TITLE Solitary fibrous tumour of pleura with malignant form and prolonged aPTT presenting as massive pleural effusion
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Abstract:
Abstract Solitary fibrous tumour of pleura Other names-Located mesothelioma, Benign fibrous mesothelioma, Benign localised fibroma. This tumour uncommon, involves age group around 5th decade usually, biologically, histologically benign appear as firm encapsulated tumour arising from visceral pleura in 70 cases and from parietal pleura in 30 cases composed of elongated spindle cells, collagen and reticulin fibres. This tumour lacks cytoplasmic, keratin expression but expresses CD34 and vimentin. Manifest two forms morphologically-1. pedunculated type—good prognosis, 2. sessile type—most consistently malignant—worst prognosis. Mostly asymptomatic but may present with chest pain, dyspnea, and cough. Transthoracic needle biopsy is the diagnosis of choice. Radiologically Solitary, sharply defined discrete mass located at the periphery of lungfield associated with pleural effusion in 20 cases. Surgery is the major mode of treatment.

Keyword: Key words Solitary fibrous tumour of pleura, Benign, Malignant Case

Chest X ray showed homogenous opacity of left hemithorax suggestive of left pleural effusion. Diagnostic pleura fluid tapping done—hemorrhagic fluid was aspirated and analysis of pleural fluid showed features of exudative pleural effusion. Cytology reported as reactive pleural effusion. Therapeutic drainage with intercostal tube done. CT Chest done and reported as large circumscribed sessile, heterogeneous mass lesion 11x10cm with intraleral vessels with geographical patterns of low attenuation within the lesion; and with bronchial cut off sign and left lower lobe collapse with minimal effusion. Hematologist opinion obtained due to persistent thrombocytopenia [platelet count more than 5 lakhs cells/cumm]. ANA, Lupus anticoagulant were negative. Blood sugar was 85mg. Prothrombin time, bleeding time, Clotting time and coagulation profile were normal. But aPTTT[activated partial thromboplastin time] was 45 as against 33 [control]- opined as inhibitor substance for plateletadhesion and started with prednisolone 1mg/kg for 8 weeks. CT guided biopsy of lung mass done; HPE—reported as fragments of neoplasm composed of spindle cells & few polyhedral cells, arranged in sheets & clusters interspersed with few congested vessels & fibrous necrotic exudate. Probably- 1. mesothelioma, 2. solitary fibrous tumour of pleura. On further Immunohistochemistry examination was CD 34 and vimentin positive and calretinin negative. Patient was referred to cardiothoracic surgeon for surgical management. Meanwhile patient developed liver metastasis and intervertebral disc extension of tumor with compression of spinal cord. Surgery deferred due to prolonged aPTT and liver metastasis. Patient expired during treatment in hospital.

Discussion: Malignant mesothelioma and solitary tumours of pleura are the main tumours of pleura producing hemorrhagic effusion. Common cause of massive pleural effusion with mediastinal shift is metastatic diseases of pleura. And pulmonary tuberculosis, empyema, cirrhosis with hepatothorax, chylothorax, hemotherax, congestive heart failure may also cause massive effusion. Malignant mesotheliomas are thought to arise from the mesothelial cells that line the pleural cavities. Individuals with a history of exposure to asbestos have a much greater risk of developing these neoplasms. Malignant mesothelioma with its dismal prognosis should be differentiated from the solitary fibrous tumor of the pleura, which has an excellent prognosis.

Fig-1 Showing massive left pleural effusion.
Solitary fibrous tumour of pleura: The term solitary fibrous tumour is preferred for several reasons: (1) although the neoplasm is usually histologically and biologically benign, malignant forms clearly exist, and, in some cases, the histologic distinction between the two is difficult, if not impossible; (2) the neoplasm often shows evidence of fibroblastic differentiation; and (3) the results of ultrastructural, immunohistochemical, and experimental studies suggest that the tumor originates in the submesothelium itself. In comparison with malignant mesothelioma, the prognosis of solitary fibrous tumors of the pleura is excellent in benign forms and poor in malignant forms. The presence or absence of an effusion apparently has no effect on the patient's prognosis.

Pathology: Grossly fibrous tumors appear as firm, encapsulated yellow tumors, which may be vascular with prominent veins over their external surfaces. About two thirds of these fibrous tumors arise from the visceral pleura, whereas one third arise from the parietal pleura. At times, these tumors invade the lung and chest wall locally. Solitary fibrous tumors are characterized histologically by uniform, elongated spindle cells and varied amounts of collagen and reticulin fibers in bundles of varying sizes. The cell of origin of this tumor is...
mesenchymal and appears to be the multipotential subpleural cell. These tumors lack expression of cytoplasmic keratins, a marker of mesothelial cells, but do expresses vimentin, another marker of mesenchymal cells. These tumors also express CD 34 which is a transmembrane cell surface glycoprotein ubiquitously observed in a novel family of interstitial spindle cells. The incidence of malignant form of solitary fibrous tumors of the pleura is 7% to 60%. De Parrot et al. have developed a classification that is based on whether the tumor is benign or malignant histologically and whether it is sessile or pedunculated. This classification is useful because it is equated with prognosis. The solitary fibrous tumors with the worst prognosis are those that are malignant and sessile, in which approximately two thirds of patients have a recurrence. Malignant pedunculated tumors recur approximately 15% of cases. Recurrences with the benign sessile and the benign pedunculated tumors occur in less than 10% of cases.

FIG-2. Showing heterogenous mass with areas of hypoattenuation

Clinical features: This tumour is evenly distributed in both sexes and median age of presentation is 57 years; 50% of patients are asymptomatic. Cough, chest pain, dyspnea are the most frequent symptoms. The two important paraneoplastic syndrome associated with the tumor are 1. hypertrophic pulmonary osteoarthropathy 2. hypoglycemia[Deoeb potter syndrome]. The mechanism responsible for the hypoglycemia appears to be the production of high levels of insulinlike growth factor II (IGF-II) by the tumor.

Radiology: These tumors are solitary, sharply defined, discrete masses located at the periphery of the lungfield or related to a fissure. At times, the mass may become very large, occupying most of the hemithorax with frequently lobulated and associated pleural effusion in 10% to 20% of cases. CT chest: The tumors are large, noninvasive, and tend to enhance with intravenous contrast and is frequently nonhomogeneous. The intense enhancement of these tumors appears to be due to their high vascularity, whereas areas of low attenuation are due to foci of myxoid or cystic degeneration and hemorrhage within the lesion,usually not associated with mediastinal lymphadenopathy.

Diagnosis: By 1.transthoracic cutting needle biopy or excision biopsy by thoracotomy or VATS. 2. Magnetic resonance imaging (MRI) is occasionally useful in evaluating potential invasion of the chest wall by a sessile tumor

Treatment: surgical excision by thoracotomy or VATS is treatment of choice. Recurrence may occur even after 10 years of resection. Due to the rarity of these tumors, there is no systematic assessment of the role of adjuvant therapy. Anecdotal reports describe long-term survivals with postoperative radiotherapy in patients with incomplete resection of the tumor and responses to ifosfamide and doxorubicin have been reported for recurrent, inoperable tumors.

FIG-3 Showing HPE- spindle cells in area of necrosis

Conclusion: Solitary tumour of pleura a rare tumor, occurs in age group 40-50 years ,with malignant transformation has poor prognosis.

References: