

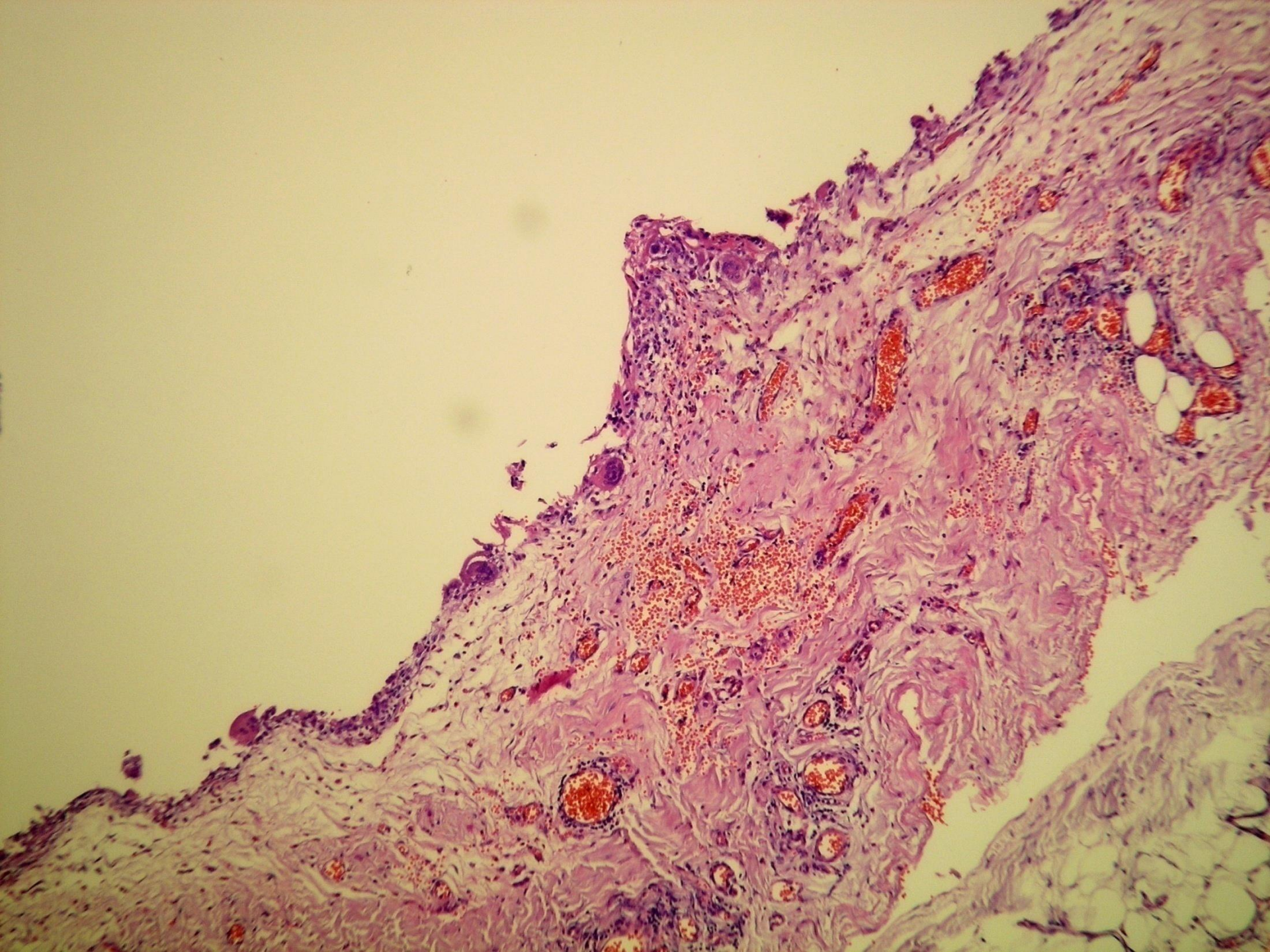
SD-IAP č. 489

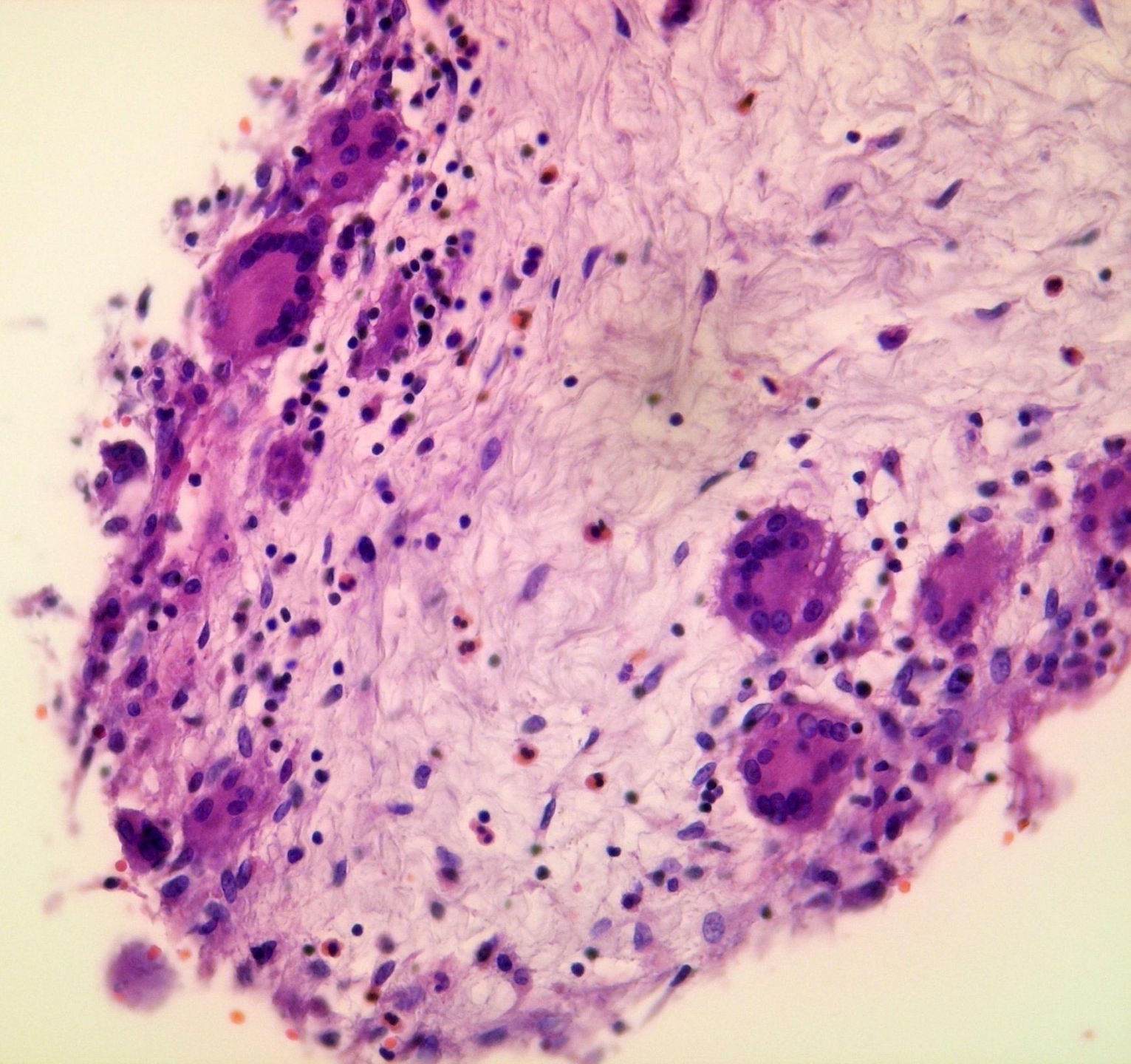
Marián Švajdler ml

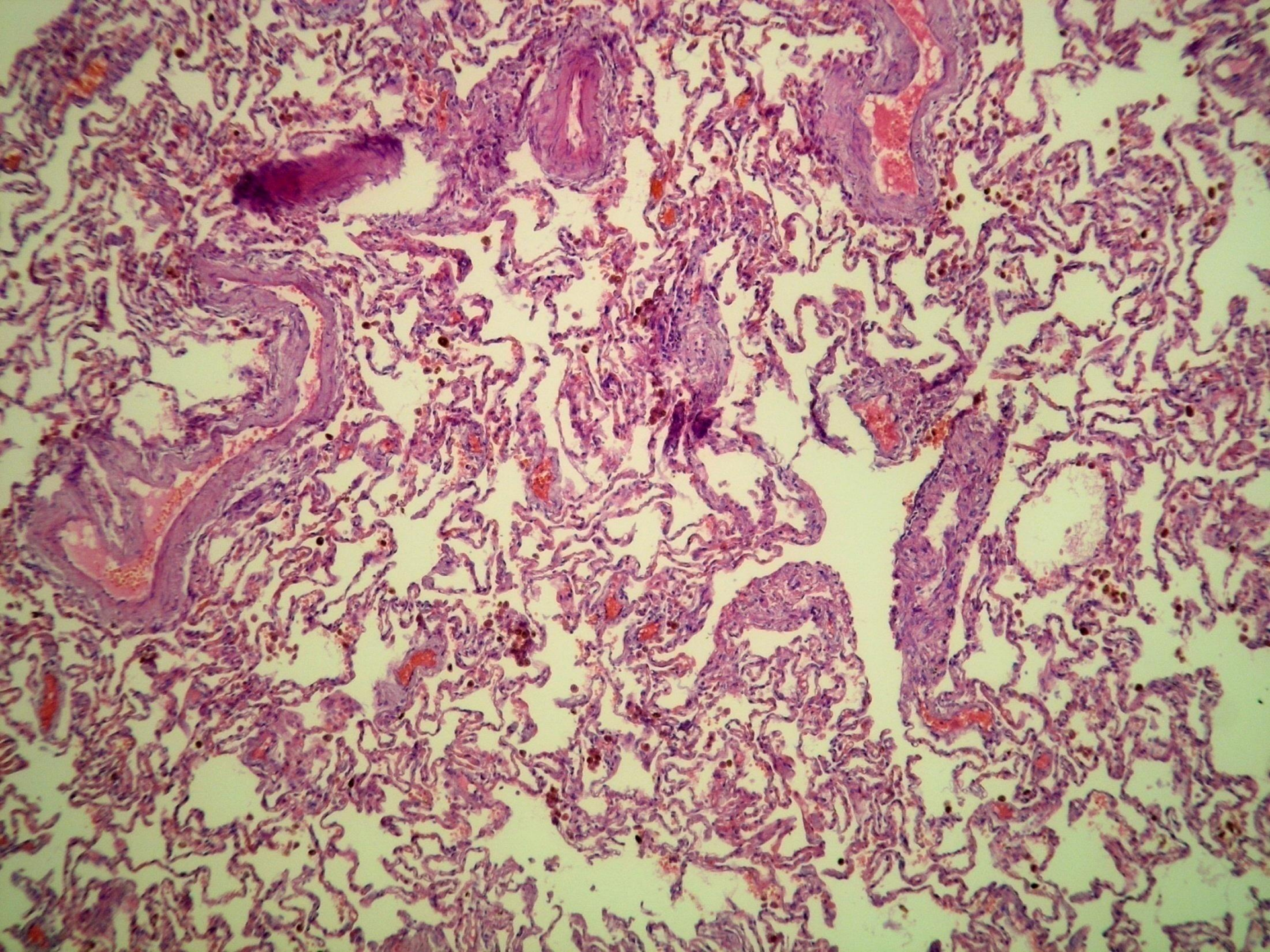
Oddelenie patológie UNLP Košice

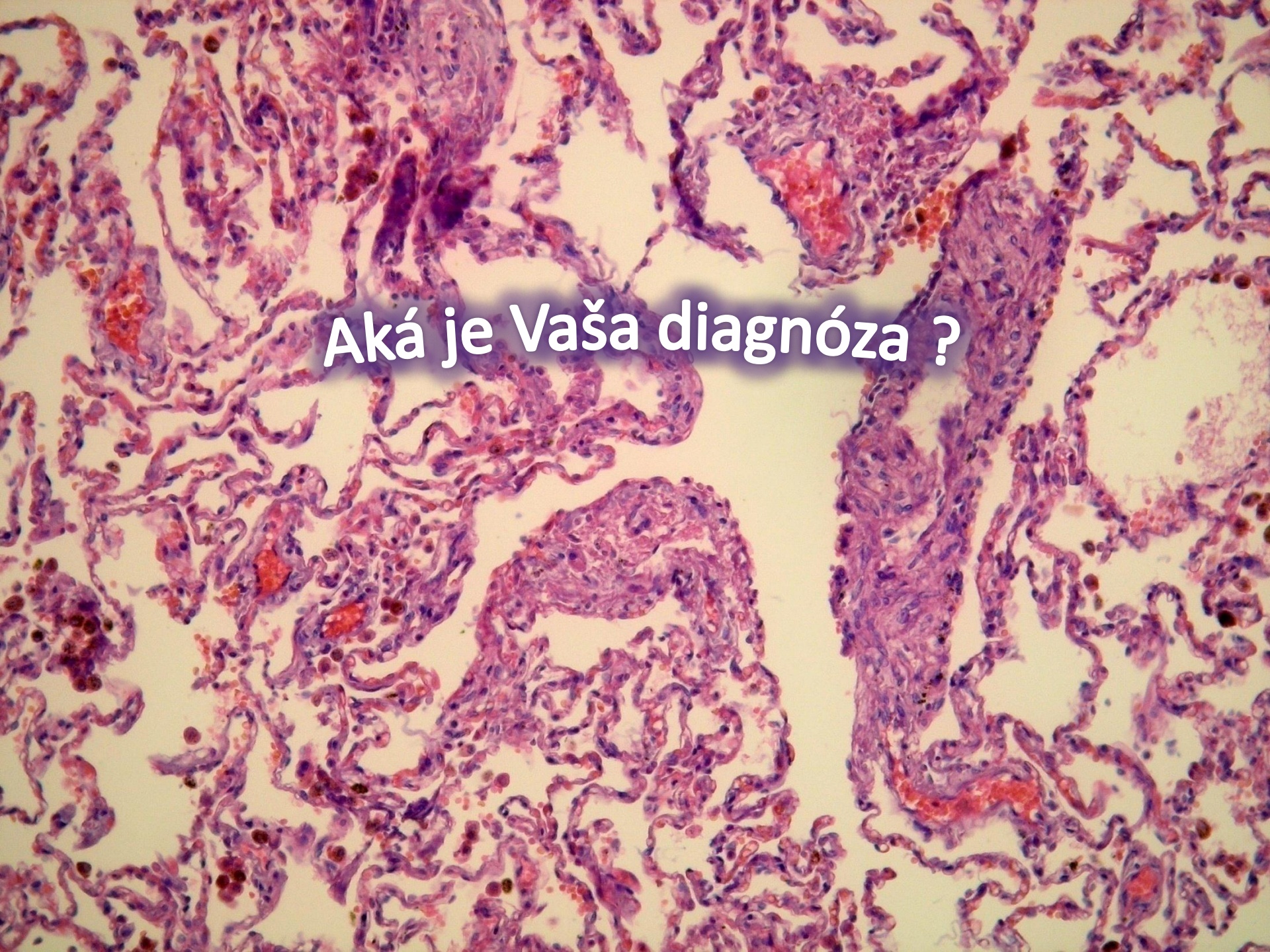
Klinické údaje

- 45 ročná žena
- J93.1 - iný spontánny pneumotorax
- na vyšetrenie zaslaná excízia parenchýmu pľúc s bulou, excízia z apexu pľúc a excízia z parietálnej pleury

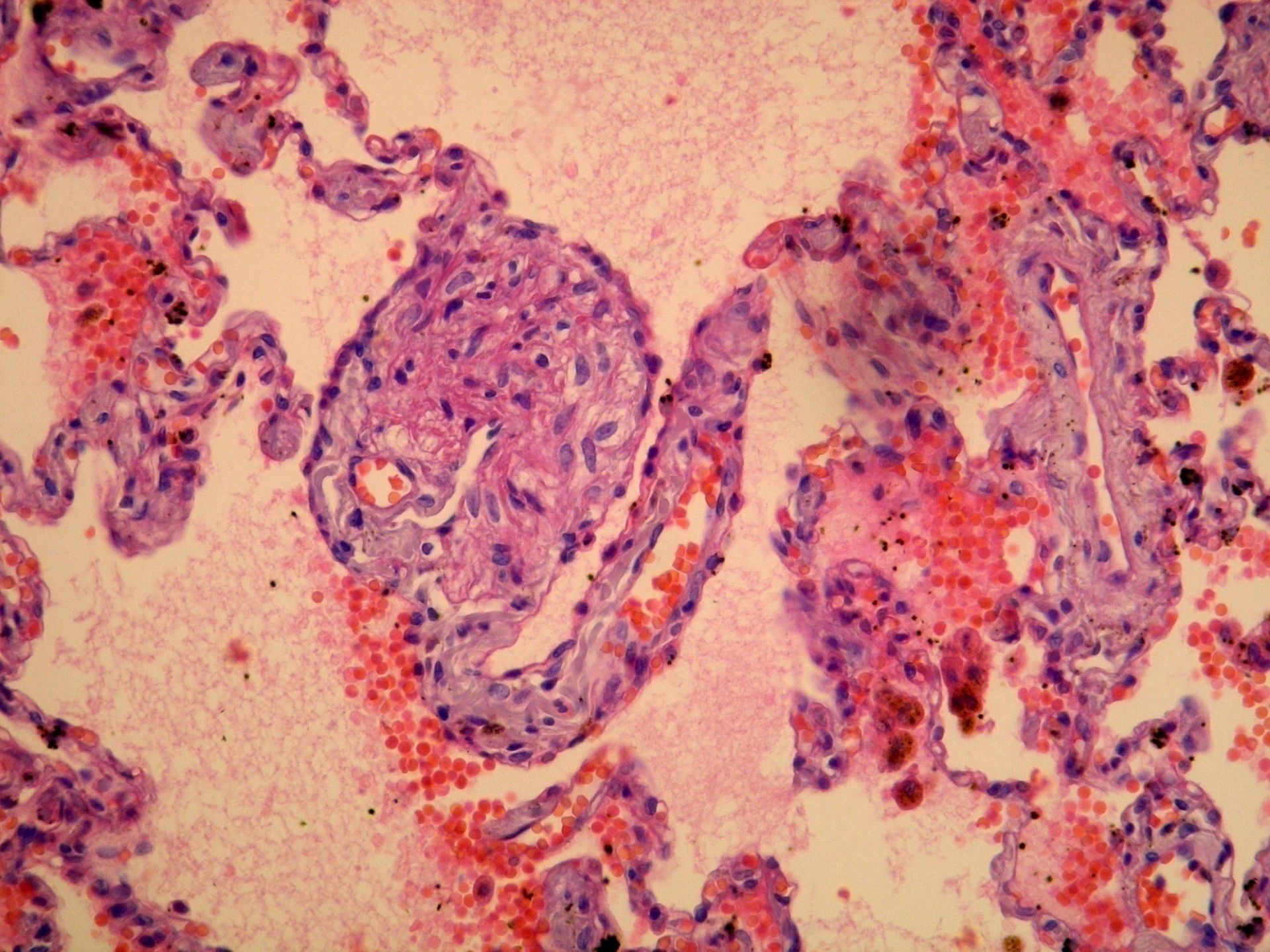


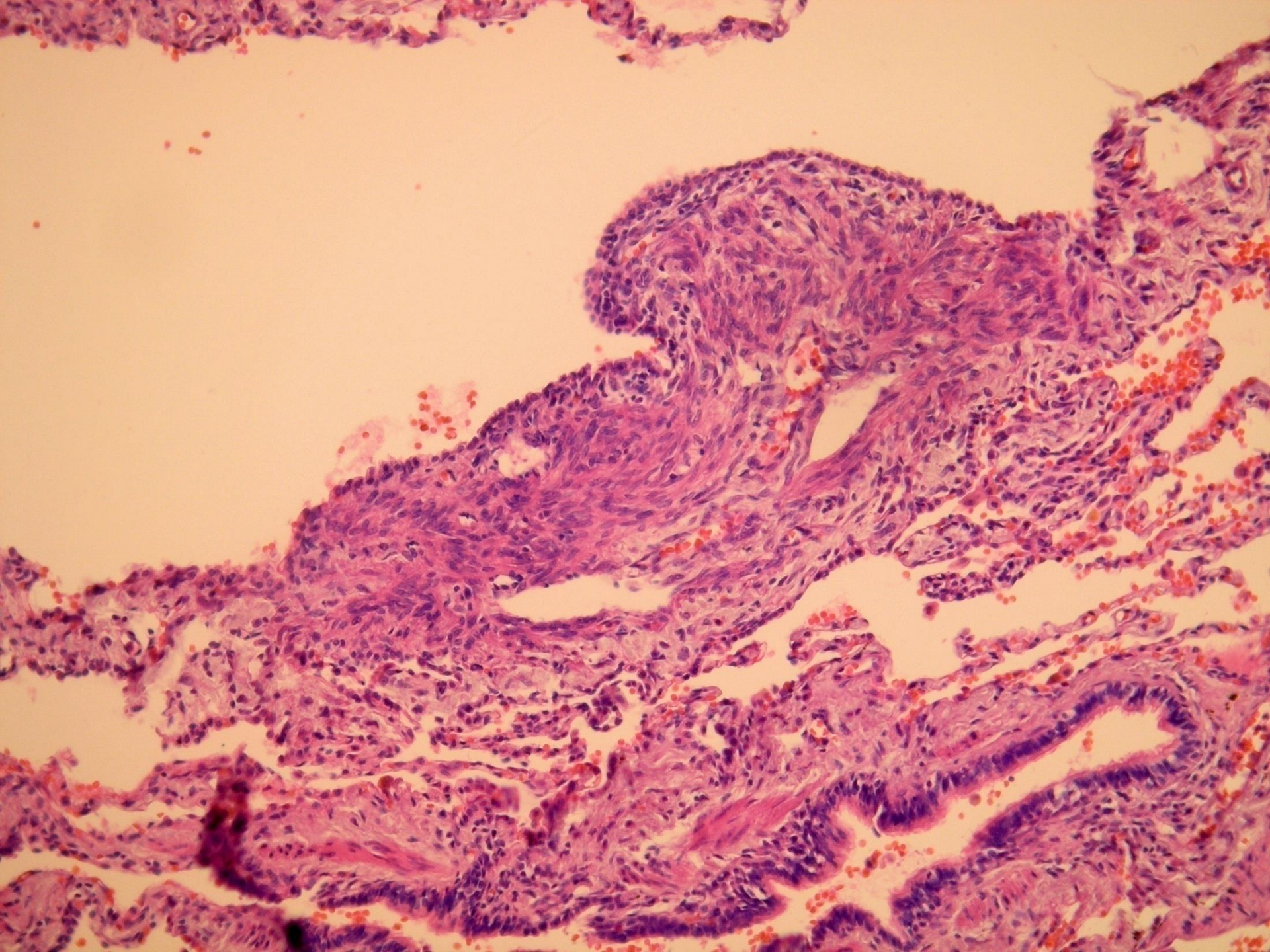


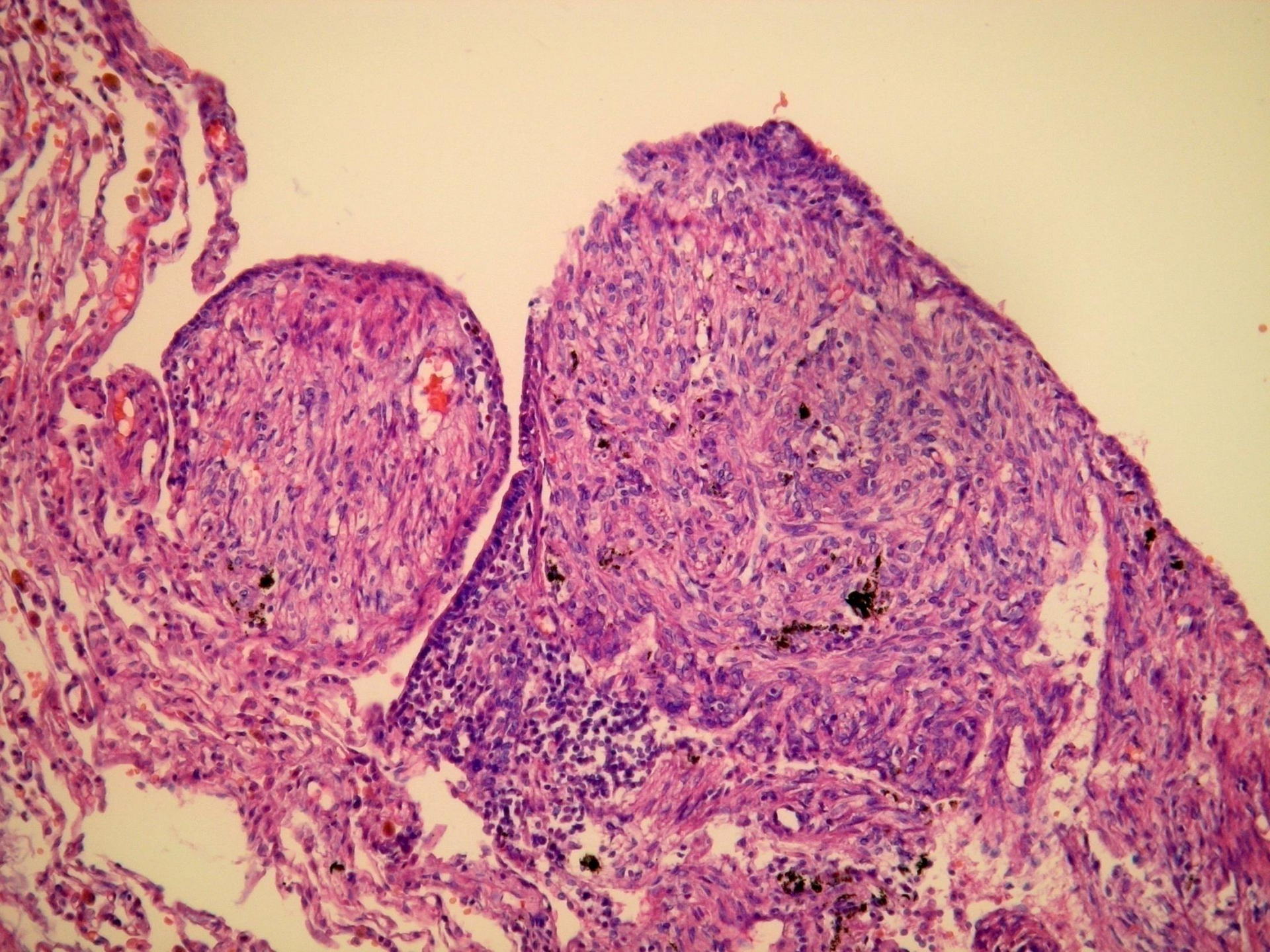


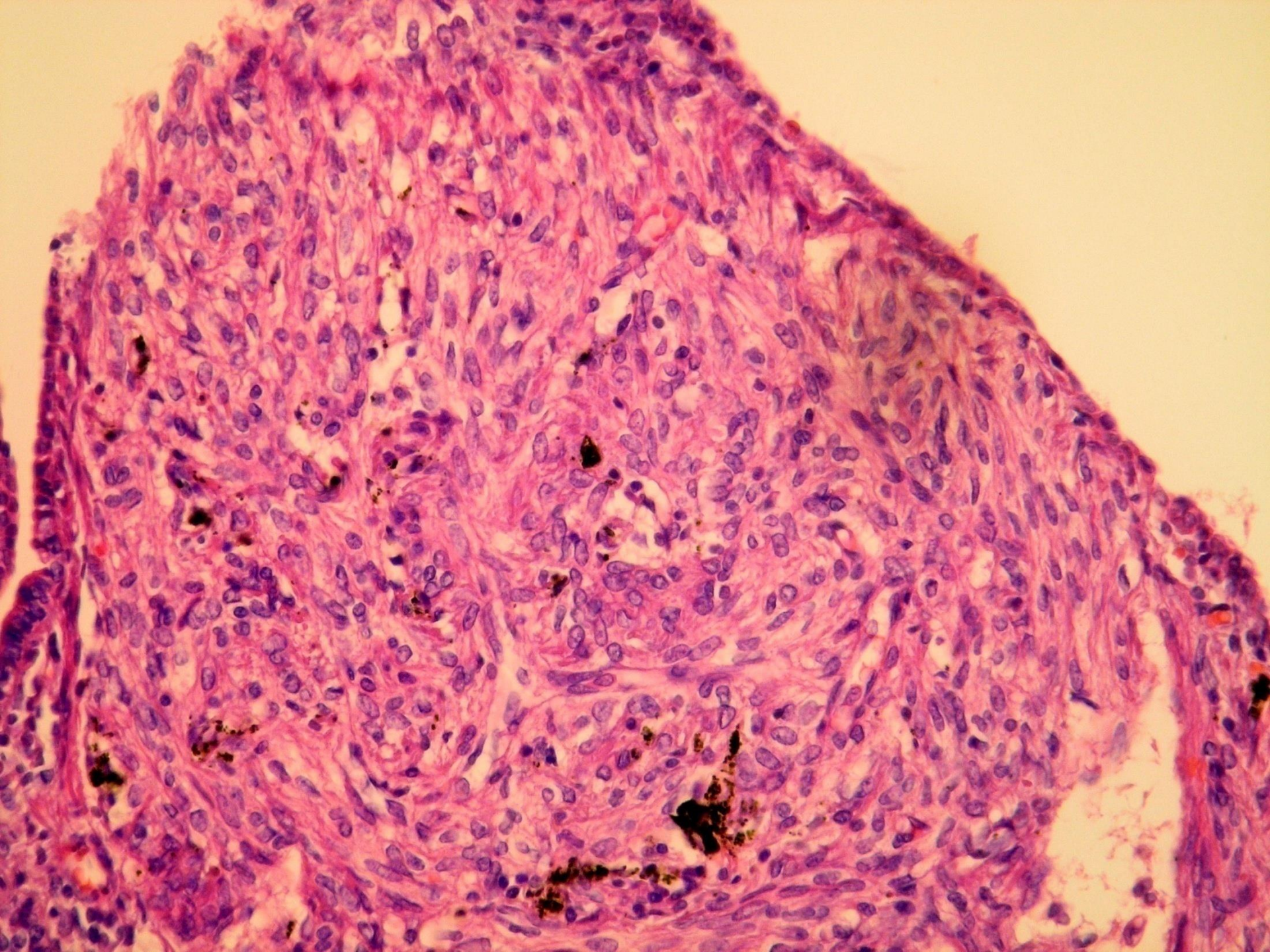
A histological section of placental tissue stained with hematoxylin and eosin (H&E). The image displays numerous chorionic villi, which are finger-like projections of the placenta. The villi are lined by a layer of trophoblastic cells, appearing as a pinkish-red outer layer. The interior of the villi contains fetal blood vessels, with red blood cells visible as small, round, red-stained cells. The surrounding decidua (the lining of the uterus) is also visible, showing a dense network of connective tissue and blood vessels. There is a prominent area of decidual hemorrhage, characterized by a large, irregular, red-stained mass in the upper right quadrant, indicating bleeding within the decidua. The overall appearance is that of a normal placental structure with some hemorrhagic changes.

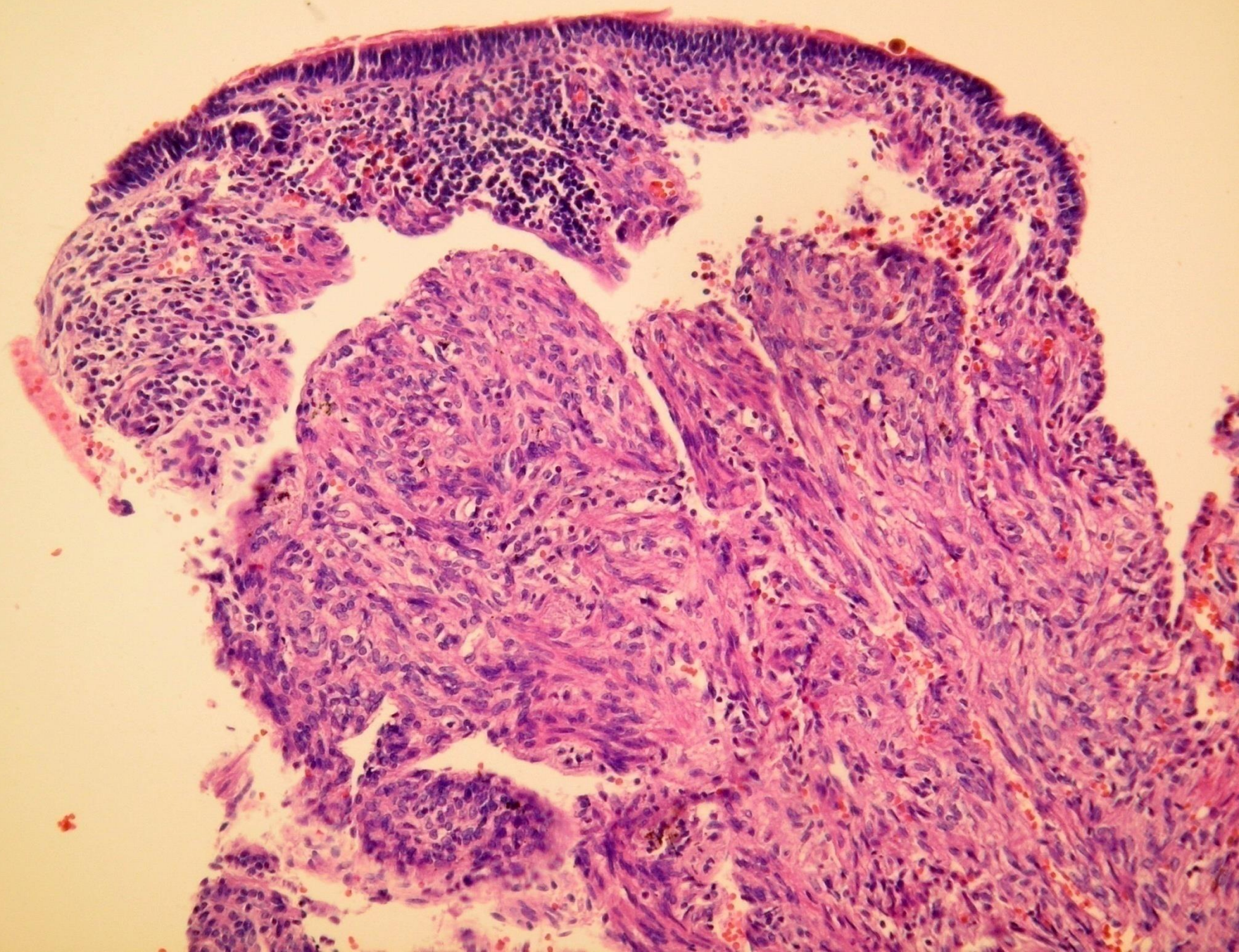
Aká je Vaša diagnóza ?

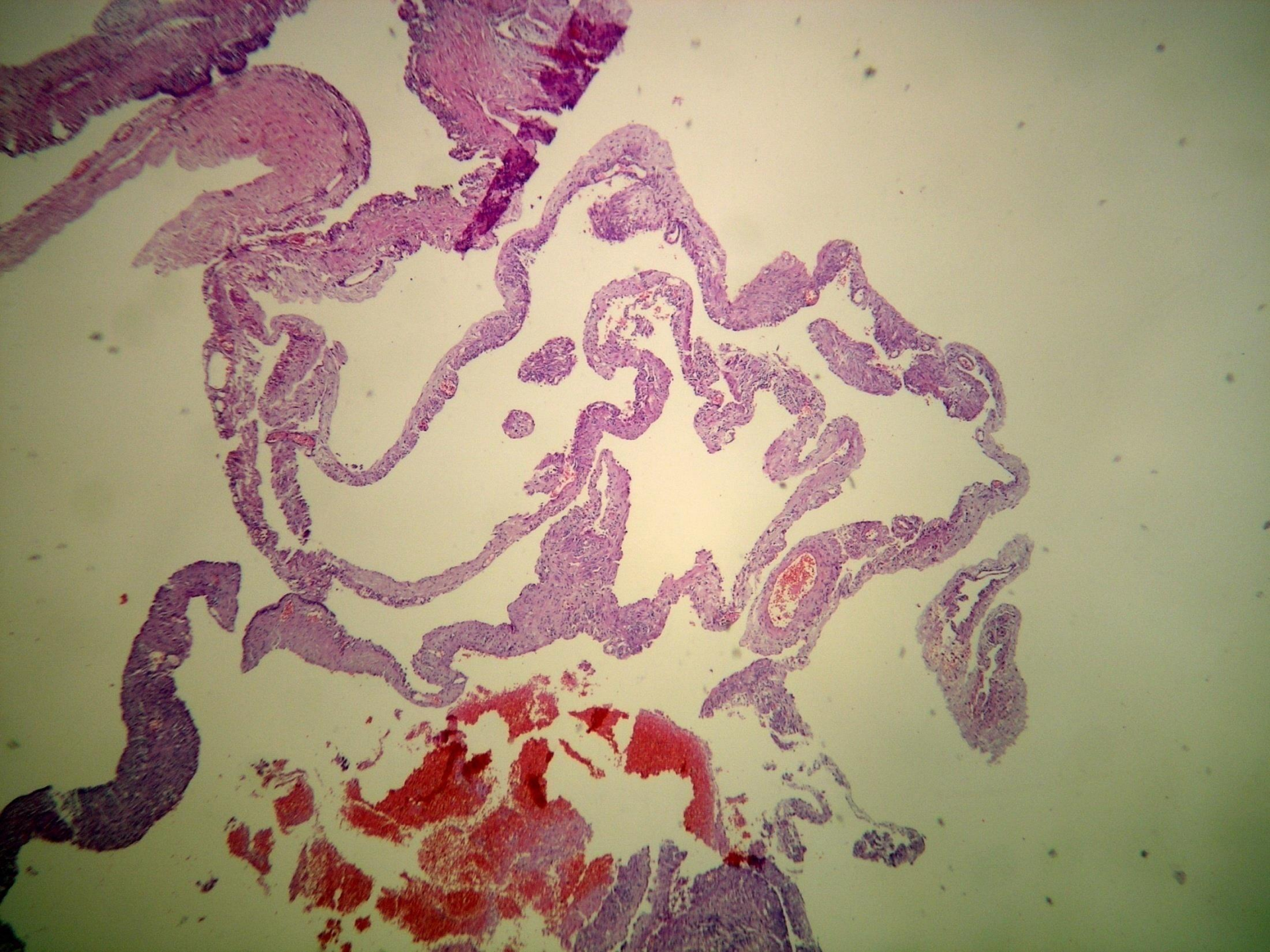


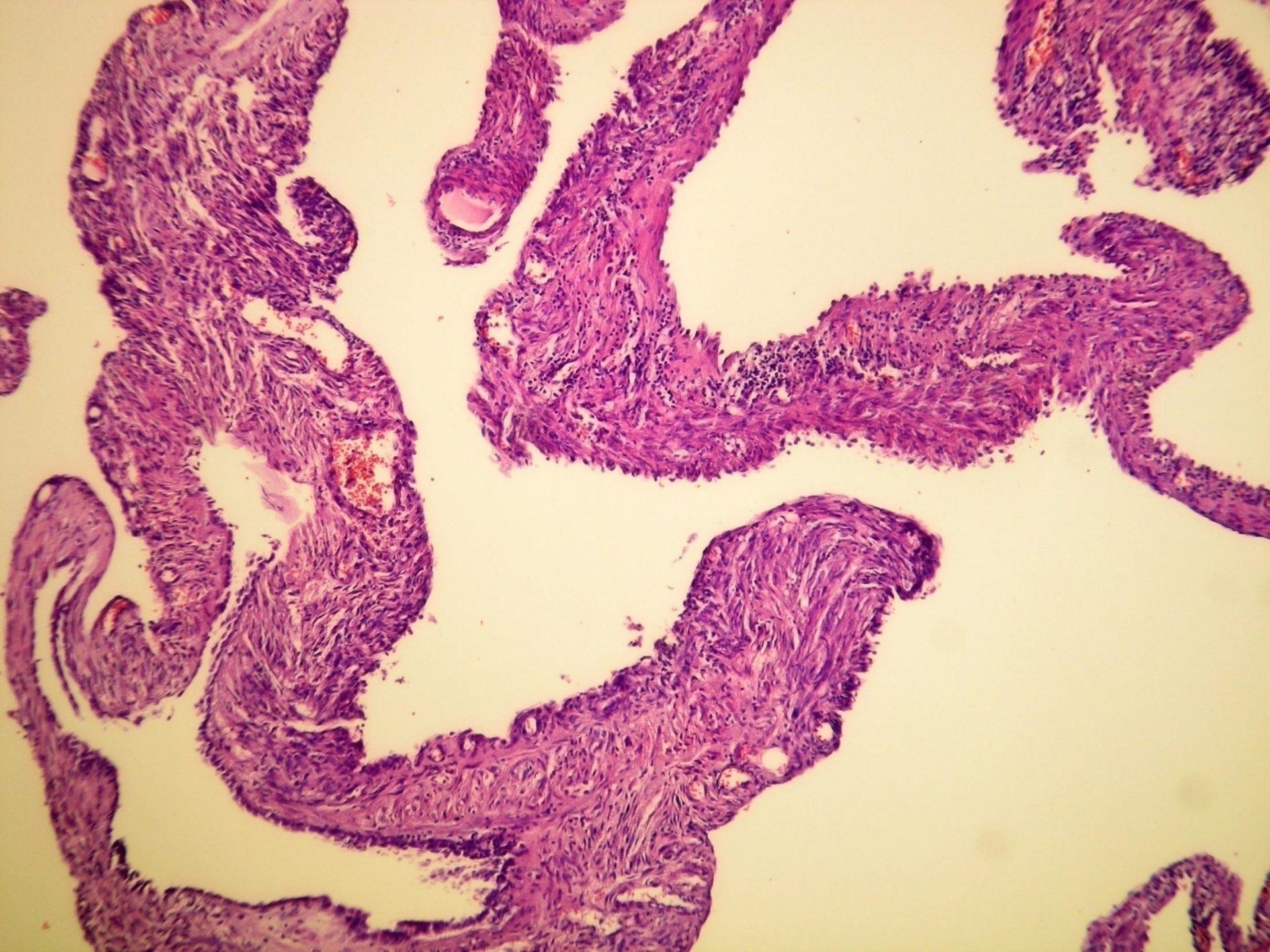


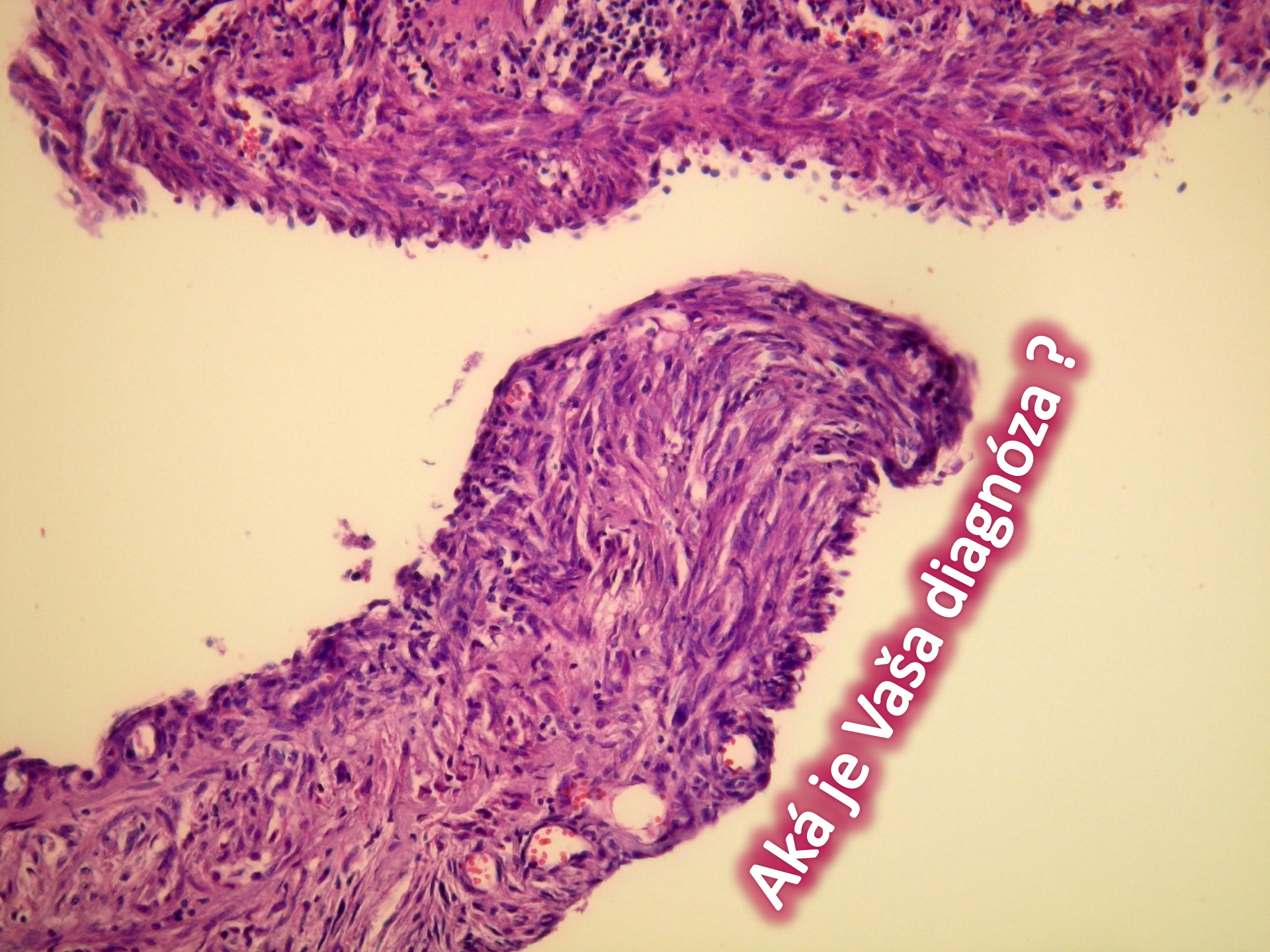






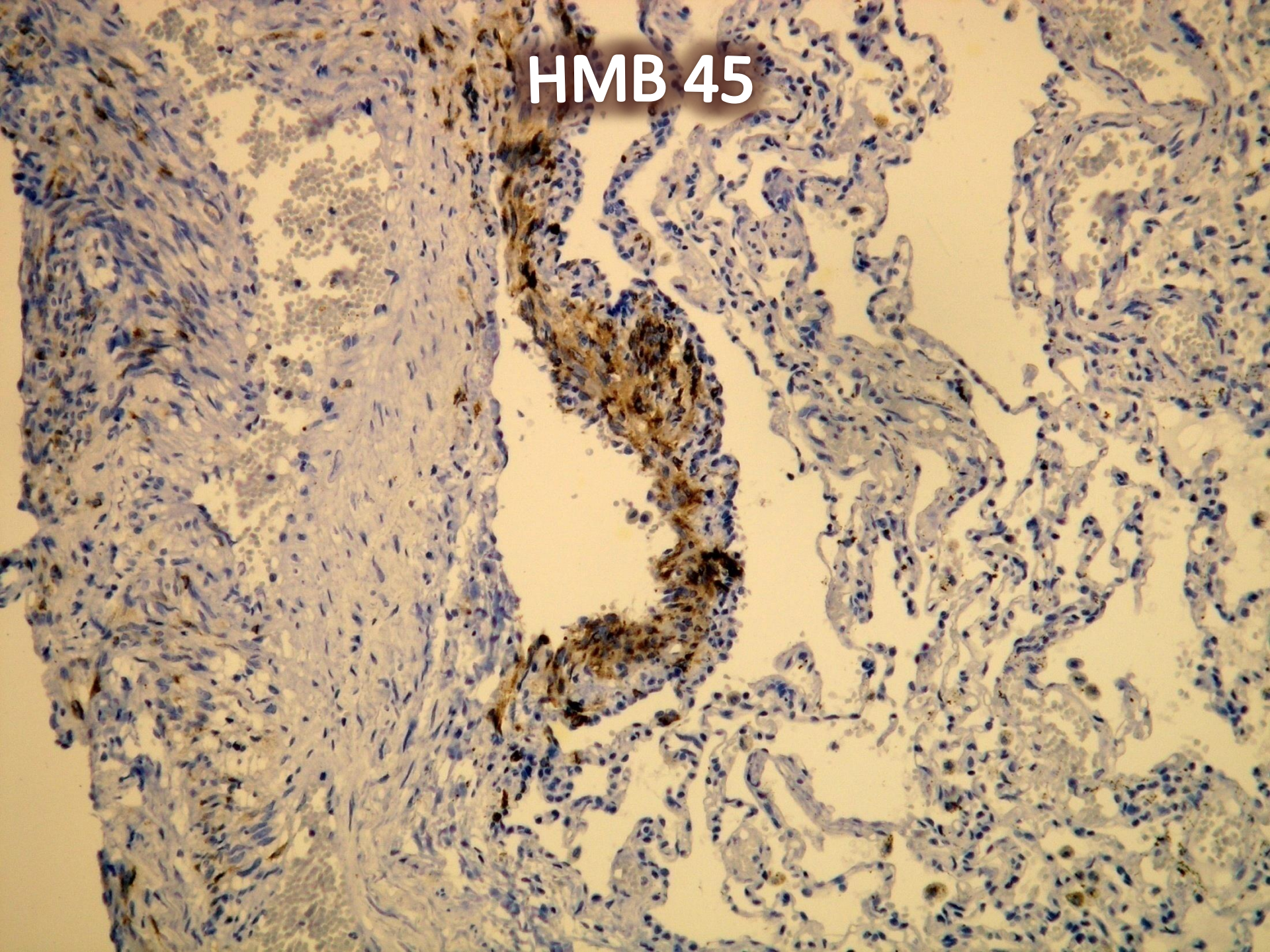




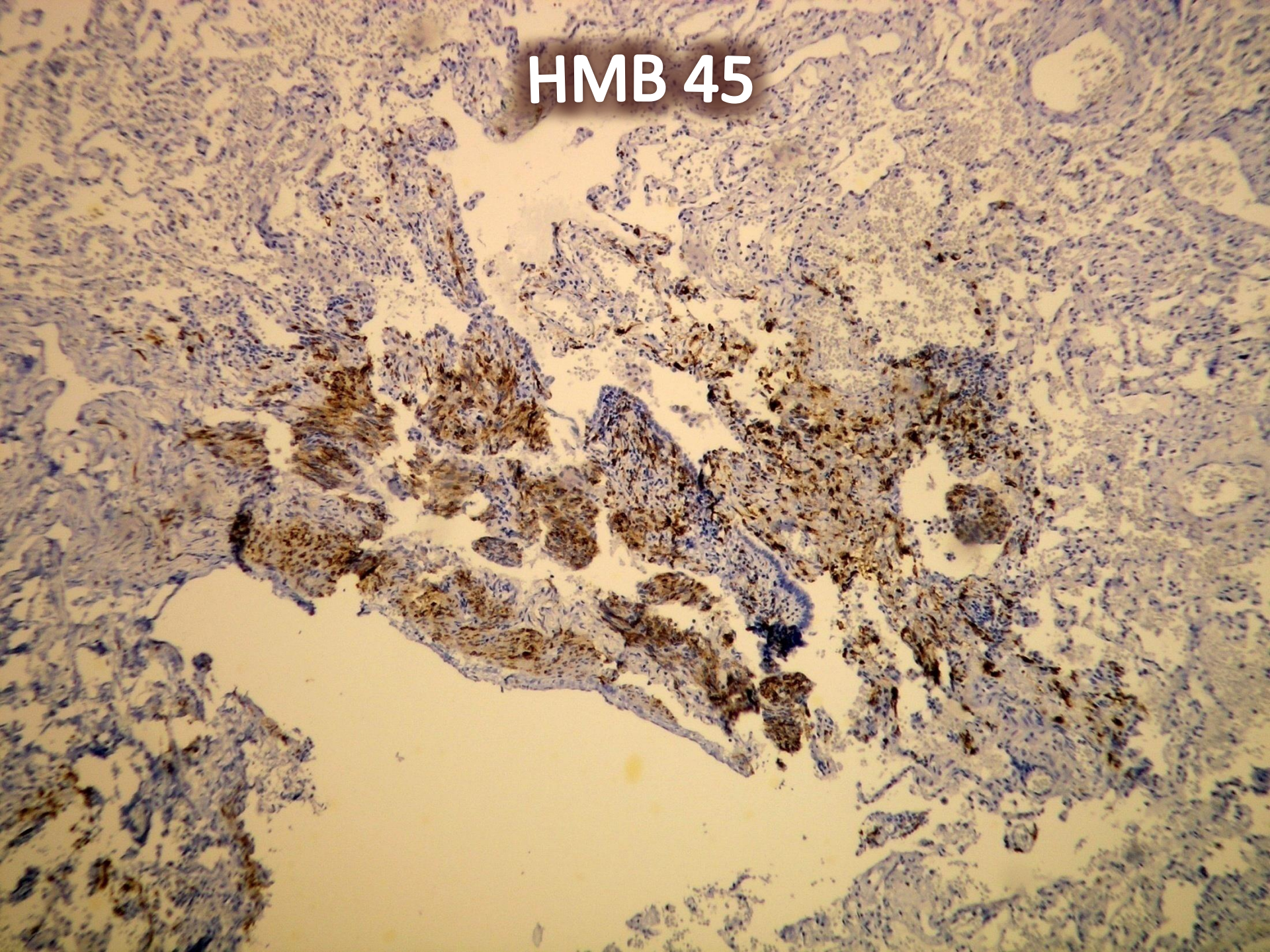


Aká je Vaša diagnóza ?

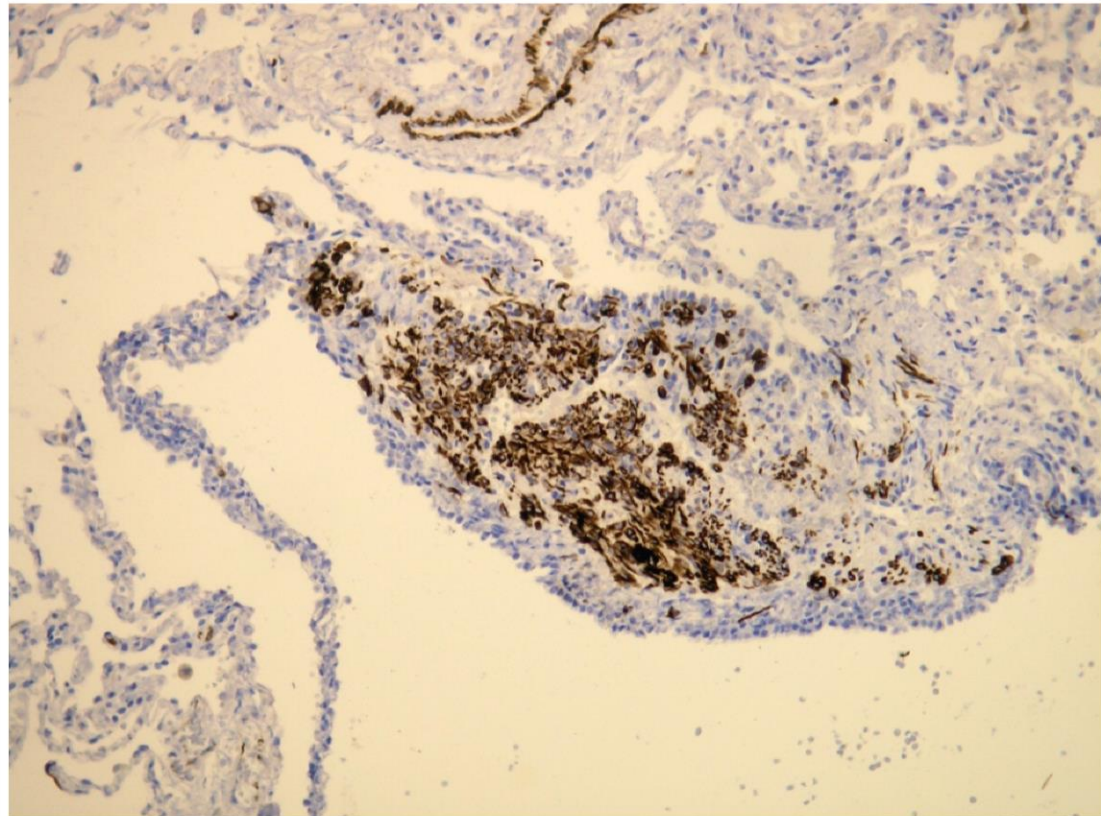
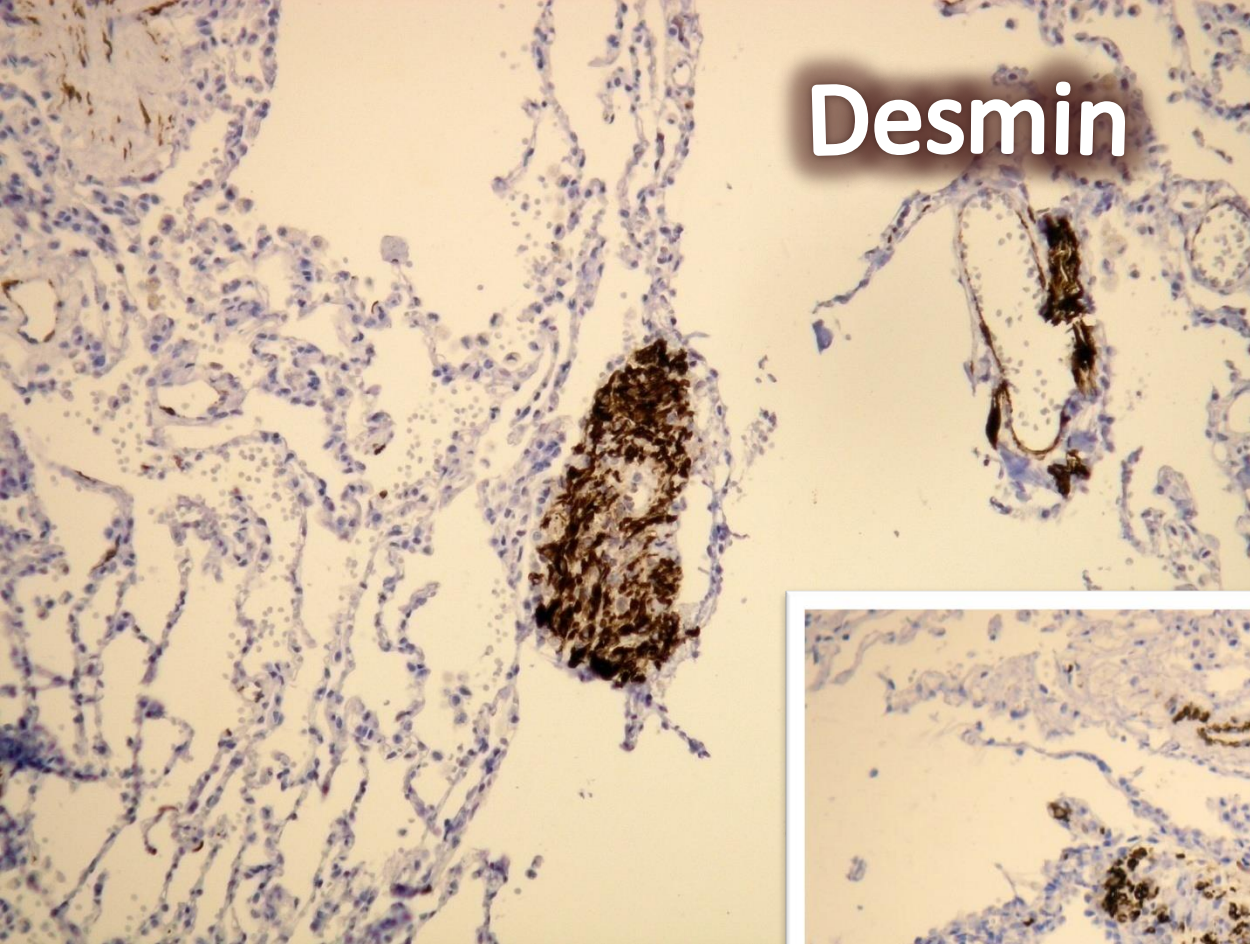
HMB 45

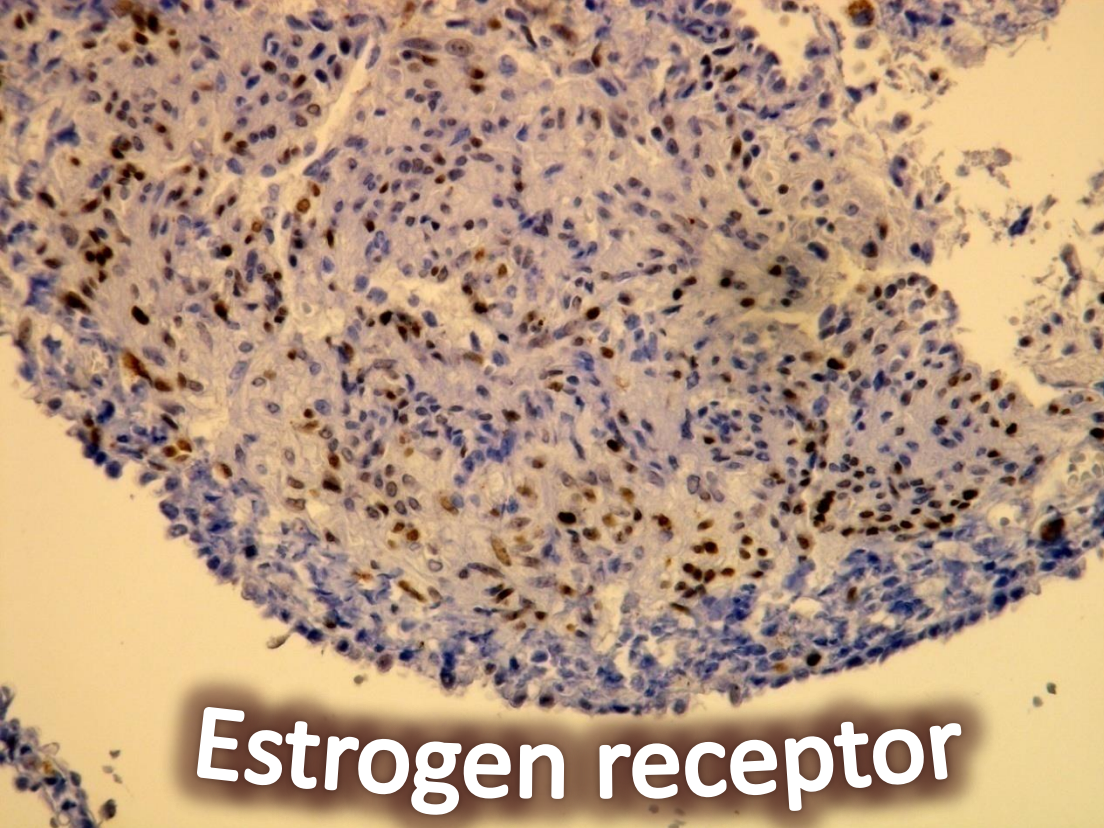


HMB 45

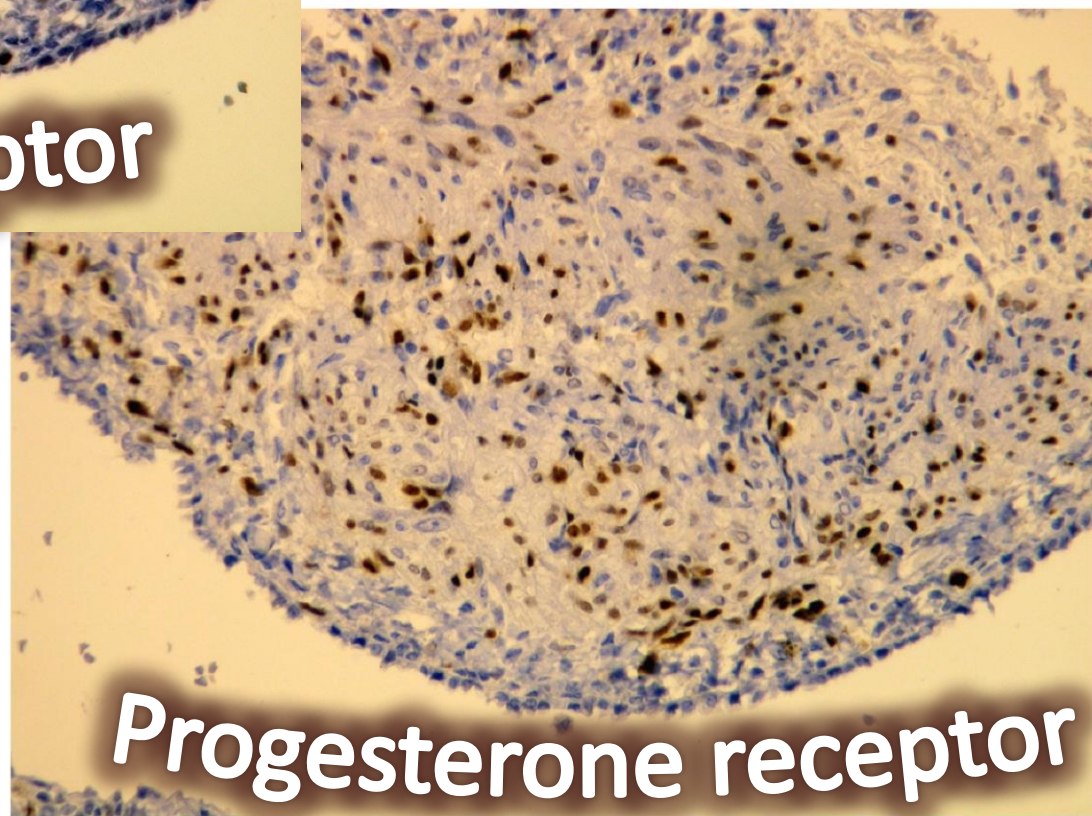


Desmin





Estrogen receptor



Progesterone receptor

Pulmonary Lymphangioleiomyomatosis

PLAM

LAM: Epidemiológia

- najčastejšie ženy v reprodukčnom veku
- incidencia 1 na 1 milión
- sporadický výskyt
- 30-40% pacientov so syndrómom TSC
- častá koincidencia s renálnymi AML (50%)
- tretia najčastejšia príčina úmrtia pri TSC
- zvýšená incidencia meningeómov

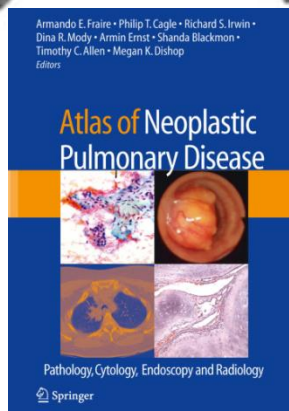
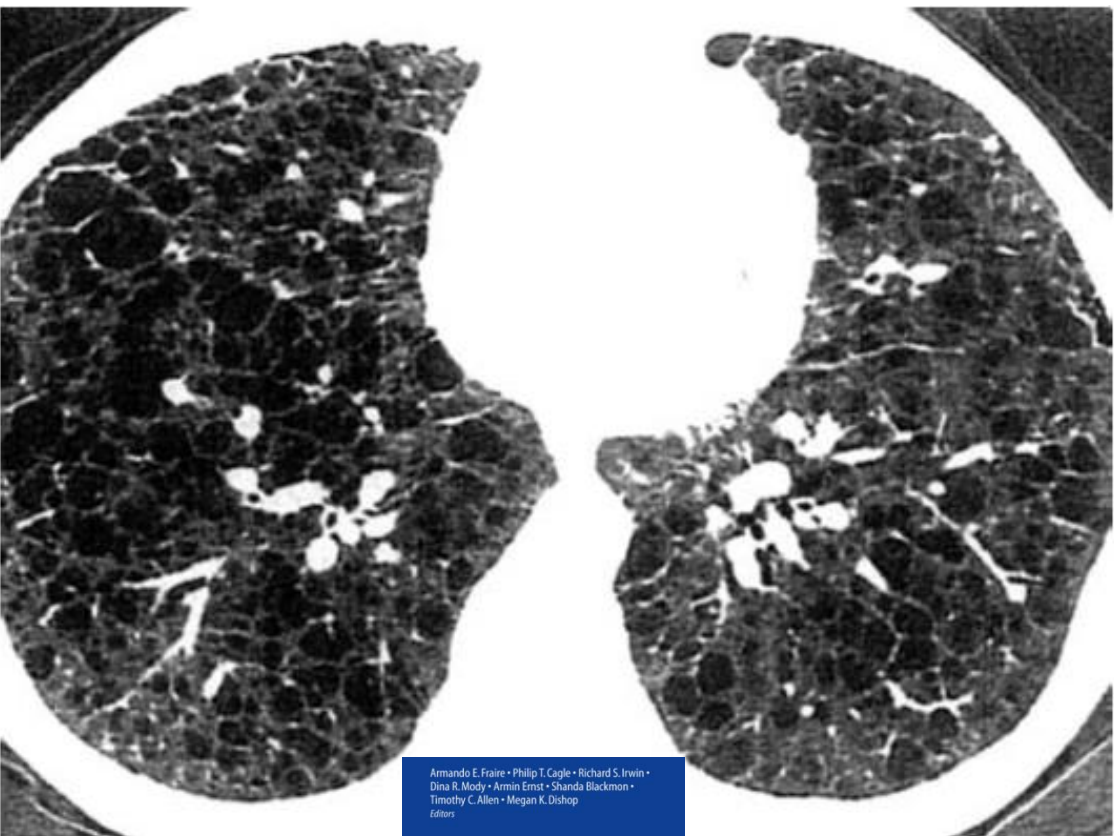
LAM: Klinické príznaky

- progresívne dyspnoe
- rekurentný pneumotorax
- hemoptýza
- chylózne výpotky

LAM: Histopatológia

- proliferácia vretenitých myoidných buniek (**proliferatívna lézia**)
- v ktorejkoľvek časti pľúc, prevažne v lymfatickej distribúcii
- subpleurálne, septá, interstícium, bronchovaskulárne zväzky, pľúcne vény, ductus thoracicus, torakálne a retroperitoneálne LU
- cystické priestory vystlané myoidnou proliferáciou (**deštruktívna lézia**)
- SMA, Desmin
- HMB-45, Melan A, MITF, tyrozináza (nie všetky bb. sú +)
- ER, PgR +/-

LAM: HRCT

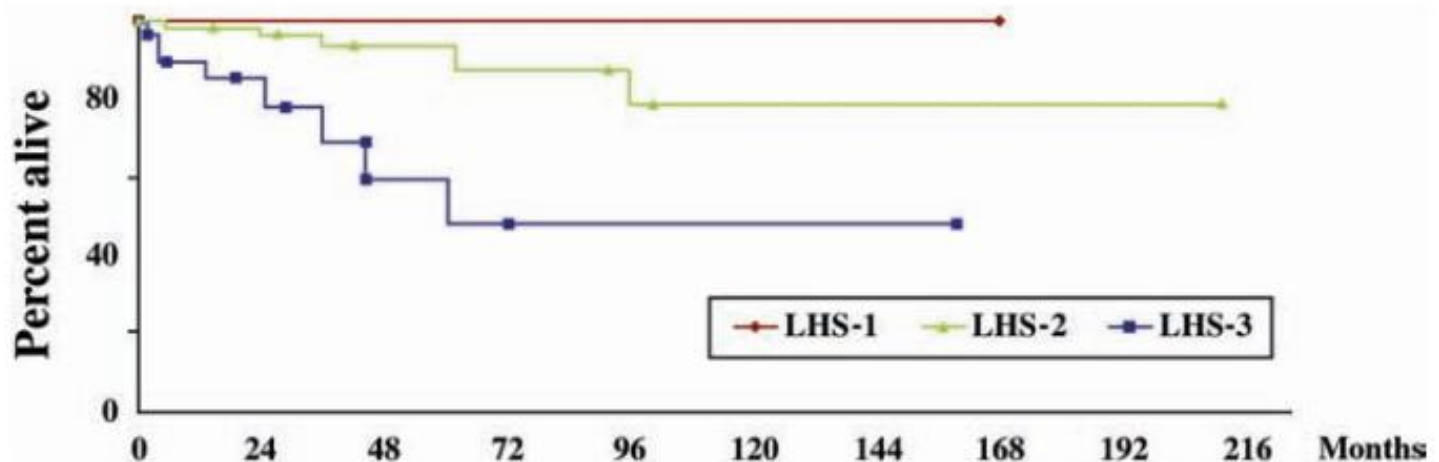


LAM: patogenéza

- lézia z rodiny tzv. PEC-ómov
 - zárodočná alebo somatická mutácia TSC1 / TSC2
 - úloha hormónov ?
 - produkcia matrix-metaloproteináz
 - lymfangiogenéza
-
- migrácia PEC buniek z renálneho AML?
 - rekurencia po transplantácii pľúc

LAM: prognóza

- medián prežívania 8-10 rokov
- Matsui K et al. Prognostic significance of pulmonary lymphangioleiomyomatosis histologic score. *Am J Surg Pathol* 2001, 25: 479



LAM: liečba

- hormonálna manipulácia (tamoxifén?, odstránenie ovárií, agonisti gonadotropin-releasing hormónu)
- transplantácia pľúc
- Experimentálna terapia
 - Rapamycin, Everolimus
 - Doxycyclin
 - Inhibítory aromatázy (Letrozol)

Ďakujem za pozornosť

svajdler@yahoo.com