ISSN No: 2456-2156

ANGIOSARCOMA: A Rare Cause of Pleural Malignancy

Mariam El yahiyaoui[1], Laila Herrak[1], Kaoutar Znati[2], Najoua El Khamlichi[1], Mustapha El Ftouh[1], Leila Achachi[1]
[1]Department of Pneumology, Ibn Sina Hospital, university Mohammed V, Rabat, Morocco,
[2] Department of Pathology, Ibn Sina Hospital, university Mohammed V, Rabat, Morocco
Correspondent author: Mariam EL YAHIYAOUI

Abstract:- Angiosarcoma is a rare tumor, developed from the endothelium of blood or lymphatic vessels with exceptional pleural localization. The clinical and radiological aspects are not very specific; its diagnosis is essentially histological. We report a case of pleural angiosarcoma in a 43-year-old woman with history of chest pain dry cough, dysnpea and weight loss, Histological study of the pleural biopsy confirmed the diagnosis. Through this observation, we propose a focus on this extremely rare location with a poor prognosis.

Keywords:- *Pleural Effusion, Angiosarcoma, Tumor.*

I. INTRODUCTION

Angiosarcomas are a subtype of soft tissue sarcomas which are aggressive, malignant endothelial cell tumors of vascular or lymphatic origin [1]. It can arise from any part of the body with skin and soft tissue being the most common sites involved. It is a rare malignant tumor which accounts for less than 1% of all sarcomas and generally 2% to 3% of all soft tissue tumors are vascular sarcomas [2], pleural localization of angiosarcoma is exceptionnel.

The clinical and radiological signs are not specific and only anatomopathology allows a formal diagnosis. His prognosis is poor in the short term. We report a case of pleural effusion that reveals angiosarcoma

II. CASE REPORT

A 43 years old women, with no notable pathological history, she had felt chest pain, dry cough, and rapidly progressive dyspnea evolving, since four weeks ago before her admission, in a context of deterioration of the general state. The clinical examination, on her arrival, found respiratory rate at 30 cycles/min, heart rate at 110 beats/min with a left fluid effusion syndrome on pleuropulmonary examination.

The chest X-ray shows homogeneous opacity occupying the totality of left thoracic feld with the presence of signs of mediastinal discharge in the right side (Figure 1). A pleural biopsy was performed with evacuation of 2 L of serohematic fluid, the biochemical and cytobactiological study found an exudative fluid with a protein level of 40g/L and a 100% lymphocyte count with a negative germ culture, pleural biopsy was inconclusive, the blood biology on admission was normal.

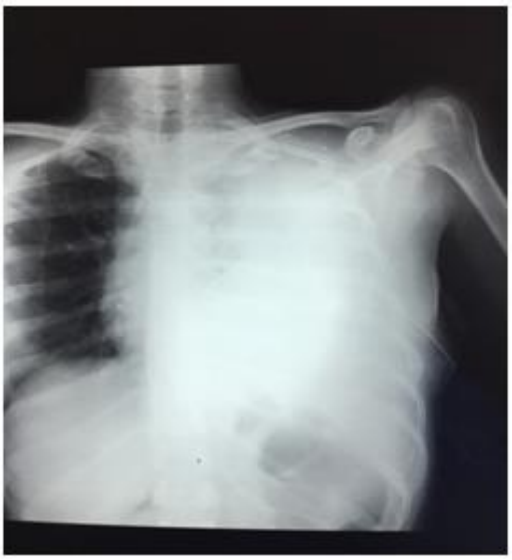


Figure 1: chest X ray showing a homogeneous opacity taking the whole hemi thoracic left feld and pushing the mediastinum towards the contralateral side.

A thoraco - abdomnopelvic computed tomography scan (figure 2-3) performed at the thoracic level showed a left pleural effusion of great abundance, with passive atelectasis and diffuse and nodular pleural thickening with individualization of a homolateral, tissue, parietal pleural nodule, measuring: 23*25*29 mm and The individualization of a pulmonary process of the ventral segment of the left upper lobe, heterogeneously enhanced, measuring: 39*62*31mm. The abdominopelvic stage was without abnormalities thus eliminating an extra thoracic origin.

ISSN No: 2456-2156

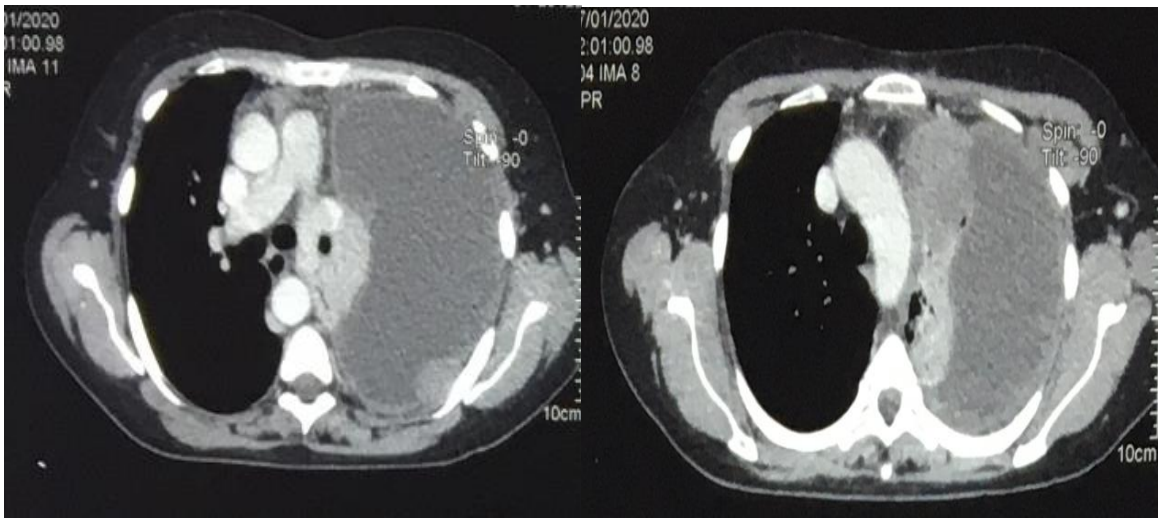
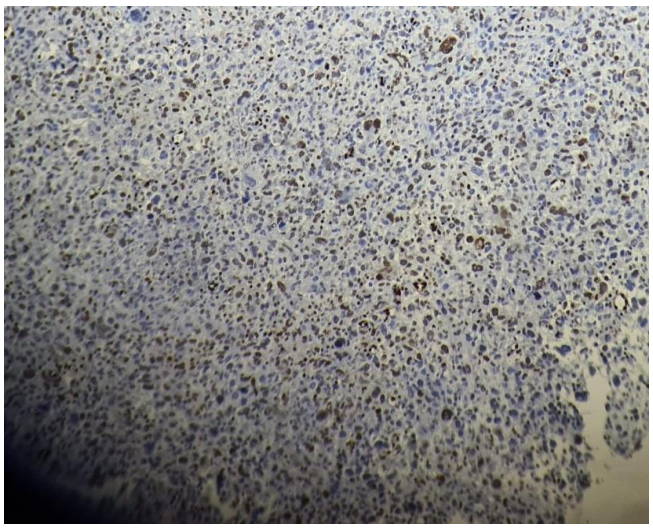


Figure 2 3: left pleural effusion of great abundance, with passive atelectasis and diffuse and nodular pleural thickening with individualization of a homolateral, tissue, parietal pleural nodule, and a pulmonary process of the ventral segment of the left upper lobe

Bronchial fibroscopy showed an extrinsic compression over the entire bronchial tree with narrowed left lower lobar orifices and the intersegmental spur of the culmen thickened. The biopsy of the thickened spurs and the study of the bronchoalveolar lavage fluid came back inconclusive.

In view of the negativity of the previous examinations, a Video-assisted thoracoscopy was performed and biopsy was done: the histological study(figure 4) found a malignant tumor proliferation of compact architecture, made up of large, pleomorphic cells with large nuclei, with angular contours, elongated, with dense and heterogeneous chromatin and highly nucoled and basophilic cytoplasm. Mitosis patterns are numerous. The tumor is traversed by a fine capillary network. Immunohistochemical analysis shows the expression by these tumor cells of the CD34 antigen (figure 5), and anti-vimentin which confirms the endothelial nature of tumor cells necessary for a diagnosis of certainty with the negativity of epithelial markers (anticytokeratin 7.20 antibodies, AE1/AE3, anti TTF1 anti CK5/6 antibodies thus confirming the diagnosis of angiosarcoma.



In view of the rapid deterioration of the general condition preventing any surgery and contraindicating any chemotherapy or radiotherapy, and at the request of the family, the patient was discharged from the hospital.

III. DISCUSSION

Angiosarcoma is a rare tumor (less than 1% of all sarcomas), developed from the endothelium of blood or lymphatic vessels, whose preferred location is cutaneous, but there are several isolated reports in the literature of mammary, visceral (liver, lung, heart, pelvis and retroperitoneum) and bony localizations [3]. Pulmonary localization is very rare, pleural localization is exceptional. An association between angiosarcoma and irradiation, environmental carcinogens (vinyl chloride, thorotrast...), or foreign bodies are frequently found. In the reported case, no specific promoting factors have been found.

Epidemiologically, the mean age at diagnosis is 52 years, and contrary to our observation, two thirds of the cases are predominantly male [4].In a large series of 161 patients, the median survival at five years is 43%, showing the aggressiveness of the tumor [5].

The clinic is not specific; hemoptysis is the most frequently reported symptom. Dyspnea, cough, chest pain, significant weight loss [6] are also noted (such is the case of our patient).

Imaging of primary pulmonary angiosarcoma is not very specific and varies depending on whether it is localized or diffuse. Compared to patients with solitary lesions, patients with multiple lesions had a poorer prognosis due to the rapid progression of multiple lesions and less effective clinical treatment [7]. Our patient had an atypical computed tomography presentation: a lung mass with nodular pleural thickening and pleural effusion of great abundance.

ISSN No: 2456-2156

Due to the non-specific respiratory manifestations, early diagnosis of pulmonary angiosarcoma remains very rare. Definitive diagnosis is established on the basis of histopathological and immunohistochemical findings. The conventional histology technique alone does not always allow a diagnosis of certainty. The use of special techniques such as immunohistochemestry makes it possible to highlight the cellular and functional characteristics of these tissues.

The choice of technique depends on the degree of differentiation of the angiosarcoma, in the case of well-differentiated angiosarcoma the conventional technique allows identification of vascular structures and therefore may be sufficient on its own to make the diagnosis[8].

In the case of poorly differentiated or undifferentiated angiosarcoma, immunohistochemistry is necessary to look for specific markers such as CD31, CD34, factor VIII and vimentin that confirm the endothelial nature of the tumor cells[9].

The prognosis of patients with primary pulmonary angiosarcoma appears to be poor. There is no specific standard treatment regimen, treatment with radiotherapy and recombinant interleukin-2, chemotherapy using ifosfamide-doxorubicin combination, pneumonectomy, lobectomy and radiotherapy have been used as treatment options with variable results but the outcome remains poor [10-11].

IV. CONCLUSION

Angiosarcoma is a rare malignant vascular tumor and its clinicopathological characteristics are not well known. Its association with pleural involvement is exceptional. Definitive diagnosis is established by histopathological and immunohistochemical findings. To date, there is no codified treatment. Surgical resection, radiotherapy and chemotherapy have all been attempted; however, these tumors have a very aggressive clinical course and a poor prognosis.

Competing interests

The authors declare no competing interest.

Authors' contributions

All authors have actively contributed to the writing and editing of this article. All the authors have read and agreed to the final manuscript.

REFERENCES

[1]. Robin Young J, Nicola Brown J, Malcolm Reed W, David Hughes, Penella Woll J. Angiosarcoma. Lancet Oncol. 2010;11(10):983-91.

[2]. Hawari M, Mohamad S. A Case of Primary Pulmonary Angiosarcoma with Pleural Metastasis. Ann Clin Case Rep. 2019; 4: 1659

[3]. Lucas DR. Angiosarcoma radiation-associated angiosarcoma and atypical vascular lesion. Arch Pathol Lab Med 2009;133: 1804—9..la

[4]. Kurian KC, et al. Primary cardiac angiosarcoma: case report and review of the literature. Cardiovasc Pathol 2006;15:110—2

[5]. Fayette J, Martin E, Piperno-Neumann S, et al. Angiosarcomas, a heterogeneous group of sarcomas with specific behaviour depending on primary site: a retrospective study of 161 cases. Ann Oncol 2007;18:2030—6

[6]. L. Herrak S. Alaziz A. Benosmane Service de Chirurgie Thoracique, CHU Ibn Sina, Rabat, Maroc. Revue des Maladies RespiratoiresVol 24, N° 1 - janvier 2007pp. 73-76

[7]. Ren Y, Zhu M, Liu Y, Diao X, Zhang Y. Primary pulmonary angiosarcoma: three case reports and literature review: primary pulmonary angiosarcoma. Thorac Cancer. (2016) 7:607–13. 10.1111/1759-7714.12376

[8]. Darré, T., Amégbor, K., Omorou, M. et al. Évaluation de l'apport de l'immunohistochimie dans le diagnostic des angiosarcomes. J Afr Cancer (2013) **5,** 37–41.

[9]. Coindre JM, Terrier P, Guillou L, et al Predictive value of grade for metastasis development in the main histologic types of adult soft tissue sarcomas. A study of 1,240 patients from the French Federation of Cancer Centers Sarcoma Group. Cancer(2001) 91:1914–26.

[10]. Ayabe T, Tomita M, Chosa E, Kawagoe K, Nakamura K. Pneumonectomy of Primary Pulmonary Angiosarcoma with Malignant Effusion and Intrapleural Hypotonic Hyperthermic Chemotherapy: Case Report and Review of the Literature. J Cancer Therapy. 2015;6(2):227-36.

[11]. Wilson R, Glaros S, Brown RK, Michael C, Reisman D. Complete Radiographic Response of Primary Pulmonary Angiosarcomas Following Gemcitabine and Taxotere. Lung Cancer. 2008;61(1):131-6.