Case Series

Primary synovial sarcoma of lung – A report of two cases

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Abstract

Primary synovial sarcoma (SS) is an uncommon lung tumor. The thigh, knee, ankle, foot, and upper extremities are the most often reported locations of origin. Sarcomas of the lung barely make up 0.1–0.5% of cases. This report presents two cases of primary pulmonary SS. Typically, the tumor manifests at a younger age. When the diagnosis of primary pulmonary SS is confirmed by clinical, histological, and immunohistochemical examinations, molecular testing is not necessary. Instead, radiological and histopathological findings are necessary for the diagnosis of SS. Early surgical intervention is recommended. The initial course of treatment for advanced SS includes ifosfamide and doxorubicin.

Keywords: Immunohistochemistry, primary synovial sarcoma, synovial sarcoma, tumor

Introduction

Solitary synovial sarcoma (SS) is an uncommon lung tumor. Primary pulmonary SS is much less prevalent than lung metastases from other organs.[1] As the name implies, the tumor originates pluripotent mesenchyme cells, not synovium. Thus, the phrase "synovial sarcoma" is not accurate. The thigh, knee, ankle, foot, and upper extremities are the most often reported sites of origin.[2] Rarely, SS can originate from the mediastinum, pleura, or lung. Just 0.1–0.5% of all initial lung cancers are pulmonary sarcomas.[3] The three most commonly documented subtypes of lung sarcomas are fibrosarcoma, leiomyosarcoma, and malignant fibrous histiocytoma.[3] Lung primary SS is uncommon. Here, we report two cases of primary pulmonary SS.

Case-1

A 63-year-old male, a current smoker (smoking index: 600) and a farmer by occupation, presented to the pulmonary medicine outpatient unit with complaints of right-sided chest pain and breathlessness for three months. The pain was insidious in onset, dull aching, and non-radiation. Breathlessness and insidious onset gradually progressed in MMRC grade-II. His history was unremarkable with no associated comorbidities. Vitals were stable at the time of presentation. Chest X-ray revealed a large homogenous opacity in the right upper, middle, and lower zones [Figure 1a]. Contrast-enhanced computed tomography (CECT) scan of the thorax and abdomen revealed an approximately 12 × 9 × 16 cm well-circumscribed, heterogeneous lesion with internal ill-abutting to the right anterior chest wall and medially its abutting the pericardium, ascending thoracic aorta, and the right main branch of the pulmonary artery [Figure 1b]. Multiple soft tissue density nodules (at least two in the right lung and five in the left lung) were seen in bilateral lung fields (the largest measuring ~ 5 mm in the left upper lobe). An ultra-sonography-guided biopsy was performed. The needle biopsy from the tumor comprised two cores which showed a relatively monotonous-looking tumor. The tumor cells had round to oval to slightly spindled morphology at different areas with scanty cytoplasm. No significant mitotic figures were noted on hematoxylin and eosin (H and E)

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stained slides. With the possibility of a soft tissue sarcoma, the following immune histochemical (IHC) markers were performed (vimentin, cytokeratin, Ki67, BCL2, SOX10, CD 34, and TLE1). CK, SOX10, and CD34 were negative, while vimentin, BCL2 and TLE1 were strongly and diffusely positive in the tumor cells [Figure 2a and b]. The morphological and IHC profile was consistent with the diagnosis of SS. Ki67 stain showed scattered positivity in the tumor cells, with a count of around 15–20% in hot spot areas, indicating a rapidly proliferating tumor.

The final diagnosis of SS with bilateral lung metastasis was made. The patient was started on chemotherapy with doxorubicin and ifosfamide. After two cycles of chemotherapy, the patient expired at home.

Case-2

A 29-year-old male presented with complaints of breathlessness and dry cough for 2 months. There was no history of fever, wheezing, hemoptysis, anorexia, or significant weight loss, but the severity of both the chest pain and dyspnea were gradually increasing. The pain was not relieved by simple analgesics and caused sleep disturbance. He was a non-smoker and a daily wage laborer. On general examination, there was no anemia, clubbing, engorged neck vein, or palpable superficial lymph nodes. His temperature was 37°C, respiratory rate was 18 breaths/min, pulse rate was 86 beats/min, blood pressure was 110/70 mmHg, and SpO2 was 97% on room air. Examination of the respiratory system revealed reduced movement of the right hemithorax, shifting of the trachea and apical impulse to the left, dull percussion note on the right side, diminished vesicular breath sounds, and vocal resonance on the right side. Examination of other systems revealed no abnormality. Complete hemogram and blood biochemistry were within normal limits. Chest X-ray PA view showed right-sided homogenous opacity involving all zones with mediastinum shifted to the other side [Figure 1c]. CECT scan of the thorax revealed a huge right-sided heterogeneous mass lesion of size 16 × 13 cm occupying almost the whole of the right hemithorax with ill-defined necrotic and cystic areas with the contralateral shifting of the mediastinum [Figure 1d]. Ultrasound-guided biopsy of the mass was performed, and histopathological examination showed spindle malignant cells with moderate pleomorphic, fine chromatin, and scanty cytoplasm, which are positive for cytokeratin, vimentin, and CD99, features suggestive of SS [Figure 2c and d]. A bone scan reveals focal osteoblastic lesions in the right high parietal bone and left 9th rib. In view of skeletal metastasis, the patient was started on chemotherapy with doxorubicin and ifosfamide and completed three cycles. The patient died due to pneumonia after 2 month of chemotherapy.

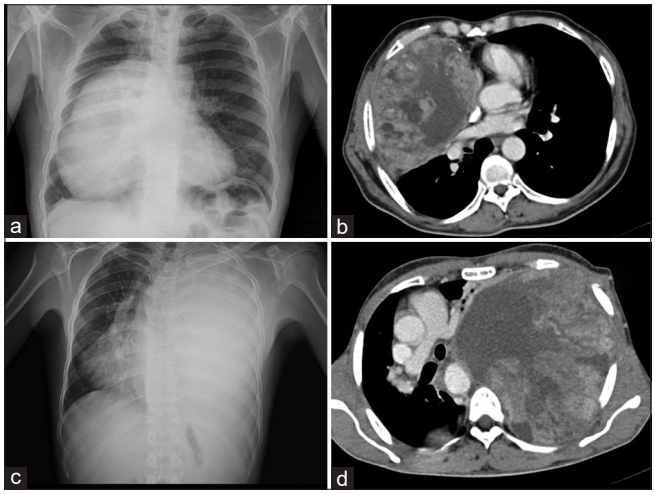

Figure 1: (a) Chest x ray of 63-year-old male showing right side well circumscribed mass. lesion in middle and lower zones. (b) Contrast enhanced computed tomography thorax & abdomen of 63-year-old male showing approximately 12 x 9 x 16 cm (well circumscribed, heterogeneous lesion with internal ill-defined necrotic areas, calcific specks. (c) Chest X-ray of 29-year-old male showing right sided homogenous opacity involving all zone with mediastinum shifted to other side. (d) Contrast enhanced computed tomography (CKCT) thorax of 29-year-old male showing huge right sided heterogeneous lesion mass lesion of size 16*13 cm mass occupying almost whole of the right hemithorax with ill-defined necrotic areas and cystic area with contralateral shifting of the mediastinum

Discussion

SS is a rare mesenchymal tumor, accounting for 10% of all soft tissue sarcomas.[4] It occurs most commonly in the soft tissues of the extremities and is very rarely observed in the lungs. Pulmonary SS comprises only 0.5% of all primary lung malignancies.[5] This rare tumor affects mainly young adults, with a mean age of diagnosis of23, and males are most commonly affected.[6] We have reported two cases of SS of the lung: one patient is at

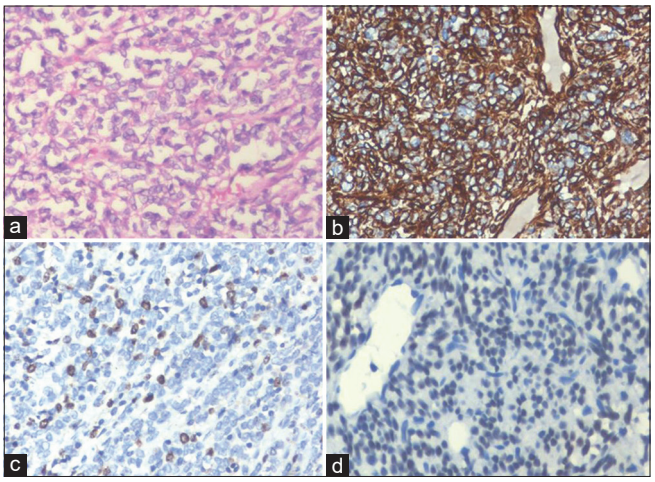

Figure 2: (a) Hematoxylin and eosin stain (400×): High-power image showing relatively monotonous tumor cells with round to oval nuclei and scanty cytoplasm. No significant mitotic figures were noted in this stain. (b) Immunohistochemistry (IHC) for bCL2 stain (400×) shows diffuse strong cytoplasmic positivity in the tumor cells, which is a consistent stain in synovial sarcoma. (c): IHC for Ki67 stain (400×) shows scattered nuclear positivity in the tumor cells, amounting from 15–20%, indicative of a high proliferative index. (d) IHC for TLE1 stain (400×) shows diffuse strong nuclear positivity in the tumor cells, confirming the tumor to be synovial sarcoma

the age of 60, and another is at the age of 29. This tumor is extremely rare in older age. The presentation of the tumor is diverse, and most of the cases have an initial presentation similar to a primary tumor of the lung. Chest pain and dyspnea are the most commonly described presenting symptoms.[7] Both of our patients presented with symptoms of chest pain, hemoptysis, and breathlessness. There are many atypical presents reported also. As per a case report by Qiu *et al.*, this tumor may mimic pulmonary embolism.[8] The diagnosis of primary pulmonary SS requires clinical, radiological, pathological, and immunohistochemically investigations.[4] A total of 66% of primary pulmonary SSs are centrally located and present with post-obstructive pneumonia, atelectasis, and hemoptysis.[9] Primary pulmonary SSs are of four subtypes – monophasic fibrous (spindle), monophasic epithelial, biphasic, and poorly differentiated, among which monophasic fibrous subtype is the most common.[4] A retrospective study from China showed that spindle cells were most commonly associated with histology types.[10] Both of our cases are of spindle cell morphology.

Primary pulmonary SSs are well-circumscribed, heterogeneously enhancing lesions with areas of necrosis on contrast-enhanced CT scans that do not invade surrounding bone or calcifications.[11] Tumors are usually not associated with mediastinal lymphadenopathy.[11]

In an immunohistochemistry study, SSs are nearly uniformly positive for cytokeratin, EMA, BCL-2, and vimentin and negative for S-100, desmin, smooth muscle actin, and vascular tumor markers.[12] Cytogenetic testing is helpful, as t (X; 18)(p11.2;q11.2) is characteristic of SS, distinguishing it from other soft tissue tumors.[13] Molecular testing is not required if the diagnosis of SS is certain or probable based on clinical, histological, and immunohistochemical evaluations. This tumor is very aggressive with poor survival rates. Centrally located tumors, high histologic grade, high mitotic rate, and necrosis have been associated with worse outcomes. Similarly, bone and neurovascular invasion are poor prognostic markers.[14] According to a retrospective study, the 2-year morality of pulmonary SS patients is 100%.[14]

Surgical excision of the tumor is the treatment of choice for patients with no metastatic, T1 (<5 cm), and superficially located tumors.[15] Radiotherapy has no role in the management of the disease. Doxorubicin and ifosfamide comprise first-line treatment for advanced SS. A combination of gemcitabine and docetaxel is reserved for patients who cannot tolerate or are resistant to standard chemotherapy. Pazopanib, a multitargeted tyrosine kinase inhibitor that predominantly inhibits vascular endothelial growth factor receptors 1, 2, and 3; PDGFR a and c-kit has been approved for the treatment of SS.[16]

Conclusion

Primary SS is an uncommon and severe lung tumor. The clinical appearance is comparable to that of other lung primary tumors. Both radiographic and histological characteristics are necessary for the diagnosis of primary pulmonary SS. The characteristic feature is a radiologically confined mass lesion with an area of necrosis. Lymphadenopathy is typically not seen in the same way as a lung primary tumor. Immunohistochemistry is another way to confirm the diagnosis. Chemotherapy and early surgery are the available therapeutic options.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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