Extra-Osseous Osteosarcoma: An Unusual Cause of Malignant Pleural Effusion and Spontaneous Pneumothorax

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Introduction:
Extra-osseous osteosarcomas (EOOs) are malignant tumors arising in the soft tissue without involving the bone or periosteum. These unusual neoplasms (comprising 1% of all soft tissue sarcomas and 2-4% of all osteosarcomas) have a very poor prognosis. This case highlights two complications of thoracic EOOs: malignant pleural effusion and spontaneous pneumothorax.

Case Presentation:
A 94-year-old Caucasian female was referred by her primary care doctor for evaluation of cough progressing over the past three months now complicated by two weeks of exertional dyspnea. Her past medical history included systemic hypertension, hyperlipidemia and hypothyroidism. Her lab work and EKG were within normal limits. A chest radiograph revealed a large, free-flowing, right-sided pleural effusion: subsequent thoracentesis yielded blood tinged fluid which was exudative by Light’s criteria. A repeat chest radiograph post-thoracentesis showed a lobulated mass obscuring the right hemidiaphragm. Follow-up computerized tomography (CT) images revealed a large (7.9x7.1x4.1 cm), partially-calcified pleural based mass.

A CT-guided biopsy of the mass revealed spindle-shaped, fibroblastic cells with islands of osteoid matrix formation consistent with extra-skeletal osteosarcoma. The patient was offered an extensive surgical resection including diaphragmatic take-down and reconstruction. Given the high perioperative mortality and her advanced age, she declined the operation.

Two weeks later, the patient presented with acutely worsened dyspnea. Chest radiographs showed recurrence of the large right-sided pleural effusion and a new pneumothorax. A thoracostomy tube was placed resulting in the resolution of her hydro-pneumothorax. Pleural fluid cytology specimens were positive for malignant cells. The patient opted for palliative care measures and succumbed to her illness four months after hospital discharge.

Discussion:
EOOs are neoplasms characterized by the production of osteoid or bone, sometimes accompanied by cartilage. EOOs have an aggressive clinical course: 70% recur locally and 80% develop metastatic disease within 2 years of resecting the primary tumor. The most frequently affected sites are the soft tissues of the limbs (67%), especially the thighs and retroperitoneum (17%). While skeletal osteosarcomas predominantly affect adolescents and young adults, EOOs usually develop after the age of 30. Cases have
been reported to have arisen in areas that have received radiation therapy or in areas with previous myositis ossificans. Osteosarcomas are associated with spontaneous pneumothoraces - even in the absence of apparent metastasis to the lungs. However, patients with lung metastases have a high incidence of spontaneous pneumothorax, especially following chemotherapy.

These neoplasms vary in their degree of histologic differentiation with widely discrepant amounts of spindle cell sarcoma and foci of malignant osteoid. The limited experience with EOOs to date suggests that the best prospect for cure is early, complete surgical excision combined with chemotherapy and/or radiotherapy. Long-term follow-up is required before the efficacy of this approach can be assessed. Median survival time is estimated at 31 months and reported 5-year survival rates are 13-22%. The presence of spontaneous pneumothorax has been shown to portend a worse prognosis.

**Conclusion:**
EOOs, while uncommon, should be considered in the differential diagnosis of pleural effusion and spontaneous pneumothorax. Early recognition of EOOs is essential as successful treatment of these tumors requires early, aggressive and multidisciplinary measures.

**References:**