Case Report

Integrative Pulmonary Medicine

The Cerebral Angiosarcoma of Pleuro-Pulmonary Origin: A Rapid Evolution and a Dark Prognosis of An Exceptional Localization

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Abstract

Introduction: Angiosarcoma is a rare malignant lesion apparently having its origin in the vasculature or precursor cells of the vessels. It comprises less than 1% of all sarcomas and may occur in any location of the body. It has predilection for skin and soft tissue. Brain metastasis from this lesion represents very extremely rare presentation.

Clinical case: We report the case of a 54-year old woman who complained of thoracic pain with subsequent haemoptysis and rapid deterioration of the general state.

Thoracic CT scan showed right pleural nodular thickening associated with ipsilateral pleural effusion with diffuse nodular and micro nodular infiltrate of both lungs. Abdominal CT scan showed a left adrenal mass. Forty eight hours later the patient presented a rapid deterioration of her neurological state. Cerebral CT scan showed a large left frontal-temporal osteolytic lesion. Decompressive craniectomy was necessary. Histological examination concluded angiosarcoma

Conclusion: Angiosarcoma is a rare malignant sarcoma with local recurrence and distal metastasis. Radical surgery is the most useful method for treating this disease. Combination treatment based on chemotherapy is strongly recommended for advanced angiosarcoma, however, there is no standard chemotherapy regimen at present.

Keywords: Angiosarcoma, Lung, Metastasis

Introduction

Angiosarcoma is a rare malignant tumor apparently having its origin in the vasculature or precursor cells of the vessels. Angiosarcoma accounts for a very small proportion of all vascular tumor and comprises less than 1% of all sarcomas. Although Angiosarcoma may occur in any location of the body, it has a decided predilection for skin and superficial soft tissue. Brain metastasis from Angiosarcoma is exceedingly rare.

Clinical Case

We report the case of a 54-year-old woman with no particular pathological history who was complaining of right basic thoracic pain that had been evolving for a month and which was initially neglected. The evolution was marked after a few days by the installation of a haemoptysis of average abundance associated with a rapid deterioration of her general state without fever, for which the patient was hospitalized in the department of pneumology where a thoracic CT scan was realized showing a right pleural thickening associated with ipsilateral pleural effusion of average abundance, nodular and diffuse nodular infiltrate of the lungs (Figure 1). Abdominal CT shows only a left adrenal mass. Fort eight hours after hospitalization the patient started with acute holocranial headache and rapid neurological deterioration. Her Glasgow coma scale was estimated at 8/15. Cerebral CT scan shows a voluminous left fronto-temporal osteolytic and heterogeneous lesion with a

significant mass effect with early temporal engagement (Figure 2). The patient underwent an emergency decompressive craniectomy with partial excision of a voluminous left vascularized fronto-parietal lesion that was hemorrhagic. Histological examination revealed vascular channel-like structure with vague lumen formations by atypical polygonal or spindle-shaped neoplastic cells (Figure 3). Immunohistochemically, the neoplastic cells are positive for FLI-1 and CD31 (Figure 4). These features associated with positivity to mesothelin and fibulin-3 contributed to angiosarcoma of pleuro-pulmonary origin.

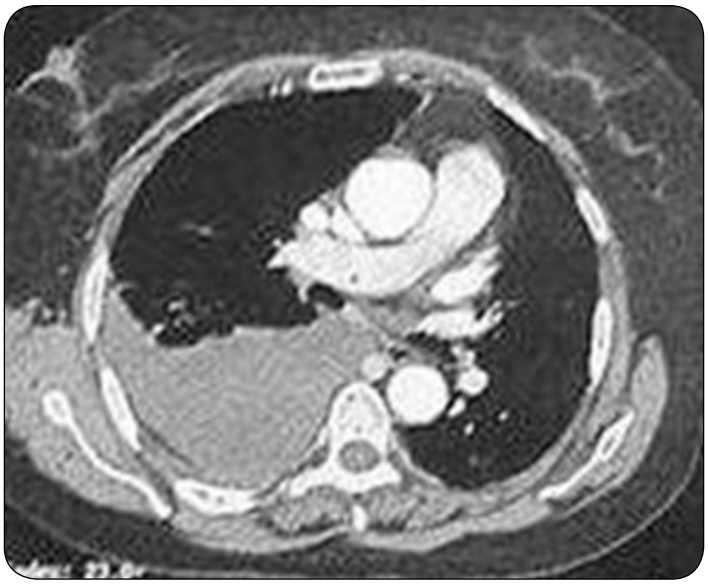

Figure 1: Thoracic CT scan showing right pleural thickening associated with ipsilateral pleural effusion of moderate abundance, nodular infiltrate and diffuse nodular micro-nodule of both lungs.

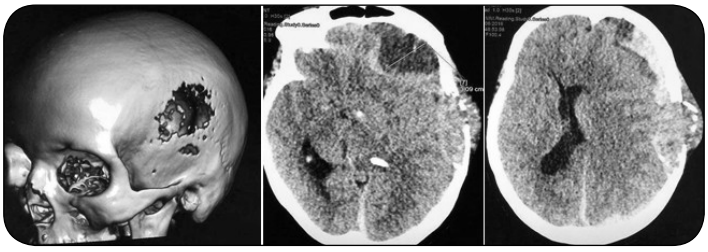

Figure 2: Cerebral CT scan showing voluminous left fronto-temporal osteolytic lesion with a significant mass effect with early temporal engagement.

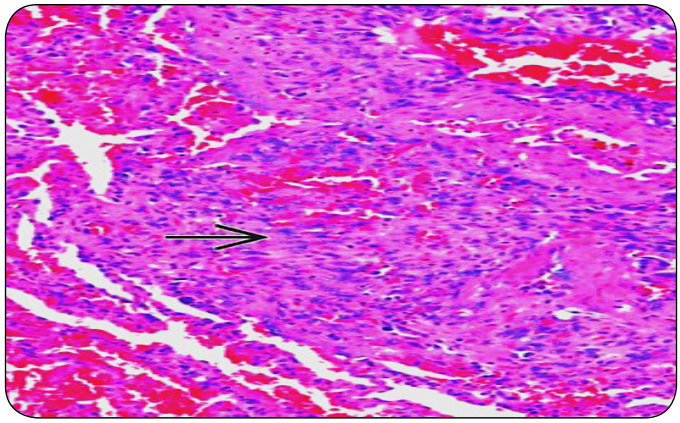

Figure 3: Histological examination showing vascular channel-like structure with vague lumen formations by atypical polygonal or spindle-shaped neoplastic cells (black arrow).

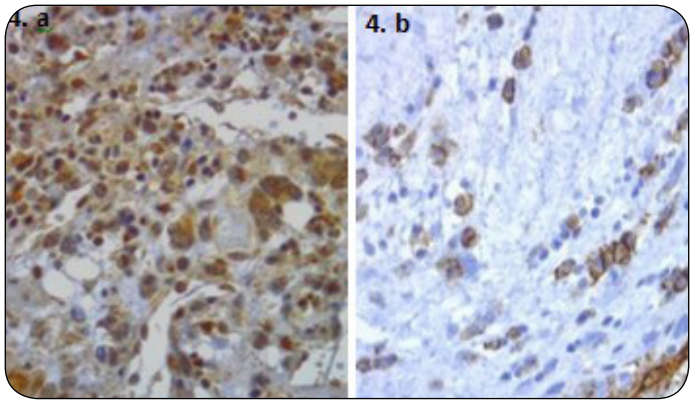

Figure 4: Immunohistochemistry showing positivity of neoplastic cells for FLI-1 (4. a) and CD31 (4. b).

Discussion

Angiosarcomas are rare malignant sarcomas derived from vascular endothelial cells, and accounting for 1–2% of all soft-tissue sarcomas [1]. Angiosarcoma seems to have a predilection for the skin in either the head or the neck. However, the tumor less commonly originates from the deep soft tissue. Angiosarcomas, regardless of their sites of origin, are particularly likely to metastasize to the brain. Other frequent sites for metastatic spread include the bone, liver, and lymph nodes [2]. In our two patients, the primary lesions were in the lung.

Angiosarcomas are subdivided into cutaneous angiosarcoma, lymphoedema-associated angiosarcoma, radiation-induced angiosarcoma, primary-breast angiosarcoma, and soft-tissue angiosarcoma, and most reports include several angiosarcoma subtypes [3].

Most angiosarcomas arise spontaneously, but there are a few reports of malignant transformation with in pre-existing benign vascular lesions [4]. Several well described risk factors exist. Chronic lymphoedema of any origin is associated with the development of angiosarcoma; a phenomenon known as Stewart-Treves syndrome. Lymphoedema is one causal aetiological factor in the development of breast angiosarcomas after treatment for breast cancer [5]. Lymphoedema caused by Milroy's disease and chronic infections, such as filariasis [3], have likewise been linked to the development of angiosarcomas.

The diagnosis of angiosarcoma mainly depends on the pathology. Under the microscope, pleomorphic and malignant endothelial cells are apparent in typical angiosarcoma tissues. In areas that are well differentiated, abnormal endothelial cells form functioning vascular sinusoids continuous with normal vascular channels. However, in areas of poor differentiation, the malignant endothelial cells form continuous sheets, usually with necrosis and hemorrhage [3]. As it is difficult to diagnose angiosarcoma by its morphology, Immunohistochemistry plays an important role in confirming the diagnosis. Typically, endothelial markers, including CD34, CD31, von Willebrand factor, Ulex europaeus agglutinin 1 and vascular endothelial growth factor are expressed [6]. Von Willebrand factor, Ulex europaeus agglutinin 1 and CD31 are the most useful markers in poorly-differentiated cases [7].

There are no symptoms specific to pulmonary angiosarcoma, as the presentation is similar to symptoms found in all lung cancers. Reported chest radiograph findings ranged from solitary lesions to multiple nodular densities, with or without pleural effusion. Early diagnosis of primary pulmonary angiosarcoma is uncommon because of the non□specific respiratory manifestations and consequent low index of suspicion. Definitive diagnosis is made on the basis of histopathological and immunohistochemical findings.

There is no standard treatment regimen specifically for pulmonary epithelioid angiosarcoma. Surgical resection, radiation, and chemotherapy have all been attempted. Surgery has been the mainstay for locally confined disease [8]. Several previous studies have shown that angiosarcoma is radiosensitive [9]. Chemotherapy has also been reported to be effective. Two chemotherapeutic combinations have demonstrated partial and full effects: doxorubicin/ifosfamide and docetaxel/gemcitabine [10]. Systemic administration of high doses of recombinant interleukin 2 also seems to have been effective [11]. The prognosis of Angiosarcoma remains poor. Only 12% of patients survived for 5 years or longer. Usually both recurrence and metastasis are noted within 2 years of diagnosis [12].

Conclusion

Angiosarcoma is a rare malignant sarcoma that is prone to local recurrence and distal metastasis. Radical surgery is the most useful method for treating this disease. Combination treatment based on chemotherapy is strongly recommended for advanced angiosarcoma, however, there is no standard chemotherapy regimen at present.

Conflicts of Interest

The authors declare that there are no conflicts of interest including financial, consultant, institutional and other relationships that might lead to bias or to a conflict of interest.

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