Spindle Cell Sarcoma Presenting as Pancoast Syndrome

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ABSTRACT

This report describes a patient who presented with pancoast syndrome, secondary to spindle cell sarcoma of the lung. A 56-year man presented with dyspnea, engorged neck veins and bilateral upper limb pitting edema. The patient also had ptosis and miosis in the right eye. Right ulnar nerve palsy with atrophy of hand muscles was seen. His chest X-ray showed bilateral pleural effusion with an opacity involving the apex of the right lung along with mediastinal widening. Echocardiography revealed a pericardial effusion which was drained. The patient's CT scan of chest strongly suspected a malignant mass in right upper lobe with extensive mediastinal lymphadenopathy, pleural metastases and pericardial involvement. He was started on oxygen inhalation, dexamethasone, and clopidogrel. Bronchoscopic biopsy confirmed the diagnosis of spindle cell sarcoma. Meanwhile, he was advised radiotherapy. The tumour was not amenable to surgery. Spindle cell sarcoma is a rare connective tissue tumor that replicates rapidly. To the best of the authors' knowledge, it is hereby reported the first case of spindle cell sarcoma of the lung presenting as Pancoast syndrome.

Key Words: Spindle cell sarcoma. Radiotherapy. Pancoast syndrome.

INTRODUCTION

Primary pulmonary sarcomas are a rare entity among the intrathoracic neoplasms. They are of connective tissue origin and seen so far in literature as leiomyosarcoma, monophasic synovial sarcoma, epithelioid sarcoma, and malignant fibrous histiocytoma. Chest physicians have a limited experience with them. They may occur anywhere in the lung, but with involvement of the apex, they may present as Pancoast syndrome, with Horner's syndrome, brachial plexus involvement, and superior vena cava obstruction. When the tumor is diagnosed, by that time local and distant metastases have already occurred, so treatment is usually palliative.

We hereby report the first case of spindle cell sarcoma of the lung presenting as Pancoast syndrome.

CASE REPORT

A 56-year school teacher presented to our ward with chief complaints of dyspnea, orthopnea and paroxysmal nocturnal dyspnea for the last 4 months. He had been to several local doctors who had treated him for chest infection but there had been only partial relief. He had a past history of excessive smoking. His only other complaint was constipation.

His blood pressure was 130/90 mmHg with pulse rate of 100/minute and body temperature of 99°F. He was having grade IV dyspnea, grade II clubbing was present. Bilateral upper limb pitting edema was also there. He had also developed central cyanosis and a non-pulsatile raised Jugular Venous Pulse. His neck veins were engorged, and Pemberton's sign was present. Seven telangiectasias were visible on the chest wall anteriorly. Respiratory system examination revealed decreased breath sounds bilaterally, increased vocal fremitus, and dull percussion notes over right upper lung zone.

The patient had ptosis and miosis in the right eye. Direct light reflex was present, but consensual reflex was absent on the right side. Neurological examination of upper limbs revealed right ulnar nerve palsy with atrophy of hand muscles. The deep tendon reflexes were normal. Musculoskeletal system examination was remarkable for proximal myopathy.

Investigations revealed a normal peripheral smear, liver and renal profile, with normal serum electrolytes and random blood sugar levels. The serum calcium was also normal. Arterial blood gases showed uncompensated respiratory alkalosis with pH = 7.52, pCO2 = 24.4 mmHg, pO2 = 88 mmHg, HCO3 = 20.3 mmol/liter, BEb = -0.8 mmol/liter, and %SO2 = 93%. His chest X-ray showed bilateral pleural effusion with an opacity involving the apex of the right lung along with mediastinal widening.

The pleural fluid routine examination gave an exudative picture with proteins = 4.75 gm/dl, leucocytes = 260/cmm, RBCs = 18,500/cmm, neutrophils = 35%, and lymphocytes = 65%. The fluid cytology was negative for Ziehl Neelsen and Gram stains.

The patient's ECG showed small QRS complexes so an echocardiography was done, which revealed a large pericardial effusion anteriorly and a small pericardial effusion posteriorly. Therapeutic pericardiocentesis was done and 325ml fluid was drained.

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The patient’s CT scan chest was done which confirmed a poorly-enhancing hypodense mass in the right upper lobe with cavities inside the lesion. Multiple enlarged lymph nodes were seen in the pre-tracheal, right para-tracheal, aorto-pulmonary recess, right tracheobronchial, hilar and para cardiac region. A lymphomatous mass in right tracheo-bronchial region was compressing the superior vena cava and azygous vein. Superior vena cava obstruction was also noticed. Bilateral pleural effusion and pericardial effusion with pericardial thickening were present. A strong suspicion of malignant mass in right upper lung lobe was made with extensive mediastinal lymphadenopathy and pleural metastases along with pericardial involvement (Figures 1a and 1b).

The patient received oxygen inhalation, 12 mg dexamethasone daily in divided doses, and clopidogrel 75 mg.

The patient was booked for bronchoscopy and biopsy which confirmed a tumour showing pleomorphic spindle cells with eosinophilic cytoplasm and brisk mitotic activity. Negative high molecular weight cytokeratin (34BE12) made sarcomatoid carcinoma unlikely. Positive desmin stain supported muscle differentiation. A diagnosis of spindle cell sarcoma was made (Figure 2).

Meanwhile, he was advised 22 cycles of radiotherapy at Institute of Radionuclear Medicine (IRNUM), Peshawar. During ward stay, he received 3 cycles with which he improved symptomatically. He was discharged and advised to return for radiotherapy. The tumour was not amenable to surgery because of extension outside the lung and involvement of the pericardium, so patient was continued on radiotherapy.

**DISCUSSION**

Spindle cell sarcoma is a mesenchymal origin cancer that has rare occurrence. The cancer is comprised of cells that appear spindle shaped under the microscope. They begin in layers of connective tissue and mostly under the skin where two or more muscles connect. Spindle cells are formed in the body in response to injury where the connective tissue cells take the shape of spindle cells to combat the injury. These spindle cells are then prone to development of spindle cell sarcoma.

Spindle cell sarcoma usually begins as a small lump in its stage I. At this stage, it can be excised along with a rim of healthy tissue and this stage carries the best prognosis. If left to progress to stage II or III, micrometastases have already occurred by then. So even if the lump is excised, complete cure is not possible. Chemotherapy or radiotherapy is required for the treatment of stage II and III sarcomas.

Spindle cell sarcoma can occur anywhere in the body. But it has never been so far reported in the lung, and that too with presentation of Pancoast Syndrome (or Pancoast Tumor; also called Superior Sulcus Tumor). This is the first case to be reported wherein a spindle cell sarcoma has led to Pancoast syndrome with involvement of the brachial plexus leading to ulnar nerve palsy; cervical ganglia leading to Horner Syndrome and Superior Vena Cava obstruction.

Other pathologies having presented as Pancoast Syndrome include mesothelioma, lymphoma, neurofibroma, leiomyosarcoma, myxomatous tumor and hydatid cysts. Spindle cell sarcoma is a rare pathology leading to Pancoast syndrome. Investigations and management are virtually the same as for other lung malignancies.
Spindle cell sarcoma presenting as pancoast syndrome

Since the tumor has spread to distant sites by the time it is diagnosed and mostly is not amenable to surgical resection, early diagnosis is the key to better management.³

To the best of the authors' knowledge, this is the first case to be reported with spindle cell sarcoma involving the lung and manifesting as Pancoast syndrome. Further studies on the nature and progression of spindle cell sarcoma of the lung are needed.

REFERENCES


