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PRIMARY PULMONARY LEIOMYOSARCOMA: A RARE PRESENTATION

*VignaKumar G P 1, Saleel P A 1, RaviKumar E 2 and MahaboobVali S 3
1Department of Pulmonary Medicine
2Department of Cardio Thoracic Surgery
3Advance Research Centre, Narayana Medical College & Hospital, Nellore-524003; Andhra Pradesh, India

*Author for Correspondence

ABSTRACT
An unusual case of primary leiomyosarcoma presenting as bilateral mass lesions on chest X ray is reported. A 22-year-old woman was found to have a massive unresectable mass lesion on the right side upon thoracotomy. On histologic examination, the resected specimen showed features consistent with primary pulmonary leiomyosarcoma. She expired 6 months after discharge.

Key Words: Pulmonary Leiomyosarcoma, Trans-Thoracic Needle Biopsy

INTRODUCTION
Primary leiomyosarcomas are extremely rare tumors of the lung arising from smooth muscle present in the bronchi and blood vessels (Miller and Allen, 1993). The first report published with four cases of pulmonary leiomyosarcomas at necropsies (Passler, 1896). There are reports of individual cases but no large series from a single institution has been published from India. Since then several more case reports have been published including recent three from our country (Gupta et al., 2002; Ramakant et al., 2007). These tumors have been found incidentally on chest radiographs and they range in behavior from indolent to very aggressive, depending on the grade of the tumor. Diagnosis is difficult and management is complete resection of the tumor.

CASES
A 22 years old house wife who is not a known case of Hypertension/Diabetes/CAD/Bronchial Asthma presented to the emergency department with a 2 day history of sudden onset of Breathlessness which rapidly progressed to breathlessness at rest and cough with expectoration. She had a history of fever on and off, but denied any weight loss/vomiting/diarrhea. Further enquiry revealed upper respiratory tract symptoms and dry cough since 2 years, with an episode of Hemoptysis 3 month’s back that made her to seek medical attention. CT was done and was reported as bilateral hydatid cyst/dermoid cyst of size 13cm and 10cm on right and left sides respectively. Past medical history is remarkable for infertility (married 7 years) with irregular menstrual history and recurrent Urinary Tract Infections. There is no history of any hereditary disorders in the family. On examination, she was in moderately severe respiratory distress and chest examination revealed features of bilateral pleural effusion. Her hematological and biochemical investigations revealed a raised ESR, leucocytosis, thrombocytosis and reversal of Albumin to Globulin ratio. Chest radiography confirmed bilateral homogenous mass shadows obscuring heart borders on either side. A computed tomography scan showed a large well defined heterogeneous mass lesion with slight contrast enhancement on the right side and 2 similar lesions which were comparatively smaller in size involving the left lung (Figure 1). Bronchoscopy showed extrinsic compression and narrowing of right upper lobe bronchus and a lobulated, smooth contoured, fragile, fleshy endobronchial mass lesion which bleeds on touch, completely occluding the right middle lobe bronchus. BAL fluid examination did not show any evidence of malignancy and was negative for AFB smear. Fine needle aspiration cytology showed features suggestive of benign cystic lesion. Ultrasonography of abdomen was significant for a hypoechoic lesion with honey comb pattern in 7th and 8th segments of right lobe of liver. Serum AFP and CEA levels were subsequently found to be within normal limits. At surgery, using a right posterolateral
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thoracotomy approach, a large growth was visualized involving the whole of right lung extending and infiltrating the right diaphragm which was heterogenous in nature, friable and bleeding on touch. A radical resection approach was not adopted since there was significant left lung involvement also by the disease as per the CT scan done preoperatively. The resected tumour was solid with cut surface smooth and grayish and histology of the resected specimen showed primary pulmonary leiomyosarcoma (Figure 2). The microscopic features depicted fascicular proliferation of spindle cells with scanty fibrillar cytoplasm and elongated blunt-ended nuclei. She was discharged on the 10th postoperative day with symptomatic improvement, but later expired 6 months after getting discharged from our hospital.

DISCUSSION

Primary pulmonary sarcoma accounts for approximately 0.5% of all lung tumors (Attanoos et al., 1996; Suster, 1995). Pulmonary leiomyosarcoma is a rare tumour mostly of adults although it has very occasionally been reported in children (Meritt and Parker, 1996). It is more prevalent among males than females (2.5:1) and is usually found incidentally on routine chest radiographs as they seldom produce symptoms (Yu, 1996). Very little is known about its clinicopathological variations. Leiomyosarcoma is the most common primary pulmonary sarcoma (Janssen et al., 1994). They grow in the parenchyma of the lung and are seen as rounded and sharply defined opacities on chest radiographs (Yu et al., 1996). Although, predisposing factors for this tumor are mostly unknown, a study has illustrated occurrence of multiple leiomyosarcoma in lung allograft associated with Epstein-Barr virus infection in an immunocompromised patient (Somers, 1998). Unlike epithelial tumors, they do not exhibit any tendency towards exfoliation and hence bronchoscopy and washings are not good diagnostic tools (Suster, 1995). Cytology of the pleural fluid may not yield much information for the same reasons. Leiomyosarcomas metastasize via the blood stream and very infrequently through the lymphatics, making mediastinoscopy inappropriate for diagnosis. Percutaneous core biopsies are necessary for definitive diagnosis and sometimes mandate thoracoscopy or thoracotomy. A frozen section is of limited use because of the similarities with some other tumors (Attanoos et al., 1996). Other tumors to be considered in the differential diagnosis include undifferentiated carcinoma, biphasic tumors, carcinosarcoma, carcinoid tumor, intrapulmonary thymoma, and lymphoma. This makes the use of special techniques such as immunohisto-chemistry and electron microscopy important in the evaluation of such lesions. It has been suggested that pulmonary leiomyosarcomas in female patients should never be considered primary neoplasm as there are reports of pulmonary leiomyosarcomas arising in patients with history of hysterectomy for benign leiomyoma several years before presentation. Most of these cases will be diagnosed either at autopsy or after surgical resection, and few were diagnosed by open lung biopsy. Occasionally the diagnosis has been made by cytological examination of bronchial secretions (Sawada et al., 1977), as appeared by isolated reports. The cytological diagnosis has limitations as interpretation of cells obtained from benign or low grade malignant neoplasm may be difficult at times. Transthoracic needle biopsy is more likely to provide diagnosis. To the best of the author's knowledge, diagnosis by transthoracic needle biopsy is probably not yet reported for pulmonary leiomyosarcoma. The present report, therefore, stress on more frequent use of this diagnostic technique among patients with mass lesion of the lung as it gives preoperative diagnosis and therefore, further management plan can be considered. The best available treatment option is complete resection of the tumor, which is associated with a reported 5-year survival of 29% to 40% (Suster, 1995; Moran et al., 1997; Travis et al., 1995). However, only a third of such patients are amenable to surgery due to patient factors or tumor extent (Suster, 1995). As these tumors are resistant to both chemotherapy and radiotherapy, adjuvant therapies are generally not considered. The most consistent predictor of long-term survival is the grade of the tumor (Attanoos et al., 1996; Janssen et al., 1994). The size of the tumor, stage at presentation, and completeness of resection are the other variables. Surgery is the mainstay of therapy for pulmonary leiomyosarcoma in limited disease and even in some cases having local spread, resection was found feasible with survival up to 7.5 years later. Role of radiotherapy is only palliative and chemotherapy is usually used for bilateral like extra-
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Thoracic dissemination and refusal for surgery. Prognosis is generally poor in tumors measuring more than 10cm diameter and more than eight mitotic figures seen per 10 high power fields, even in surgically resected cases (Yellin et al., 1984).

Figure 1: Computed tomography scan of the chest showing the cystic mass with bilateral pleural effusion

Figure 2: A. Specimen of the leiomyosarcoma measuring 13cm excised along with a sliver of lung. B. Photomicrograph showing cells with mitotic figures (hematoxylin and eosin stain, original magnification x40). Light microscopy shows interlacing fascicles of malignant spindle shaped cells. Individual cells are large having a high nuclear-cytoplasmic ratio.

REFERENCES


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