Malignant Primary Pulmonary Hemangiopericytoma/Solitary Fibrous Tumor (HPC/SFT) with Extensive Endobronchial Involvement: A Case Report

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Background

The (HPC/SFT) is a rare mesenchyme neoplasm; represent less than 1% of vascular tumors. It originates from the cells that surround endothelial tissue. The most common sites were the head and neck, lower extremities and retroperitoneum; its primary localization in the lung and endobronchial is extremely rare. Are considered malignant when they have more than four mitoses per high power field, cellular atypia, necrosis and hemorrhage. We report the case of a patient with a malignant primary pulmonary (HPC/SFT) with extensive endobronchial involvement.

Results

This is a woman of 40 years old with a history of dyspnea, chest pain and hemoptysis, who was a lung mass in the right hemithorax with loss of volume and endobronchial lesions evident on the CT. In biopsies taken by bronchoscopy diagnosis was inconclusive, then open biopsy was carried in which document (HPC) with high mitotic activity and necrosis. Figure 1 shows CT, endobronchial lesions and endoscopic resection. She was not surgical candidate for the extension of the disease, and their oncologist decided start chemotherapy with doxorubicin and cisplatin in the ICU make possible to retire mechanical ventilation support, after she started bevacizumab plus temozolomide with excellent clinical and radiological response. For the management of endobronchial lesions therapeutic bronchoscopic was done using electrocautery and argon plasma coagulation, with permeability of the airway and control of episodes of hemoptysis. Endobronchial lesions subsequently progressed and she died one year after diagnosis.

Bibliography


Figure 1, A. CT with mass in right hemithorax with endobronchial lesion; B. C, D. Appearance of the lesions and endoscopic electrocoagulation procedure for resection

Figure 2, A. bronchial-invasive lesions with spindle cell and epithelial cells. B. biopsies vascularized with thin channels between tumor cells. C. High mitotic activity. D. Aspect of tumor necrosis

Methods

We review the clinical history data

Conclusions

HPC/SFT is a rare tumor; surgical resection is the treatment of choice. Chemotherapy with doxorubicin-based schemes has been used with ifosfamide or gemcitabine and docetaxel, with only a modest disease control. Combination therapy with temozolomide and bevacizumab appears to provide clinical benefit with a low rate of major toxicities. Interventional pulmonology offers an effective palliative treatment to reduce symptoms, the risks of bleeding and improving the quality of life