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PULMONARY EPITHELIOID ANGIOSARCOMA: A CASE REPORT

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ЭПИТЕЛИОИДНАЯ АНГИОСАРКОМА ЛЕГКИХ: КЛИНИЧЕСКОЕ НАБЛЮДЕНИЕ

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Primary pulmonary epithelioid angiosarcoma is an extremely rare condition with a rapidly progressive course and poor prognosis. Currently, there is no standard treatment regimen for pulmonary angiosarcoma. The primary treatment for this type of cancer is radical surgery followed by chemotherapy. The article focuses on a 5-year follow-up in a 38-year-old patient with pulmonary epithelioid angiosarcoma. Since the regional lung resection, the patient has received chemotherapy with a positive effect for more than four years. Every three months, the patient undergoes positron emission tomography (PET) / computed tomography of the chest and internal organs to assess the disease course. A sustained absence of PET signs of tumor metabolic activity has been noted. The patient's condition is satisfactory. He leads a relatively active lifestyle.

Keywords: primary pulmonary epithelioid angiosarcoma, chemotherapy

Первичная эпителиоидная ангиосаркома легких встречается крайне редко, характеризуется быстро прогрессирующим течением и неблагоприятным прогнозом. В настоящее время отсутствуют стандарты лечения ангиосаркомы легких. Основным методом терапии этого опухолевого процесса является радикальная операция с последующей химиотерапией. Описывается 5-летнее наблюдение 38-летнего пациента с эпителиоидной ангиосаркомой легкого. С момента регионарной резекции легких больной уже более 4 лет получает химиотерапию с положительным эффектом. Каждые 3 месяца пациенту проводят позитронно-эмиссионную томографию (ПЭТ) / компьютерную томографию органов грудной клетки и внутренних органов для оценки течения заболевания. В настоящее время отмечается стойкое отсутствие ПЭТ-признаков метаболической активности опухоли. Состояние больного удовлетворительное. Он ведет относительно активный образ жизни.

Ключевые слова: первичная эпителиоидная ангиосаркома легких, химиотерапия

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ANCA – anti-neutrophil cytoplasmic antibodies
ARF – acute respiratory failure
AS – angiosarcoma
BAL – bronchoalveolar lavage
CT – computed tomography
DNA – deoxyribonucleic acid
HIV – human immunodeficiency virus
PET – positron emission tomography

Angiosarcoma (AS) is one of the rarest soft tissue tumors, accounting for a small proportion of vascular tumors and less than 1 % of all sarcomas [1]. Pulmonary AS is even less common (<10 % of all AS cases) and has a poor prognosis [2]. The average life expectancy in patients with primary pulmonary AS is about nine months [3]. Such a low prevalence of this condition makes the following primary pulmonary epithelioid AS in a male patient extremely interesting.

Case history. Patient M., 38 years old, presented to the clinic in November 2015 after the first onset of hemoptysis. He mentioned having a viral respiratory infection a month before. He had a history of a stab wound to the right lung, due to which, in 2004, partial resection of the lower lobe was performed. Chest computed tomography (CT)

performed on November 16, 2015, showed hemorrhagic foci in both lungs.

He complained of a cough with bloody sputum, moderate shortness of breath, weakness, and fatigue during hospitalization. ANCA test results include rheumatoid factor, lupus anticoagulant, antinuclear factor, anti-glomerular basement membrane antibodies, anti-double stranded DNA antibodies, human immunodeficiency virus (HIV), and tumor markers, and procalcitonin were negative. Bronchoscopy showed signs of catarrhal endobronchitis and traces of blood from basal bronchi on the left. In the bronchoalveolar lavage (BAL) cells, there were signs of inflammation, proliferation, and hyperplasia of the bronchial epithelium. Repeated sputum and BAL tests did not reveal Mycobacterium tuberculosis. Chest CT with intravenous contrast showed signs of hemorrhage in the lung parenchyma. Abdominal and kidney CT did not reveal any pathological changes. After the examination, the patient was diagnosed with: «Community-acquired bilateral polysegmental viral-bacterial pleuropneumonia of moderate severity and prolonged course. Vasculitis? Recurrent hemoptysis. Acute respiratory failure (ARF).»

On combination therapy (antibacterial therapy, prednisone 40 mg/day per os), the patient's condition improved: hemoptysis resolved; follow-up chest CT showed a partial decrease in the volume of hemorrhagic foci. However, in mid-December 2015, blood streaks in sputum and profuse hemoptysis reappeared. On December 29, 2015, chest CT showed signs of recurrent hemorrhage in the lung parenchyma (S7 of the right lower lobe and S3 of the left upper lobe), polysegmental pulmonary fibrosis of both lungs, and disk-shaped atelectasis of the right lower lobe (Fig. 1).

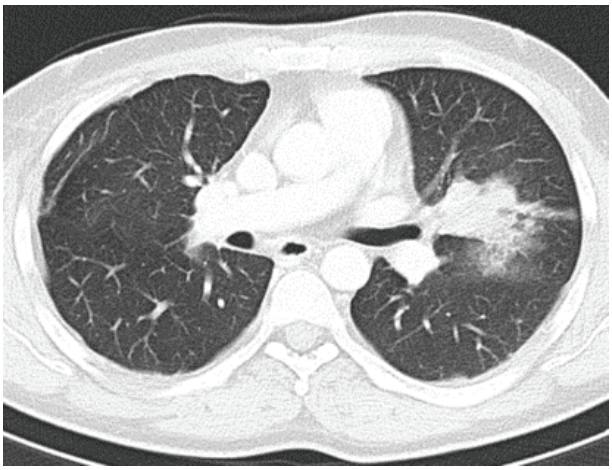

Fig. 1. Chest CT of December 29, 2015. Relapse of hemorrhage into both lungs parenchyma

The patient was re-hospitalized in January 2016. Based on the examination results, he was diagnosed with «Pulmonary vasculitis. ARF.» The prednisone dose was increased up to 60 mg/day per os, with a recommendation to reduce the dose following the disease stabilization. However, with a decrease in the prednisone dose to 40 mg/day, in February 2016, hemoptysis episodes reoccurred without fever. The patient was consulted at the Research Institute of Pulmonology, diagnosed with pulmonary vasculitis (ANCA-negative), and lung biopsy was recommended.

In March 2016, chest CT with contrast showed negative changes during the subsequent hospitalization: focal lesions appeared, with granuloma formation in both lungs and an increase in right-sided paracostal and interlobar hemothorax. Based on the results of bronchoscopy with bronchial mucosa histology, the patient was diagnosed with edematous-catarrhal endobronchitis. Angiography of the bronchial and pulmonary arteries did not reveal pulmonary vascular malformation. The patient received the following treatment: prednisone 60–40–30 mg/day per os, antibiotic therapy, and plasmapheresis. During this period, «streaks of blood in the sputum» appeared from time to time. Due to the therapy failure, on April 27, 2016, a diagnostic thoracotomy and lung biopsy were performed at the Department of Thoraco-Abdominal Surgery of N. V. Sklifosovsky First Aid Research Institute to refine the diagnosis. Resection of the lower lobe of the right lung with neoplasms was also performed (Fig. 2).

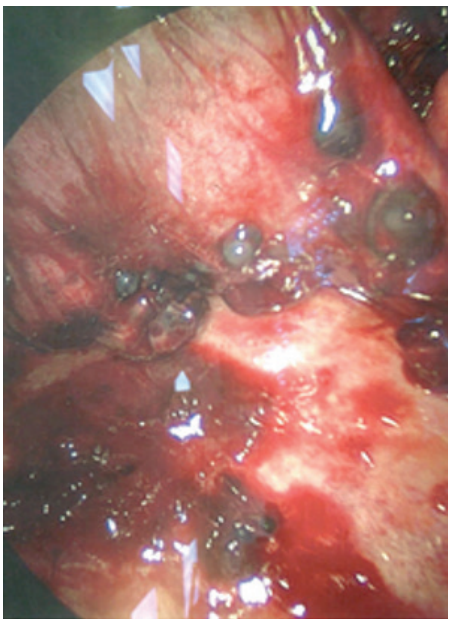

Fig. 2. Video-assisted thoracoscopy of the right lower lobe with neoplasms

Histopathological examination was performed at N. N. Blokhin Russian Cancer Research Center. The following conclusion was made on May 18, 2016: the specimen consists of lung tissue fragments with grade III AS tumor node, with an extensive necrosis zone of more than 50 %. Additionally, immunohistochemistry was performed using anti-CD31, -CD34, -Fli1, -HHV8, and -Ki67 antibodies. CD31, CD34, and Fli1 expression in tumor cells was revealed, which suggests their endothelial differentiation in the absence of HHV8 expression by tumor cells. The proliferation index in Ki67 tumor cells was 40 %. The analysis made it possible to establish the diagnosis: epithelioid pulmonary AS, grade III. Mts to the lungs.

After the diagnosis verification, in agreement with the oncologists of N. N. Petrov Oncology Research Institute and N. N. Blokhin Russian Cancer Research Center, the patient received the first chemotherapy course according to the following scheme: docetaxel 190 mg, gemcitabine 1700 mg No. 2. Bevacizumab (Avastin) 560 mg was added to the scheme starting from the second chemotherapy course. In total, seven chemotherapy courses were performed according to this scheme.

Follow-up chest CT of October 18, 2016, performed during treatment showed positive changes (no new hemorrhages were detected, existing lesions in S3 of the left lung resolved completely, the foci sizes in S1, 2, 6, and 8 decreased compared to the previous examination) (Fig. 3).

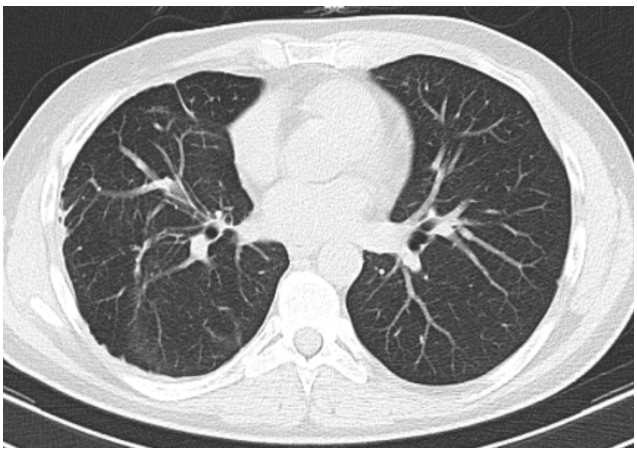

Fig. 3. Chest CT of October 18, 2016. Positive changes on chemotherapy. Fibrosing changes in both lungs

On October 26, 2016, positron emission tomography (PET) with fluorodeoxyglucose was performed at N. P. Bekhtereva Institute of Human Brain. Conclusion: condition after combination therapy of pulmonary angiosarcoma without PET signs of metabolic activity. The patient was transferred to maintenance therapy with bevacizumab 600 mg monthly with regular PET/CT every three months at N. P. Bekhtereva Institute of Human Brain. The following conclusion was made during the last examination on December 17, 2020: condition after combination therapy of epithelioid pulmonary angiosarcoma with a sustained absence of PET signs of tumor metabolic activity.

Discussion. This is a rare case of a 5-year follow-up from the onset of first symptoms of primary epithelioid pulmonary AS. Difficulties in early diagnosis are associated with a low prevalence of this condition, lack of awareness among practitioners about it. Chest CT as well as histopathological and immunohistochemical examinations, are the most valuable tools in diagnosing pulmonary AS [1]. No standard treatment regimens for pulmonary AS have yet been developed [3]. The literature describes surgical resection, vascular embolization, radiation therapy, immunotherapy, and chemotherapy for this condition [2]. Most authors believe that the primary treatment is radical surgery followed by chemotherapy [4, 5].

Conclusions. This case confirmed the effectiveness of combination therapy for epithelioid pulmonary AS, taking into account the patient's high compliance, which significantly increased his life expectancy after the start of treatment, compared with the cases described in the literature worldwide.

Disclosures: The authors declare no conflict of interest.

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