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Case Report

Metastatic pulmonary synovial sarcoma: A double coincidence: Case report ☆,☆☆

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ABSTRACT

Synovial sarcomas are considered as one of the most aggressive neoplasms that account for approximately 8% of all soft tissue sarcomas; they are mainly localized in soft tissues of the extremities and joints and rarely occur in the thorax. In this case report, we describe a 34-year-old woman presenting a chest pain with a chest radiography showing a mass lesion occupying two-thirds of the right hemi-thorax. A malignant pulmonary tumor was suspected after CT imaging revealing a bilateral renal metastasis, and then a spindle-cell carcinoma was thought-about. The post-operative pathological analysis of the main mass confirmed the diagnosis of a pulmonary synovial sarcoma.

Introduction

The lung hosts a variety of primary and secondary malignancies. Synovial sarcoma, although extremely uncommon, can also occur in this location [1,2].

Synovial sarcoma is a mesenchymal tissue cell tumor that displays epithelial differentiation. While a primary occurrence in the pulmonary tissue is extremely uncommon, with only a few cases reported in the published literature [3]. It is even more scarce to have it associated with a bilateral renal metastasis.

A pathological examination is essential to make a diagnosis, as the clinical manifestations and imaging characteristics are frequently nonspecific and confusing [4,5]. We present a case of a primary pulmonary synovial sarcoma diagnosed in a 34-year-old patient for which computed tomography proved useful in assessing the size and extent of the tumor and the diagnosis of accompanied metastases.

Case report

We present a case of a 34-year-old woman with no prior surgical, medical or familial history, who experienced during her pregnancy respiratory symptoms such as hemoptysis, mild chest pain and wet cough treated symptomatically with

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no imaging exploration. After the delivery, the patient manifested a persistence of her symptoms. Physical examination did not reveal anything significant. Her blood investigations showed a hemoglobin level of 12.0 g/dL (normal range at 12-18 g/dL), white body counts of 9490 cells/mm^3 (normal range 4000-10,000 cells/mm^3), and platelet counts of 305,000/mm^3 (normal range 150,000/mm^3-450,000/mm^3). Her serum electrolytes, renal function tests were within normal range. Germ cell tumor markers such as lactate-dehydrogenase and ß-human chorionic gonadotropin levels were higher than the reference range except the alpha-fetoprotein which was normal.

The chest X-ray showed a large opacity in the right hemithorax (Fig. 1A). Then, the patient underwent a computed tomography with i.v. contrast medium, manifesting the presence of a 149 mm pulmonary mass in the right superior and medium lobes, well limited, hypodense, heterogeneously enhanced and delimiting areas of necrosis without bone involvement (Fig. 1B and C). The lesion came in contact with the trachea, the right stem bronchus, the SVC, the right pulmonary artery which were compressed but remained patent. It also reached the ascending aorta over <180° with loss of fatty separation line. The mass encompassed the right superior lobar artery and the superior pulmonary vein which are laminated but remained permeable. In addition, there were numerous mediastinal lymphadenopathy, pleural and pericardiac effusion of low abundance. The CT also showed 2 renal metastases, hypodense and non-enhanced after the injection of the contrast medium suggesting secondary metastases (Fig. 1D).

The patient underwent retroperitoneal exploration by way of a midline incision with bilateral simultaneous partial nephrectomy. Dissection confirmed involvement of only the cortical or capsular portion of the kidney. The estimated parenchymal sparing was 95% and 90% for the right and left kidneys, respectively.

Pathologic examination of the specimens revealed high-grade sarcomatoid malignancy (grade III) with biphasic features and bilateral negative margins. Resection of the left pulmonary lesion was done 1 month later with negative margins. A final diagnosis of biphahasic synovial pulmonary sarcoma with bilateral renal metastasis grade III was eventually made.

Based on the R0 status after surgical resection, our patient didn't receive systemic chemotherapy neither a radiation therapy.

The patient was disease free during her last follow-up visit at 12 months postoperatively.

Discussion

The term synovial sarcoma is misleading because the tumor does not emerge from the synovium and only resembles to synovial tissue on light microscopy. It appears to develop from multipotent stem cells that lack synovial differentiation and can differentiate into mesenchymal and/or epithelial structures [6,7].

In 5%-10% of all soft tissue sarcomas, synovial sarcomas most frequently occur in the deep soft tissues of the extremities [8,9]. Aside from soft tissues, synovial sarcomas have been found in almost every anatomic region, including the thoracic cavity [10–12]. Synovial sarcoma is most common between the ages of 20 and 40, but it can appear at any age. In our case, the patient is 34-year-old. There is a minor masculine predominance in the literature [8,14].

Chest X-rays are not very helpful to determine the characteristics of the tumor, including its volume, limits and location in the chest wall, pleura, or lung parenchyma. It detects calcifications in more than 25% of cases [15,16]. In our case, the chest X-ray revealed an opacity that nearly filled the entire right lung and showed no signs of calcification.

Eventual metastasis is common, with the lungs the most common site, up to 80% in some series, followed by dissemination to the bones and bone marrow. We report an uncommon pattern of metastasis involving bilateral renal involvement.

Thoracic CT scan provides a better evaluation of the tumor's site, endo- and exo-thoracic extension, and allows the identification of signs of malignancies such as heterogeneous appearance with central necrosis, pleurisy, and mediastino-pulmonary invasion [15]. Accurate disease staging is critical for appropriate patient management and necessitates the evaluation of the primary tumor as well as assessment for distant disease. The standard imaging method for evaluating soft tissue masses has been magnetic resonance imaging; however, more recently, soft tissue sarcomas have been recognized as having enhanced 18F-FDG uptake in positron emission tomography [17].

The diagnosis of pulmonary synovial sarcoma is confirmed based on pathological, immunohistochemical, and cytogenetic analysis of the tumor tissue. Morphologically, synovial sarcomas can be categorized into monophasic and biphasic subtypes. The biphasic variant is characterized by the proliferation of bland looking spindle-shaped cells, along with evidence of epithelial differentiation. Contrarily, monophasic subtypes may exclusively exhibit spindle cells or, less frequently, epithelial components [8]. A poorly differentiated form of synovial sarcoma is also recognized [18]. Furthermore, genetic and immunochemistry investigations are critical in distinguishing the monophasic form from other stromal tumors such as fibrosarcoma, hemangiopericytoma, leiomyosarcoma, and the spindle cell variant of squamous cell carcinoma [19,20].

The cytogenetic hallmark of synovial sarcoma is the t (X; 18) (p11; q11) chromosomal translocation, leading to the rearrangement of the SS18 and one of the SSX genes [13].

The cornerstone of therapy is surgery, followed by radiotherapy at the tumor location [21].

Pre-operative chemotherapy was not mentioned in any previous case studies [20], or is the place of chemotherapy for this tumor clearly specified [22].

Good prognostic factors include: Tumor diameter less than 5 cm, low mitotic index (less than 10 mitoses per 10 fields at high magnification), low proliferation index (Ki-67 < 10%), absence of tumor necrosis, and absence of residual tumor after surgical resection [8]. The prognosis is still poor despite the use of modern chemotherapeutic medicines. Five-year survival ranges from 76% to 35%, while 10-year survival ranges from 63% to 10% [23].

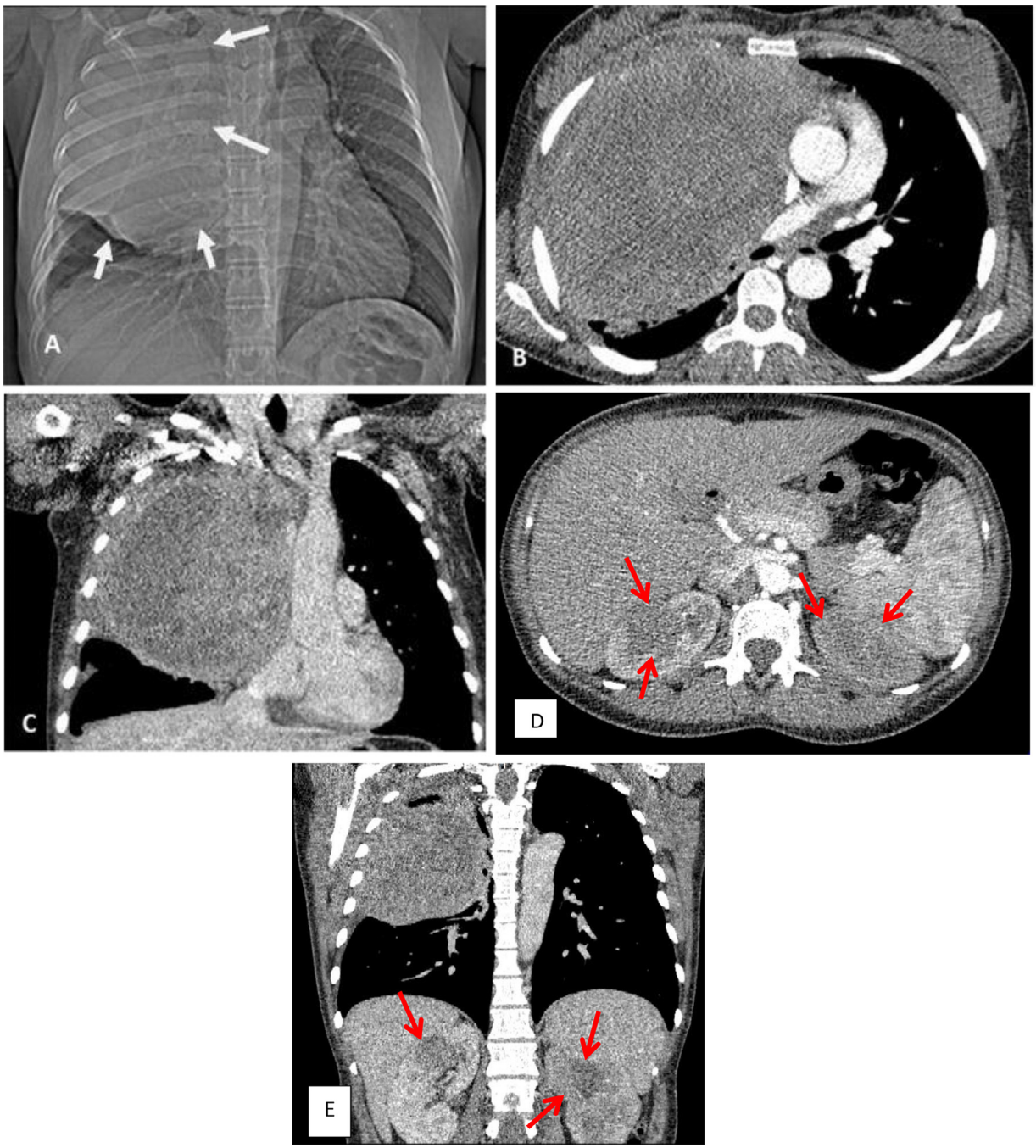

Fig. 1 – (A) Chest X-ray showed a large lump in the right thoracic cavity. (White arrows). (B,C) Axial and coronal contrast-enhanced computed tomography scans of the chest in the mediastinal parenchyma window, revealing a 149mm pulmonary mass hypodense, heterogeneously enhanced and delimiting areas of necrosis with a clear mediastinal deviation. (D,E) Axial and coronal contrast-enhanced computed tomography scan of the abdomen; showed two renal metastasizes, hypodense and non-enhanced after the injection of the contrast medium suggesting secondary metastases. (Red arrows).

Conclusion

Pulmonary synovial sarcoma is a rare tumor that can pose significant diagnostic challenges because of the uncommon location of the tumor. The main symptoms observed are chest pain, dyspnea, and cough. Radiologically, the lesion appears most frequently heterogeneous well limited with areas of necrosis on the chest scan. Pathology studies are required to confirm the diagnosis of pulmonary sarcoma in the presence of non-specific radiological images. This type of tumor requires an aggressive therapeutic approach and the progno-

sis is poor and linked to presence and location of the metastases to the quality of the surgical excision and the histological grade.

Patient consent

Written informed consent for publication was obtained from patient.

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