A RARE PRESENTATION OF MALIGNANT MESOTHELIOMA AS ENCYSTED HEMOTHORAX, WITHOUT ASBESTOS EXPOSURE

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ABSTRACT: Malignant mesothelioma is an asbestos-induced, highly aggressive tumor. Malignant mesothelioma is a tumor of the serosal cavities most commonly involving the pleura and tends to present at relatively advanced stages. We describe a case of (epithelial) type of malignant mesothelioma of pleura in an elderly lady from a rural background with no history of exposure to asbestos, presented with left encysted hemothorax initially suspected to be due to trauma which later turned out to be due to malignant mesothelioma.

KEYWORDS: malignant mesothelioma, encysted hemothorax.

INTRODUCTION: Malignant mesothelioma is an asbestos-induced, highly aggressive tumor.

Amphibole fibers, as compared with chrysotile, are generally more toxic in part because amphibole fibers accumulate more readily in the distal lung parenchyma, are not cleared as effectively, and are more durable.¹

Malignant mesothelioma is a tumor of the serosal cavities and tends to present at relatively advanced stages. It most commonly affects the pleura, with patients usually presenting with symptoms of chest pain or discomfort, dyspnea and cough. The 3 major histological types of mesothelioma are sarcomatous, epithelial, and mixed.²

Most malignant mesotheliomas have complex karyotypes, with extensive aneuploidy and rearrangement of many chromosomes. A loss of a single copy on chromosome 22 is the most common abnormality. Other chromosomal changes include 1p, 3p, 9p, and 6q. Several changes in the tumor suppressor gene p16 (CDKN2A) and p14 (ARF) and loss of function of neurofibromin 2 (NF2) or merlin and inactivation of INK4 α /ARF are key events in tumorigenesis. Numerous studies over the past several years have examined whether biomarkers in asbestos-exposed workers are useful indicators of increased risk (eg, mesothelin, megakaryocyte potentiating factor, osteopontin, soluble mesothelin-related protein, and others).

We describe a case of (epithelial) type of malignant mesothelioma of pleura in an elderly lady from a rural background with no history of exposure to asbestos, presented with left hemothorax initially suspected to be due to trauma was later found to be due to malignant mesothelioma.

CASE DESCRIPTION: A 62yr old house wife, hard manual agriculture laborer was admitted with complaints of breathlessness and left sided chest pain of 2 months duration. There was history of trauma to the chest twice, 4 months ago when she had fallen over a stone and sustained blunt injury over left side of her chest, with dull ache which she ignored and again 2 months prior to admission she fell down and sustained injury to front of the chest. The old chest pain worsened and new symptom of breathlessness appeared, for which she came to hospital.

No fever, chronic cough, weight loss, night sweats, reduced appetite, angina, syncope, palpitation, swelling of legs, hoarseness of voice, wheeze, back pain or past history of tuberculosis. She was not a diabetic or hypertensive. She was not on any regular medications.

On examination, moderately built and nourished lady with no pallor, Icterus, cyanosis, clubbing, pedal edema or lymph node enlargement or raised JVP, with pulse $90/\min$, regular and blood pressure of 120/80 mm Hg.

Respiratory system examination showed drooping of left shoulder, scoliosis of spine to left, tracheal shift towards left, decreased movements and crowding of ribs on left side, stony dull to percuss with diminished breath sounds on left side. Other systemic examination was within normal limits.

A provisional diagnosis of Left sided pleural effusion with collapse of the left lung due to blunt trauma was made.

Laboratory investigations showed hemoglobin-9.2 gm.%, WBC- 15, 700 cells/mm3, N70, L30, E0, Normochromic Hypochromic anemia, mild neutrophilia., ESR- 54mm in 1hr., RBS- 98mg/dl, urine routine was normal, blood urea- 22mg/dl and serum creatinine- 0.1mg/dl.

ECG was within normal limits. Chest x-ray revealed left sided haziness in all zones, ultrasound chest revealed massive fluid collection in left pleural space but pleural tap done through ultrasound guidance was dry tap.



Figure 1

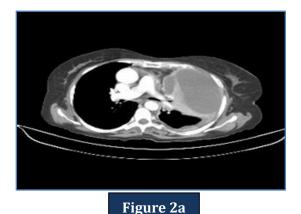
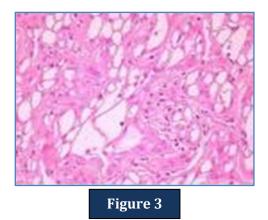


Figure 2b

Contrast CT-thorax showed 2 thick walled loculated pleural effusions in left hemithorax with collapse compression of left lung with mild mediastinal shift to right side and no obvious mass. Intercostal drainage tube was inserted which drained around 2 liters of frank blood following which patient felt better within next 48 hours and breath sounds were heard in left lung areas with reduced intensity.

Pleural fluid analysis showed hemorrhagic fluid with protein of 1.5gm%, sugar-45mg%. Smear showed dispersed mesothelial cells in hemorrhagic back ground, neutrophils and no malignant cells were found.

Since intercostal drainage tube continued to drain about 250 ml of hemorrhagic fluid every day for more than a week, thoracotomy was done, which showed thickened parietal pleura adherent to visceral pleura with no space between parietal pleura and lung. Plane could not be created between parietal pleura and lung surface, and intercostal drainage tube was continued.



Histopathological report of parietal pleura confirmed malignant mesothelioma of microglandular pattern.

Other management included antibiotics, analgesics and 2 units of blood was transfused.

Post-operatively50 ml to 100 ml of hemorrhagic fluid continued to drain every day. Decortication, chemotherapy with radiotherapy was planned but patient did not agree for any interventions and was lost for follow up.

DISCUSSION: Malignant mesothelioma is pleural malignancy strongly associated with exposure to crocidolite, chrysotile, amosite and all other type of asbestos. Persons working in ship breaking, gas mask production, asbestos insulation work or people living near asbestos factories have increased risk of developing malignant mesothelioma. Onset of symptoms varies from 20-40 yrs. after exposure to asbestos. Malignant mesothelioma is more common in men, with a male-to-female ratio of 3:1.When there is no history of exposure to asbestos, other causes like infections (simian virus 40, HIV, CMV, yersinia enterocolitica) genetic (inactivation of the neurofibromatosis-2 gene and INK4 α /ARF), familial and nutritional causes (carotenoid fruits and vegetables)⁶⁻⁹ are to be considered.

Clinically pleura, peritoneum or pericardium may be involved and patients may present with symptoms of pleural effusion, breathlessness and dull chest pain.

In advanced stages of the disease there may be weight loss, distant metastases, cardiac tamponade, superior vena caval obstruction, chest wall infiltration and esophagus obstruction. Tumor grows through thoracotomy scars and needle tracks. Mediastinal shift is unusual, clubbing and hypertrophic osteoarthropathy are rare.

Chest x-ray shows pleural effusion and opposite lung may show pleural plaques (asbestosis)

Diagnosis is by history of exposure to asbestosis, confirmed by pleural biopsy. Pleural fluid will be hemorrhagic with presence of hyaluronic acid.

Malignant mesothelioma grossly appears as thick white or grey yellow tissue involving both layers with pleural space obliteration and infiltration of surrounding lung tissue. Histopathologically can be differentiated to epithelial cells (cuboidal/flattened) forming tubular and papillary structure separated by matrix (d/d adenocarcinoma) and diffuse spindle cell variety. Sometimes both types can co-exist. Asbestos bodies may be found in lungs but not in tumor.

Malignant mesothelioma has a steady deterioration over 1 to 2 years.

Management is palliative, surgical decortication with radiotherapy and chemotherapy (doxorubicin, cyclophosphamide) are still not of proven value. Studies using proteosome inhibitor bortezomib, monoclonal antibodies bevacizumab in combination with pemetrexed and cisplatin are ongoing.¹¹⁻¹³

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