

## MENINGOTHELIOID NODULES COEXISTING WITH PULMONARY HYDATID CYST: A RARE CASE REPORT

### *Pulmoner Hidatid Kist ile Birlikte Bulunan Meningotelioid Nodüller: Nadir Bir Vaka Sunumu*

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#### ABSTRACT

Meningothelioid nodules are rare and usually benign pulmonary nodules. We present this case report because it represents the first reported case of an association between meningothelioid nodules and a pulmonary hydatid cyst.

A 48-year-old Turkish female patient presented with a cough and hemoptysis lasting for a week. Chest computed tomography images revealed a well-circumscribed, unilocular cystic lesion in the left lower lobe of the lung, displaying the typical “double-wall sign” of a hydatid cyst. It was decided that the patient would undergo thoracoscopic wedge resection of the hydatid cyst in the left lower lobe. Microscopic examination revealed multiple perivenular nodules composed of epithelioid cells with round to oval nuclei, a moderate amount of eosinophilic cytoplasm, and finely granular chromatin in hematoxylin-eosin sections. Whorling of tumor cells was observed focally in most areas. Some cells exhibited pseudonuclear inclusions. The surrounding lung parenchyma contained hydatid cysts. Based on these histopathological and immunohistochemistry findings, a diagnosis of a meningothelioid nodule and a hydatid cyst was made.

We encountered a rare case of coexisting meningothelioid nodules and a pulmonary hydatid cyst. Careful pathological, clinical, and radiological examination are required for the definitive diagnosis of meningothelioid nodules, and they can provide an excellent prognosis with surgery.

**Keywords:** Meningothelioid nodule, pulmonary chemodectoma, pulmonary hydatid cyst

#### ÖZ

Meningotelioid nodüller nadir görülen ve genellikle benign pulmoner nodüllerdir. Bu olgu sunumunu, meningotelioid nodüller ile pulmoner hidatid kist ilişkisini rapor eden ilk olgu olması nedeniyle sunduk.

48 yaşında Türk kadın hasta, bir haftadır süren öksürük ve hemoptizi şikayetiyle başvurdu. Toraks bilgisayarlı tomografi görüntülerinde akciğerin sol alt lobunda hidatid kist için tipik “çift kontür bulgusu” gösteren iyi sınırlı, uniloküler kistik lezyon görüldü. Hastaya sol alt lobdaki hidatid kist nedeniyle torakoskopik wedge rezeksiyon yapılmasına karar verildi. Mikroskopik değerlendirmede hematoksilen-eozin kesitlerinde yuvarlak-oval nükleuslu, orta genişlikte eozinofilik sitoplazmalı ve açık granüler kromatinli epitelioid hücrelerden oluşan çok sayıda perivenüler nodül görüldü. Çoğu bölgede fokal olarak girdaplanmış tümör hücreleri mevcuttu. Hücrelerin bazılarında psödonükleer inklüzyonlar izlendi. Çevredeki akciğer parankiminde hidatid kist görüldü. Bu histopatolojik ve immünohistokimyal bulgulara dayanarak meningotelioid nodül ve hidatid kist tanısı konuldu.

Meningotelioid nodüller ve pulmoner hidatid kistin bir arada bulunduğu nadir bir olguyla karşılaştık. Meningotelioid nodüllerin kesin tanısı için dikkatli bir patolojik, klinik ve radyolojik inceleme gereklidir ve cerrahi ile mükemmel bir prognoz sağlanabilir.

**Anahtar Kelimeler:** Meningotelioid nodül, pulmoner kemodektoma, pulmoner hidatid kist



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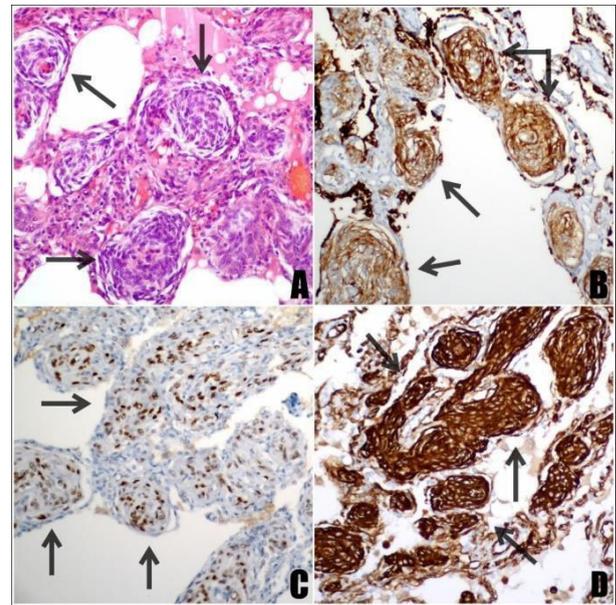
## INTRODUCTION

Meningothelioid nodules are rare pulmonary nodules that are usually benign and silent, often discovered incidentally at autopsy or in surgical specimens (1). They may mimic adenocarcinoma in situ, tumorlet, carcinoid tumor, and paraganglioma. The etiology of meningothelioid nodules remains unclear. This case report highlights the first documented instance of an association between meningothelioid nodules and pulmonary hydatid cysts. The potential link between these conditions is not well-understood, prompting further investigation. We present this case to bring attention to this unique association and to underscore the need for awareness and further research into the relationship between meningothelioid nodules and pulmonary hydatid cysts.

## CASE REPORT

A 48-year-old Turkish female patient with a history of heavy smoking visited our hospital complaining of cough and hemoptysis lasting for a week. Although the patient had hypertension, she denied any other symptoms. Physical examination and laboratory test results, including tumor markers, were normal. The patient exhibited no respiratory failure during the pulmonary function testing. Chest computed tomography images revealed a well-circumscribed, unilocular cystic lesion in the left lower lobe of the lung, displaying the typical “double-wall sign” indicative of a hydatid cyst. It was decided that the patient would undergo thoracoscopic wedge resection of the hydatid cyst in the left lower lobe. The resected material measured 6x5x3 cm, with the cut surface revealing well-circumscribed spherical cysts of 30 mm in diameter or larger. The cyst had a fibrous border and contained several daughter cysts. Additionally, several millimetric white nodules were randomly distributed throughout the lung parenchyma. The surgical specimens were fixed in 10% neutral formaldehyde and embedded in paraffin. Subsequent staining with hematoxylin-eosin and examination under an optical microscope revealed multiple perivascular nodules composed of epithelioid cells with round to oval nuclei, a moderate amount of eosinophilic cytoplasm, and finely granular chromatin. Whorling of tumor cells was seen focally in most areas (Figure 1A). Some cells displayed pseudonuclear inclusions, but there was no evidence of necrosis or mitotic figures. The surrounding lung parenchyma showed cysts with inner (protoscolices), middle (germinal membrane) and outer (acellular laminated membrane) layers, with no evidence of interstitial fibrosis or dysplastic changes in the bronchial or alveolar epithelium. Immunohistochemical profiling indicated that the nodules were positive for epithelial membrane antigen, progesterone receptor, and vimentin,

and negative for cytokeratin AE1 /AE3, synaptophysin, chromogranin, CD56, and S100 protein (Figure 1B,C,D). Based on these histopathological and immunohistochemical findings, a diagnosis of meningothelioid nodule and hydatid cyst was made. The patient has remained free from recurrence one year after surgery.



**Figure 1:** (A) Histopathologic examination findings. Photomicrograph obtained by thoracoscopic wedge resection shows epithelioid cells arranged in a whorled pattern (arrows) (H&E staining). Immunohistochemical staining for (B) epithelial membrane antigen (EMA), (C) progesterone receptors (PR), and (D) vimentin shows positive staining of meningothelial cells (arrows). (Original magnification, X200.)

## DISCUSSION

Korn et al. described these nodules as tumors resembling chemodectomas, due to their characteristic microstructure, cytological features, and presence adjacent to vessels (2). Later, immunohistochemical staining of these cells, showing positivity for epithelial membrane antigen, progesterone receptor, and vimentin, supported the notion of meningeal derivative rather than the neuroendocrine origin (3). More recently, Lonescu et al. found that meningothelioid nodules and intracranial meningiomas were unrelated (4). Further investigations by Niho et al. and Lonescu et al. suggested that meningothelioid nodules were reactive rather than neoplastic lesions (4,5). Currently, the significance or origin of meningothelioid nodules remains unknown. This case represented the first reported instance of coexisting meningothelioid nodules and pulmonary hydatid cyst, leaving it unclear whether there is a relationship between meningothelioid nodules and pulmonary hydatid cysts. Most patients are asymptomatic, with the reported incidence of these nodules is 7-13.8% at surgical resection (3,6).

Meningothelioid nodules are observed more often in females, affecting patients aged 22 to 84 years (mean age 62) (3). The underlying pathogenesis is unclear, and they are more frequently noted in patients with underlying chronic lung disease (3). Asakawa et al. found that among patients with meningothelioid nodules, 64% had adenocarcinoma, 7% had squamous cell carcinoma, 7% had atypical adenomatous hyperplasia, and 21% had metastatic lung tumors from colorectal cancer (7). Additionally, meningothelioid nodules were found more frequently in patients with malignant lung tumors (7.3%) than in those with benign tumors, according to Mizutani et al. (6). The lesions are most often solitary or few, with diffuse bilateral nodules being extremely rare (1). Meningothelioid nodules tend to be located in the peripheral zone and even on the interlobar pleura (8). They can occur in all pulmonary lobes, with a similar incidence frequency between lobes (6). Usually benign, surgery can provide an excellent prognosis for meningothelioid nodules. Microscopic examination and differential diagnosis considerations include tumorlet, carcinoid tumor, meningioma, and paraganglioma. Tumorlet and carcinoid tumors exhibit higher nuclear-cytoplasmic ratios, a more stippled chromatin pattern, and more elongated shapes. Immunohistochemically, tumorlets and carcinoid tumor show weak cytoplasmic reactivity for cytokeratin and are also immunoreactive with chromogranin, synaptophysin, and CD56. Lesions smaller than 5 mm are termed tumorlets, while those larger than 5 mm are termed carcinoid tumors. These histopathological and immunohistochemical features distinguish tumorlets and carcinoid tumor from meningothelioid nodules. Pulmonary meningiomas are clonal neoplastic lesions, usually presenting as lung masses rather than minute nodules. Unlike paraganglioma, sustentacular cells are absent in the meningothelioid nodule, and there is no expression for S100 protein.

In conclusion, we encountered a rare case of coexisting meningothelioid nodules and pulmonary hydatid cysts. Neuroendocrine tumors should be excluded before diagnosing a meningothelioid nodule, as misdiagnosis can lead to irreversible results due to differences in treatment strategies. Careful pathological, clinical, and radiological examination is required for a definitive diagnosis of meningothelioid nodules, which can provide an excellent prognosis with surgery.

*Conflict of Interest:* All authors declare that there is no conflict of interest.

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