



DEPARTMENT OF PATHOLOGY

Case of the Week

Thoracic Pathology: Pulmonary Meningioma

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History

Prior to undergoing a cataract extraction, a 75-year-old male was found to have an incidental right lung nodule on a routine pre-operative chest x-ray. A positron emission tomography-computed tomography (PET-CT) scan subsequently demonstrated a 1.7 x 1.6 cm hypermetabolic pulmonary nodule in the middle lobe of the right lung. The patient then underwent video-assisted thoracoscopic surgery (VATS) and a lung wedge resection was performed. On gross examination of the lung, there was a single, tan-yellow, firm, well-circumscribed nodule identified in the lung parenchyma.

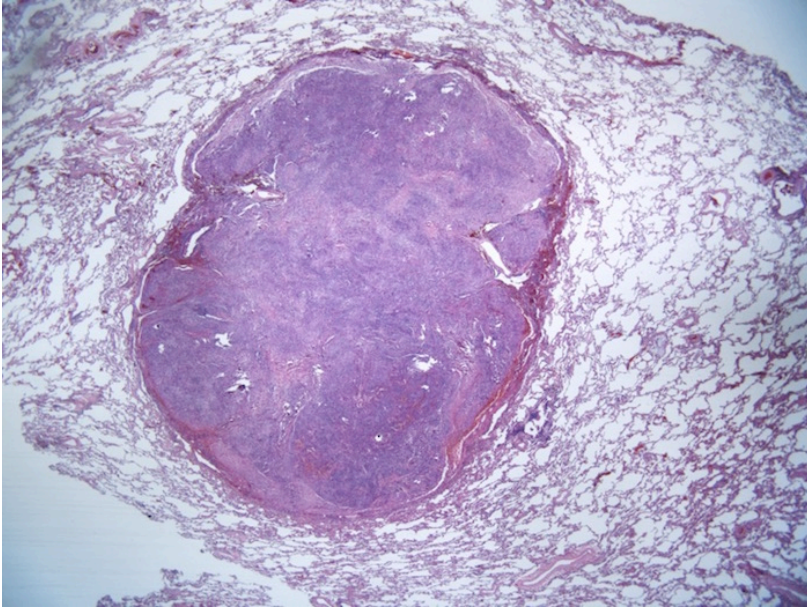


Figure 1: Hematoxylin and eosin stain, 20x magnification

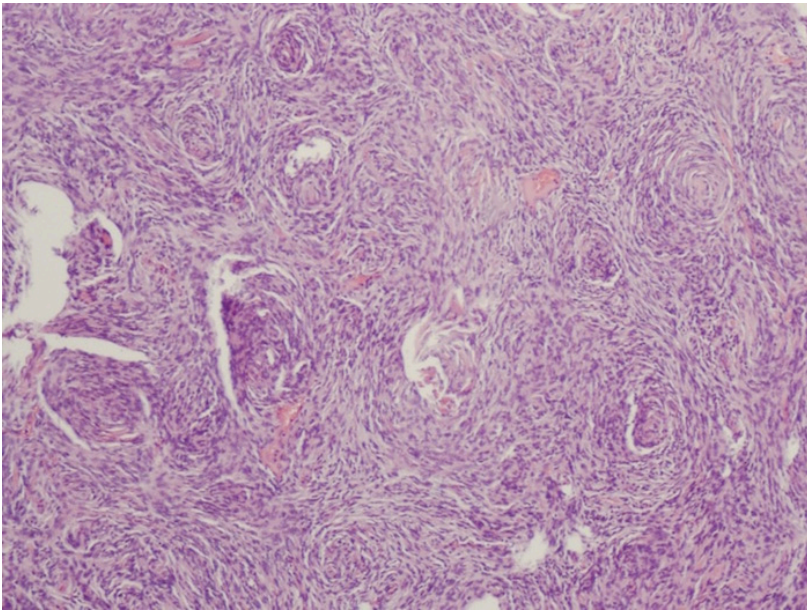


Figure 2: Hematoxylin and eosin stain, 100x magnification

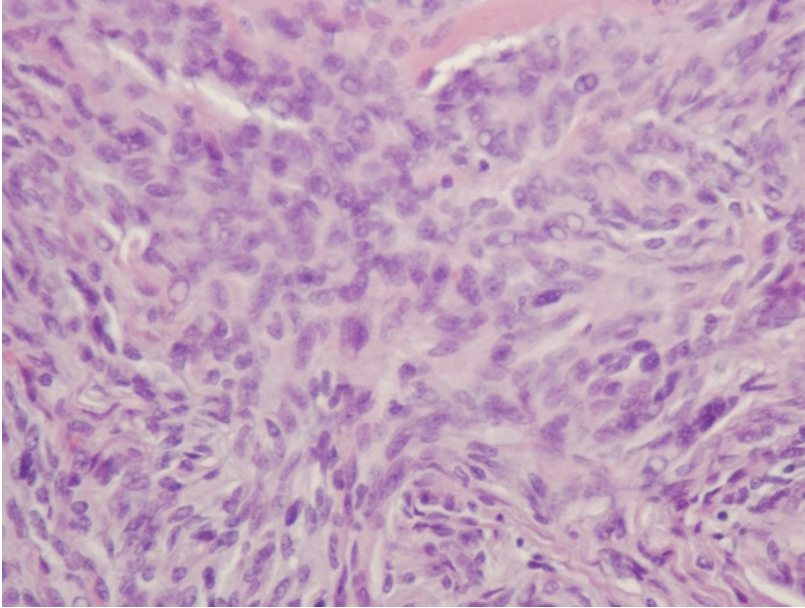


Figure 3: Hematoxylin and eosin stain, 400x magnification

Figure 1-3

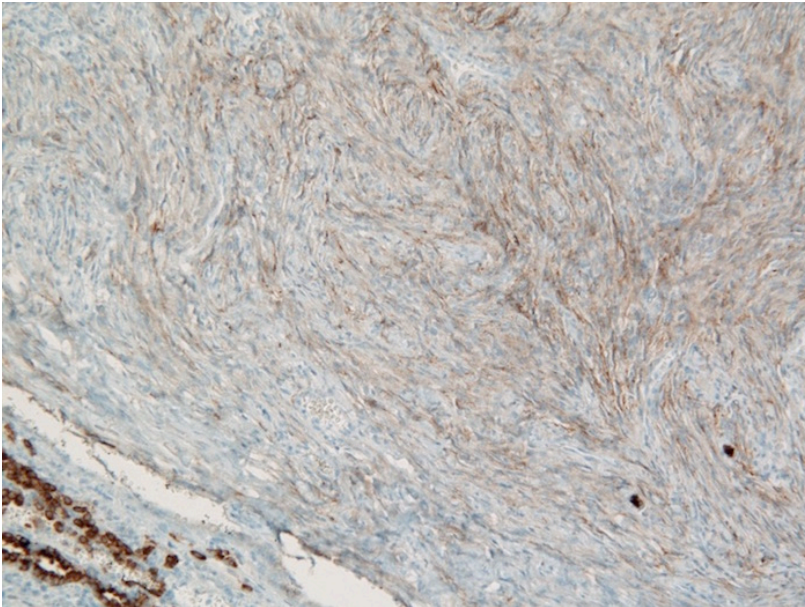


Figure 4: Epithelial membrane antigen (EMA) immunohistochemical stain, 100x magnification

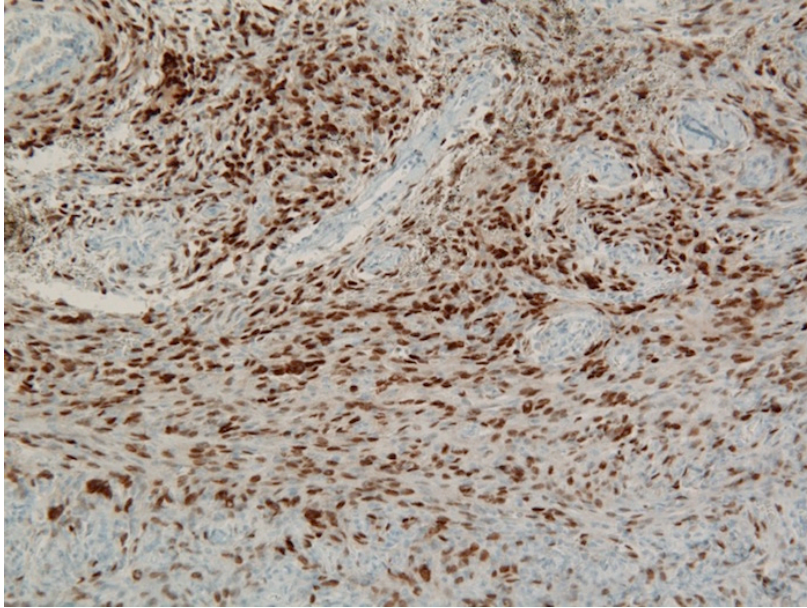


Figure 5: Progesterone receptor (PR) immunohistochemical stain, 100x magnification

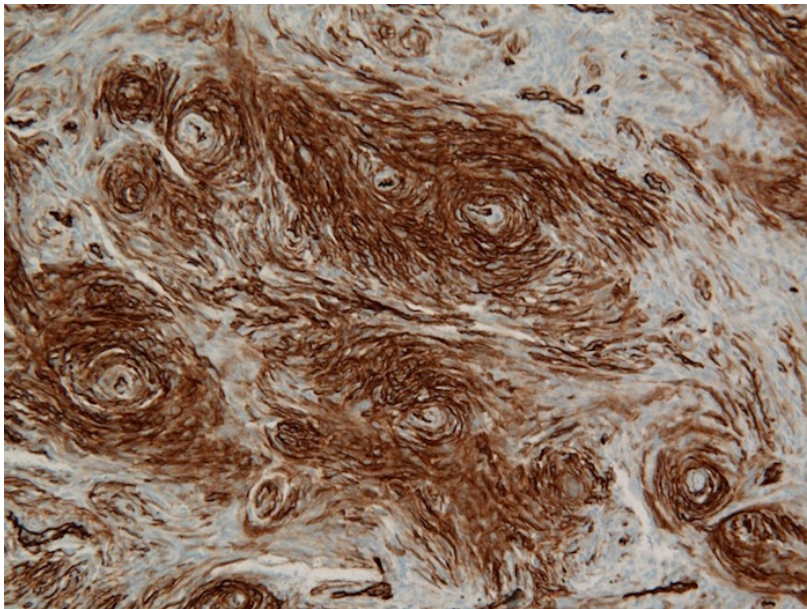


Figure 6: CD34 immunohistochemical stain, 100x magnification

Figure 4-6

Figures 4-6. Immunohistochemistry. Fig. 4: Representative area of tumor demonstrating weak patchy EMA expression (cytoplasmic and membranous; 100x magnification). Fig. 5: There was positive PR expression (nuclear; 100x magnification). Fig. 6: Representative area of tumor demonstrating patchy but strong positive CD34 expression (cytoplasmic and membranous; 100x magnification).

Other immunohistochemical stains performed (not pictured): Immunohistochemical stains for bcl-2 and CD99 show weak positive reactivity in tumor cells. Synaptophysin, chromogranin, cytokeratin AE1/AE3 (pancytokeratin cocktail) and S-100 demonstrate negative expression in the tumor cells. MIB-1/Ki-67 proliferation index shows labeling of approximately 2% of tumor cells.

Diagnosis

Pulmonary meningioma

Discussion

Introduction

Primary pulmonary meningiomas are remarkably rare; less than 50 cases have been reported in the English literature ¹. A radiologic study of the central nervous system (preferably an MRI) should first exclude an occult primary intracranial or spinal meningioma ². For this case, a brain MRI did not show evidence of an intracranial mass lesion in the patient. Most primary pulmonary meningiomas are incidentally detected in asymptomatic patients as a “coin lesion” on chest x-ray ¹.

The true cell of origin for primary pulmonary meningiomas still remains undetermined. Hypotheses have been put forward that suggest these tumors are derived from either: (1) pluripotential subpleural mesenchymal cells or (2) heterotopic embryonic rests of arachnoid cells dispersed during embryonic development ¹.

Microscopic findings

Histologically, primary pulmonary meningiomas resemble their central nervous system counterparts ¹. They are often characterized by lobules of plump spindle-shaped cells arranged in whorls (as seen in meningotheial CNS meningiomas) and/or spindled cells with surrounding collagenous stroma (as seen in fibrous CNS meningiomas) ³. Intra-nuclear pseudoinclusions and staghorn-like vessels can also be seen. By definition, pulmonary meningiomas are nodular lesions that replace the pulmonary parenchyma ¹. This is in contrast to minute pulmonary meningotheial-like nodules which are interstitial lesions that preserve the lung architecture ¹. It has been speculated that minute pulmonary meningotheial-like nodules can progress to primary pulmonary meningiomas ¹.

Immunohistochemistry

It has been shown that primary pulmonary meningiomas share immunohistochemical features with their counterparts in the central nervous system ³. Meningiomas typically show positive reactivity for epithelial membrane antigen (EMA) and progesterone receptor (> 50%) with limited or no cytokeratin expression ⁴. The immunohistochemical profile of meningiomas is non-specific and there may be overlap with other entities such as solitary fibrous tumor. However, STAT-6 is a new immunohistochemical antibody that has been found to be sensitive and specific for solitary fibrous tumors and may be used as a diagnostic tool when appropriate ⁵.

Genetics

Recently, deletion of the neurofibromatosis (NF) 2 gene on chromosome 22 and chromosomal gains of 22q have been identified in both minute pulmonary meningothelial-like nodules and primary pulmonary meningiomas¹. Meningiomas of the CNS have been known to exhibit loss of the NF2 gene¹. These results give some insight in the relationship between these three entities and provide some evidence that they may arise from the same precursor cell.

Management

Primary pulmonary meningiomas are generally benign, however, two malignant cases have been reported². Treatment is surgical resection, where wedge resections are typically performed for peripheral lesions and lobectomies are reserved for centrilobar lesions^{2,3}. For benign cases, there have been no reported relapses after complete resection².

References

1. Weissferdt A, Tang X, Suster S, et al. Pleuropulmonary meningothelial proliferations: evidence for a common histogenesis. *Am J Surg Pathol*. 2015 Aug 18.
2. Incarbone M, Ceresoli GL, Di Tommaso L, et al. Primary pulmonary meningioma: report of a case and review of the literature. *Lung Cancer*. 2008;62:401-407.
3. Moran CA, Hochholzer L, Rush W, et al. Primary intrapulmonary meningiomas. A clinicopathologic and immunohistochemical study of ten cases. *Cancer*. 1996;78:2328-2333.
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5. Schweizer L, Koelsche C, Sahm F, et al. Meningeal hemangiopericytoma and solitary fibrous tumors carry the NAB2-STAT6 fusion and can be diagnosed by nuclear expression of STAT6 protein. *Acta Neuropathol*. 2013;125:651-658.