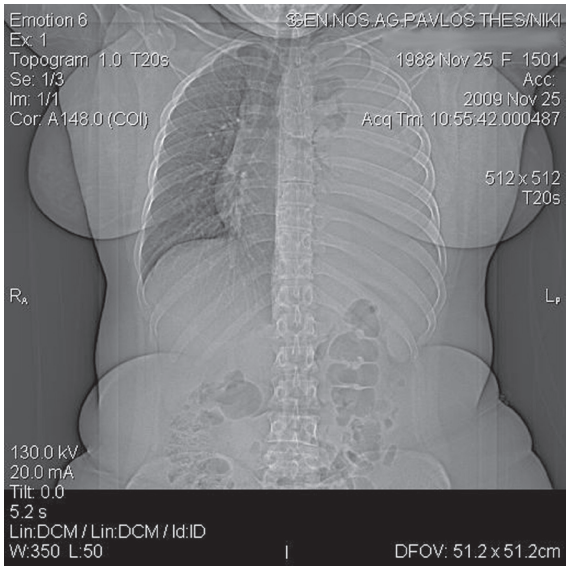
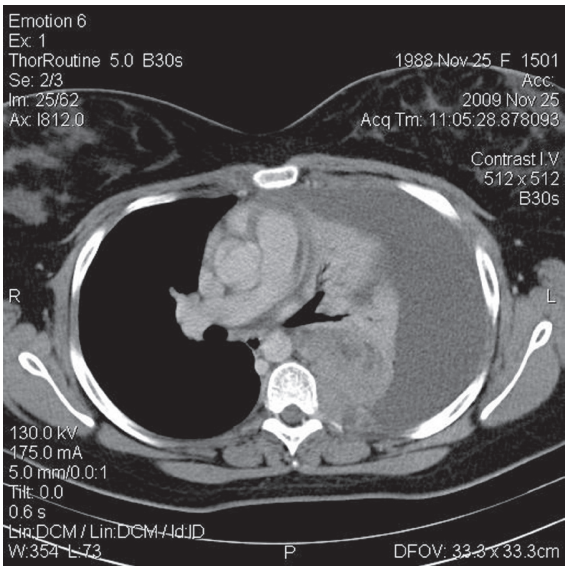
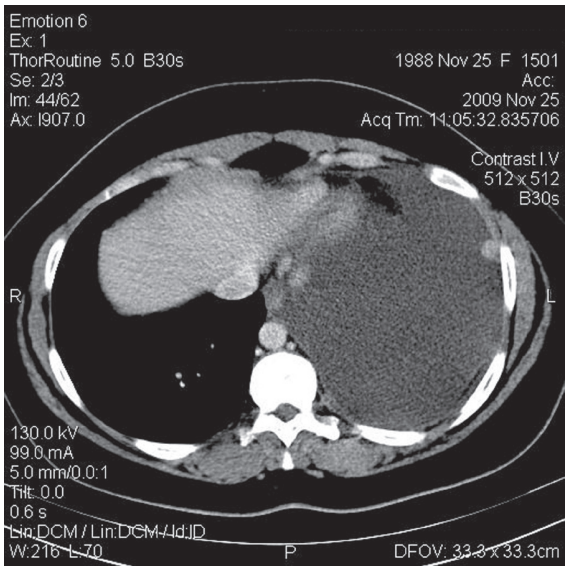


Figure 1: Chest Scanogram demonstrating the opacification of the left hemithorax and the mediastinal shift to the right.





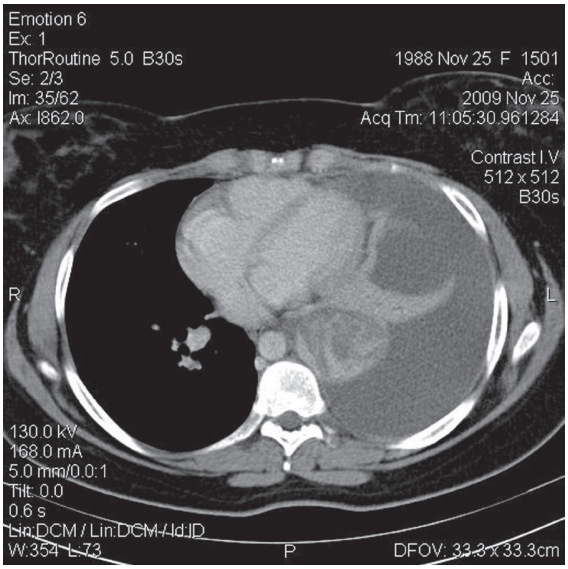


Figure 2:CT examination after contrast administration revealing a large pleural effusion and a heterogeneous, enhancing mass located at the left pleural cavity (a, b).

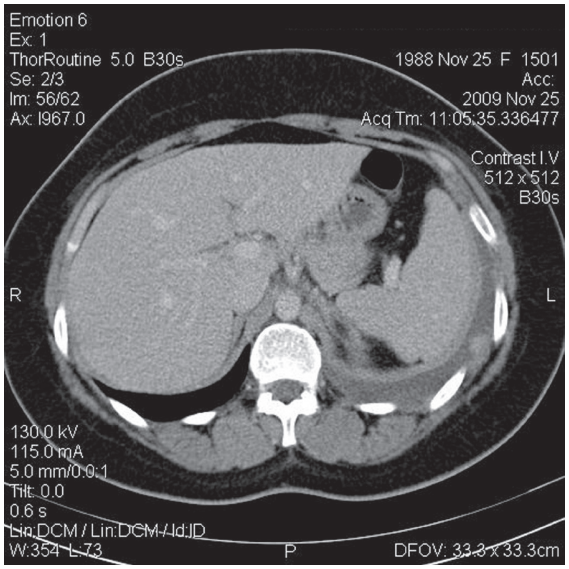


Figure 3:CT examination after contrast administration demonstrating multiple enhancing nodules at the lateral wall of the pleura (a, b).

al. described a series of 5 patients who presented with a soft tissue tumor in the para vertebral region, related to an unusual Ewing Sarcoma that didn't seem to affect the adjacent bony structures[3].

Extra skeletal Ewing Sarcoma (EES) arises in the soft tissues of the trunk or the extremities and although is a primary soft tissue tumor it can invade the adjacent bones. The most frequent sites of occurrence are the chest wall, the para vertebral region and the lower extremities. EES has also been reported to occur in the pelvis, the hip region, the upper extremities, the mediastinum, the retroperitoneal cavity, the diaphragm, the duodenum, the perigastric space, as well as in the parapharyngeal space, the parotid gland and the hard palate[1-8].

EES is usually found in adolescents and young adults (younger than 30 years). A handful of cases affecting patients older than 50 years have been reported in the literature[4]. There is no significant difference in the prevalence of EES between the genes. EES is associated with reciprocal translocation between chromosomes 11 and 22[9, 10].

EES morphologically is indistinguable from osseous Ewing Sarcoma. EES is histologically characterized by the presence of small blue round or oval cells containing solid material separated by fibrous septations. It must be distinguished from the other tumors of the Ewing family: osseous Ewing Sarcoma and Primitive Neuroectodermal Tumor (PNET). It should also be differentiated from embryonal rhabdomyosarcoma, neuroblastoma and lymphoma. The differential diagnosis should be based on clinical features, immunohistochemical staining and cytogenetic

analysis. It has been reported that in patients with Ewing Sarcoma immunohistochemical staining is positive for CC99, CC56 but is negative for S-100, demin, factor 8 cytoceratin and neurofilament as well as for T- cells, B- cells, LCA, CD43 and CD68. Ewing Sarcoma can be definitely diagnosed by detection of the EWS gene and rearrangement of ETS related cancer genes[2].

At CT examination EES usually appear as homogeneous masses when they are small and heterogeneous masses when they are large. On T-1 weighted MR images tumors have signal intensity equal to or greater than that of a muscle. Large tumors appear heterogeneous because of the presence of foci of cystic degeneration, necrosis and hemorrhage. On T-2 weighted images the masses tend to have inhomogeneous high signal intensity. Tumors show marked enhancement after contrast medium administration[10].

Treatment should include aggressive surgical resection and multiagent chemotherapy. Adjuvant radiation therapy should also be considered. A patient age < 16 is associated with a favorable prognosis[3]. Other major prognostic factors include the size of the tumor, complete resection of the tumor with wide surgical margins, the location of the mass (favorable prognosis in extremity lesions), the presence of metastases, the extent of necrosis, the initial response to chemotherapy and the presence of EWS/FL 11 transcripts[2, 3].

As a conclusion EES should be considered in the differential diagnosis of any patient, of any age especially adolescents or young adults, with a soft tissue mass of the trunk or the extremities. All patients even those who present metastases at the time of diagnosis should be treated and may benefit from an aggressive multimodal management strategy[1, 3].

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