Primary Epithelioid Sarcoma of the Pleura: An Intricate Diagnosis

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A 33-year-old man with recurrent episodes of breathlessness and hemoptysis over the previous year presented with right lung collapse and pleural effusion. A cavitating right upper lobe lesion thought to be non-malignant was identified on the chest roentgenogram (Fig 1) and on tomography. Video-assisted thoracoscopic surgery (VATS) revealed an entrapped lung, blood clots, and thickened pleura, and the surgery was converted to thoracotomy. Intraoperative fresh frozen sections suggested a mesothelioma, and an extensive decortication was performed. Three pathology departments were involved in the diagnosis. Conventional microscopy showed a diffuse epithelioid neoplasm with occasional mitotic figures and focal necrosis. Immunohistochemical analysis demonstrated uniform positivity for cytokeratin AE1/AE3 and CD34. Additionally, the cells were positive for EMA and MNF-116 with widespread cytoplasmic and focal nuclear immunoreactivity for WT1. There was negative staining for CD31, ERG, INI1, TTF1, cytokeratin 7, cytokeratin 5/6, and calreinin mesothelin (Fig 2). The pattern of positivity for cytokeratins and CD34 with negative staining for INI1 is very characteristic of epithelioid sarcoma. Fluorescence in situ hybridization studies for WWTR1 were negative for gene rearrangement, providing strong evidence for the exclusion of epithelioid hemangiendothelioma. Alternative primary sites for his tumor were not found. A diagnosis of primary epithelioid sarcoma of the pleura was made. Post-operatively, the patient received 5 cycles of ifosfamide and doxorubicin, and received oncologic follow-up.

Fig 1.

Fig 2.