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A Case of Primary Pleural Synovial Sarcoma

Primer Plevral Sinovyal Sarkom Olgusu

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Abstract

We present here the case of a 29-year-old male with primary pleural synovial sarcoma. The patient had no complaints, no significant medical history and did not smoke. A well-circumscribed giant mass, adjacent to the chest wall was observed on a PA chest X-ray, with a solid mass lesion in the lateral right hemithorax with diffuse amorphous calcifications noted on thorax computed tomography. An F18-FDG uptake with a SUVmax of 5.0 was detected in the mass defined by PET/CT. A Tru-cut biopsy revealed monophasic as histological subtypes composed of a dense cellular and interlaced fascicular proliferation of spindle cells, and SYT-SSX1 fusion (X;18)(p11.23;q11) consistent with synovial sarcoma. The patient was followed-up in the oncology center after surgery and chemoradiotherapy, and has been stable for 1 year. This paper draws attention to a diagnostic approach to primary pleural synovial sarcoma, which is a rare and aggressive tumor, and the need for the prompt initiation of treatment for an improved outcome.

Key words: *Lung cancer, pleura, synovial sarcoma.*

Öz

Primer plevral sinovyal sarkom tanılı 29 yaşında erkek hastanın, gelişinde herhangi bir şikayeti yoktu, sigara içmiyordu ve özgeçmişinde özellik yoktu. PA akciğer grafisinde, göğüs duvarına bitişik düzgün sınırlı dev kitle izlendi. Toraks BT'sinde; sağ hemitoraks lateralinde, diffüz amorf kalsifikasyon içeren solid kitlesel lezyon tespit edildi. PET/BT'de, tanımlanan lezyonda SUVmax 5.0 olan F18-FDG tutulumu saptandı. Tru-cut biyopsi sonucunda, histolojik olarak monofazik, yoğun hücresel ve birbiri üzerine geçen fasiküllerin oluşturduğu iğsi hücreler, SYT-SSX1 füzyon (X;18)(p11.23;q11) saptanması üzerine sinovyal sarkom tanısı kondu. Cerrahi ve kemoradyoterapi sonrasında 1 yıldır stabil olarak onkoloji merkezinde takip edilmektedir. Bu yazı, nadir görülen ve agresif bir tumor olan primer plevral sinovyal sarkomun tanı basamakları ve iyi sonuçlar alınabilmesi için hızlıca tedavisinin başlanmasına dikkat çekmeyi amaçlamaktadır.

Anahtar Sözcükler: *Akciğer kanseri, plevra, sinovyal sarkom.*

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Although synovial sarcoma is relatively rare, it accounts for 5–10% of all soft tissue sarcomas. It is frequently seen in adolescents and young adults, and is diagnosed equally in both sexes (1). It is often found in the lower extremities, and primarily in the hip or knee-joint regions in periarticular soft tissue, and can rarely be of bone origin (2). Solid tumors of pleural origin other than malignant mesothelioma include desmoid type fibromatosis, neurogenic tumors, liposarcoma, epithelioid hemangioendothelioma, solitary fibrous tumors and synovial sarcoma (3). Primary synovial sarcoma is very rare in the lung, accounting for 0.5% of all lung cancers (4). Fewer than 50 cases of pleural synovial sarcoma have been reported in literature (5). Immunohistochemical staining for transducine-like enhancers of split 1 (TLE1), cytokeratins (CK), epithelial membrane antigens (EMA), PANCK (cytokeratin AE1/AE3), B cell lymphoma 2 (BCL2), beta catenin, calponin, CD 56 (cluster of differentiation), CD57, CD99 and calretinin and SS18-SSX fusion-specific antibodies are used for definitive diagnosis (6). Here, we present the case of a 29-year-old male with primary pleural synovial sarcoma.

CASE

A 29-year-old male patient was referred to our outpatient clinic after a lesion was observed in a PA chest X-ray taken for a workplace health check-up. The patient had no complaints and did not smoke. He had no significant disease history and no comorbid disease. White blood count: 10980 mm^3, hemoglobin: 14.5 g/dL, hematocrit: 42.9%, platelet count: 291000 mm^3, C-reactive protein (CRP): 33.3 mg/L, international normalized ratio (INR): 1.12, lactate dehydrogenase: 405 U/L and creatinine: 1.05 mg/dL were detected. There was no pathology in a postero-anterior (PA) chest X-ray taken 3 years earlier. A well-circumscribed giant mass, adjacent to the chest wall, forming a wide angle with the chest wall, almost completely filling the upper and middle zone of the right lung was observed in the current PA chest X-ray (Figure 1). A 10x13x14 cm solid mass lesion in the lateral right hemithorax with its wide base sitting on the pleura and a regular free contour causing compression on the lung and with diffuse amorphous calcifications was detected on thorax computed tomography (CT) (Figure 2). No mediastinal or hilar lymph nodes were observed on CT. Heterogeneous fluorine-18-fluorodeoxyglucose (F18-FDG) uptake with a standardized uptake value (SUV) max of 5.0 was detected in the mass, defined from positron emission tomography (PET)/CT taken for metastasis evaluation, and pathological FDG uptake with a SUVmax of 5.0 was detected in the 1 cm parenchymal nodular lesion posterior to this lesion and adjacent to mild pleural effusion. There was no other primary focus other than the lung on PET/BT. Ultrasound guided tru-cut biopsy was performed on the giant mass through interventional radiology. Monophasic histological subtypes composed of dense cellular and interlacing fascicular proliferations of spindle cells were seen. CK7 (focal), CK19 (focal), beta catenin, TLE1 and CD99 were positive; SRY-related HMG-box 10 (SOX10), calretinin, Wilm's tumor 1 (WT1), S100 protein, CD34, CD117, Desmin, smooth muscle actin (SMA) and human melanoma black-45 (HMB45) were negative in immunohistochemical (IHC) staining, and the Ki67 proliferation index was 10% (Figure 3). Rearrangement was detected in the SS18 (SYT) gene by a fluorescence in-situ hybridization (FISH) technique. Fusion of the SYT gene located on chromosome 18 to the synovial sarcoma X-1 (SSX1) gene in the Xp11 region (SYT-SSX1 fusion) was detected in an investigation of reciprocal chromosomal translocations (X;18)(p11.23;q11) using a reverse transcriptase-polymerase chain reaction (RT-PCR) technique.

A diagnosis of synovial sarcoma was made based on the available histopathological findings. The patient was referred to surgery, but opted to undergo surgery in another thoracic surgery clinic unconnected to our hospital. The patient's medical records revealed that he underwent a mass excision and visceral and parietal decortication, and since no palpable nodule was detected during surgery, no lung parenchyma resection was performed. A postoperative control PET/CT revealed that the parenchymal nodule had regressed spontaneously, as well as the pleural effusion. However, since there was suspicion about R0 resection and a 1 cm sized nodule was detected in the lower lobe, the patient was referred to medical oncology for chemoradiotherapy. The mitosis rate was lower than 10/10 high-power field (HPF), and the necrosis rate was less than 50%.

DISCUSSION

Primary pulmonary synovial sarcoma is a very rare, highly aggressive malignant tumor that is detected equally in both sexes and mostly in young adults, and accounts for 0.5% of all lung cancers (7). The METASARC study reported 5-year survival in only 8.5% (16/188) of patients with a diagnosis of metastatic synovial sarcoma following diagnosis (8). Clinical features, radiological findings, histopathological features and genetic parameters are

important in the diagnosis and differential diagnosis of synovial sarcoma, which is a rare disease associated with high mortality.

Although primary pulmonary synovial sarcoma can involve any lobe radiologically, it is reported mostly in the upper lobes, and can be located centrally or peripherally in the lung. Chest pain is the most common complaint, as well as symptoms of cough, hemoptysis, weight loss, fever, and shortness of breath. Patients may also be completely asymptomatic, with radiological masses detected only incidentally (9,10). In the present case, the radiological lesion was detected incidentally in a non-smoking asymptomatic young adult patient.

There was no radiological finding of calcification in Hartel et al.'s (11) primary pulmonary synovial sarcoma study of 60 patients, while diffuse amorphous calcification areas were observed in our case. Concurring with literature, a mild F18-FDG uptake was detected in the mass lesion in our case. In the study by Lan et al. (12), in four of the 26 patients studied, the pleurapulmonary and mediastinal synovial sarcoma were of pleural origin The authors also reported detecting an ipsilateral pleural effusion in 12 of 21 (57.1%) patients. In our case, an ipsilateral pleural effusion was detected at the time of diagnosis, and complete regression was observed postoperatively.

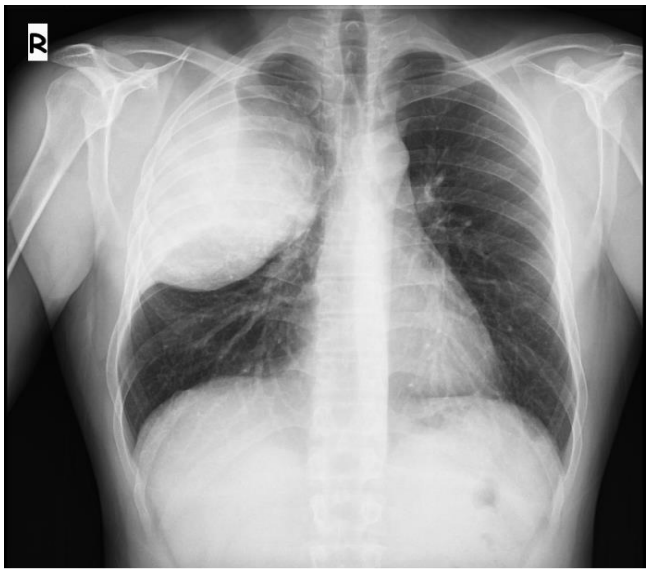

Figure 1: A well-circumscribed giant mass adjacent to the chest wall forming a wide angle with the chest wall, almost completely filling the upper and middle zone of the right lung on a PA chest X-ray

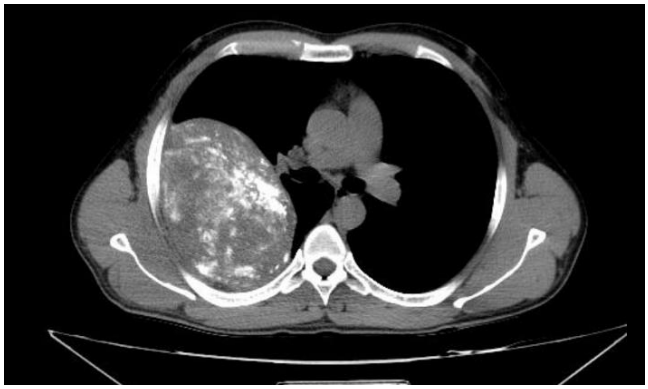

Figure 2: A 10x13x14 cm solid mass lesion in the lateral right hemithorax with its wide base sitting on the pleura and with a regular free contour, causing compression in the lung and with diffuse amorphous calcifications on thorax computed tomography

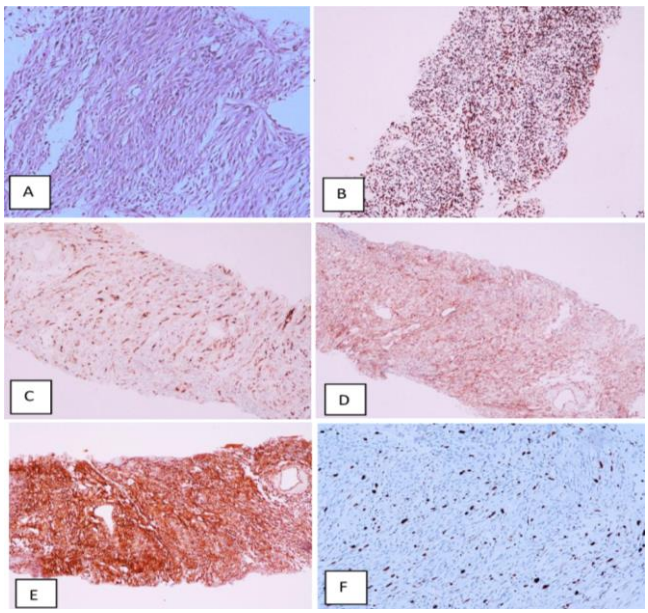

Figure 3: Histologic features of synovial sarcoma. Synovial sarcoma is one of the most cellular spindle-cell neoplasms of the lung, and is characteristically composed of densely cellular interlacing fascicles (A). Tumor cells showing diffuse nuclear staining with TLE-1 (B). Cytokeratin positivity is critical for diagnosis and can be minimal (C). Moderate intensity CD99 staining (D) and strong beta-catenin staining have shown (E). Slightly high Ki-67 proliferative index (F)

Soft tissue tumors present with quite different clinics due to their histological differences, variable pathological findings and genetic changes. The histological subtypes of synovial sarcoma are monophasic, biphasic and poorly differentiated. Among these, the synovial sarcoma monophasic subtype can be confused with other soft tissue tumors, and so IHC examinations are important in a differential diagnosis (13,14). The monophasic subtype was detected histologically in this case report.

The combination of EMA and cytokeratin positivity and CD34 negativity has been stated as the most useful marker in the diagnosis of synovial sarcoma in IHC staining (15). In the presented case, CK7, CK19, PANCK, beta catenin, TLE1 and CD99 were positive; and SOX10, calretinin, WT1, S100 protein, CD34, CD117, desmin, SMA and HMB45 were negative. The optimum approach to the definitive diagnosis of synovial sarcoma is the demonstration of SS18-SSX (2/3 SSX1, 1/3 SSX2 and rarely SSX4) fusion by RT-PCR or FISH techniques. In a recent study investigating the diagnostic efficacy of the SS18-SSX fusion-specific antibody, 93 masses, including 10 synovial sarcomas metastasizing to the lung, and five intrathoracic solitary fibrous tumors, 39 primary lung cancers, and 49 non-synovial sarcomas metastasizing to the lung were compared. While SS18-SSX was positive in all 10 synovial sarcomas, it was negative in all 93 non-synovial sarcoma masses (100% sensitivity, 100% specificity) (16). A definitive diagnosis was made by demonstrating SS18-SSX1 fusion with the FISH technique in the presented case.

Although there is no clear recommendation for optimal treatment, the leading treatment option is surgery, involving the complete removal of the mass, lobectomy or pneumonectomy. Synovial sarcomas are relatively chemotherapy-sensitive tumors. Adriamycin alone or in combination with ifosfamide can be used, while radiotherapy can be used for the local control of the disease (11,17). In the presented case, tumor excision and decortication were performed, and the patient was then referred to oncology for chemoradiotherapy.

Advanced age, female gender, R1 and R2 resection, tumor size >5 cm, extensive tumor necrosis, high mitotic activity and neurovascular invasion have been reported as poor prognostic criteria. Although SYT-SSX1 fusion has also been stated to be a poor prognostic factor, other studies reported opposite findings (4,18). The present case was a 29-year-old male with a >5 cm tumor, an unclear R0 resection, mild tumor necrosis and low mitotic activity, and SYT-SSX1 fusion was detected. The patient has maintained a stable condition for 1 year on follow-up in the oncology center.

CONCLUSION

Primary synovial sarcoma is very rare in the lung, and is a highly aggressive malignant tumor that is detected equally in both sexes, and mostly in young adults. We presented this case to draw attention to the approaches to the diagnosis and treatment of this rare disease.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - K.C., I.Y., T.C., O.A., O.O., Z.K., N.K.T.; Planning and Design - K.C., I.Y., T.C., O.A., O.O., Z.K., N.K.T.; Supervision - K.C., I.Y., T.C., O.A., O.O., Z.K., N.K.T.; Funding -; Materials - K.C., I.Y., T.C., O.A., N.K.T.; Data Collection and/or Processing - K.C., I.Y., T.C., O.A., N.K.T.; Analysis and/or Interpretation - K.C., I.Y., T.C., O.A., N.K.T.; Literature Review - K.C., T.C., O.O., Z.K., N.K.T.; Writing - K.C., T.C., O.A., O.O., Z.K.; Critical Review - K.C., I.Y., T.C., O.A., O.O., Z.K., N.K.T.

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