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Case report

Primary mediastinal angiosarcoma: A rare observation in a patient with 8-year-survival

M. MLIKA[1], A. BERRAIES[1], M.S. BOUDAYA[2], A. HAMZAOUI[2], F. EL MEZNI[1]
[1] Department of Pathology, Abderrahman Mami Hospital; [2] Department of Pulmonoly, Abderrahman Mami Hospital; [**] Department of Thoracic Surgery, Abderrahman Mami Hospital, Research Unit: 02/UR/08/08, University of Medicine, Tunis El Manar, Tunisia

Key words

Mediastinum • Angiosarcoma • Hemangioendothelioma • Surgery

Summary

Background. Vascular tumours of the mediastinum are rare, accounting for 1-2% of all mediastinal tumours in this location. Angiosarcomas are most often encountered as sporadic lesions, typically in the scalp or face of elderly patients. However, they can occur in any anatomic site. Mediastinal angiosarcomas (MA) are very rare with less than 50 cases reported.
Case report. The authors describe the case of a 38-year-old woman whose past medical history was consistent for a MA that was diagnosed in 2003. This tumour was treated by complete surgical resection followed by radiation therapy and chemotherapy. Diagnosis was based on histologic examination. In 2011, the patient presented a pleural localisation of the angiosarcoma and died one month after admission, 8 years after diagnosis of the MA.
Conclusion. MA is a very rare tumour causing a diagnostic dilemma. Clinical and radiologic findings are non-specific, and final diagnosis is based on histologic examination. The case described is unusual considering the long period of survival, which may be explained by the treatment modalities associating complete surgical resection, chemotherapy and radiation therapy.

Background

Primary mediastinal sarcomas are very rare, representing 2-8% of the malignant mediastinal tumours and 1.4% of soft tissue sarcomas [1 2]. These sarcomas are characterized by their poor behaviour that is essentially related to their development next to vital organs. Angiosarcomas are very rare, accounting for 1-2% of all mediastinal tumours. These tumours cause a diagnostic dilemma and necessitate multi-modal treatment procedures. We describe a new case of mediastinal angiosarcoma that us unusual due to the long-term survival of the patient. This case was documented by radiologic and histologic features.

Case presentation

The authors describe the case of a 38-year-old woman whose past medical history consistent of cholecystectomy performed in 2009 and a mediastinal angiosarcoma diagnosed in 2003. This tumour was treated by a complete surgical resection followed by radiation therapy and chemotherapy. Diagnosis was based on histologic examination. Microscopic features consisted in a tumour composed of multiple epithelioid cells with atypical and focally mitotic nuclei. Focal areas of necrosis were present (Fig. 1a). In June 2011, the patient presented with acute respiratory distress. Laboratory tests and tumoural markers (ACE, CA 27-29, CA 125) were within normal values. Physical examination was normal. A CT scan showed a right pleural effusion with pneumothorax. Many lesions of fibrosis were also noticed in the right lung (Fig. 2). Fine needle aspiration of the liquid was performed, and microscopic study concluded a mesothelial localisation of a sarcoma. Because of the increase of the dyspnoea, a talcage associated with surgical biopsy of the parietal pleural nodules through thoracoscopy was performed. Microscopic examination showed tumour proliferation consisting of sheets of epithelioid

Correspondence
Mona Mlika, Department of Pathology, Abderrahman Mami Hospital, Tunisia - Tel. +216 98538862 - Fax +216 71710780 - E-mail mlika.zorgati.mona@hotmail.com

Fig. 1. **a**. Microscopic findings of the primary mediastinal angiosarcoma: Multiple anastomosing vascular structures lined by large rounded cells (HE x250). **b**. Pleural localisation of the mediastinal angiosarcoma: large rounded cells arranged in rudimentary vascular channels (HE x250). **c**. Immunohistochemical findings: cytoplasmic expression of CD 31 antigen by tumour cells (HE x400).

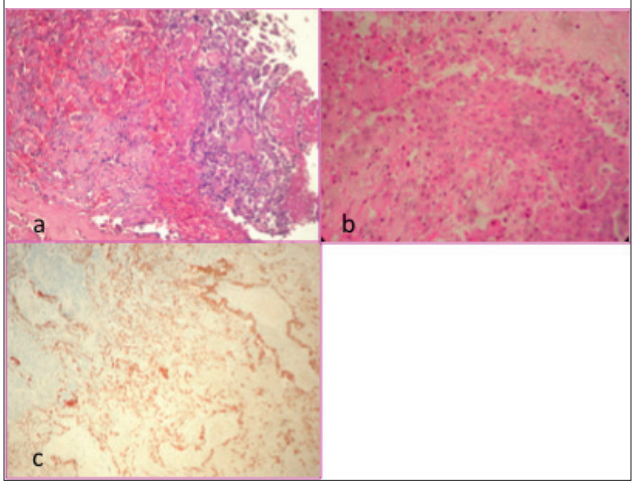
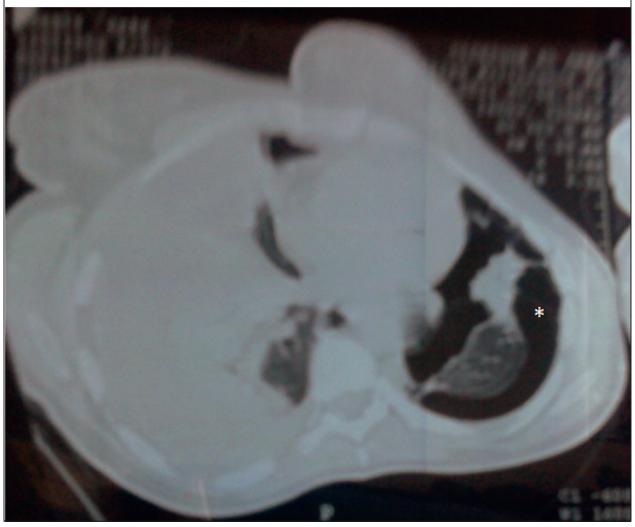


Fig. 2. CT findings: Pleural effusion with pneumothorax (asterisk).



tumour cells with atypical nuclei (Fig. 1b). Immunohistochemical study showed expression of vimentin, CD 31 and CD34 by tumour cells, revealing their vascular nature (Fig. 1c). A diagnosis of pleural localisation of angiosarcoma was retained. The evolution of the patient was marked by the increase of the dyspnoea, and exacerbation of thoracic chest pain followed by death one month after admission and 8 years after diagnosis of the mediastinal angiosarcoma.

Discussion

Vascular tumours of the mediastinum are rare, accounting for 1-2% of all tumours in this location [1]. Mediastinal sarcomas are very rare. In our department, 16 mediastinal sarcomas have been diagnosed over a 16-year-period, and this is the first case of angiosarcoma diagnosed during the same period. Angiosarcomas are most often encountered as sporadic lesions, typically in the scalp or face of elderly patients. However, they can occur in any anatomic site, including the deep soft tissue, breast, visceral organs and bone [3]. Important predisposing conditions have been reported including radiation exposure, chronic lymphoedema, exposure to toxins such as vinyl chloride or thorotrast, or foreign bodies [2]. Primary angiosarcomas of the mediastinum are rare with less than 50 cases reported in the English literature. Most tumours were located in the anterior mediastinum [4 5]. Chest pain is the main presenting symptom [3]. The patient age ranged from 25 to 62 years. Radiologically, tumours presented as a nonspecific anterior mediastinal mass. Diagnosis is based on histologic examination, which may show typical features ranging from low-grade neoplasms with definitive vasoformation to high grade lesions with a more solid growth pattern and vast areas of necrosis. Immunohistochemical study demonstrates a vascular phenotype with immunoreactivity for factor VIII-related antigen, CD31 and CD34. Many differential diagnoses must be ruled out such as haemangioma or carcinoma, but the distinction is quite easy when based on histologic and immunohistochemical features. The most relevant mimickers of MA is epithelioid haemangioendothelioma, which is a vascular neoplasm of low to intermediate malignant potential that follows a nonaggressive clinical course. The distinction between both neoplasms is based only on microscopic findings without immunohistochemical study, which is unable to make any distinction [4-6]. Another important consideration is the possibility that angiosarcoma may occur in association with a mediastinal germ cell tumour, which is considered as a sarcomatous transformation of these tumours and with very poor prognosis. This distinction is dependent on thorough sampling of the specimen [7 8]. There is no consensus regarding management of MA due to its rarity, but surgical resection represents a mainstay. Thoracotomy has been used in almost all cases, but some authors report that, in patients with limited disease, endoscopic transthoracic approaches can reduce approach-related soft-tissue morbidity and facilitate recovery by preserving normal tissues of the chest wall [9]. In our opinion, since MA is a malignant disease, it is rarely localised, resulting in the necessity of thoracotomic approaches. Other authors have recommend radical excision followed by post-operative radiotherapy, especially in cases where the tumour has been partially excised as the treatment of choice [10]. It is extremely difficult to draw any conclusions about the value of adjuvant radiotherapy and/or chemotherapy, mainly because the numbers of patients are small. In our case, we can only suppose that the adjuvant therapy is implicated in the long survival of the patient. Many authors have reported that angiosarcomas of the

mediastinum seem to pursue a less aggressive clinical course [3]. The 5-year overall survival has been estimated to be 43%. On the other hand, angiosarcomas of the liver, bone and heart have a particularly poor outcome, with 5- year survival rates ranging from 0-36% [3]. The present case is unusual due to the 8-year survival, which may be explained by the efficacy of combined radiotherapy and chemotherapy. In addition, the occurrence of mediastinal metastases is relatively rare.

Conclusion

Mediastinal angiosarcoma is a very rare tumour causing a diagnostic dilemma, especially with haemangioendothelioma and carcinoma. Clinical and radiological findings are non-specific, and final diagnosis is based on histologic examination. There is little consensus regarding treatment modalities because of its rarity of the cases, although radical excision followed by post-operative radiotherapy appears to be the treatment of choice. The case described herein is unusual due to the long period of survival, which may be explained by the treatment modalities consisting in complete surgical resection, chemotherapy and radiation therapy.

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