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Primary Pulmonary Chordoid Meningioma

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Primary pulmonary meningioma is a rare disease, and chordoid meningioma is an uncommon variant of meningioma in the central nervous system (CNS) with a high recurrence rate. We report a case of primary pulmonary chordoid meningioma that presented as a solitary pulmonary nodule (SPN). The SPN was resected by thoracoscopic wedge resection and was revealed to have characteristics of chordoid meningioma. After confirming the absence of a meningioma in the CNS by brain imaging, the nodule was diagnosed as a primary pulmonary chordoid meningioma. The patient remained disease-free after 26 months postoperatively. To our knowledge, this is the third case of primary pulmonary chordoid meningioma to be reported.

Key words: 1. Primary pulmonary meningioma
2. Chordoid meningioma
3. Thoracoscopy/VATS
4. Lung wedge resection

Case report

A 43-year-old woman was found to have a 17-mm solitary pulmonary nodule (SPN) as an incidental finding on a chest X-ray. She had no symptoms related to the SPN. She had multiple underlying diseases: systemic lupus erythematosus (SLE), Graves disease, diabetes mellitus, and antiphospholipid antibody syndrome. In a retrospective review of serial chest X-rays, a 9-mm SPN in the same location (previously undetected due to the normal hilar structure) was discovered on a chest X-ray performed 15 months earlier. Chest computed tomography (CT) showed a 19-mm, oval-shaped, well-enhanced soft tissue mass in the left upper lobe of the lung. A carcinoid tumor was suspected, and an 18F-fluorodeoxyglucose (18F-FDG) positron emission tomography (PET)-CT scan showed increased uptake in the tumor (maximum standard uptake value, 2.48) with no other hypermetabolic lesions (Fig. 1). A percutaneous needle biopsy was performed and showed fibromyxoid tissues, suggesting the possibility of pulmonary hamartoma (Fig. 2). However, its relatively rapid tumor doubling time (138 days), increased 18F-FDG uptake on a PET-CT scan, and the slightly increased risk of malignancy in SLE patients meant that malignancy could not be excluded. Therefore, surgical resection was performed, both with curative intention and to make an accurate diagnosis. Preoperative laboratory findings and pulmonary function tests suggested no particular challenges for surgery.

The patient underwent video-assisted thoraco-

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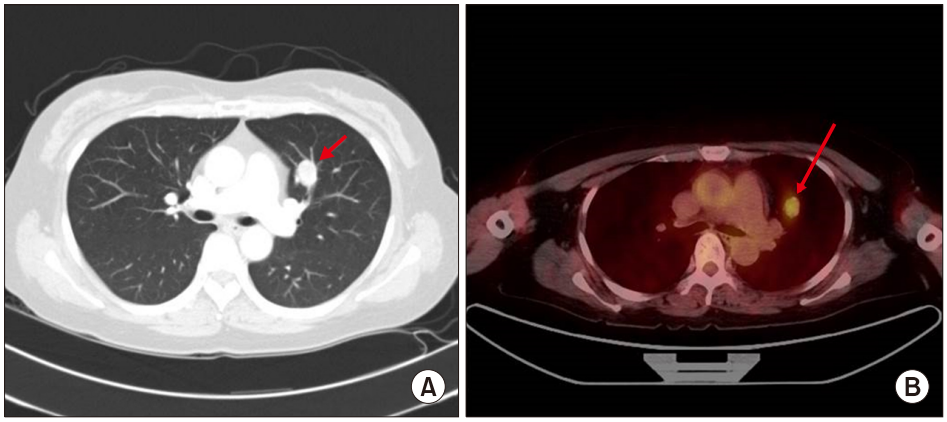

Fig. 1. (A) Chest CT revealed a 1.9-cm, well-demarcated, oval-shaped, homogenous, well-enhanced soft tissue mass in the anterior segment of the left upper lobe of the lung (arrow). (B) On 18F-fluorodeoxyglucose positron emission tomography-CT, the mass showed hypermetabolic activity with a maximum standardized uptake value of 2.48 (arrow). There were no other areas of abnormal hypermetabolic activity. CT, computed tomography.

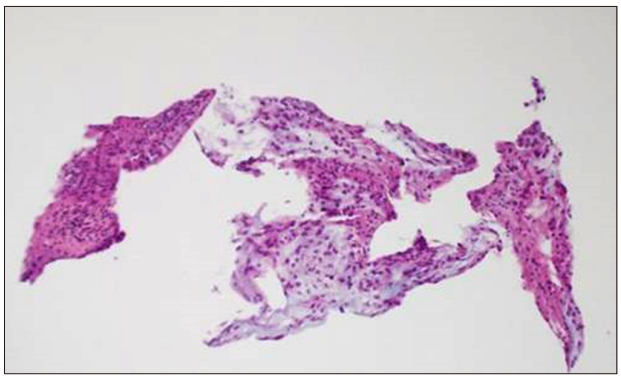

Fig. 2. The needle biopsy specimen showed bland-looking, round-to-ovoid tumor cells and inflammatory cells in the fibrous and myxoid stroma (H&E, ×400).

scopic surgery (VATS) with 3 ports. An incision was made for the working window via the fifth intercostal space (ICS) mid-axillary line with a small wound protector for nodule palpation and another 2 incisions via the seventh ICS anterior axillary line with a 12-mm trocar and the ICS posterior axillary line with a 10-mm trocar for the instrument and thoracoscope. The tumor was not identified on the visceral pleural surface, while it was easily detected by instrumentation and finger palpation. The tumor was removed by wedge resection with three 60-mm endoscopic staplers (Echelon Flex Powered Endopath Stapler, green; Ethicon, Cincinnati, OH, USA) without any hilar vascular or bronchial resection. When performing wedge resection, we took the time for a totally collapsed lung, which we simulated with Mixter right angle forceps to prevent vascular injury. The diagnosis made by intraoperative frozen section biopsy was hamartoma, the same result obtained by the preoperative biopsy. The distance from the tumor edge to the resection margin was 5 mm, and we decided to complete the operation without further resection. A chest tube was removed on the fourth postoperative day because of a minimal air leak. The patient was discharged without any complications.

On pathologic evaluation using permanent sections, the tumor showed epithelioid features with vacuolated cytoplasm forming a cord-like or trabecular arrangement and abundant myxoid stroma. The tumor cells showed diffuse positivity for epithelial membrane antigen and vimentin immunohistochemical staining, which is characteristic of meningioma. Based on histology and immunohistochemistry, the pathologic report was revised to chordoid meningioma (Fig. 3). To exclude metastatic meningioma, brain magnetic resonance imaging (MRI) was performed; it confirmed the absence of an intracranial lesion. Therefore, the final diagnosis became primary pulmonary chordoid meningioma. Through a multidisciplinary discussion, the decision was made to conduct careful follow-up without adjuvant therapy. Follow-up chest CT examinations were performed and evaluated every 6 months. The patient has remained free of disease for 26 months after surgery.

The Institutional Review Board of Hanyang University Seoul Hospital has waived the IRB review and the requirement for individual patient consent for this case study.

Discussion

Primary pulmonary meningioma is a highly uncommon subtype of the rare category of primary ex-

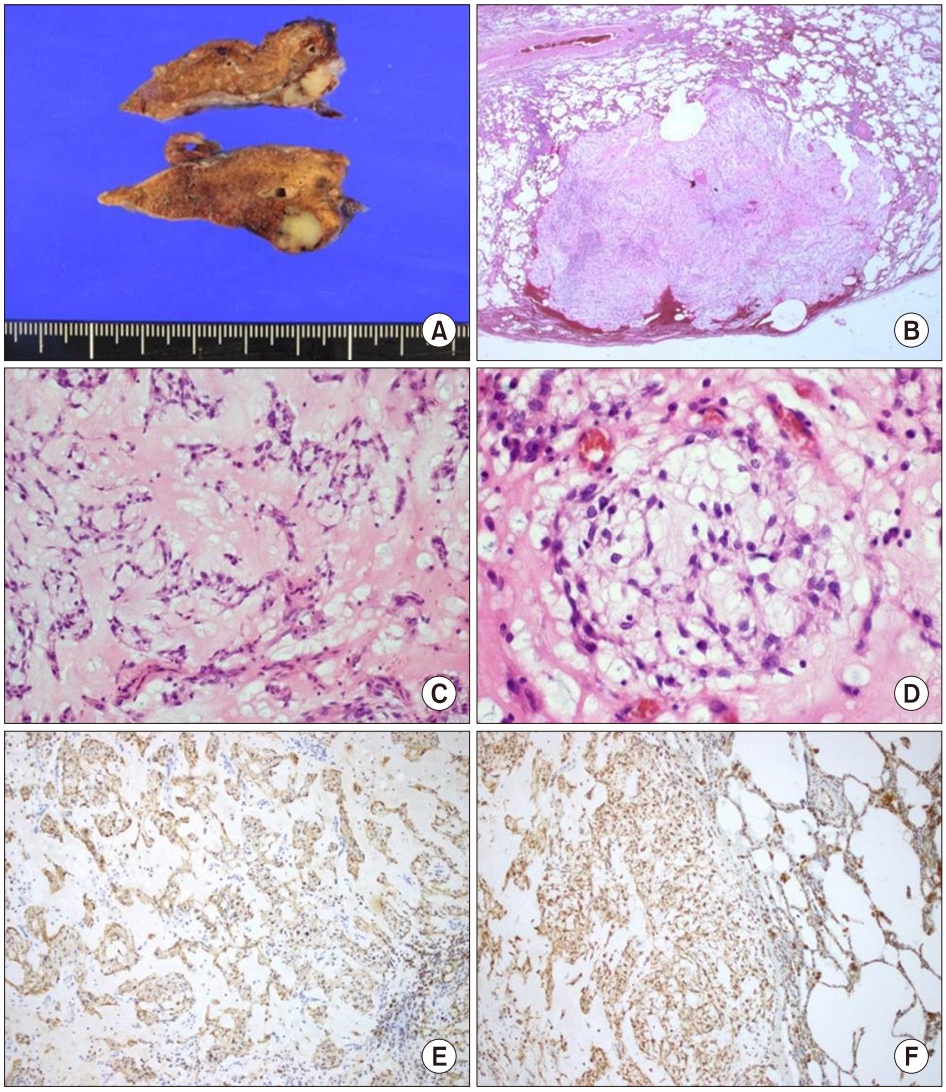

Fig. 3. Histopathologic findings of chordoid meningioma of the lung. (A) Gross examination revealed a well-demarcated, gray-white solid mass with a hard consistency. (B) On microscopic examination, the tumor was sharply demarcated (H&E, ×12.5), and (C) it showed cord-like and solid tumor cell nests within abundant mucous-rich stroma (H&E, ×200). (D) The tumor cells often showed epithelioid features with vacuolated cytoplasm (H&E, ×400). The tumor cells had diffuse immunoreactivity for epithelial membrane antigen (E, ×100) and vimentin (F, ×100).

tracranial meningioma [1]. Ectopic primary meningioma, which accounts for 1% to 2% of all primary meningiomas, is typically seen in the head and neck region, and primary extracranial meningiomas are rare outside the head and neck region. Fewer than 50 cases of primary pulmonary meningioma have been reported in the medical literature. The major subtypes of primary pulmonary meningioma, which are regarded as benign meningiomas, are typical syncytial and transitional meningiomas. Chordoid meningioma is an uncommon subtype of meningioma that constitutes 0.5% of all surgically resected meningiomas [2]. To the best of our knowledge, this is the third reported case of primary pulmonary chordoid meningioma [2,3].

The pathogenesis of primary pulmonary meningioma is unknown. Hypotheses about the cellular origin of primary pulmonary meningioma include the possibilities that it may derive from pluripotential subpleural mesenchyme or from heterotopic embryonic remnants of arachnoid cells [4]. However, this is still debated. In the present case, the patient had SLE. The prevalence of meningioma in SLE patients has been reported to be 1.65%, which is higher than the rate of 0.5% in the general population [5]. The relationship between SLE and meningioma has been hypothesized to be rooted in hormonal influences, but this might be a pure coincidence.

Making the correct preoperative diagnosis of primary pulmonary meningioma is somewhat difficult.

Chest CT showed a mostly well-circumscribed solitary nodule. Some cases show lobulated margins, but this is not a specific finding. PET-CT scans cannot differentiate primary pulmonary meningioma from other types of lung cancer because of the possibility of false-positive PET-CT scans [1]. Moreover, preoperative needle biopsy is generally not diagnostic. In a study of 10 misdiagnosed cases of primary pulmonary meningioma, a histological analysis of preoperative transthoracic needle biopsies revealed 9 false negatives and 1 false positive as papillary thyroid carcinoma [1]. Such misdiagnoses can lead to incorrect treatment. Primary pulmonary meningioma patients with a history of breast or colon cancer have incorrectly received chemotherapy [1]. In one case, a primary pulmonary meningioma patient who was misdiagnosed as having papillary adenocarcinoma underwent a lobectomy and mediastinal lymph node dissection, which might have been replaced by wedge resection if the preoperative diagnosis had been correct [6]. In the diagnostic criteria, chordoid meningioma has cords of small epithelioid tumor cell that contain eosinophilic or vacuolated cytoplasm embedded in a basophilic, mucin-rich matrix [7]. However, vacuolated cytoplasm is sometimes hard to distinguish from ice crystals, which are an artifact of freezing. Because of this, the intraoperative frozen biopsy sample in our case was misdiagnosed as hamartoma, which was the same misdiagnosis as in the preoperative needle biopsy. Although we did not know which operation is optimal for primary pulmonary chordoid meningioma, the operation was completed as wedge resection based on the incorrect frozen biopsy result.

Primary pulmonary meningioma should be distinguished from metastatic disease. Because meningioma in the central nervous system can metastasize to the lung, albeit rarely, radiologic studies of the brain and spine should be evaluated to rule out metastatic pulmonary meningioma. In our case, postoperative brain MRI showed no intracranial lesion. However, spinal MRI was not performed. The preoperative PET-CT scan showed the pulmonary nodule and no definitive lesion in the spine. We utilized the PET-CT scan instead of spine MRI to assess whether a lesion was present in the spine.

Chordoid meningioma is classified as World Health Organization (WHO) grade II (International Classification of Diseases for Oncology-3.1 code: 9538/1) due to its high recurrence rate [7]. A recent meta-analysis of 221 patients with chordoid meningioma reported that the 3-, 5-, and 10-year progression-free survival rates were 76.0%, 67.5%, and 54.4%, respectively, in the central nervous system [8]. Poor prognostic factors for tumor recurrence were subtotal resection and a relatively high MIB-1 label index ($\geq$5%), while adjuvant radiotherapy was not a prognostic factor [8]. It seems to be reasonable to perform a standard lobectomy for primary pulmonary chordoid meningioma because of the high rate of recurrence despite complete resection in the central nervous system. However, the surgical extent and completeness differ between neurosurgery and thoracic surgery. In thoracic surgery, a greater margin of safety can be obtained than in neurosurgery. There were no available analytic data for predicting recurrence in primary pulmonary chordoid meningioma. The 2 previous case reports of primary pulmonary chordoid meningioma described patients who underwent lobectomy. One reported presented 1-year follow-up data with no recurrence, and the another report did not show any follow-up data [2,3]. Twenty-three cases of primary pulmonary benign meningioma (WHO grade I) showed no recurrence. However, 2 patients with primary pulmonary malignant meningioma (WHO grade III) relapsed after lobectomy and pneumonectomy [1]. In our case, a limited wedge resection was performed because the preoperative needle biopsy and intraoperative frozen biopsy samples were misdiagnosed as hamartoma. The distance from the tumor edge to the resection margin was 5 mm. Wedge resection without nodal dissection might be an incomplete surgical procedure for these histological findings in view of the high recurrence rate in the central nervous system. However, data were lacking regarding the primary lung origin of this tumor. A multidisciplinary team fully discussed this issue and decided to perform a chest CT evaluation every 6 months without any adjuvant therapy. A further central nervous system evaluation was not performed because this case was regarded as primary lung cancer with a rare histological pattern. During the 26-month follow-up period, there was no recurrence on chest CT. The authors therefore make the careful suggestion that limited resection with a sufficient margin could be a viable surgical option for primary

pulmonary chordoid meningioma.

In conclusion, primary pulmonary chordoid meningioma is an extremely rare entity that is difficult to diagnose preoperatively. The treatment of primary pulmonary chordoid meningioma has not been well studied. We report the case of a patient with primary pulmonary chordoid meningioma who underwent complete resection by VATS wedge resection and showed no recurrence during a 26-month follow-up period. Further close, long term follow-up is needed due to the aggressive nature of chordoid meningioma.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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