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Primary Pleural Angiosarcoma: 2 Report Cases

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Summary:
Angiosarcoma is a rare, severely cancerous tumor that develops from the endothelial cells of tiny blood arteries. They typically affect the liver, breast, deep soft tissues, and skin. The only case reports of pleural angiosarcomas in the medical literature are extremely rare.
The clinical and radiological aspects are not very specific; its diagnosis is essentially histological.
Immunohistochemistry is valuable in making the diagnosis, showing negative reactivity for mesothelial markers and positivity for vascular markers.
We report 2 cases of pleural effusion that reveals angiosarcoma.
Key words*: Angiosarcoma, malign, pleural effusion.*

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I. Introduction:

Angiosarcomas are uncommon malignancies of the vascular endothelial origin. It includes about 1% of all soft tissue malignancies and most commonly involves skin, soft tissue, liver, bone, breast, and spleen **[1]**. They are extremely rare in the pleura and other serous membranes like the pericardium and peritoneum **[2].** We report 2 cases of pleural effusion that reveals angiosarcoma.

II. Cases Report:

A 43 years old woman, 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.

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.

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 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 3), 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.

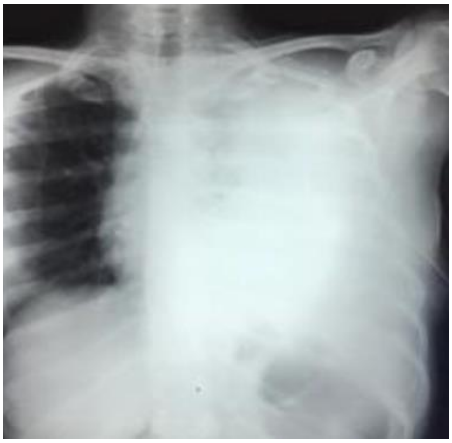

Figure 1: chest X-ray showing a homogeneous opacity taking the whole hemi thoracic left feld and pushing the mediastinum towards the contralateral side.

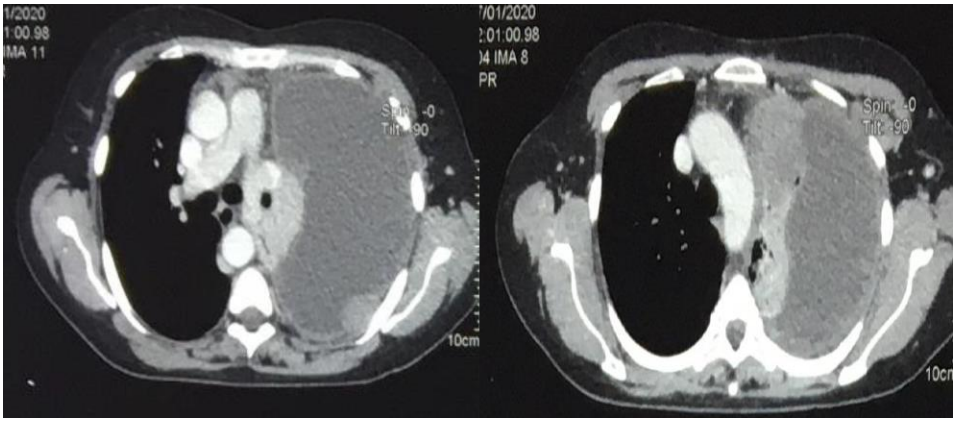

Figure 2: left pleural effusion of great abundance, with passive atelectasis and diffuse and nodular pleural thickening with individualization of a homolateral, tissue, parietal pleural nodule, and a pulmonary process of the ventral segment of the left upper lobe.

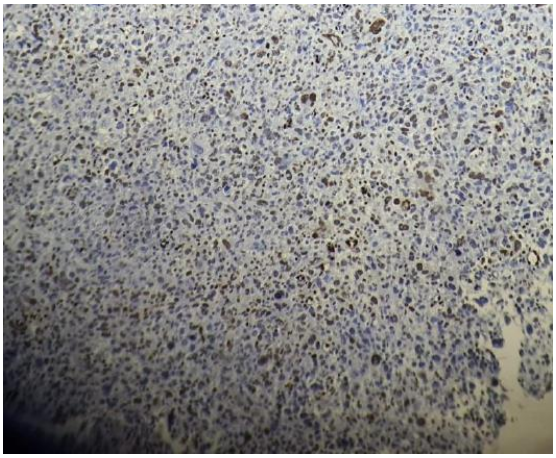

Figure 3: Immunohistochemical analysis shows the expression by the tumor cells of the CD34 antigen

Case 2:

74-year-old patient, never treated for tuberculosis, no recent tuberculosis contagion, no known diseases, no toxic habits, who has presented for three weeks with rapidly worsening dyspnoea progressing to mMRC stage IV, associated with intermittent dry cough, all evolving in a context of apyrexia and non-quantified weight loss.

Clinical examination revealed a polypneic patient at 24 cycles/min, with a right fluid effusion syndrome.

The patient had a thoraco-abdomino-pelvic CT scan, which revealed a multifocal, circumferential, irregular, nodular right pleural thickening, raised in places after injection and measuring a maximum of 28 mm at the level of the anterolateral cul-de-sac. A pleural effusion of great abundance is associated with atelectasis of the entire lung and deviation of the mediastinum towards the contralateral side. There was also a secondary hepatic lesion in segments IV and VIII (Figure 4).

Pleural puncture was performed, with 100% lymphocyte-predominant exudative fluid; pleural biopsy was subsequently performed, but was inconclusive.

Bronchial fibroscopy revealed extrinsic compression of the right middle and lower lobar with thickened spurs. Bronchial biopsy was inconclusive.

A 2nd pleural biopsy was performed with anatomo-pathological and immune-histochemical studies revealed the development of a malignant tumor with compact architecture, big, pleomorphic cells with large nuclei, angular outlines, and elongated cells with dense and heterogeneous chromatin, as well as highly nucleated and basophilic cytoplasm. Numerous mitosis patterns exist. A delicate capillary network encircles the tumor. The CD34 antigen and anti-EMA are expressed by these tumor cells (Figure 5).

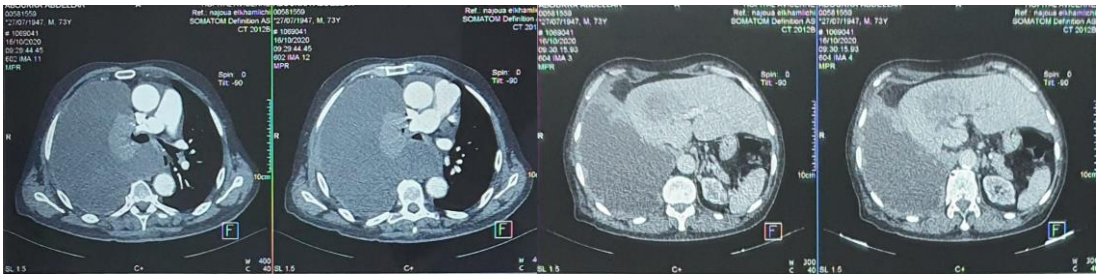

Figure 4: A. multifocal, circumferential, irregular, nodular right pleural thickening, measuring a maximum of 28 mm at the level of the anterolateral cul-de-sac associated to also a secondary hepatic lesion in segments IV and VIII. B. pleural effusion of great abundance is associated with atelectasis of the entire lung and deviation of the mediastinum towards the contralateral side.

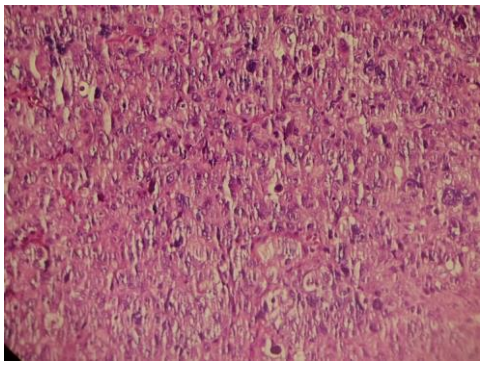

Figure 5: a malignant tumor with compact architecture, big, pleomorphic cells with large nuclei, angular outlines, and elongated cells with dense and heterogeneous chromatin, as well as highly nucleated and basophilic cytoplasm. Numerous mitosis patterns exist. A delicate capillary network encircles the tumor.

III. Discussion:

Primary angiosarcoma of the chest wall and pleura is an extremely rare and aggressive malignancy. It represents 1%–2% of all soft tissue tumors. [1, 3] It originates from the vascular endothelial cells of small blood vessels and affects retroperitoneum, subcutaneous tissue, liver, heart, spleen, and breast [4].

These tumors' etiology and pathogenesis are unclear. Trauma, persistent lymphedema, radiation, foreign bodies, thorium, viral infection, and chronic pyothorax are some of the etiological causes that have been linked. All of the documented Japanese patients [5] had a history of persistent tuberculous pyothorax before developing pleural angiosarcomas, and this appears to be the disease's strongest link [6].

Some [7] hypothesize that serosal angiosarcomas could represent a peculiar differentiation of a malignant mesothelioma along an abnormal angioblastic pathwayor alternatively a lesion arising from the native subserosal vessels or a vascular malformation. Most serosal angiosarcomas are located in the pericardium and some are thought to represent tumors extending locally from the heart and great vessels [8].

According to epidemiology, the average age at diagnosis is 52 years old, and males make up the majority of cases (around two thirds) [9].The median five-year survival rate in a large dataset of 161 individuals is 43%, demonstrating the tumor's aggressiveness. [10]

Pleuritic chest pain is the most typical symptom, followed by shortness of breath, hemoptysis, coughing, and constitutional symptoms like weight loss.

When radiologic assessment is performed, focal or diffuse pleural thickening and unilateral or bilateral pleural effusion are frequently seen, simulating the mesothelioma presentation [11]. In roughly half of the reported instances, mass lesions are present [2].

According to the literature, the lymph nodes, bone, brain, liver, spleen, adrenals, skin, oral cavity, and spinal cord are the most frequent metastatic sites [2].

Primary pulmonary localization of angiosarcoma is exceptional, and often represents a secondary localization. The primary form has been reported in only a few cases [14].

The tumor's histologic features include vascular gaps lined by atypical tumor cells that are intracytoplasmic and vasoformative spaces containing red blood cells. Malignant cells with epithelioid neoplasm are also present. A biphasic pleural tumor might also have additional differential diagnoses, such as mesothelioma and sarcomatoid cancer [11].

The diagnosis of angiosarcoma requires the presence of at least one positive immune-histochemical marker (CD31, CD34, Factor VIII, or FLI-1), which is important for confirming the diagnosis.

The most precise and sensitive immunohistochemistry marker is CD31 [12]. The epithelioid form of angiosarcoma expresses epithelial markers [12]. In a few case cases, positive cytokeratin and immunoreactivity to CAM5.2, CK7, CK8, or CK18 are documented [1].

Pleural angiosarcoma is treatable with surgery, radiation, and/or chemotherapy. Surgery may be beneficial when a localized tumor is present. Endovascular vascular embolization can help maintain perioperative hemostasis by lowering tumor size and vascularity [13]. Chemotherapy has a limited use, while adjuvant radiation has some benefits. With very few exceptions, the disease's clinical history progresses quickly and typically results in death within a few months of diagnosis.

IV. Conclusion:

Angiosarcoma is an uncommon type of malignant vascular tumor and its clinic-pathological characteristics are not well known.

It has a peculiar association with pleural involvement. Histopathological and immune-histochemical results determine the final diagnosis. There isn't a codified treatment as of now. Although surgical excision, radiation, and chemotherapy have all been tried, the prognosis is not good for these tumors because of their aggressive clinical history.

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