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Case Report

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Bilateral Spontaneous Recurrent Hemothorax As a Complication of Primary Angiosarcoma of the Pleura

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RELEVANCE Pleural angiosarcoma is a rare type of malignant tumors. The diagnosis of primary pleural angiosarcoma is extremely difficult and is based on the results of immunohistochemical examination of tumor biopsies. The etiology and treatment methods of this disease have been poorly studied.

AIM Demonstration of a rare case of pleural angiosarcoma as a cause of bilateral spontaneous hemothorax.

CONCLUSION Primary epithelioid pleural angiosarcoma is a rare tumor with a high degree of malignancy and an unfavorable prognosis, which does not have pathognomonic symptoms. Spontaneous hemothorax may be the only primary clinical manifestation of the disease. Diagnostic thoracoscopy with pleural biopsy performed at an early stage to clarify the cause of hemothorax allows morphological verification of the tumor structure, which can affect the outcome of the disease in case of timely treatment.

Keywords: hemothorax, spontaneous hemothorax, malignant diseases of the pleura, angiosarcoma, angiosarcoma of the pleura

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PPA — primary pleural angiosarcoma
MSCT — multislice computed tomography

INTRODUCTION

The most common causes of hemothorax are chest trauma or medical procedures (central vein catheterization via subclavian access, thoracentesis, etc.). Due to the relatively low incidence and diversity of causes of spontaneous hemothorax, most publications available in the medical literature are presented in the form of individual clinical cases or small series of observations [1, 2]. Spontaneous hemothorax is most often a complication of spontaneous pneumothorax [3]. According to the classification proposed by D. Patrini et al. (2015), spontaneous hemothorax is divided into four categories by etiology: coagulopathic, vascular, tumor, and other [4, 5].

Angiosarcoma is a rare type of malignant tumor, accounting for 1 to 4% of all sarcoma cases, with morphological and immunohistochemical features of endothelial cells [6]. Angiosarcomas are a heterogeneous group of tumors. The primary localization of angiosarcoma is diverse (skin, soft tissues, internal organs), which affects the prognosis of the disease. Overall five-year survival rates range from 28.0% for tumors located in the soft tissues of the head and neck to 87.5% for tumors located in the mammary gland [6–8].

Diagnosis of primary pleural angiosarcoma is extremely difficult and is based on the results of immunohistochemical examination of tumor biopsy specimens. The morphological structure of the tumor can be spindle-shaped, which corresponds to the classic variant, or epithelioid type. The epithelioid variant accounts for 75% of pleural angiosarcomas. It is often misdiagnosed as mesothelioma or pleural adenocarcinoma. The epithelioid type of primary pleural angiosarcoma is a variant with a higher malignant potential compared to the classic spindle-shaped type [9–11]. Classic primary pleural angiosarcoma (PPA) has an irregular vascular pattern consisting of anastomosing vessels bordered by pleomorphic atypical endothelial cells. Conversely, this vascular structure is poorly represented in the epithelioid variant of the tumor, which is characterized by a continuous nodular pattern, neoplastic epithelioid cells with abundant eosinophilic cytoplasm and large pleomorphic nuclei with prominent nucleoli. Mitosis, hemorrhage and necrosis may be present in varying proportions [12, 13].

Treatment options for pleural angiosarcoma, like most neoplasms, include surgery, chemotherapy, and radiation therapy. However, the potential of any

given treatment method is limited. Surgical treatment is the most radical method in patients with localized lesions. Chemotherapy shows low efficiency, and radiation therapy is used as an additional treatment method [14]. Endovascular interventions such as embolization of vessels supplying the tumor can reduce tumor size and control pleural bleeding [15, 16].

Primary pleural angiosarcoma is a rare, highly malignant neoplasm. Despite modern medical capabilities, the prognosis of the disease is usually unfavorable, most patients die within a few months of diagnosis, which requires further improvement of early diagnostic methods and treatment of this pathology.

We report our own clinical observation of a patient with primary pleural angiosarcoma complicated by bilateral spontaneous recurrent hemothorax, which expands our understanding of this rare type of tumor lesion and its clinical manifestations.

Clinical observation

Patient S., 46 years old, was admitted with complaints of dyspnea with minimal physical exertion for a month. The patient's medical history does not indicate trauma or previous respiratory diseases. He was hospitalized in one of the Moscow hospitals. During the examination, bilateral large hydrothorax was diagnosed, in connection with which drainage of the left pleural cavity was performed with the evacuation of 3800 ml of hemorrhagic contents. For further treatment, he was transferred to the Department of Thoracic Surgery of the N.V. Sklifosovsky Research Institute for Emergency Medicine with drainage in the left pleural cavity.

The general condition was moderate, hemodynamics were stable, breathing was independent, without oxygen support.

Multispiral computed tomography (MSCT) of the chest organs on admission: right-sided hemothorax with a volume of 1500 cm3, fibrous changes in the left lung, foci in the upper and middle lobes of the right lung (Fig. 1). Drainage of the right pleural cavity was performed with evacuation of 1600 ml of blood. Laboratory examination of discharge from the pleural cavity: Hb - 72 g / l (blood Hb - 66 g / l), hematocrit - 23%. Bilateral hemothorax without an established cause or source was diagnosed. There were no clinical signs of ongoing intrapleural bleeding.

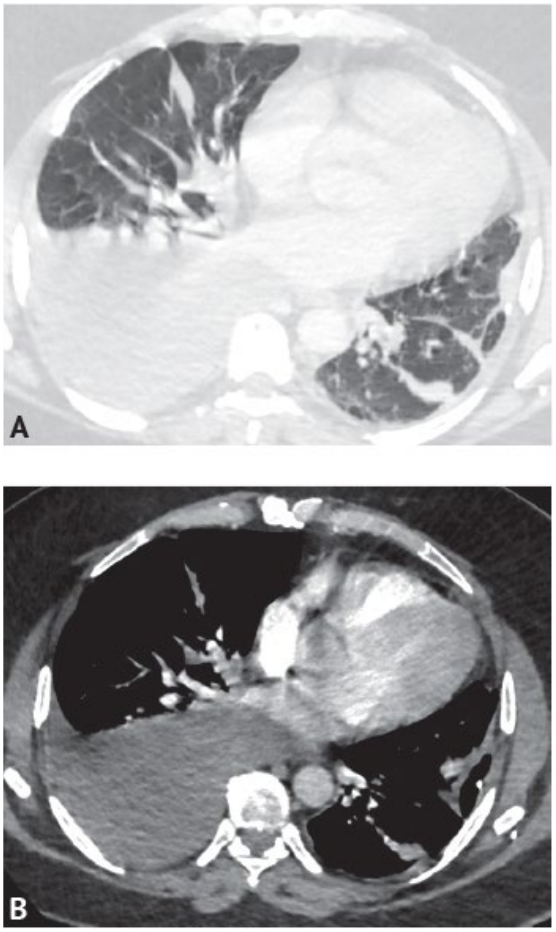

Fig. 1. MSCT of the chest upon admission. In the right pleural cavity along the interlobar fissure, heterogeneous contents with a density of 20–40 UH, a volume of 1500 cm^3 are determined

Considering the unspecified cause of hemothorax, the patient underwent diagnostic and therapeutic thoracoscopy on the left on the second day after hospitalization. Examination of the pleural cavity revealed multiple small dark cherry-colored foci on the visceral pleura of the upper lobe of the left lung, on the parietal pleura in the pericardial tissue zone with smooth, clear contours, 0.3 to 0.6 mm in size, without signs of bleeding (Fig. 2). The lung parenchyma was unchanged. There was a minimal amount of liquid blood in the pleural cavity. Tumorous pleural lesion was suspected as the cause of hemothorax. A biopsy of pathological areas of the pleura was performed.

The early postoperative period was uneventful. The drainage from the left pleural cavity was removed on the seventh day after surgery. Subsequently, repeated accumulation of hemorrhagic exudate was noted, which required repeated drainage of the pleural cavities. Laboratory examination of the exudate: Hb — 58 g/l (blood Hb — 75 g/l), no atypical cells were detected. The patient's condition remained severe, anemia progressed, requiring repeated transfusions of blood components. Daily hemorrhagic contents continued to flow through the pleural drainage in a volume of up to 500 ml per day. The patient's condition worsened, dyspnea increased. A right-sided clotted hemothorax was diagnosed. Thoracotomy on the right, sanitation of the pleural cavity, chemical pleurodesis were performed. Against the background of intensive therapy, the patient's condition progressively worsened. A day after the repeated operation, biological death was confirmed.

Intravital histological examination of tumor biopsy: in the visceral and parietal pleura there are areas of malignant tumor growth with pronounced circulatory disorders in the form of large focal extensive hemorrhages; the tumor is represented by clusters of rounded epithelioid-like cells with abundant cytoplasm and a vesicular nucleus with pronounced nucleoli; some epithelioid cells have cytoplasmic vacuoles - a primitive manifestation of the formation of vessel lumen, areas with the presence of irregular vascular channels lined with atypical endothelial cells. In the lumen of the vessels, there are tumor emboli from the above-described atypical cells. In the thickness of the lung tissue, mainly subpleurally, there is a tumor growth site, identical in histological structure to the tumor in the pleura (Fig. 3, 4). The histological picture may correspond to epithelioid mesothelioma of the pleura, however, subsequent immunohistochemical examination verified the tumor as epithelioid angiosarcoma

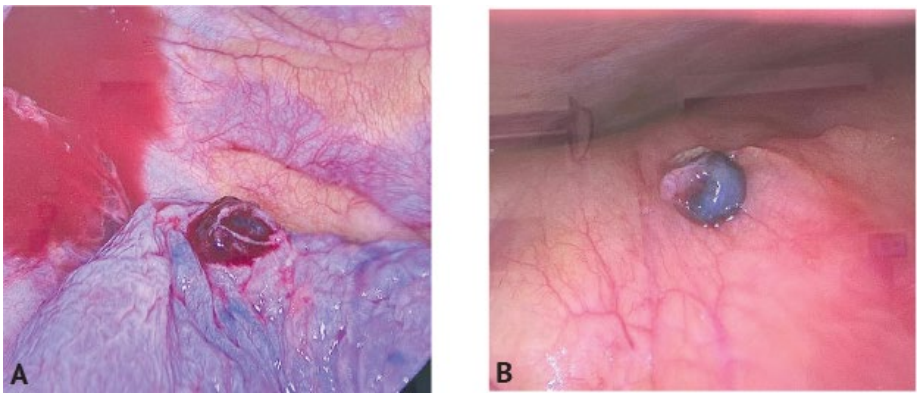

Fig. 2. Multiple small dark cherry-colored foci on the visceral pleura of the upper lobe and the parietal pleura in the pericardial tissue zone with smooth, clear contours ranging in size from 0.3 to 0.6 mm

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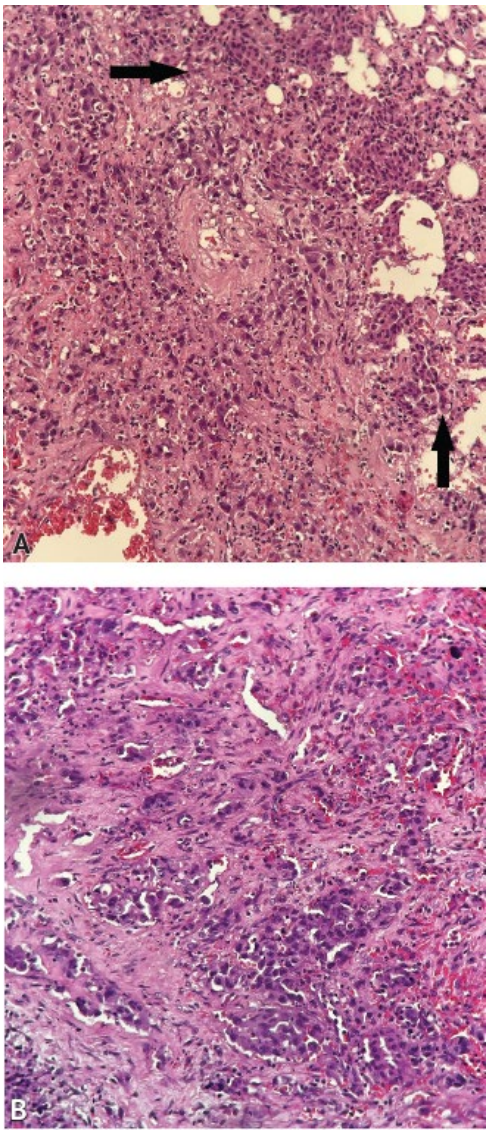

Fig. 3. Infiltrative growth of the malignant tumor in the thickness of the parietal pleura, represented by clusters of rounded epithelioid-like cells with abundant cytoplasm and a vesicular nucleus with pronounced nucleoli; some epithelioid cells have cytoplasmic vacuoles — a primitive manifestation of the formation of the vessel lumen, areas with the presence of irregular vascular channels lined by atypical endothelial cells. Magnification of figure A — 10x, figure B — 20x

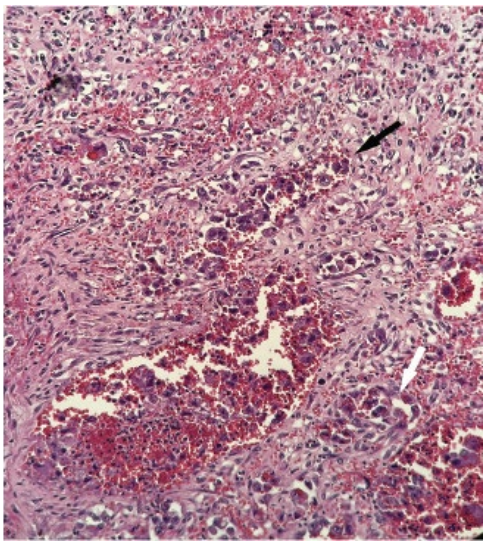

Fig. 4. In the thickness of the lung tissue, mainly subpleurally, there is a focus of tumor growth, identical in histological structure to the tumor in the pleura, with the presence of numerous tumor emboli in the lumens of the vessels

DISCUSSION

PPA is an extremely rare malignant disease of endothelial origin, characterized by a high incidence of local recurrence and metastasis, as well as an extremely poor survival prognosis. According to public data from the National Electronic Library of Medicine, 50 cases of primary pleural angiosarcoma have been published in the literature over the past 28 years. The etiology and treatment methods of this disease are poorly understood. The primary clinical manifestation of PPA is characterized by the appearance of nonspecific symptoms such as cough, dyspnea, chest pain, and hemoptysis. Recurrent exudative or hemorrhagic pleural effusion may develop. The most common clinical signs described in the literature are as follows: chest pain, hemoptysis, anemia, and recurrent hemothorax [7, 8].

The radiological semiotics of the disease have no specific signs, which does not allow differentiating PPA from other primary or secondary tumor lesions of the pleura. Chest X-ray may show localized pleural thickening and unilateral pleural effusion. MSCT examination shows a lobular pleural formation with clear boundaries, heterogeneously accumulating contrast agent.

Diagnosis of primary pleural angiosarcoma is based on the results of immunohistochemical examination of the tumor.

CONCLUSION

Primary epithelioid pleural angiosarcoma is a rare tumor with a high degree of malignancy and an unfavorable prognosis, which does not have pathognomonic symptoms. Spontaneous hemothorax may be the only primary clinical manifestation of the disease. Diagnostic thoracoscopy with pleural biopsy performed early to clarify the cause of hemothorax allows morphological verification of the tumor structure, which may affect the outcome of the disease if treatment is started in a timely manner.

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