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Primary chest wall angiosarcoma

Primer göğüs duvarı anjiyosarkomu

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ABSTRACT

In this article, we report a unique case of angiosarcoma with multiple pleura-pulmonary metastases that involved the chest wall. Ultrasonography guided fine needle aspiration biopsy and immunohistochemical staining led to the diagnosis. Distant metastases were detected on thyroid and lymph nodes. Vascular neoplasms must be considered in the differential diagnosis of chest wall tumors.

Keywords: Fine needle aspiration biopsy; immunohistochemical staining; soft tissue tumors; ultrasonography.

ÖZ

Bu makalede göğüs duvarında yerleşim gösteren ve multipl plevra ve pulmoner metastazları yapmış nadir bir anjiyosarkom olgusu sunuldu. Ultrason eşliğinde ince iğne aspirasyon biyopsisi ve immünohistokimyasal boyama ile tanıya ulaşıldı. Tiroid ve lenf nodlarında uzak metastazlar tespit edildi. Göğüs duvarı tümörlerinin ayırıcı tanısında vasküler neoplazmlar göz önünde bulundurulmalıdır.

Anahtar sözcükler: İnce iğne aspirasyon biyopsisi; immünohistokimyasal boyama; yumuşak doku tümörleri; ultrasonografi.

Angiosarcoma is a rare tumor originating from endothelial cells of small blood vessels. It is highly malign and aggressive. Distant metastasis is usually found at the beginning of admission. Skin, liver, soft tissues, spleen, and bones are the most common sites that the tumor spreads. Lung, pleura, mediastinum, chest wall, and thyroid are the rare localizations.[1,2] The etiology may involve radiation exposure, lymphedema, and some carcinogens.

In this article, we report an asphalt worker diagnosed with primary chest wall angiosarcoma along with distant metastases mimicking other primary tumors of those tissues.

CASE REPORT

A 48-year-old male, non-smoker asphalt worker with no significant prior medical illness presented with hemoptysis (100 mL/day) and dyspnea which started a month ago. He had constitutional symptoms like fatigue and weight loss. Before admission, he was treated for pneumonia. However, his clinical signs and symptoms did not improve.

Past medical history did not include tuberculosis, malignancy, drug use or radiation exposure. However, he may have been exposed to toxic gases in his workplace during his working period.

Physical examination revealed fatigue, cachexia, and low performance status graded as 2-3 according to Eastern Cooperative Oncology Group. A few coarse crackles were audible on each hemithorax with bilateral dullness of sinuses. No palpable peripheral lymphadenopathy or hepatosplenomegaly were detected. However, on his anterior chest wall, above manubrium sterni, an immobile fluctuated mass was palpable with nearly 4x5 cm diameters. Moreover, left lobe of thyroid was hard and diffusely palpable with double size enlargement.

The hemoglobin level was 5 mg/dL and four units of erythrocyte suspension were transfused. Erythrocyte sedimentation rate and C-reactive protein were 84 mm/h and 112 mg/L, respectively. Biochemical parameters were normal. Chest X-ray showed enlargement of mediastinum, multiple coin

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lesions scattered on both hemithorax and pleura along with obliteration of both costophrenic angles. Thoracic computed tomography demonstrated the destructive mass behind sternum infiltrating retrosternal fat tissue and multiple parenchymal nodules with a mean size of 13 mm, such like metastasis with mediastinal and hilar lymphadenopathies (Figure 1a, b). A positron emission tomography scan verified the computed tomography findings by showing high 18F-fluoro-2-deoxyglucose involvement at these lesions especially for the masses on sternum (maximum standardized uptake value: 3.6) (Figure 1c) and thyroid (Figure 1d). Positron emission tomography also revealed metastases on the liver and bone marrow. Approximately 3 liters of hemorrhagic pleural fluids were aspirated on both sides showing malignant epithelial cells on cytology. Flexible bronchoscopy was unremarkable other than slight outer compression at both lung base. Pleural fluid cytology showed atypical mesothelial cell proliferation, arousing suspicion of malignant epithelial tumor most probably suggesting malignant mesothelioma. Fine needle aspiration biopsy on left thyroid lobe revealed malignant epithelial tumor again. For a definitive diagnosis, ultrasonography guided fine needle aspiration biopsy was performed on sternal mass. After the procedure, it needed pressure dressing to control bleeding. Once again, malignant epithelial tumor was detected in histopathology and cell block examination (Figure 2a, b). Immunohistochemical staining (IHS) was performed on all cytologic specimens. CD31, CD34, fligmentin and Factor VIII (FVIII) antigens were used to confirm endothelial origin of the tissues. CD31 and FVIII-related antigen expressions were shown on Figure 2c.

After the diagnosis of angiosarcoma is made, chemotherapy and radiotherapy were initiated. Despite three regimens of paclitaxel + carboplatin, the mass enlarged reaching 17x12 cm in size. As the disease was progressive despite this therapy, the regimen was switched to ifosfomide + mesna + adriamycin (IMA) with 20% dose reduction in addition to synchronous palliative thoracic radiotherapy (3000Gy). After the first regimen

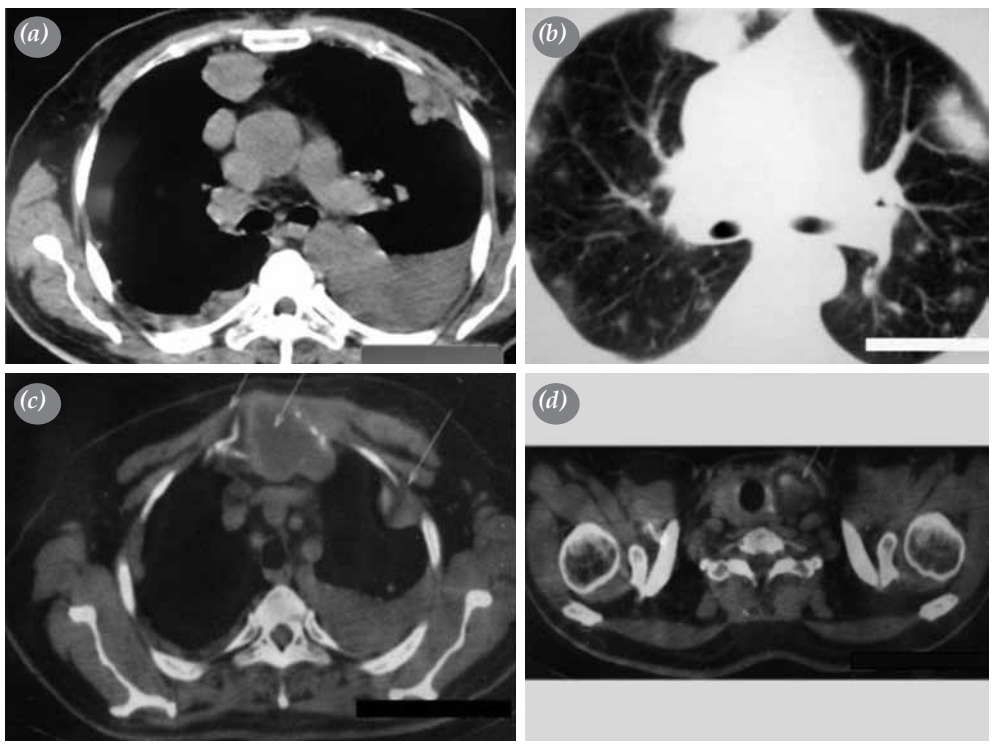

Figure 1. (a) Thoracic computed tomography showed enlargement of mediastinum with enlarged lymph nodes, bilateral pleural collection, and a subpleural opacification on left hemithorax. **(b)** Multiple coin lesions were seen in both lungs. **(c)** Positron emission tomography demonstrated mass behind sternum causing bone destruction showing high 18F-fluoro-2-deoxyglucose involvement that infiltrated retrosternal fat tissue and multiple parenchymal nodules including large mass on the left chest wall with pathological mediastinal and hilar lymph nodes. **(d)** Enlarged thyroid left lobe with high 18F-fluoro-2-deoxyglucose involvement was shown on positron emission tomography.

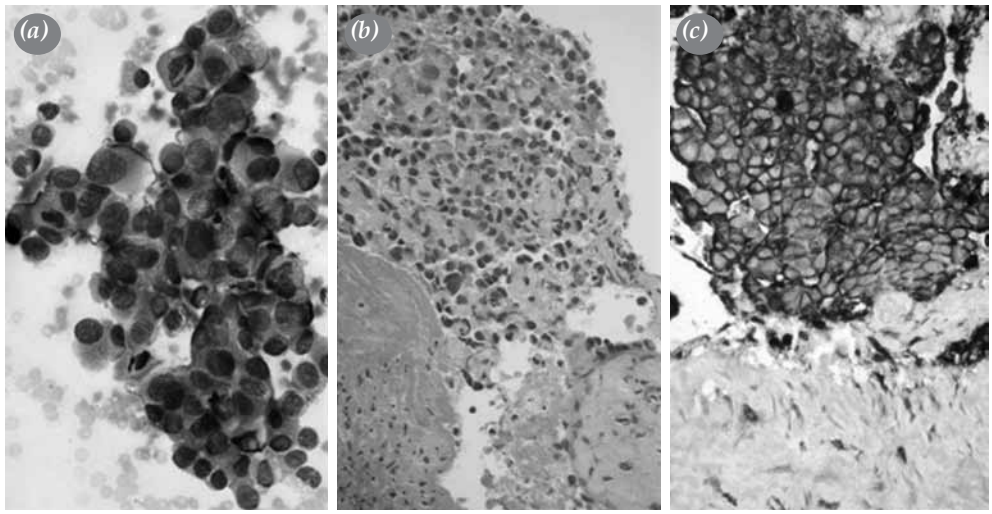

Figure 2. (a) Fine needle aspiration biopsy disclosed pleomorphic epithelioid tumor cells show gland-like structures mimicking adenocarcinoma (MGG stain x 400). **(b)** Cell block section shows an epithelioid solid tumor island in a fibrinous and hemorrhagic background (H-E x 200). **(c)** Diffuse and strong CD31 positivity in epithelioid cell tumors characteristic for angisarcoma. MGG stain: May-Grünwald-Giemsa staining, H-E: Hematoxylin and eosin.

of IMA, tumor size reduced to 6x8 cm. However, unfortunately, the patient passed away due to massive hemoptysis in the 13th month of follow-up.

DISCUSSION

Angiosarcoma accounts for 4.1% of soft tissue sarcomas. Chronic lymphedema, irradiation and exposure to vinyl chloride, thorotrast, arsenic, foreign bodies or anabolic steroids are main risk factors.[3,4] We believe that our patient may have been exposed to some of these risk factors due to the presence of irritant fumes and gases his workplace.

Bones are the most common sites for angiosarcomas.[1,2] In our patient, first we thought that the tumor originated from sternum and then invaded adjacent soft tissues. However, invasion of adjacent structures is not common for this type of tumor. Moreover, ribs and sternum were affected with distant metastases to thyroid tissue, lungs, and pleura. Therefore, a diagnosis of chest wall angiosarcoma was established.

Cytological features are extremely variable and difficult to distinguish from other sarcomas such as poorly differentiated carcinomas like mesothelioma. Common features of angiosarcoma are small clusters of cells; round, oval to spindle cell morphologies, and bloody background and single atypical cells.[5] Diagnosis of angiosarcoma depends on correlation of morphologic and clinical findings with neoplasm site, patient history and immunohistochemical findings. CD31, CD34, friend leukemia integration 1 transcription factor, vimentin, and FVIII are highly specific and sensitive markers. However, S100 protein is negative for this tumor.[6] In a recent study, two cases mimicked follicular center lymphoma.[7] Therefore, atypical lymphoid neoplasm might be misinterpreted as angiosarcoma.[8] In our patient, thyroid and sternal mass and pleural fluid cytologies showed malignant epithelial differentiation. However, endothelial origin of the tumor was only clarified by IHS. Although there were some other mesenchymal markers which could have been studied, once CD31 positivity was shown, there was no need for further evaluations.

Angiosarcoma may be accepted as an aggressive tumor among all sarcomas as it always recurs even after resection with negative margins.[9] So, the outcome with angiosarcoma is poor for those patients in whom aggressive surgery is not possible. Suggested chemotherapy generally consists of the combination of anthracyclines and ifosfamide. It usually responds well to paclitaxel or adriamycin.[10,11] According to a 14-year retrospective study including 125 patients, overall five-year survival rate of patients with angiosarcoma was 31%. Our patient was inoperable at first admission due to metastases and survived only 13 months despite chemo and radiotherapy.[10]

To conclude, vascular neoplasms should be considered when a chest wall tumor shows high vascularization and the patient had exposure to toxic

gases/fumes, and IHS should be performed for a definitive diagnosis.

Declaration of conflicting interests

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REFERENCES

1. Alexiou C, Clelland CA, Robinson D, Morgan WE. Primary angiosarcomas of the chest wall and pleura. Eur J Cardiothorac Surg 1998;14:523-6.
2. Pramesh CS, Madur BP, Raina S, Desai SB, Mistry RC. Angiosarcoma of the pleura. Ann Thorac Cardiovasc Surg 2004;10:187-90.
3. Del Frate C, Mortele K, Zanardi R, Hunsaker AR, Nikpoor N, Cibas ES, et al. Pseudomesotheliomatous angiosarcoma of the chest wall and pleura. J Thorac Imaging 2003;18:200-3.
4. Fury MG, Antonescu CR, Van Zee KJ, Brennan MF, Maki RG. A 14-year retrospective review of angiosarcoma: clinical characteristics, prognostic factors, and treatment outcomes with surgery and chemotherapy. Cancer J 2005;11:241-7.
5. Falconieri G, Bussani R, Mirra M, Zanella M. Pseudomesotheliomatous angiosarcoma: a pleuropulmonary lesion simulating malignant pleural mesothelioma. Histopathology 1997;30:419-24.
6. Boucher LD, Swanson PE, Stanley MW, Silverman JF, Raab SS, Geisinger KR. Cytology of angiosarcoma. Findings in fourteen fine-needle aspiration biopsy specimens and one pleural fluid specimen. Am J Clin Pathol 2000;114:210-9.
7. Rongioletti F, Albertini AF, Fausti V, Cinotti E, Parodi A, Fraitag S. Pseudolymphomatous cutaneous angiosarcoma: a report of 2 new cases arising in an unusual setting. J Cutan Pathol 2013;40:848-54.
8. Requena L, Santonja C, Stutz N, Kaddu S, Weenig RH, Kutzner H, et al. Pseudolymphomatous cutaneous angiosarcoma: a rare variant of cutaneous angiosarcoma readily mistaken for cutaneous lymphoma. Am J Dermatopathol 2007;29:342-50.
9. Enzinger FM, Weiss SW, editors. Soft Tissue Tumors. 3rd ed. St. Louis: Mosby; 1995.
10. Hart J, Mandavilli S. Epithelioid angiosarcoma: a brief diagnostic review and differential diagnosis. Arch Pathol Lab Med 2011;135:268-72.
11. Skubitz KM, Haddad PA. Paclitaxel and pegylated-liposomal doxorubicin are both active in angiosarcoma. Cancer 2005;104:361-6.