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CASE REPORT
ΕΝΔΙΑΦΕΡΟΥΣΑ ΠΕΡΙΠΤΩΣΗ

Synovial sarcoma
Report of a case of diffuse metastasis to the lungs

Synovial sarcoma is a rare aggressive type of cancer that accounts for a very small percentage of lung malignancies. It affects the soft tissues, which connect, support, and surround the bones and organs in the body, including muscles, fat, blood and lymph vessels, nerves, tendons, and the lining of joints. Here, we report the case of a 43-year-old woman, who presented with shortness of breath, severe cough and hemoptysis and was found to have synovial sarcoma of the thigh that had metastasized to the lung.

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Σάρκωμα του αρθρικού υμένα: μια σπάνια περίπτωση διάχυτων μεταστάσεων στους πνεύμονες

Περίληψη στο τέλος του άρθρου

Key words

Lung sarcoma
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Synovial sarcoma is one of the rare types of cancers, affecting connective tissues such as bones, muscles, tendons, cartilages, nerves, fat and blood vessels in the breasts and legs. It can also occur in other parts of the body. Synovial sarcoma typically involves periarticular tissue, but it has been documented in all sites that malignancy occurs. The lung is an uncommon place for primary sarcoma,[1,2] but sarcoma often metastasizes to the lung. Sarcoma has many different types, including lymphosarcoma, leiomyosarcoma, Kaposi's sarcoma and synovial sarcoma, in which the chances of survival are low.[3,4]

Synovial sarcoma is rare, aggressive type of sarcoma of soft tissue origin[5,6] that presents in the soft tissues of the extremities; for example, the site that has been most commonly reported is near the large joints.[4] Synovial sarcoma is diagnosed most commonly in the third decade of life.[7] The tumor size is usually greater than 5 cm. The resection margin status, presence of metastasis and degree of histological differentiation are commonly used prognostic factors.[8] Two histological subtypes of synovial sarcoma that are recognized are biphasic and monophasic. The biphasic variant includes both epithelial and spindle cells and the monophasic spindle

cells only.[10] Synovial sarcoma metastasizes mostly to the lung. Radiotherapy and chemotherapy are commonly used forms of treatment for patients with synovial sarcoma, but there are not enough relevant case reports to draw conclusions on their efficacy.[6,7] We report the case of lung metastasis of synovial sarcoma from a primary lesion in the left thigh.

CASE PRESENTATION

A 43-year-old woman with lung metastasis and shortness of breath was referred for chemotherapy. Her primary symptoms were severe cough with sputum and hemoptysis, and lung metastatic involvement was diagnosed by computed tomography (CT) scan (fig. 1). The patient had no other symptoms (including fever, shivering, weight loss, appetite loss, nausea, vomiting, diarrhea, or constipation). She had no history of heart disease but she had hypertension because of taking corticosteroid drugs for arthritis. She was not a smoker. Her medication history was otherwise negative and there was no history of food and medication allergies. There was no family history of cancer. At the time of admission, the patient was not obviously ill or toxic. Her vital signs were stable. Her laboratory tests were in the normal range.

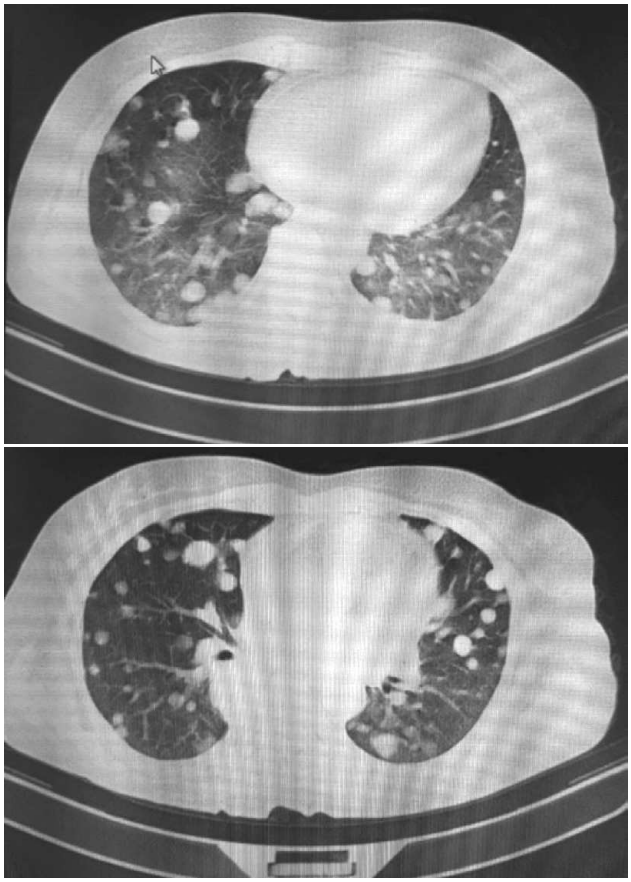

Figure 1. Lung metastasis of synovial sarcoma in a 43-year-old woman. Computed tomography (CT) scan showing multiple masses in the lungs.

In immunoassays, only CPA was positive, which appears to be due to the history of rheumatoid arthritis. On ultrasonography imaging (U/S), a solid tumor was detected in the posterior region of the left thigh with the dimensions 94×140×135 mm. Magnetic resonance imaging (MRI) of the left thigh showed a large soft tissue mass in the posterior muscle of left thigh measuring about 140×100×130 mm. Intense enhancement was reported extending to the vastus intermedius muscle and these signs were first considered to be consistent with a diagnosis of leiomyosarcoma. On histopathological of a nodule from the right lung, microscopy and immunohistochemistry showed a malignant neoplasm composed of atypical spindle cell proliferation positive to PanCK, TLE1 and Bcl2 immunostaining, focal equivocal Calretinin and Synaptophysin staining, but no reaction by TTF1, CD34, P40, and WT1. Overall, these findings were compatible with synovial sarcoma. Chemotherapy was administered, with satisfactory response, and the patient is well at present.

DISCUSSION

Synovial sarcomas are aggressive neoplasms, but are rare among the soft tissue sarcomas that occur most commonly in adolescents and young adults. Synovial sarcoma metastasizes mostly to the lungs. Almost 20% of cases of extremity sarcoma will give rise to pulmonary metastatic disease during the course of the disease,[8,9] and the pulmonary metastasis may produce the presenting symptoms. Patients with this type of sarcoma may present with chest wall pain, cough, shortness of breath, and hemoptysis.[2] Synovial sarcoma is slightly more frequent in males and is diagnosed mostly in young adults (median age 35 years) although the reported age range is 5–85 years.

Synovial sarcomas include two major histological subtypes, biphasic and monophasic, of which the monophasic is the most common and consists of spindle cells only. The biphasic type contains both spindle and epithelial cells.[10,11]

Clinical investigations, conventional x-ray and CT of the chest can be used for the detection and diagnosis of metastatic synovial sarcoma, and immunohistochemical investigations are required to confirm metastatic sarcoma.[12]

On CT scans, these lesions are observed as sharply demarcated with no calcification and a heterogeneous appearance.[13] The chest CT image of our patient, showed multiple metastases that were clearly visible widely spread through the lungs and abundant. The distribution of the tumors was unusual. In our patient, the primary tumor was identified in the left thigh.

Primary synovial sarcoma of the lung is a very rare tumor, with a poor prognosis. Due to the rarity of this disease, standard treatment protocols are not available. Surgery,

radiotherapy, chemotherapy can all play a part in treatment. The most appropriate management of the primary tumor is surgical excision with negative margins, whenever feasible.

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ΠΕΡΙΛΗΨΗ

Σάρκωμα του αρθρικού υμένα: μια σπάνια περίπτωση διάχυτων μεταστάσεων στους πνεύμονες

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Το σάρκωμα του αρθρικού υμένα είναι ένας επιθετικός και σπάνιος τύπος καρκίνου που ευθύνεται για ένα πολύ μικρό ποσοστό κακοηθειών του πνεύμονα. Προσβάλλει τους μαλακούς ιστούς, οι οποίοι συνδέουν, υποστηρίζουν και περιβάλλουν τα οστά και τα όργανα του σώματος, όπως μυς, λίπος, αιμοφόρα ή λεμφικά αγγεία, νεύρα, τένοντες καθώς και την επένδυση των αρθρώσεων. Το σάρκωμα του αρθρικού υμένα είναι ένας επιθετικός και σπάνιος τύπος σαρκώματος. Περιγράφεται μια περίπτωση 43χρονης γυναίκας, η οποία παρουσίαζε δυσκολία στην αναπνοή, έντονο βήχα και αιμόπτυση και βρέθηκε ότι είχε σάρκωμα του αρθρικού υμένα με μεταστάσεις στον πνεύμονα.

Λέξεις ευρετηρίου: Μεταστάσεις, Σάρκωμα αρθρικού υμένα, Σάρκωμα αρθρικού υμένα στον πνεύμονα, Σάρκωμα πνεύμονα

References

1. MILLER KD, SIEGEL RL, LIN CC, MARIOTTO AB, KRAMER JL, ROWLAND JH ET AL. Cancer treatment and survivorship statistics, 2016. *CA Cancer J Clin* 2016, 66:271–289
2. RAJEEV L, PATIDAR R, BABU G, SURESH BABU MC, LOKESH KN, PATIL OKALY GV. A rare case of primary synovial sarcoma of lung. *Lung India* 2017,34:545–547
3. KIM JH, PARK HS, HEO SJ, KIM SK, HAN JW, SHIN KH ET AL. Differences in the efficacies of pazopanib and gemcitabine/docetaxel as second-line treatments for metastatic soft tissue sarcoma. *Oncology* 2019, 96:59–69
4. DENNISON S, WEPPLER E, GIACOPPE G. Primary pulmonary synovial sarcoma: A case report and review of current diagnostic and therapeutic standards. *Oncologist* 2004, 9:339–342
5. HERZOG CE. Overview of sarcomas in the adolescent and young adult population. *J Pediatr Hematol Oncol* 2005, 27:215–218
6. LADANYI M, ANTONESCU CR, LEUNG DH, WOODRUFF JM, KAWAI A, HEALEY JH ET AL. Impact of SYT-SSX fusion type on the clinical behavior of synovial sarcoma: A multi-institutional retrospective study of 243 patients. *Cancer Res* 2002, 62:135–140
7. VLENTERIE M, LITIÈRE S, RIZZO E, MARRÉAUD S, JUDSON I, GELDERBLOM H ET AL. Outcome of chemotherapy in advanced synovial sarcoma patients: Review of 15 clinical trials from the European Organisation for Research and Treatment of Cancer Soft Tissue and Bone Sarcoma Group; setting a new landmark for studies in this entity. *Eur J Cancer* 2016, 58:62–72
8. MADABHAVI I, BHARDAWA V, MODI M, PATEL A, SARKAR M. Primary synovial sarcoma (SS) of larynx: An unusual site. *Oral Oncol* 2018, 79:80–82
9. BILLINGSLEY KG, BURT ME, JARA E, GINSBERG RJ, WOODRUFF JM, LEUNG DH ET AL. Pulmonary metastases from soft tissue sarcoma: Analysis of patterns of diseases and postmetastasis survival. *Ann Surg* 1999, 229:602–610
10. NILSSON G, SKYTTING B, XIE Y, BRODIN B, PERFEKT R, MANDAHL N ET AL. The SYT-SSX1 variant of synovial sarcoma is associated with a high rate of tumor cell proliferation and poor clinical outcome. *Cancer Res* 1999, 59:3180–3184
11. VEZERIDIS MP, MOORE R, KARAKOUSIS CP. Metastatic patterns in soft-tissue sarcomas. *Arch Surg* 1983, 118:915–918
12. GAERTNER E, ZEREN HE, FLEMING MV, COLBY TV, TRAVIS WD. Biphasic synovial sarcomas arising in the pleural cavity: A clinicopathologic study of five cases. *Am J Surg Pathol* 1996, 20:36–45
13. AYDOĞDU K, SAHIN F, FINDIK G, KAYA S. Pulmonary synovial sarcoma. *Asian Cardiovasc Thorac Ann* 2014, 22:92–94

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