Prípad SD-IAP č. 690

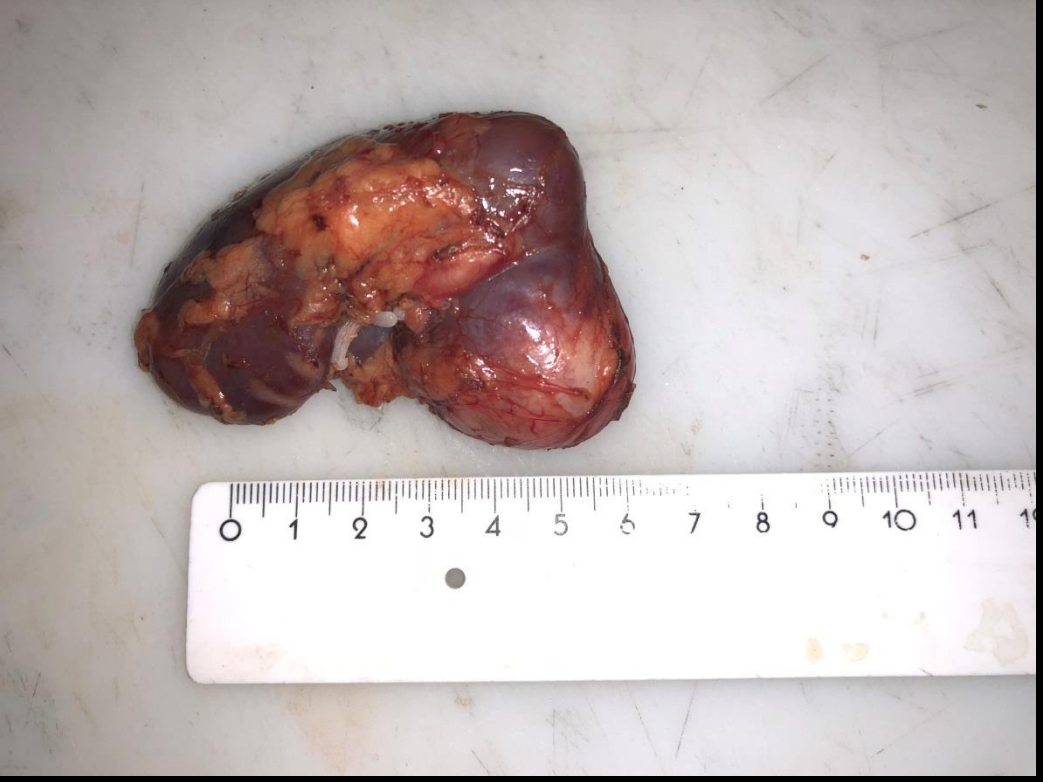
B. Rychlý
Cytopathos, Bratislava

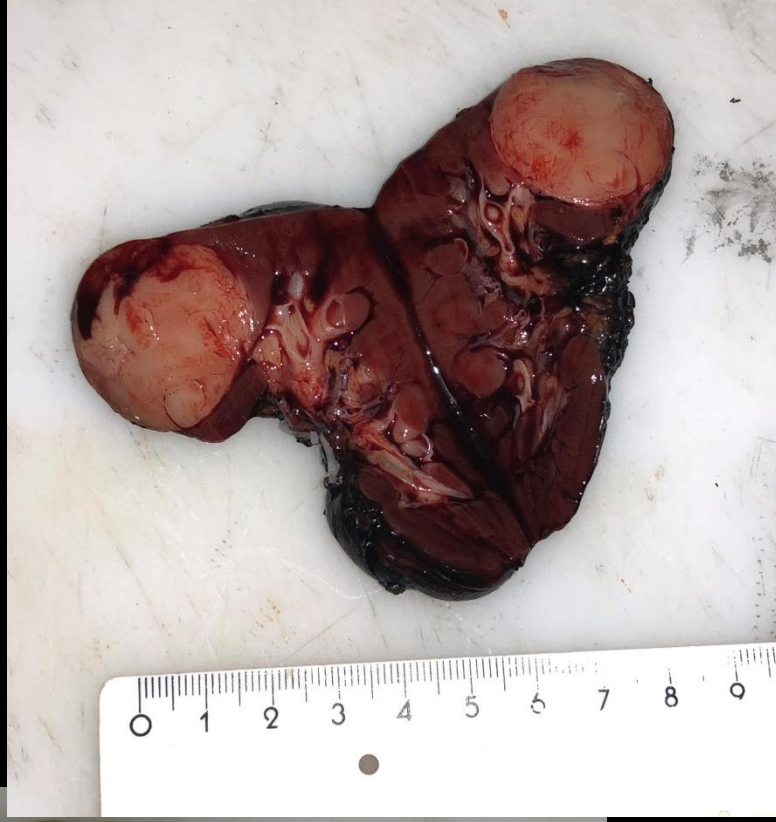
Klinické údaje

- 4-ročné dievča
- bolesti brucha, susp. akútna apendicitída
- USG a potom CT TU dolnej tretiny pravej obličky s kalcifikátmi
- susp. Wilms
- predoperačná CHT podľa SIOP protokolu
- čiastočný efekt (14,5 – 10ml)
- operácia

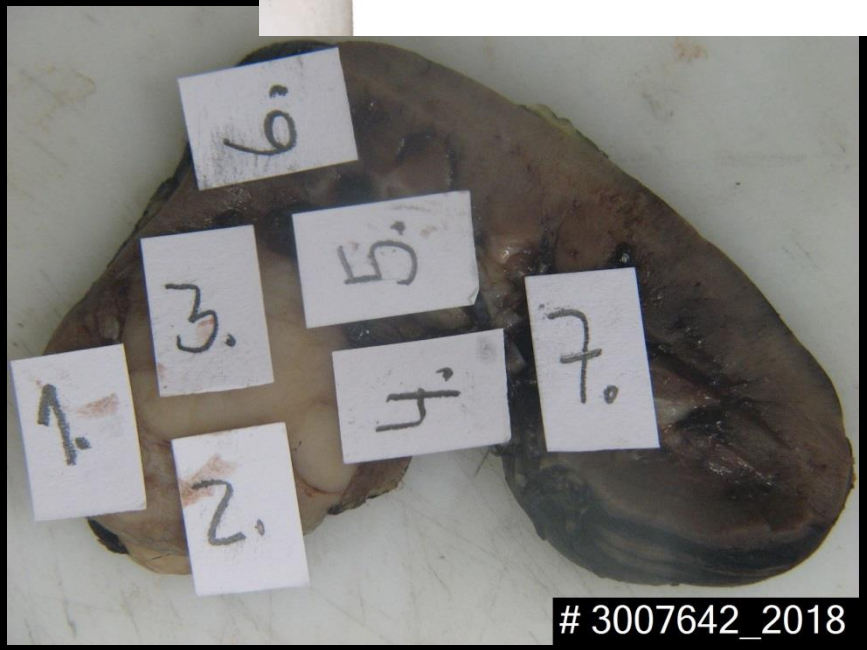
Makro

- pri dolnom póle unifokálny tumor
- 3cm v najväčšom priemere
- makroskopicky bez nekrózy, bez známok ruptúry
- limitovaný na obličku





3007642_2018



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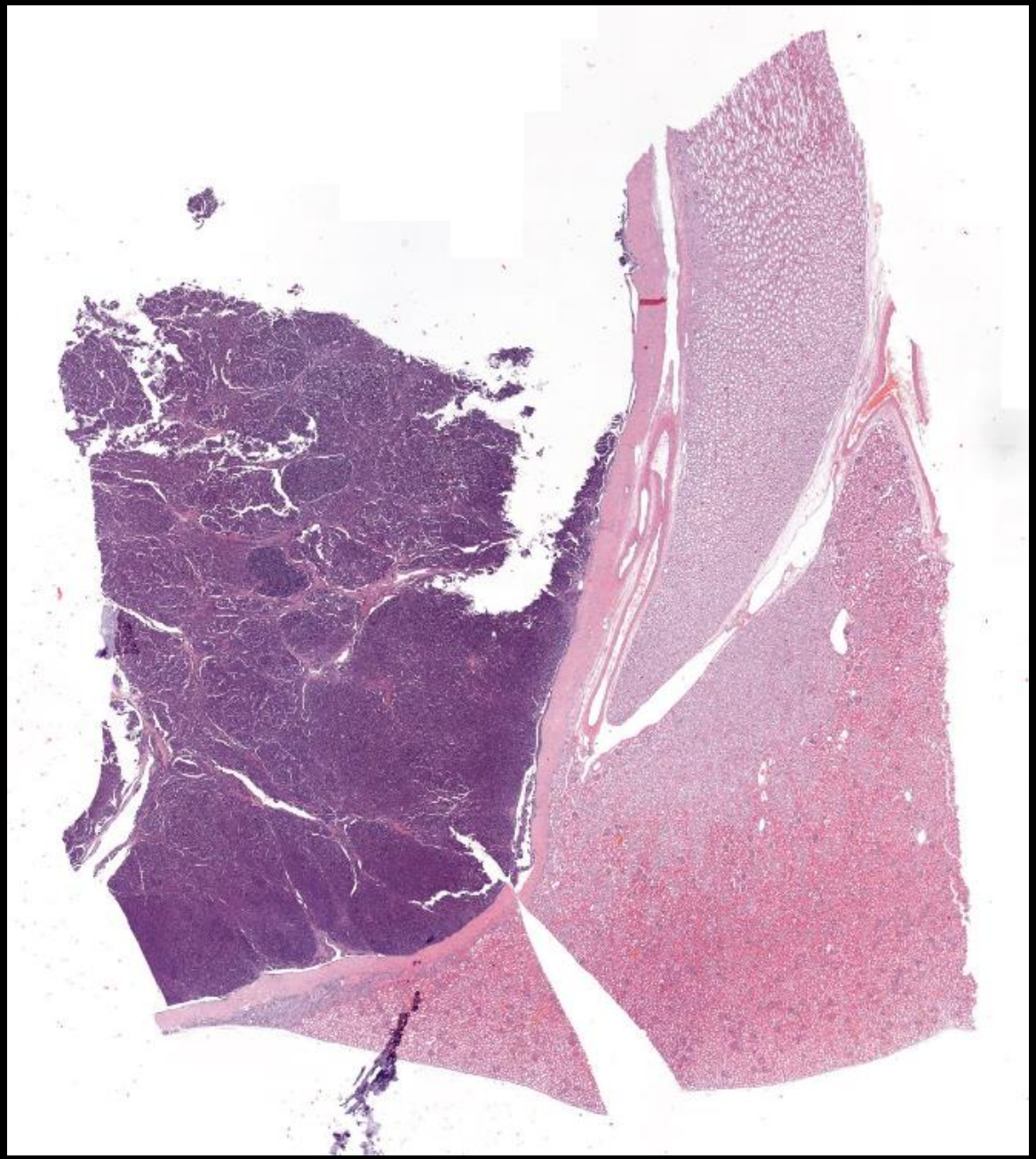
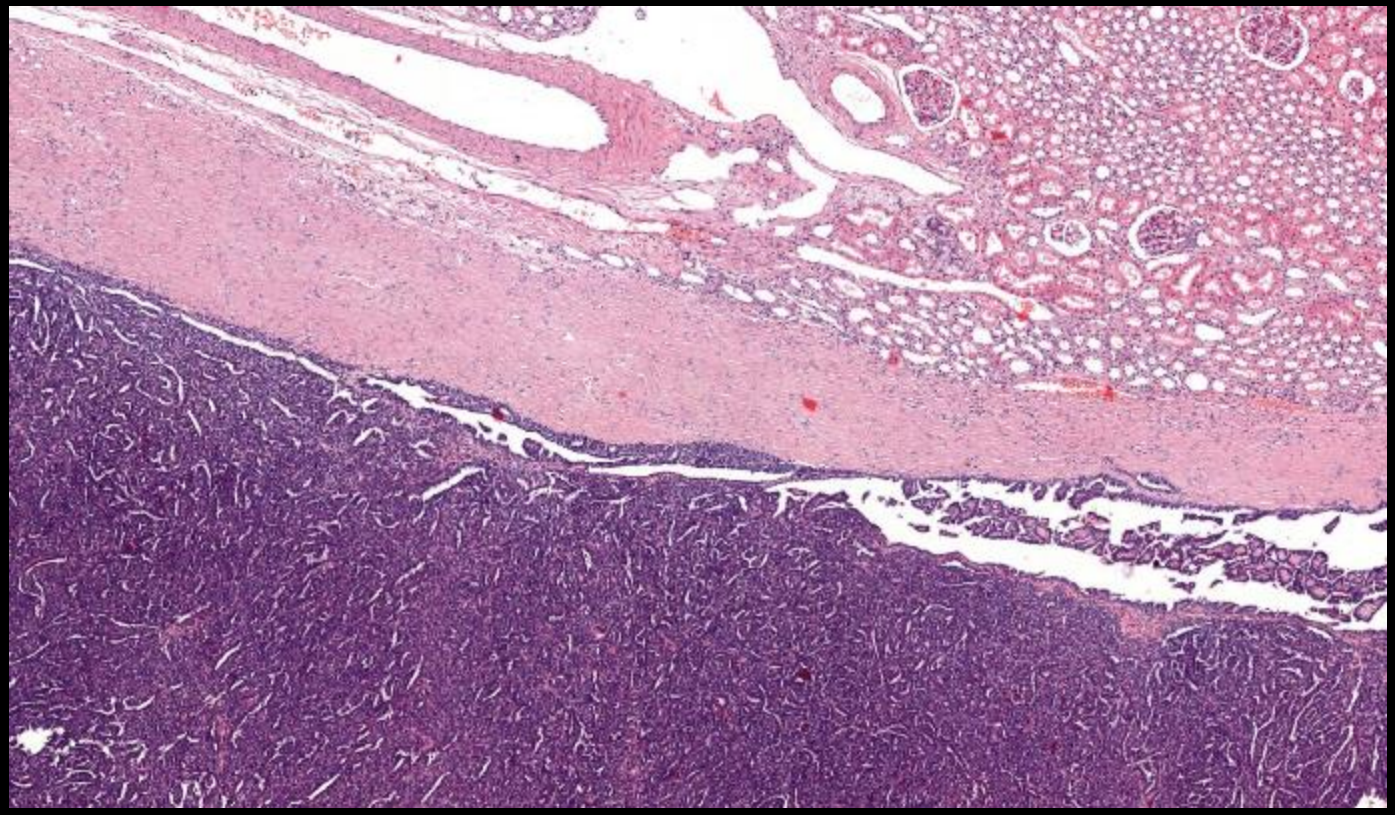
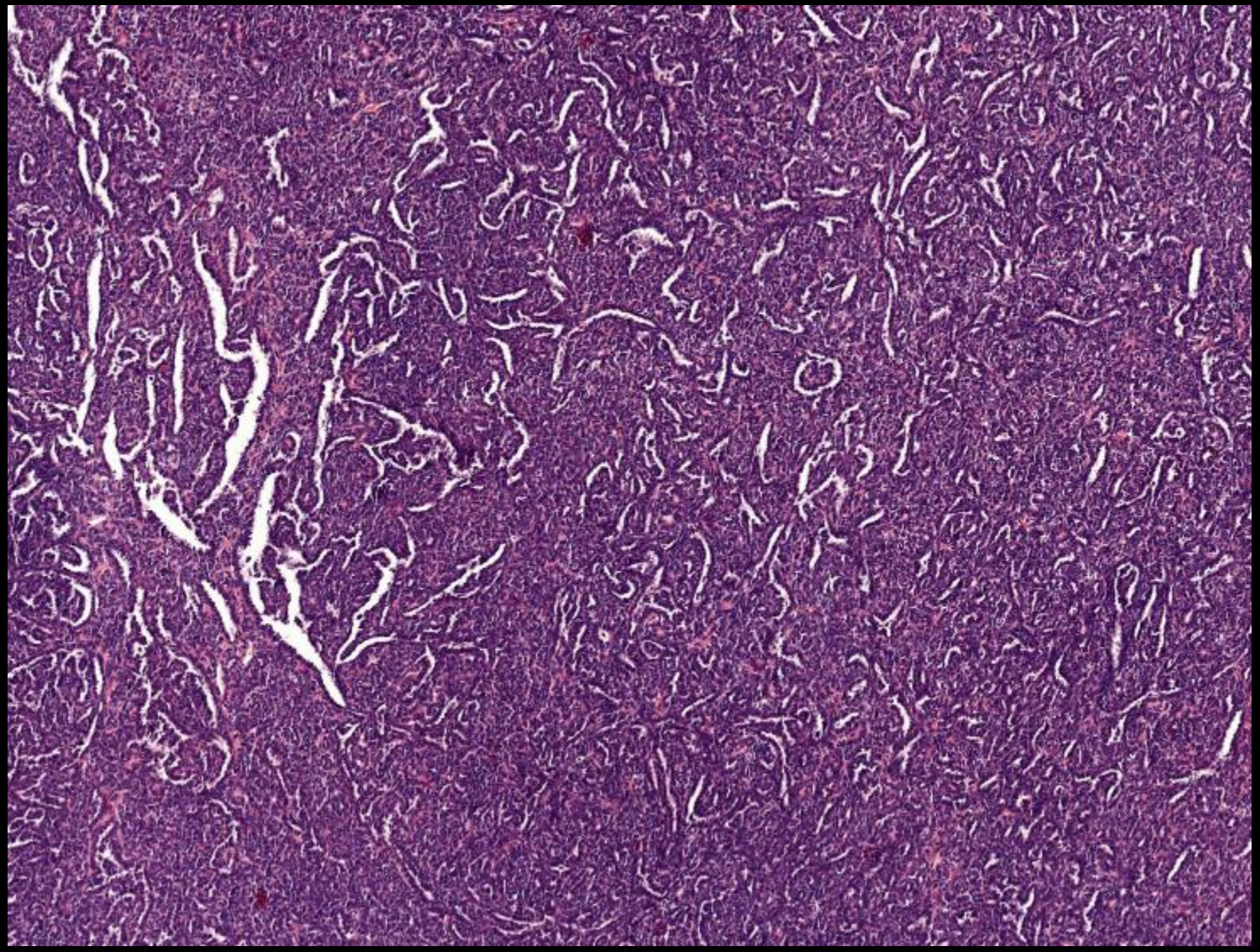
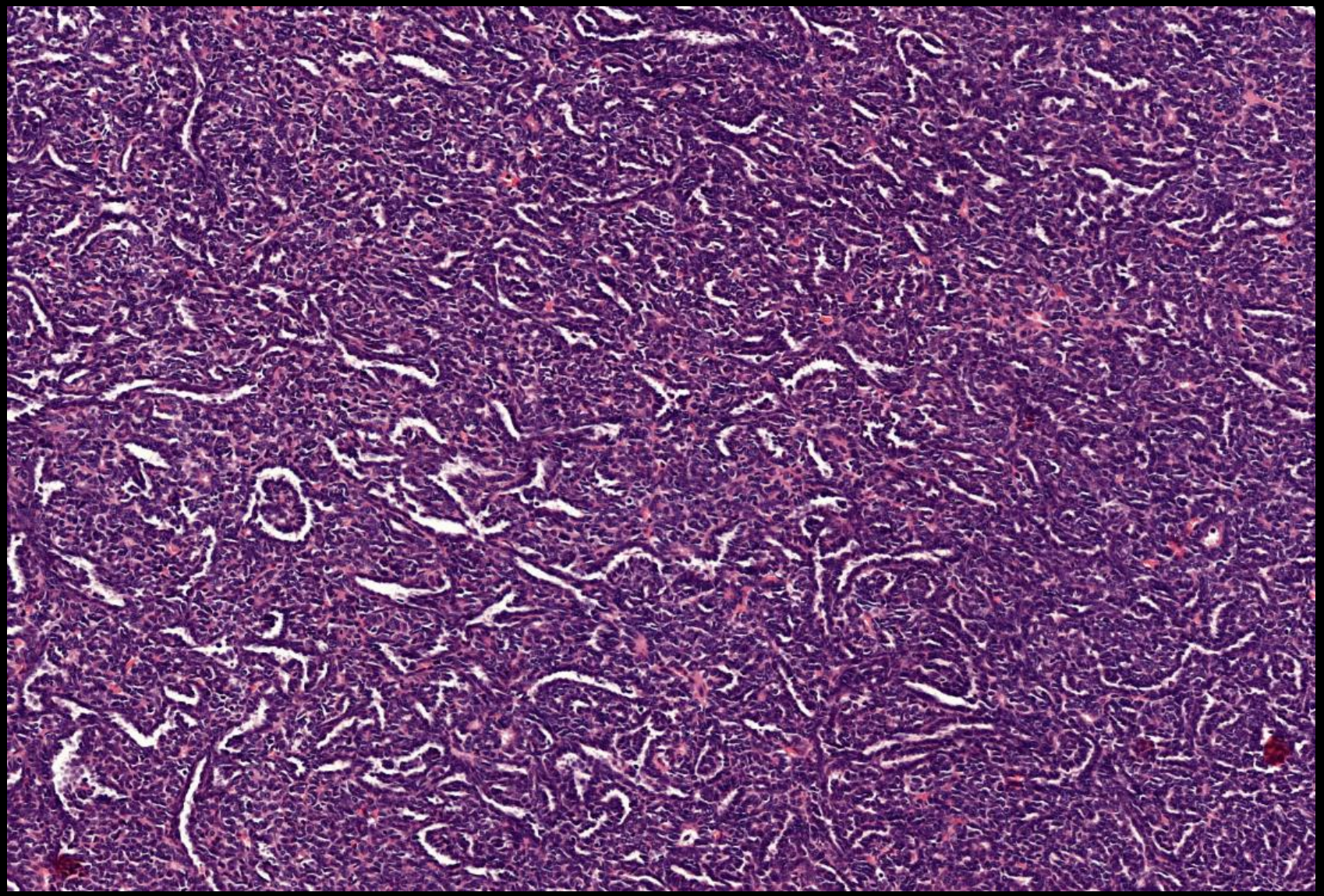
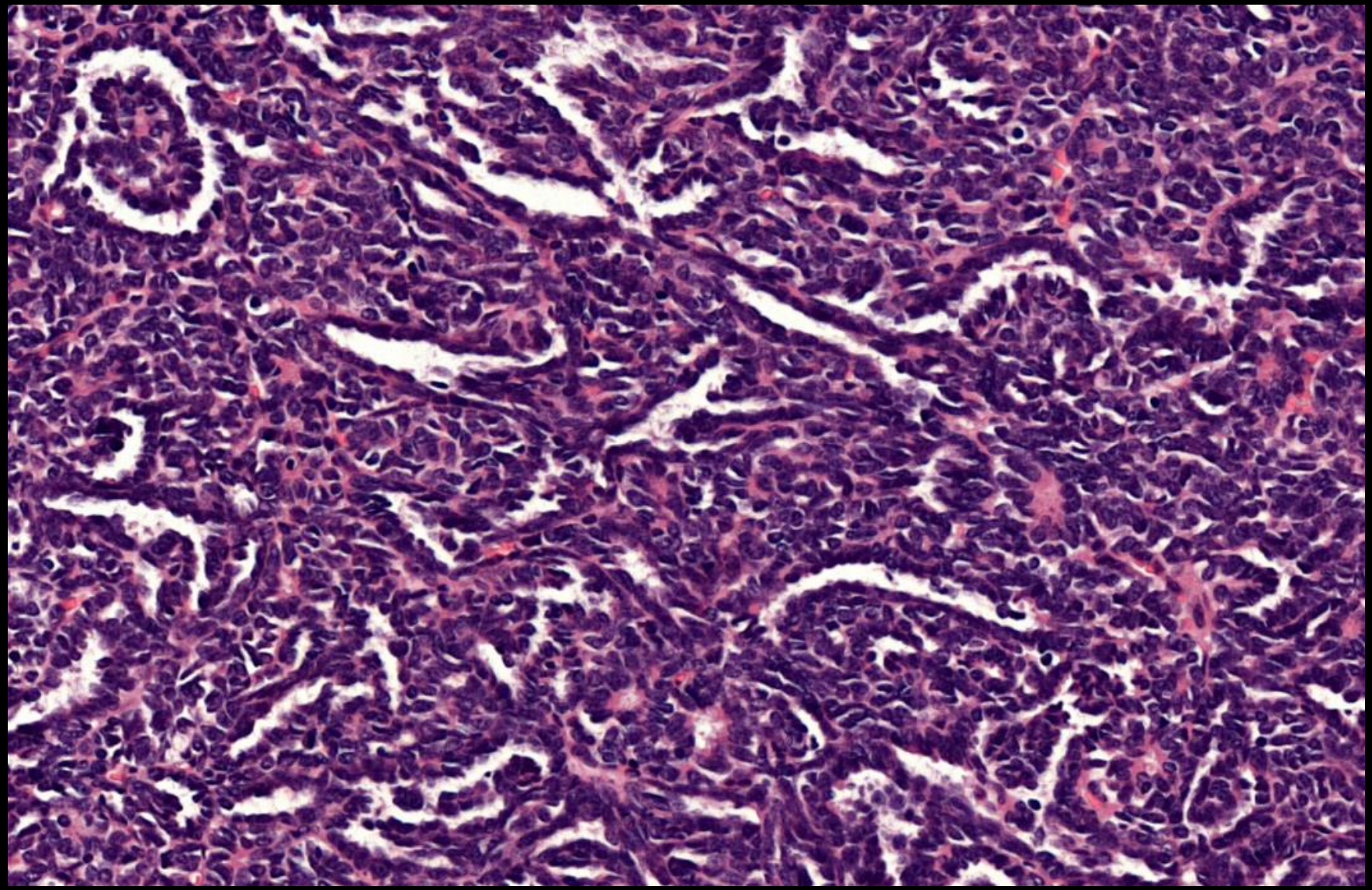
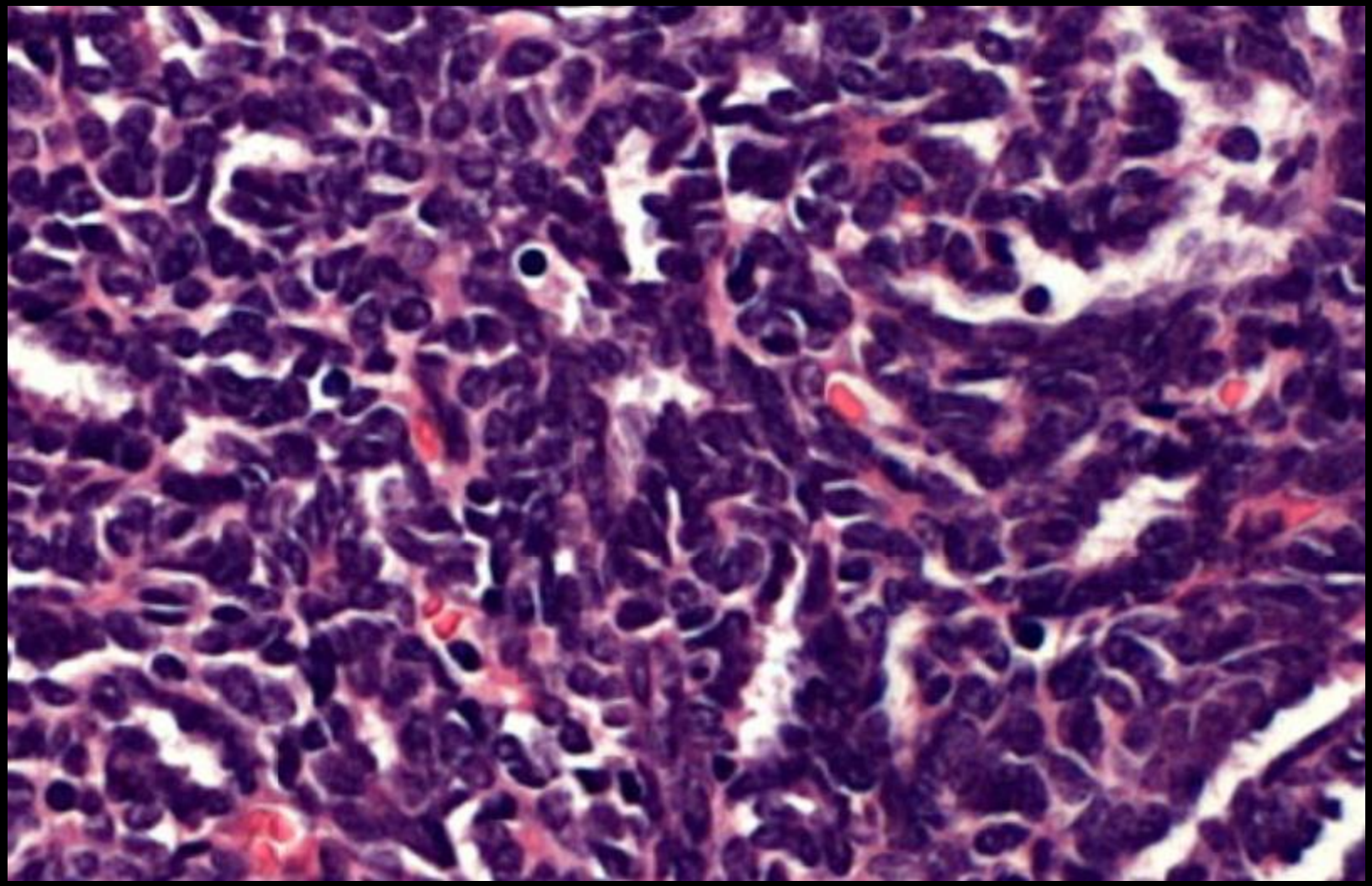


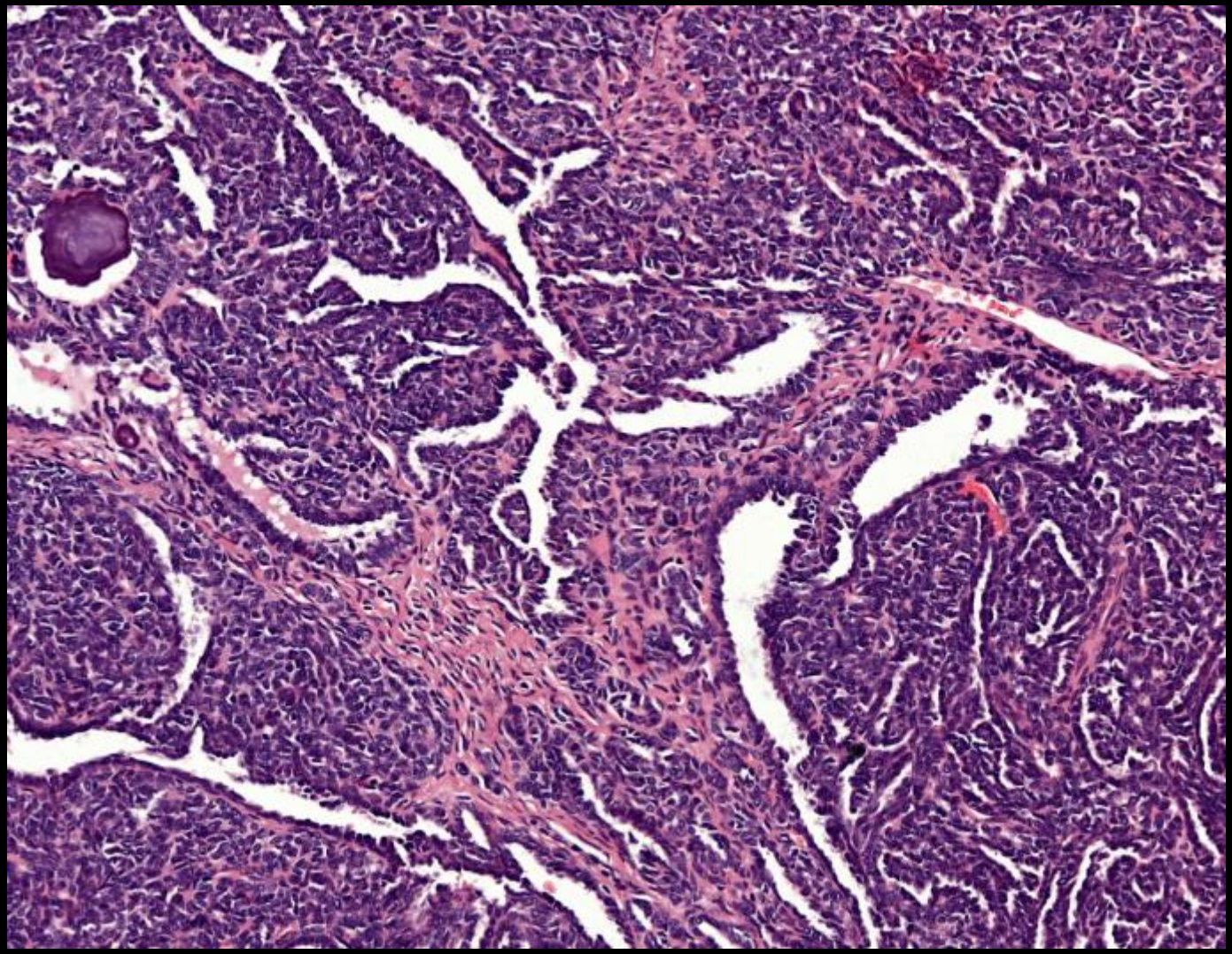


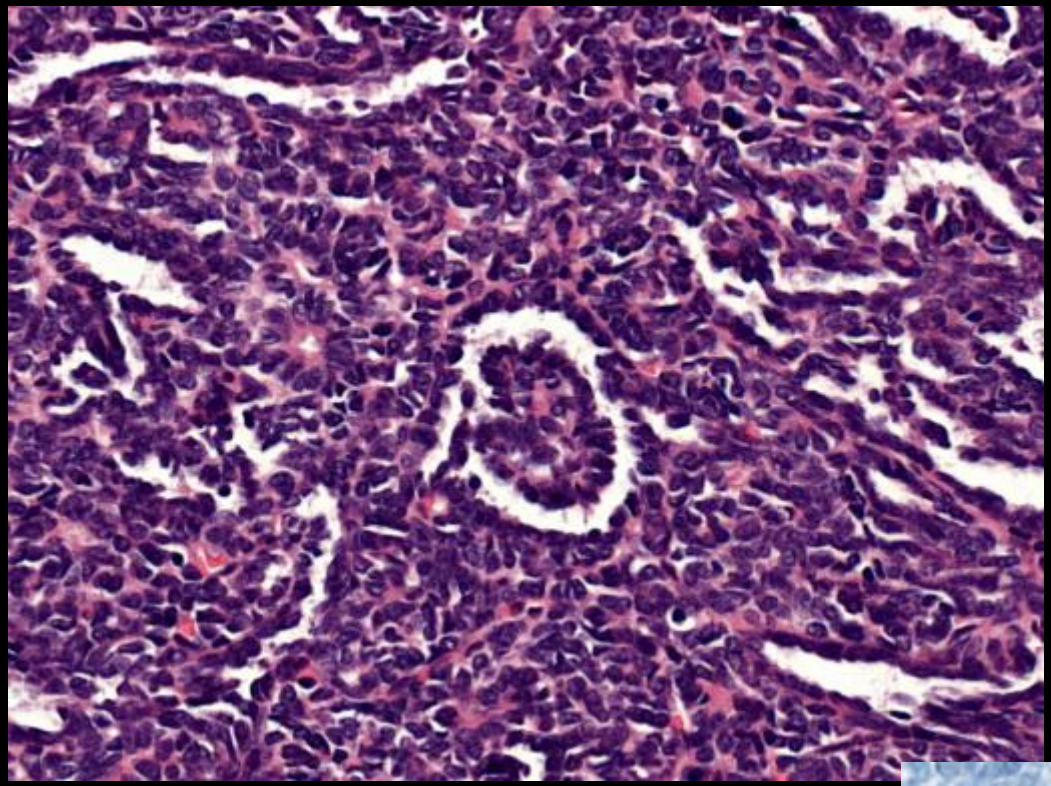




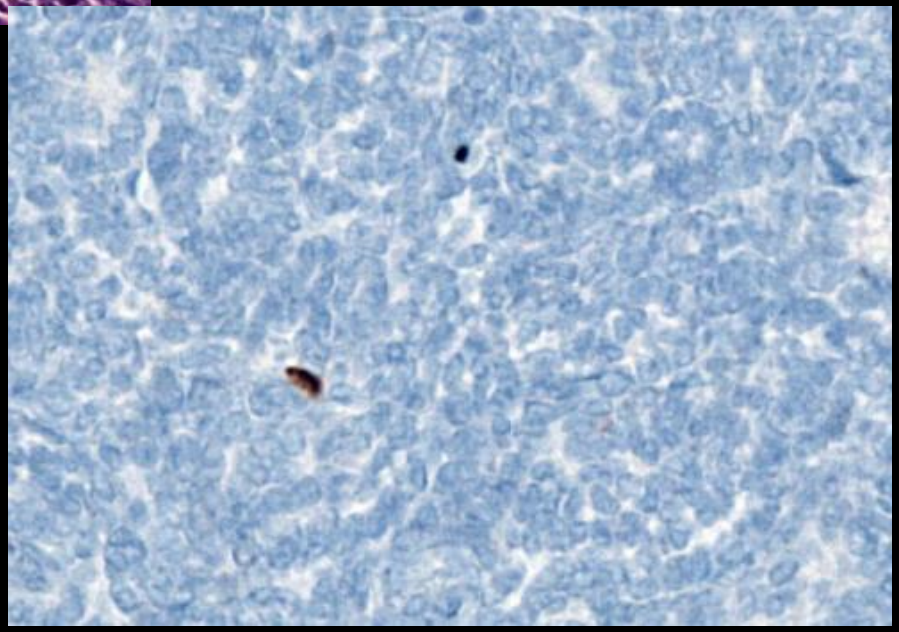




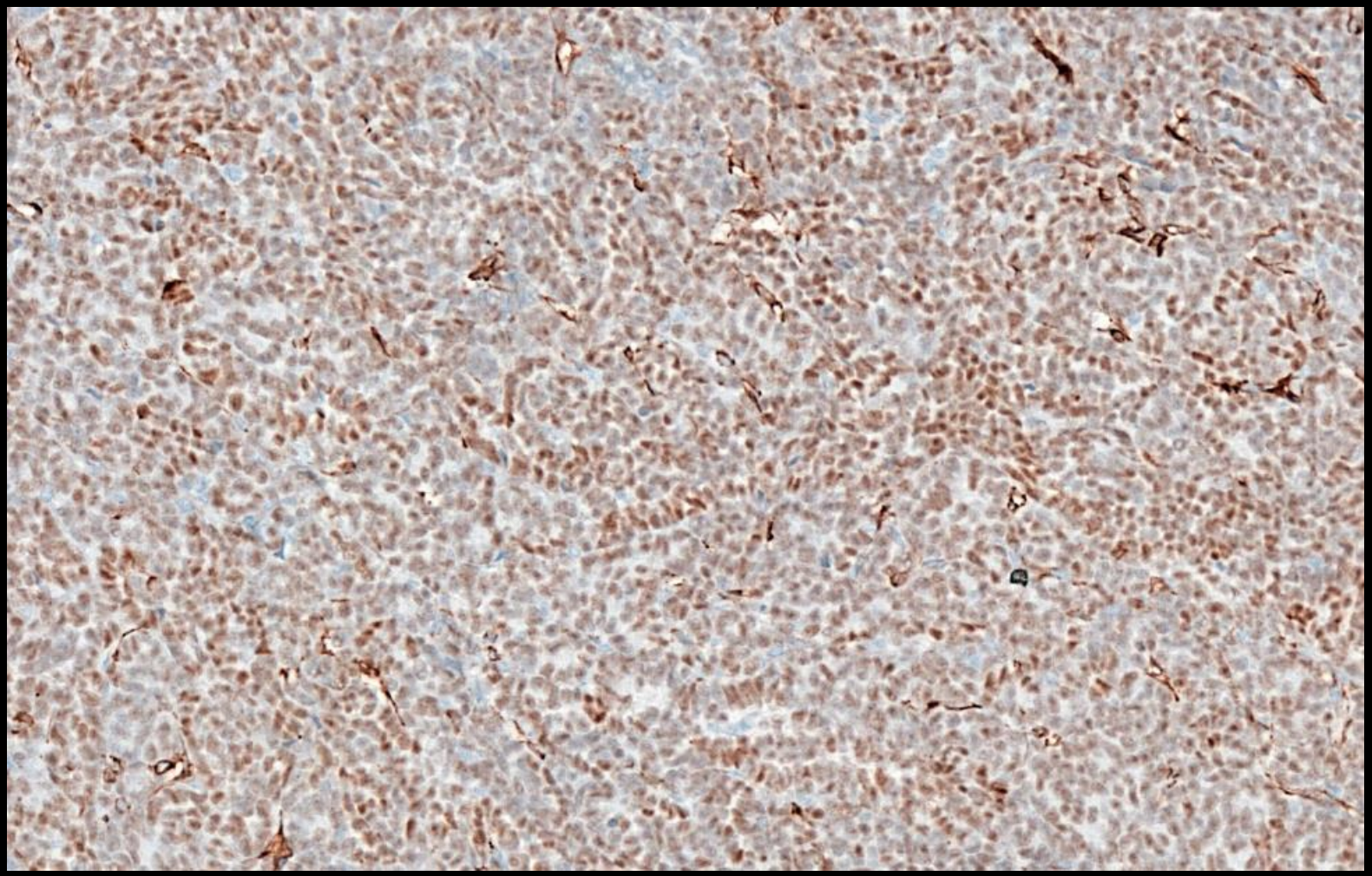




PHH
mierna mitotická aktivita



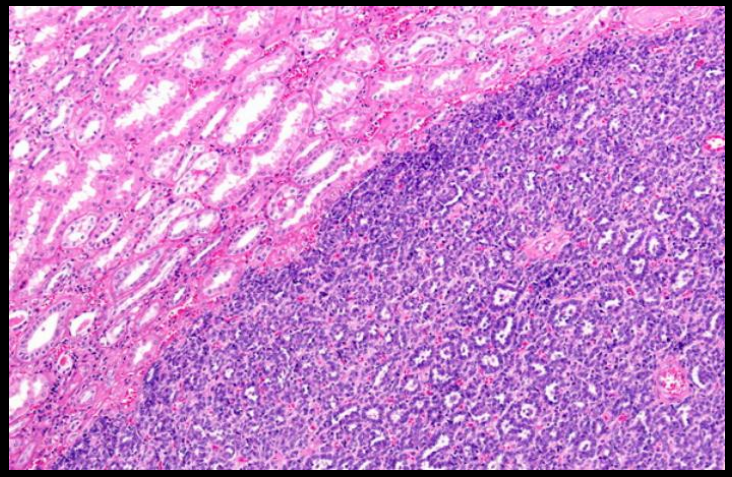
WT1



negat: CD99 negat., P53 negat., CK7, P504S
INI1 pozit., Ki67 cca 20%

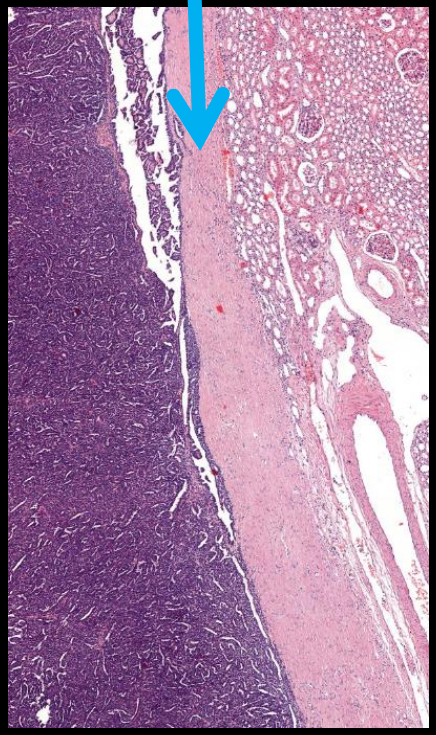
Epitelový Wilms?

- je nejaká skutočná dif. dg.?
- detský papilárny CA (CK7 a P504S+, WT1 negat., stromálne makrofágy)
- metanefrický adenóm (bez puzdra)



← metanefrický adenóm

Wilms →



Genetika, FISH

- 1pq negat. 200/200
- MYCN(2p24) negat. 200/200
- 16q (CBFB) negat. 200/200
- TP53(17p13) negat. 200/200

ZÁVER

Wilmsov tumor (nefroblastóm) epitelový typ, riziková skupina II (intermediate risk), stage I (limitovaný na obličku, SIOP)

Dear Boris

I've received your case and looked at it... it is a rather tricky case... but I think it is more likely to be a Metanephric adenoma than an epithelial Wilms tumour. I'd need to do some immuno and probably BRAF testing, which might take a week or so to do (and I am away from 2 to 11 March), and I presume that in the meantime, the child is being treated as stage 1 WT (4 weeks of VA therapy), which is not too harmful, so by the time we have the definitive diagnosis, treatment will be over, but it will still important to us to now.

I will write when I get more information.

Best regards

Gordan

PUZDRO

- metanefrický adenóm predliečený CHT podľa SIOP protokolu vytvára puzdro
- a teda je nepoužiteľné ako dif. dg. znak oproti epitelovému Wilms TU

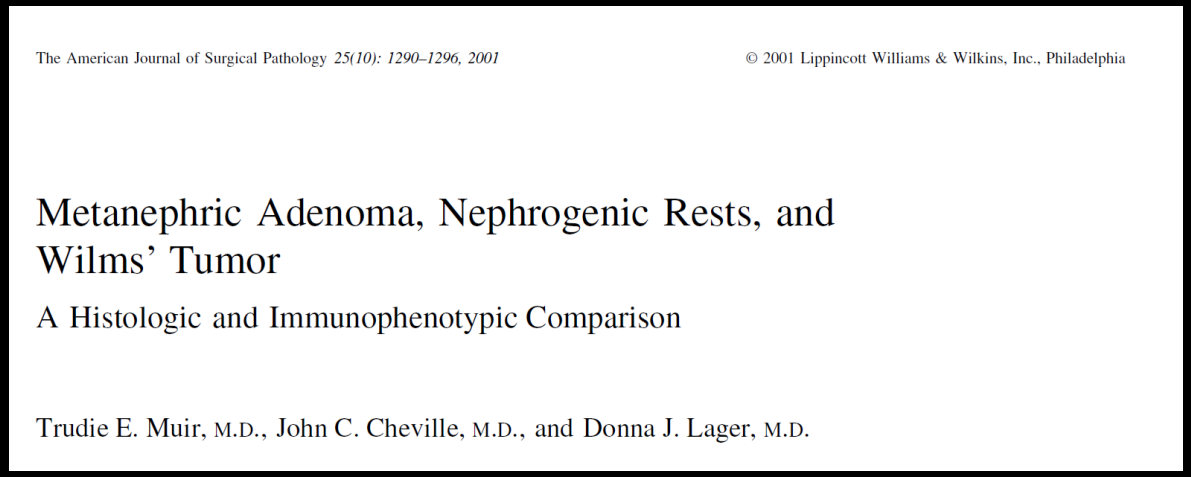
Nakoľko je dôležité odlíšiť MA a Wilms?

The American Journal of Surgical Pathology *24(4): 570–574, 2000*

Metastatic Metanephric Adenoma in a Child

Andrew A. Renshaw, M.D., David R. Freyer, D.O., and
Yuki A. Hammers, M.D.

we feel that the distinction between metanephric adenoma and a pure epithelial Wilms' tumor may be more semantic than biologic



The American Journal of Surgical Pathology *25(10): 1290–1296, 2001*

Metanephric Adenoma, Nephrogenic Rests, and Wilms' Tumor

A Histologic and Immunophenotypic Comparison

Trudie E. Muir, M.D., John C. Cheville, M.D., and Donna J. Lager, M.D.

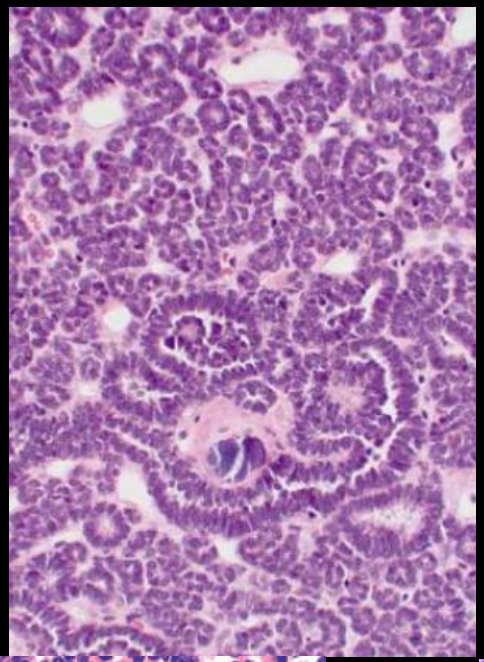
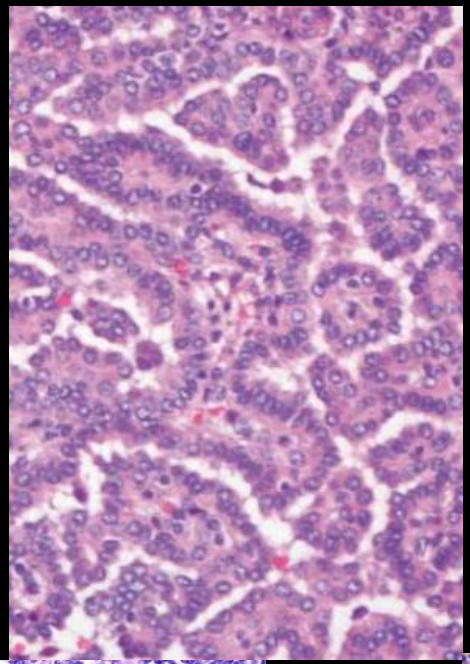
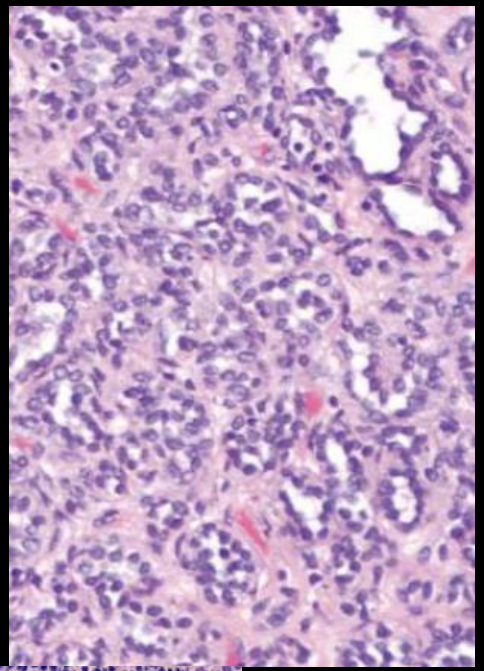
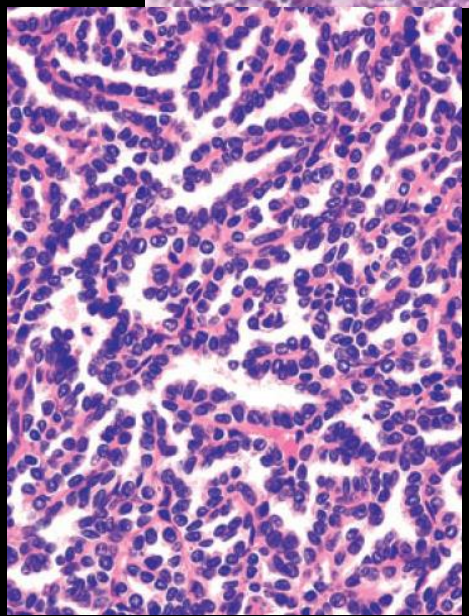
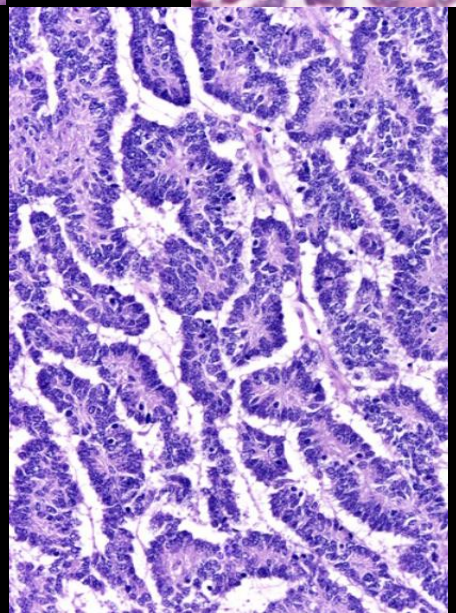
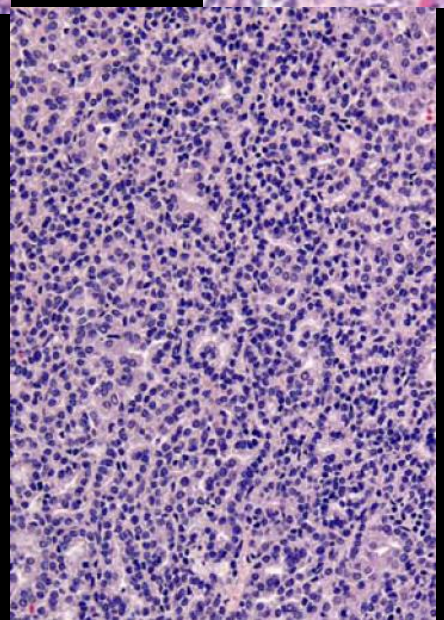
- *Metanephric adenoma is histogenetically related to WT and is morphologically and immunophenotypically identical to maturing WT and nephrogenic rests.*

Dôležité to je

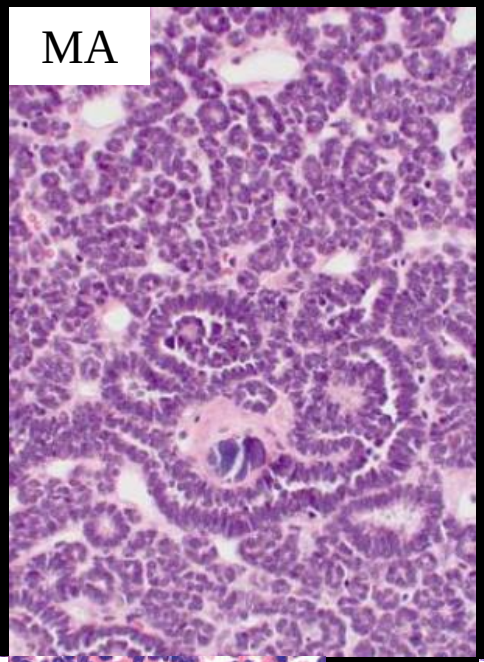
- MA – považovaný za benígny nádor (napriek zopár publikovaným MTS prípadom), po OP bez liečby, ak by bola predoperačná dg., bola by snaha o obličku zachovávajúcu OP

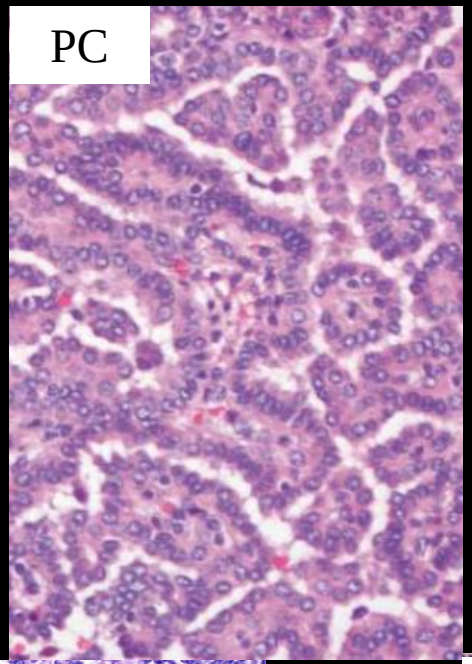
- Wilms – malígny nádor, 14. dní po OP druhá línia CHT – podľa rizikovej skupiny a stage

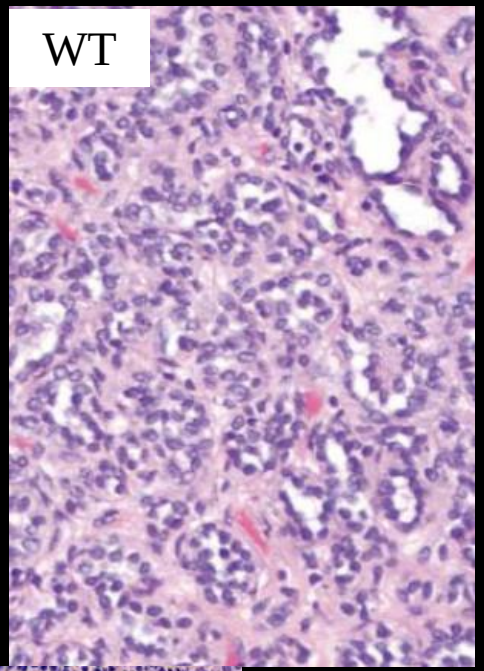
Epitelový Wilms, metanefrický adenóm, papilárny CA ?

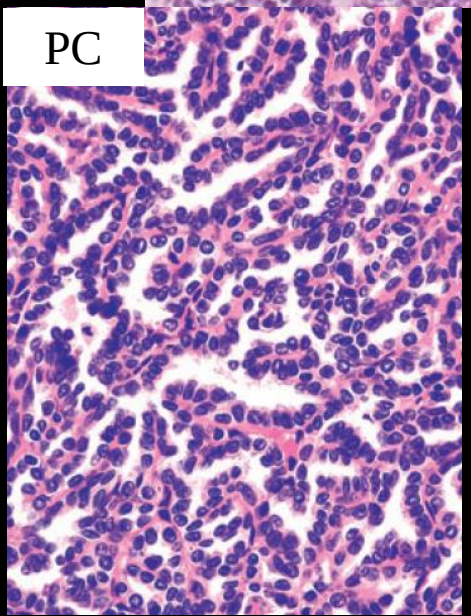







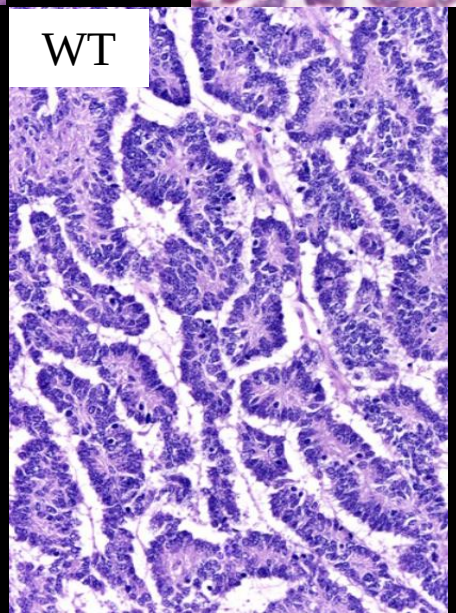
Epitelový Wilms, metanefrický adenóm, papilárny CA ?

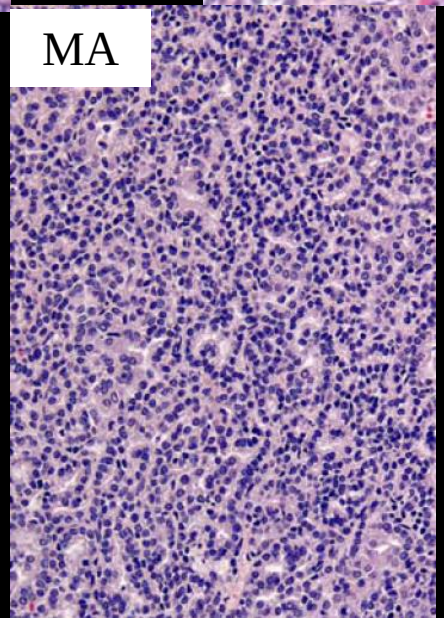




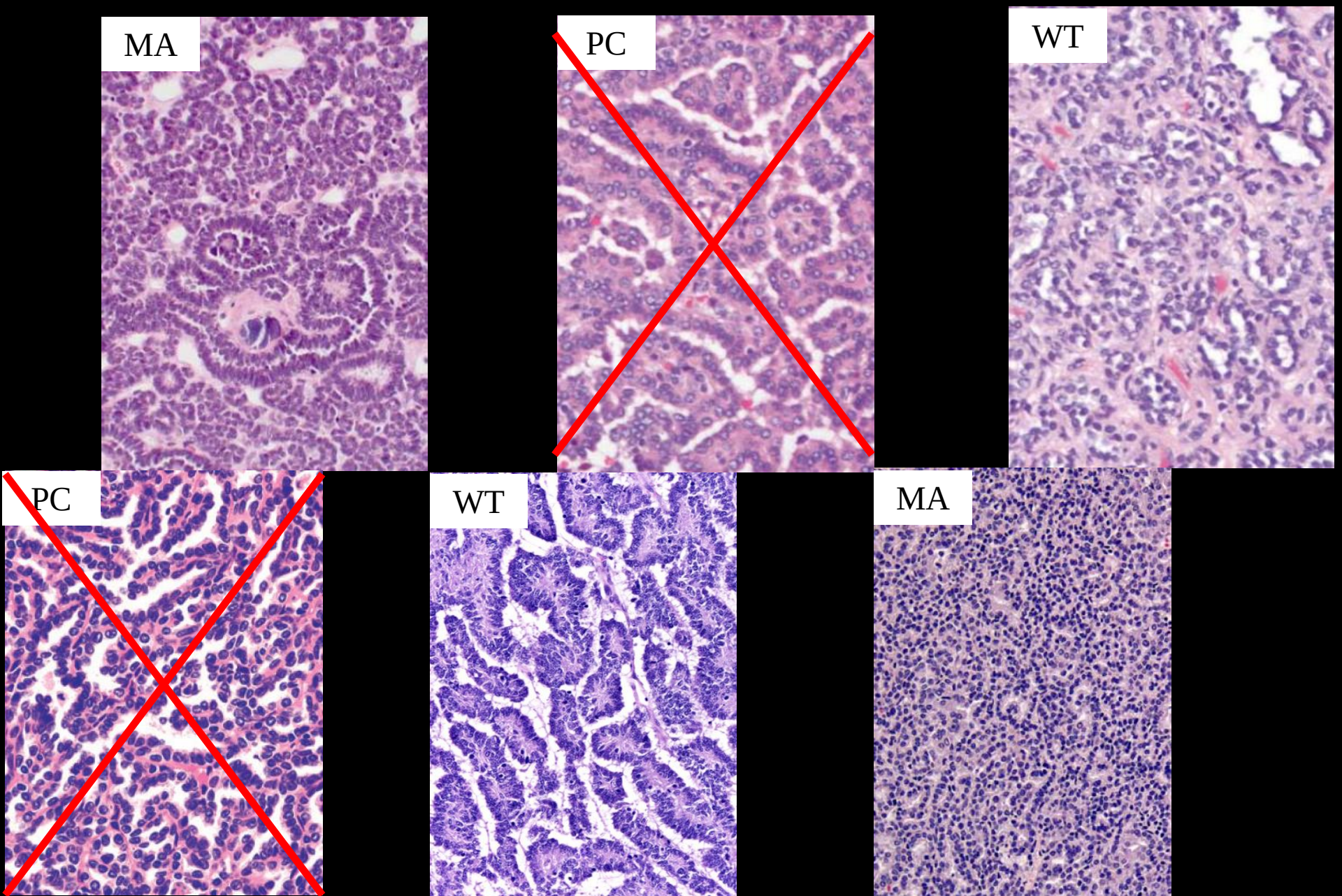




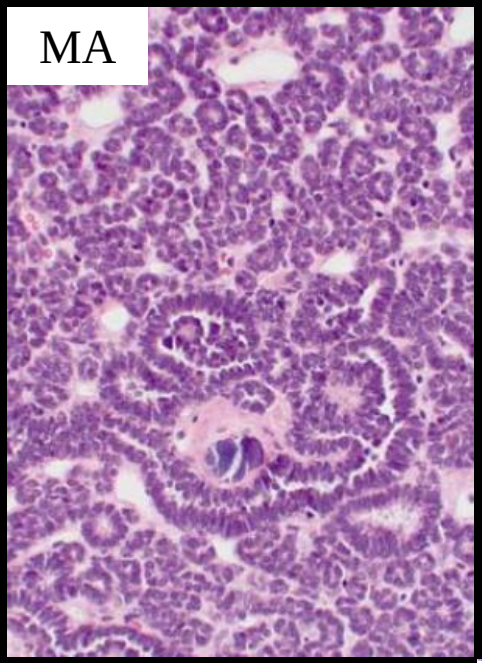


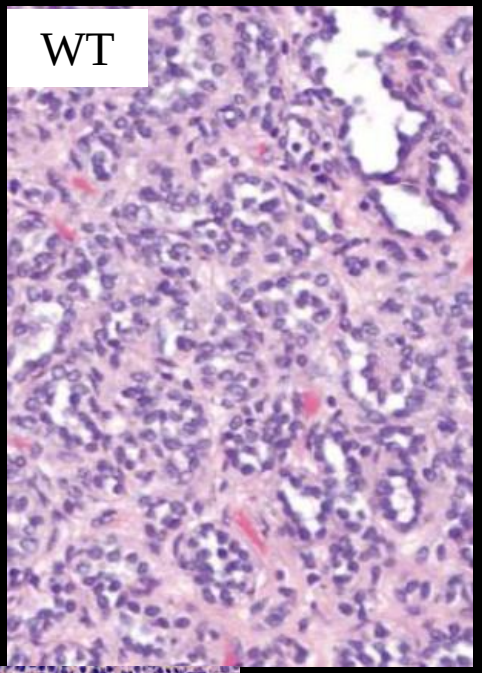


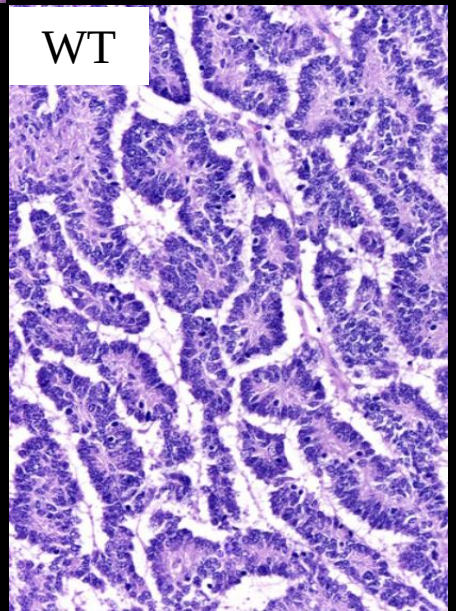
Epitelový Wilms, metanefrický adenóm, papilárny CA ?

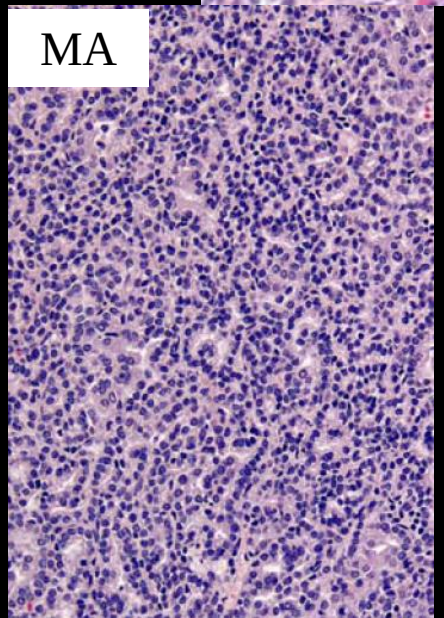


Epitelový Wilms, metanefrický adenóm?

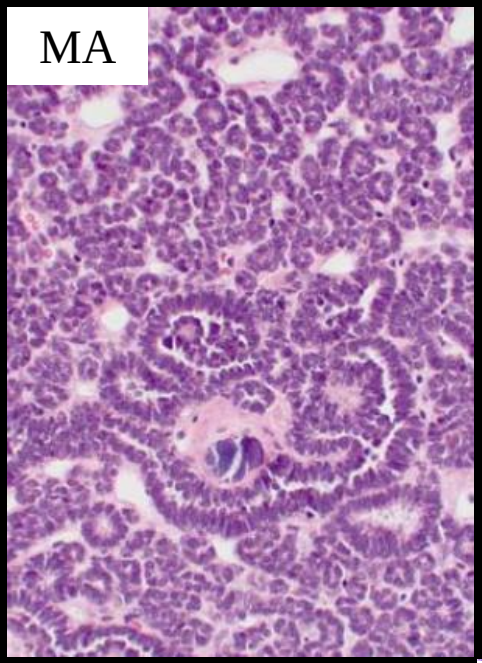




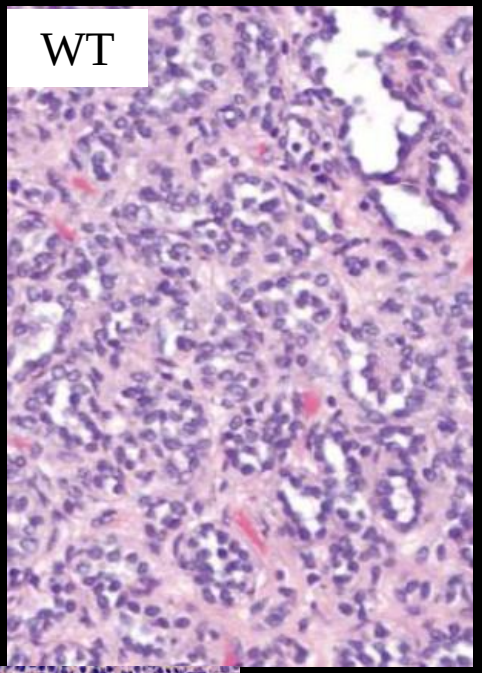


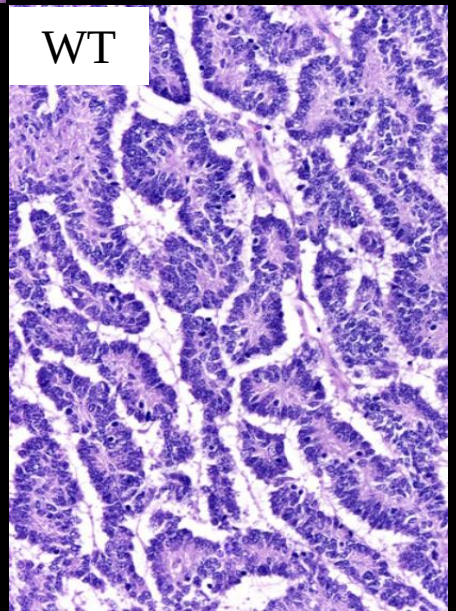


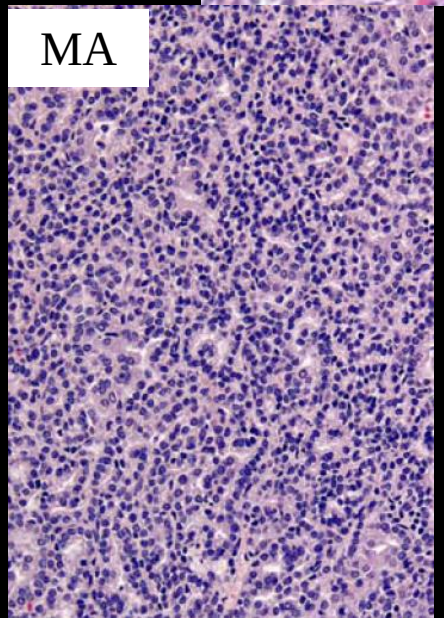
Epitelový Wilms, metanefrický adenóm, papilárny CA ?



← imuno?
CD57,
Cadherin 17







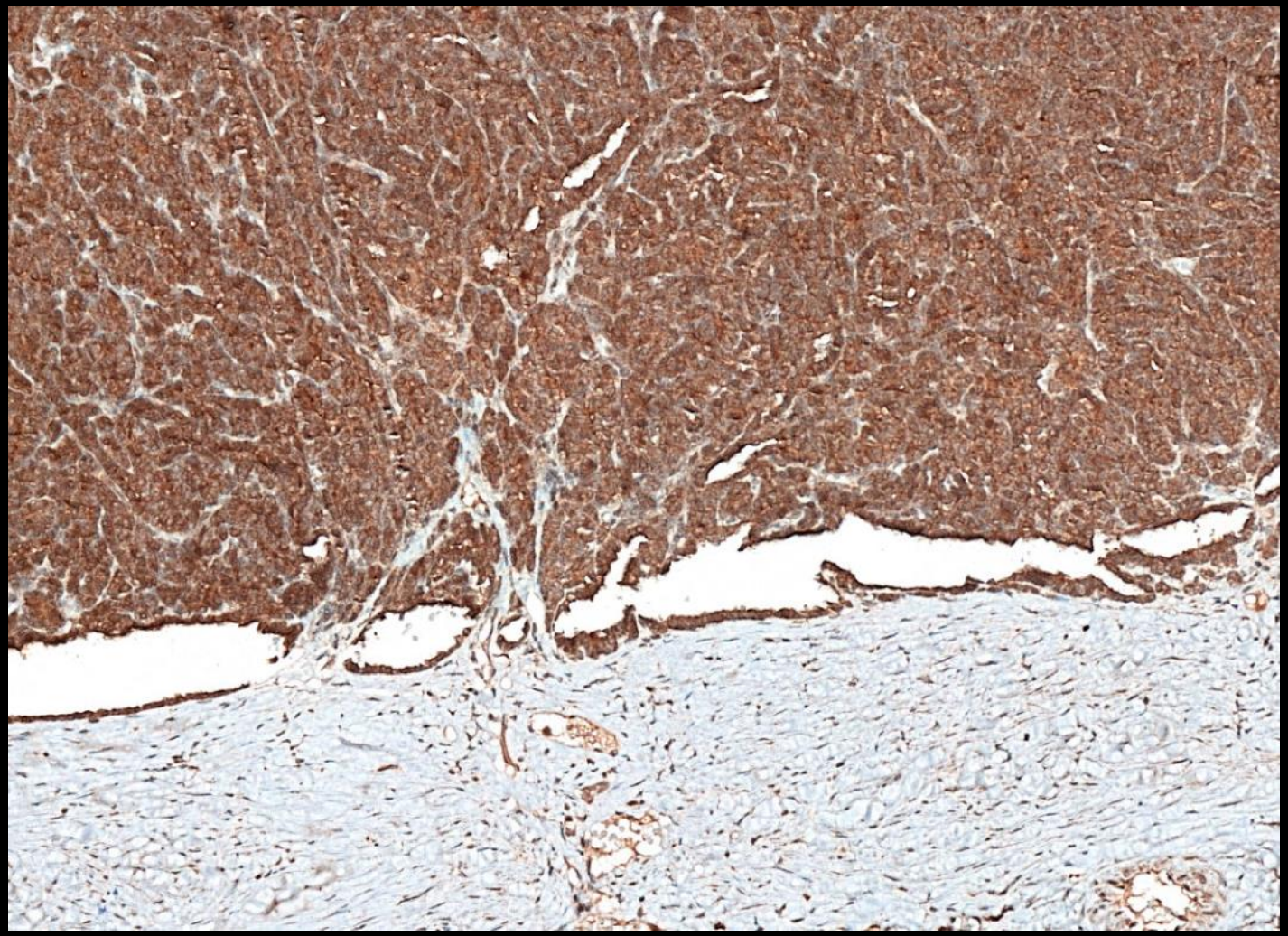
Je niečo, čo naozaj pomôže v dif. dg.?

Metanefrický adenóm, genetika:

Choueiri TK, Cheville J, Palescandolo E, et al. BRAF mutations in metanephric adenoma of the kidney. *Eur Urol*. 2012;62:917–922.

- 90% metanefrických adenómov má BRAF V600
- v kontexte renálnych nádorov pomerne špecifická

- BRAF V600



- aj geneticky

Thank you for sending me 8 HE slides (and corresponding 8 paraffin blocks), gross pictures and a covering letter with a summary of your findings in the above named girl's renal tumour. I understand that tumour measured 3 x 2.5 x 2.5 cm, with a well-formed capsule which separated it from the renal parenchyma.

Sections show a tumour composed of well-differentiated, small, closely packed tubules lined with hyperchromatic cells, and areas with papillary structures. In many areas there are numerous psammoma bodies. Tumour is separated from the renal parenchyma with a thick fibrous capsule. Tumour is not infiltrating the renal sinus or perirenal fat.

In the differential diagnosis one has to consider two entities: epithelial type Wilms tumour and metanephric adenoma. Despite the well-formed capsule which, by definition, is not present in metanephric adenoma, I think that this criterion is not applicable in this particular case because the child received pre-operative chemotherapy as per SIOP Wilms tumour protocol, and in our experience, it very often causes the formation of the (pseudo)capsule as, for example, in treated nephrogenic rests.

I will be doing some immuno to support my opinion, but on the basis of the present features, I think that this is a **Metanephric adenoma, local pathological stage 1**.

Thank you for sharing this interesting case with me. I hope you will find my opinion of some help.

Best wishes

Professor Gordan M Vujanic
Chair of the SIOP-RTSG Pathology Panel

... I showed the case at the SIOP Panel review meeting in Paris ...

Metanefrický adenóm

- pripomína skorú metanefrickú tubulárnu dif.
- bol považovaný za hyperdiferencovaný / benígny koniec epitelového Wilmsa
- metanefrický adenóm, adenofibróm, fibróm
- prevažne u dospelých (medián 41)
- M:F = 1:2
- benígny (niekoľko raritných reportov MTS)
- bez kapsuly, bez útlaku okolia (po CHT neplatí)
- WT1, CD57, Cadherin 17 pozit., CK negat., P504S (môže)

Epitelový Wilms vs metanefrický adenóm

- kapsula (oproti nepredliečeným MA)
- hyperchrómnejšie prekrývajúce sa jadrá
- mitózy (viac)
- stromálna zložka a blastém (ak prítomné)
- nefrogénne zvyšky
- negat. CD57, Cadherín 17
- bez BRAF V600 mutácie

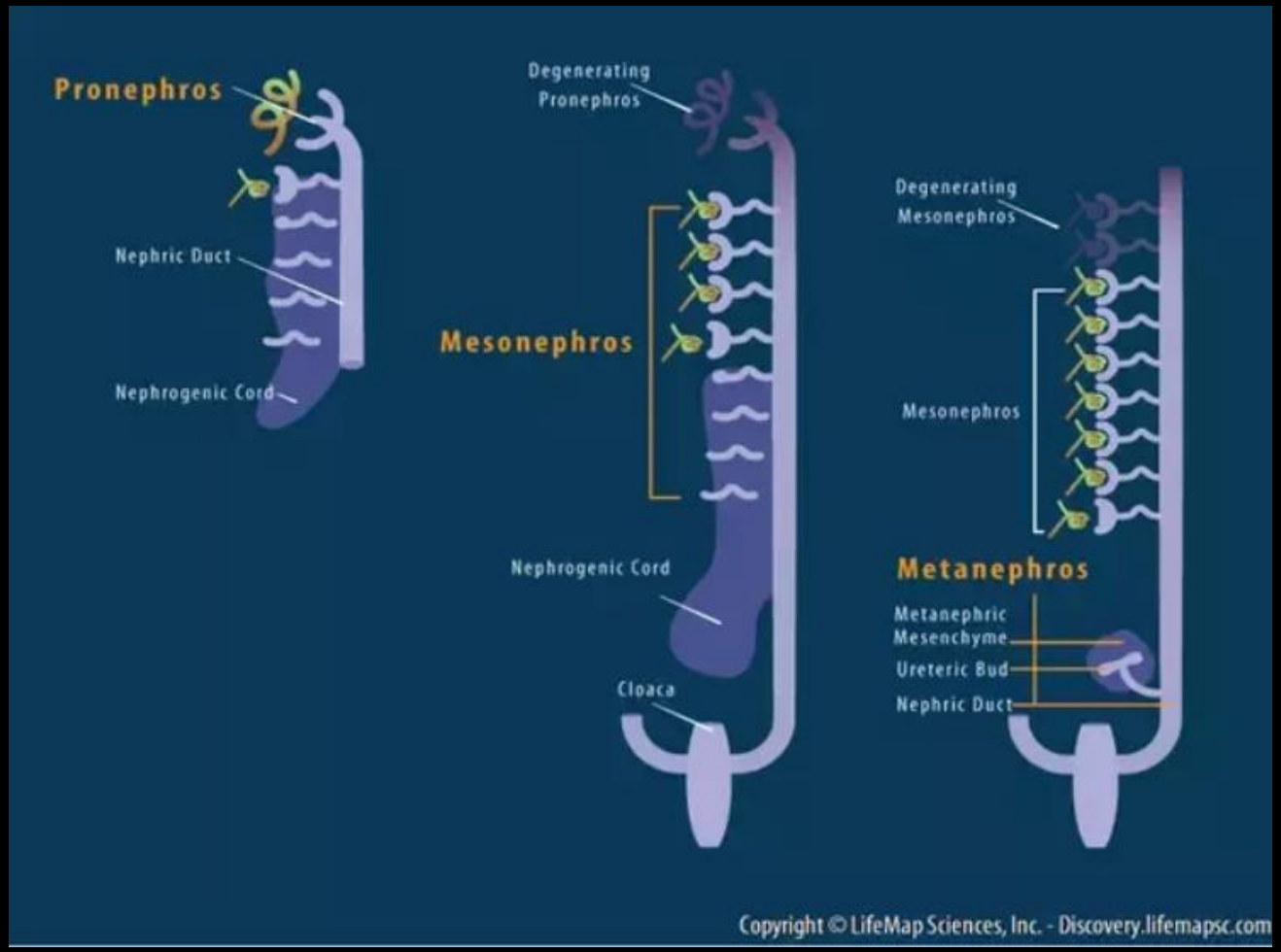
- niektoré metanefrické adenómy ale nečítajú separátky (nemajú typicky popisovaný IHC prof.)

Metanefrický a mezonefrický?

- metanefrický adenóm

- mezonefrické zvyšky
- mezonefrická hypeplázia
- mezonefrický adenóm
- mezonefrický karcinóm

Metanefrický a mezonefrický



Záver

Slovenské deti s Wilmsom si zaslúžia centrále
čítanie patologického nálezu SIOP / Wilms patológom..

ďakujem za pozornosť