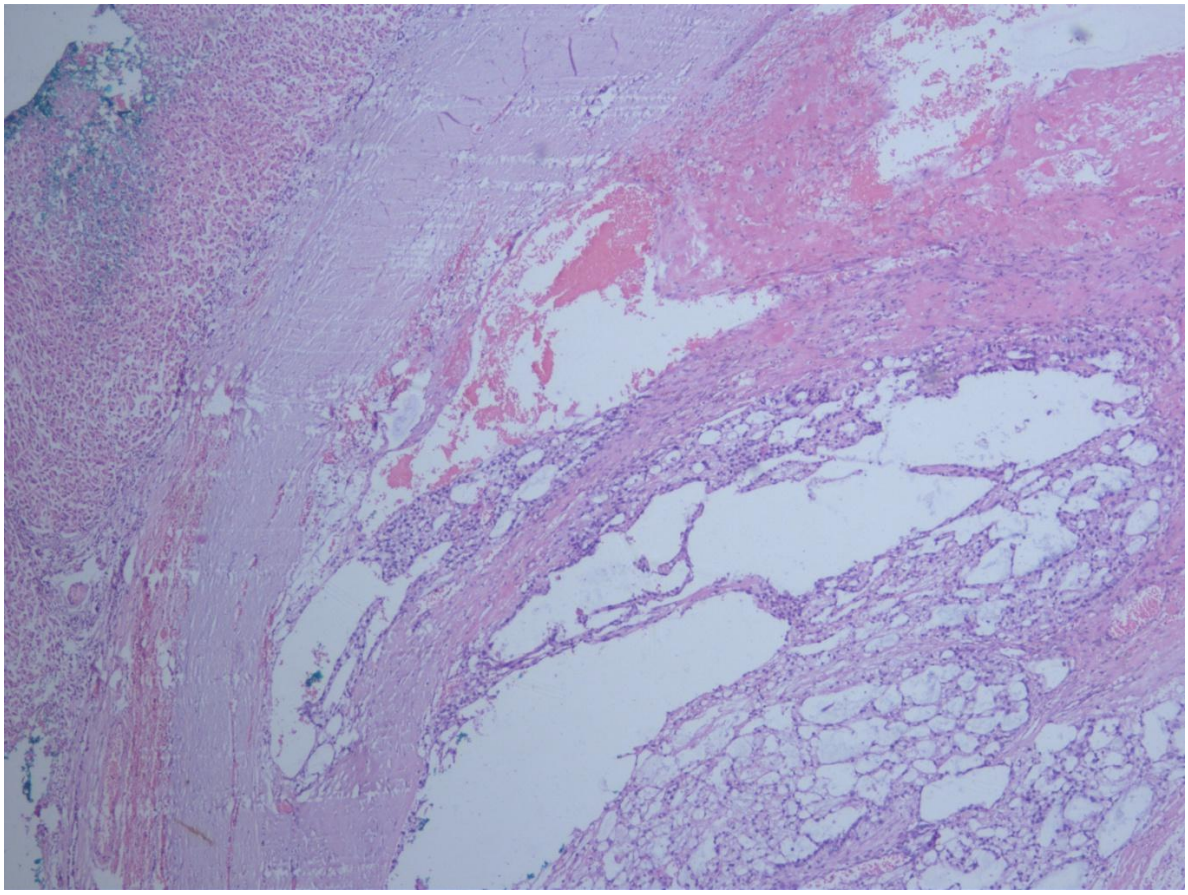
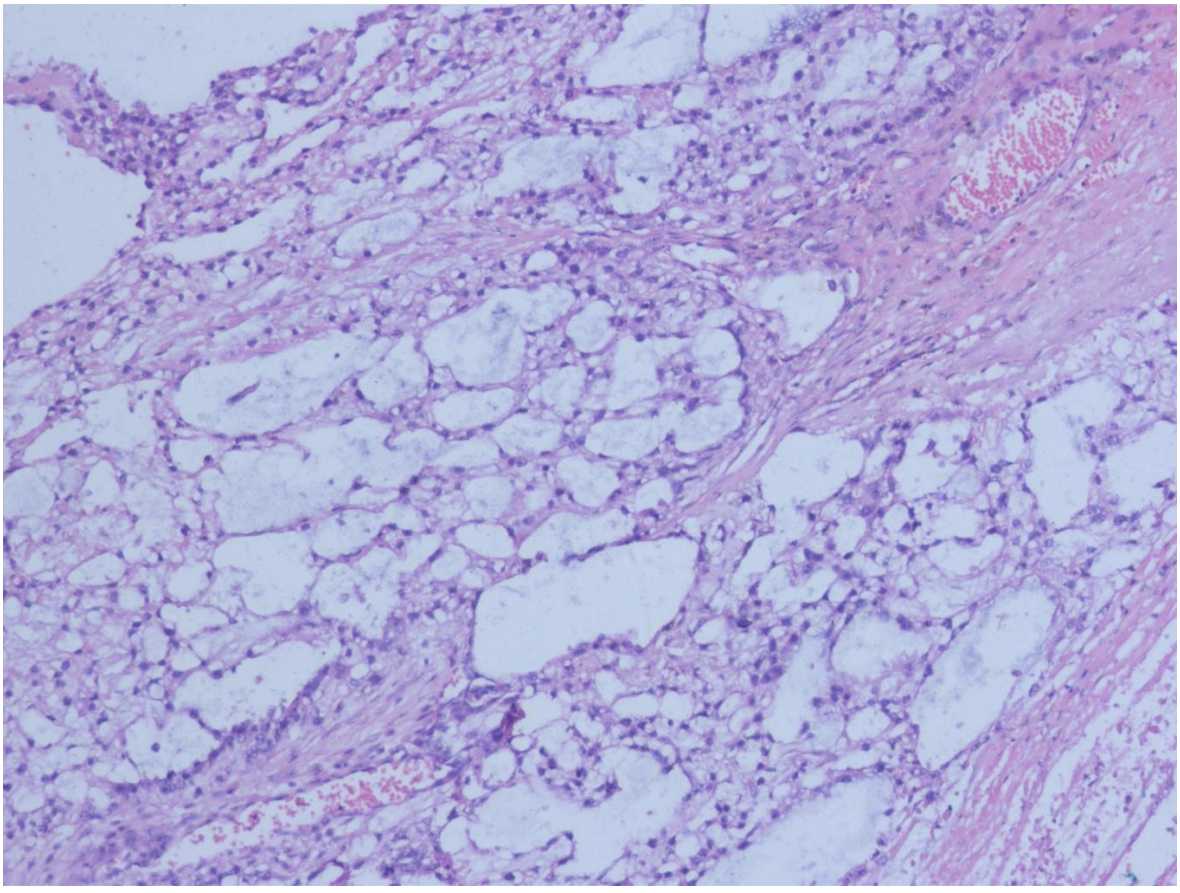
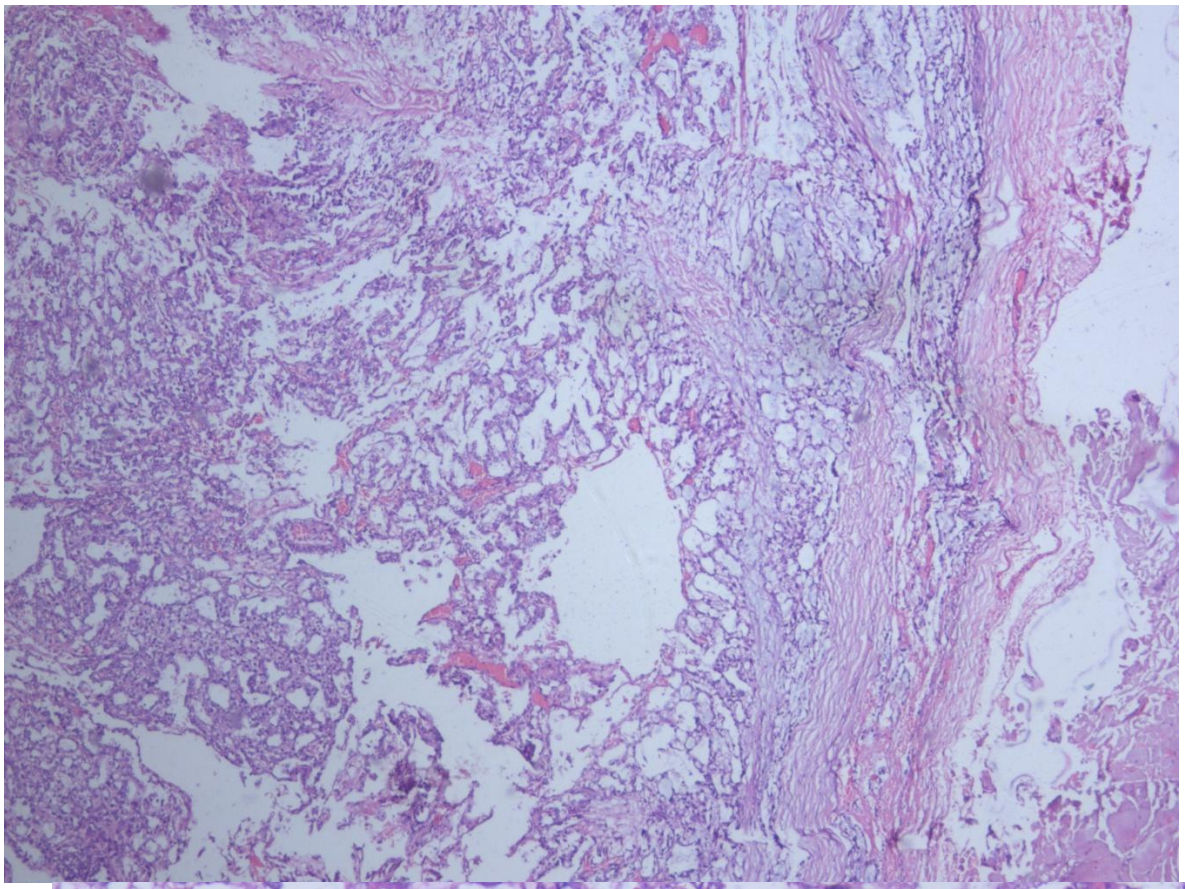
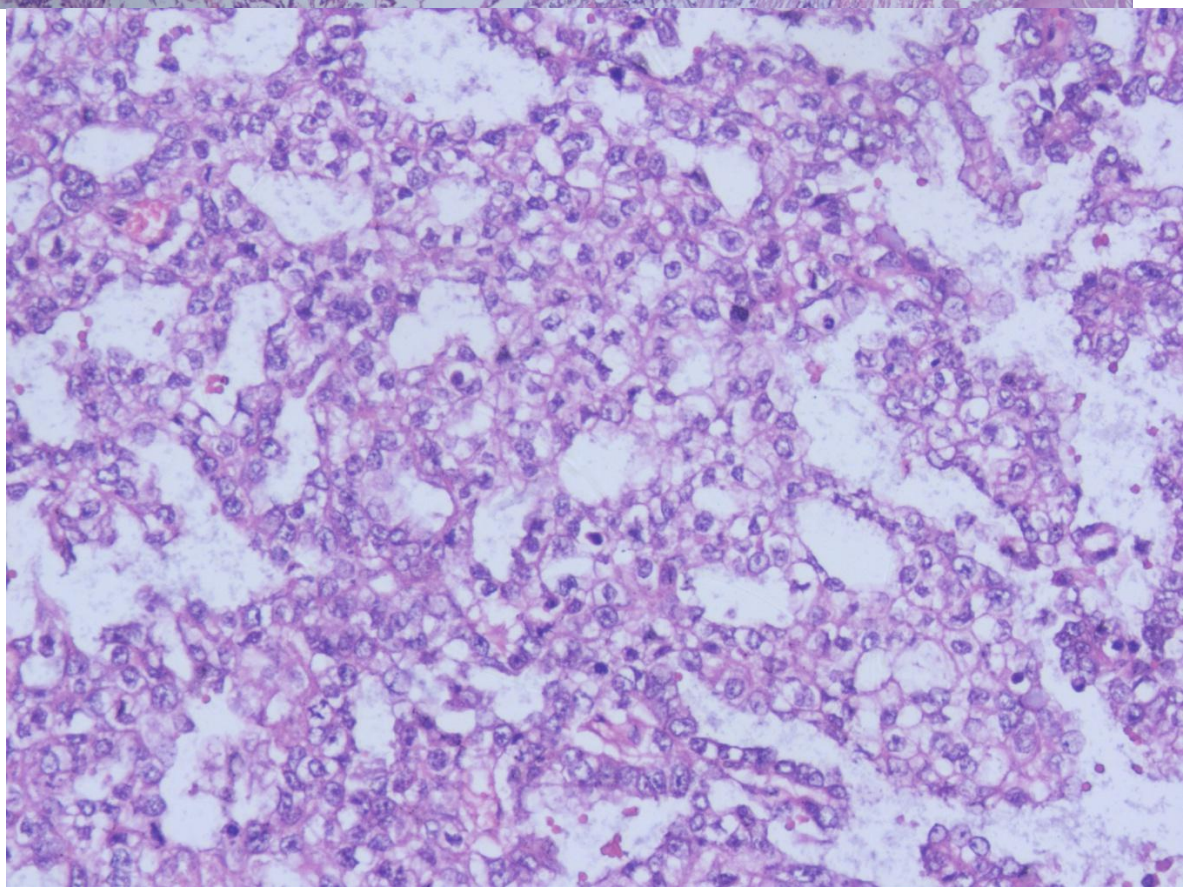
Case of the Fortnight: By Dr. Mithraa Devi S, Senior Resident and Dr. Srinivas BH Additional Professor. JIPMER. Puducherry

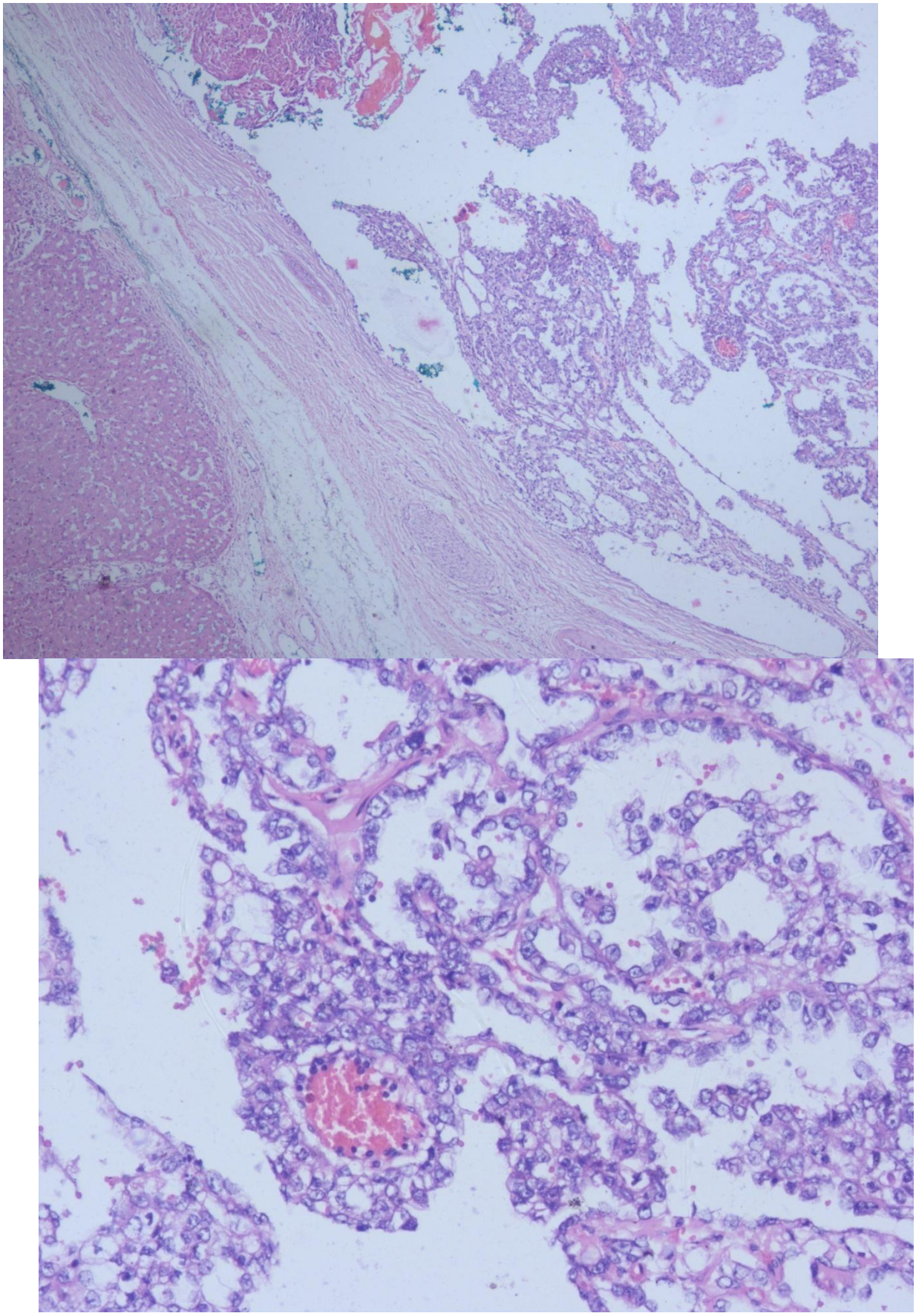
2 years old girl child presented with complaints of abdominal distension for 4 days duration. No other relevant clinical complaints were identified. On CECT abdomen, a large well defined heterogeneous mass of size 10.4x 9.9x 9.5cm with internal vascularity and multiple cystic/ anechoic areas were noted within the mass in the right lobe of the liver predominantly involving the 7th segment extending into 6 and 8th segments. AFP levels- >3000ng/ml. Trucut biopsy performed, diagnosed as hepatoblastoma. Liver resection specimen received.





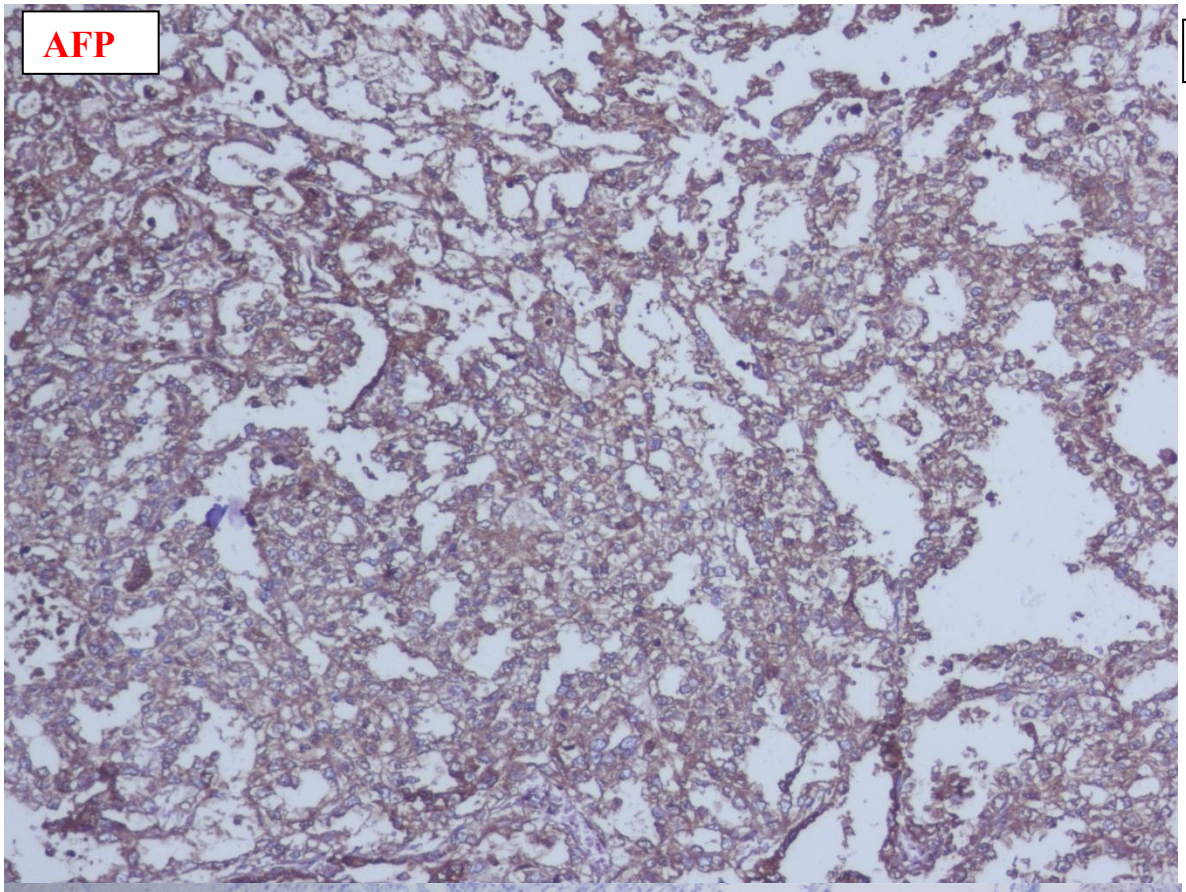


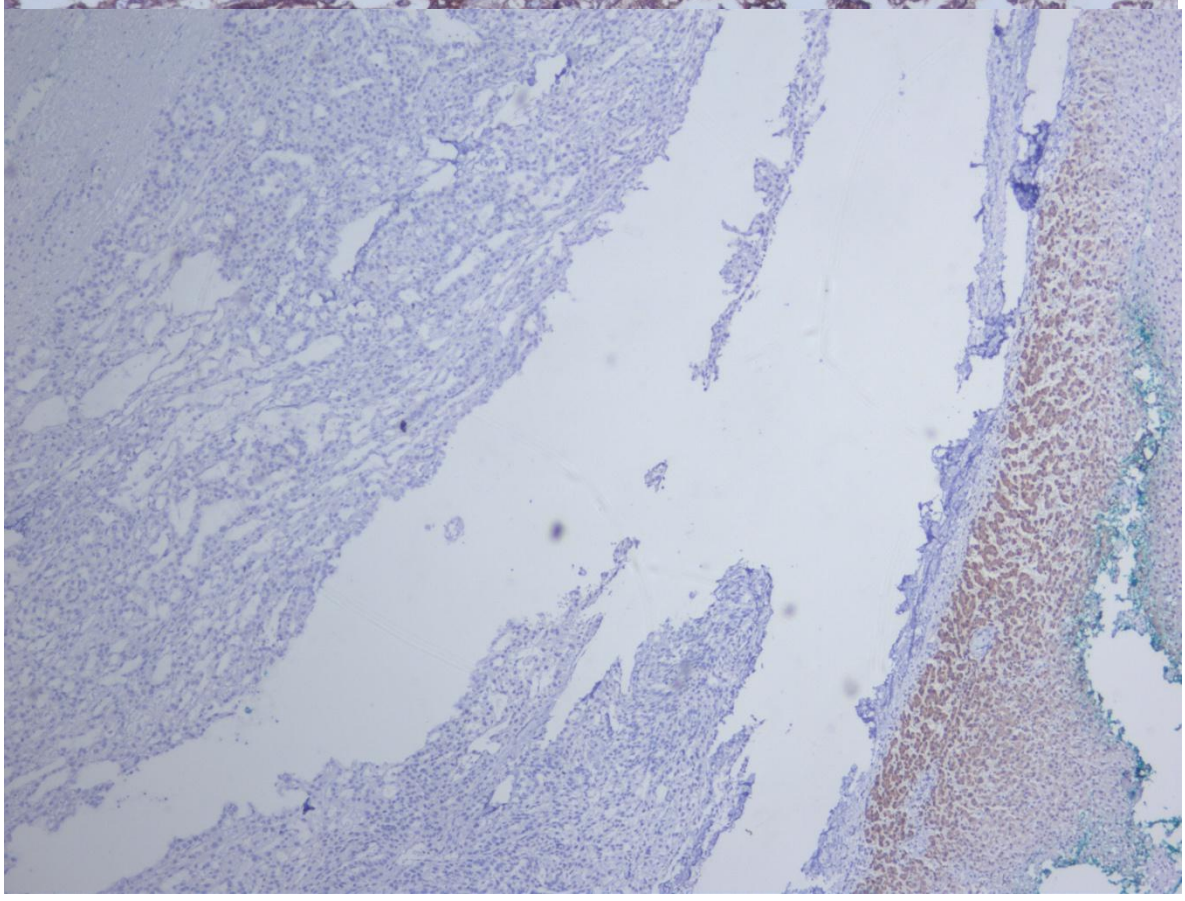




H&E sections shows a normal hepatic parenchyma with adjacent area showing tumor proper . Tumor cells arranged in microcystic, reticular pattern with focal myxoid background.

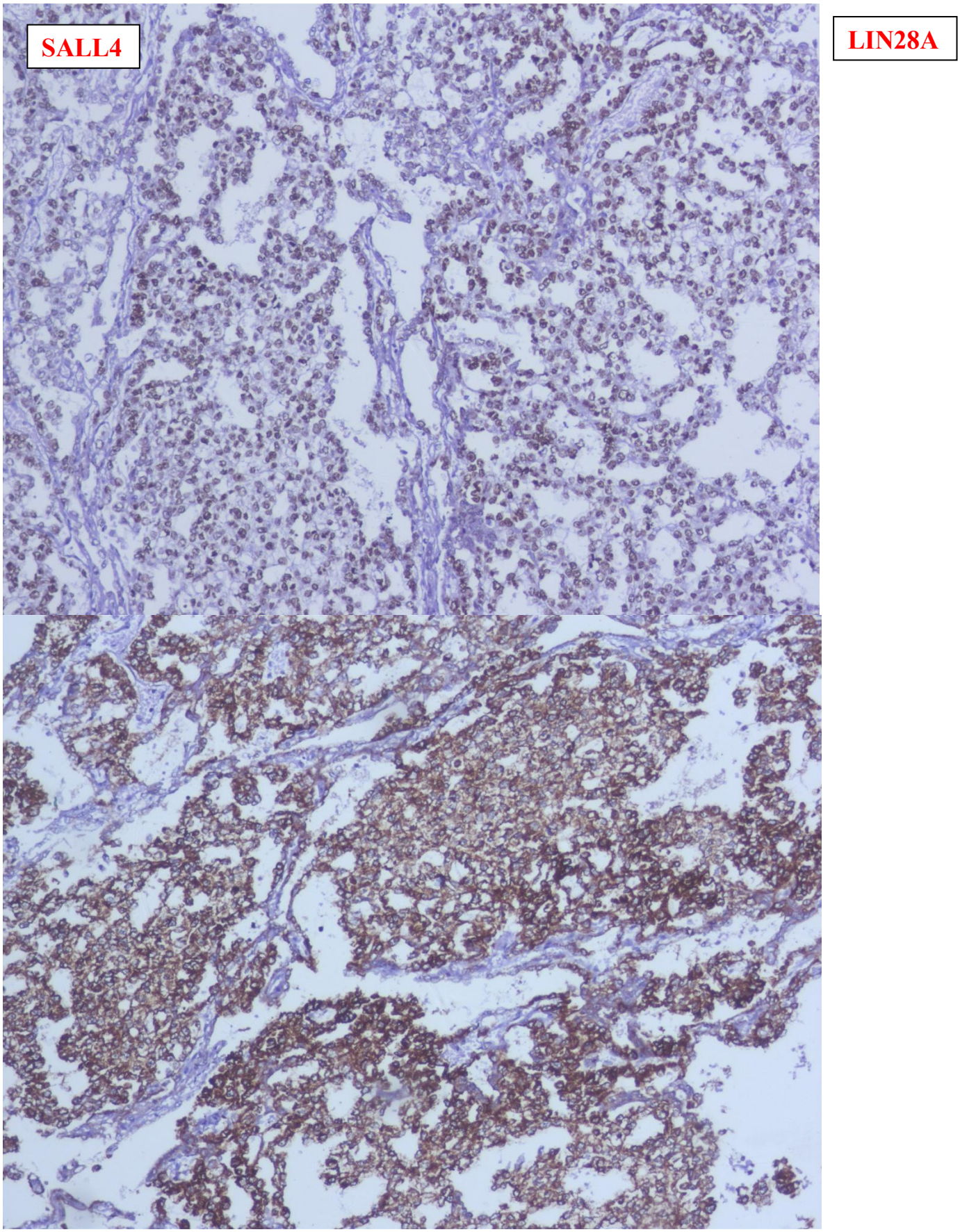
Characteristic Schiller-Duval bodies also seen. Tumor cells exhibits moderate nuclear pleomorphism.





AFP shows strong positivity in tumor cells.

Hepar 1 ne
parenchyma


SALL4
LIN28A

SALL4 shows strong nuclear positivity in the tumor cells.	LIN28A

Final Diagnosis

Primary Hepatic yolk sac tumor (YST)

Learning Points

1. Primary hepatic YST is a rare tumor.
2. Anterior mediastinum, retroperitoneum, sacrococcygeal region, vagina, and less commonly central nervous system &pineal gland are the commonest sites for extragonadal YST.
3. Characteristic histological features are microcystic or reticular pattern, Schiller-Duval bodies, PAS-positive and diastase resistant cytoplasmic hyaline globules.
4. YSTs do get misdiagnosed as hepatoblastomas or as hepatocellular carcinomas (HCC) especially in trucut biopsies as morphology is deceptive and same time shows elevated AFP levels.
5. Hepatoblastoma is the most common (accounting for 37%) primary hepatic tumor in children, presents as a large liver mass with hemorrhagic and necrotic areas, and with elevated serum AFP levels commonly. Histologically, the embryonal subtype often shows primitive tumor cells that can resemble YST.
6. SALL4 helps in diagnosing GCTs especially at extra-gonadal sites including YST.
7. LIN28A IHC is considered as a novel sensitive, and relatively specific diagnostic marker for pediatric YSTs, and it might have an advantage over AFP in diagnosing pediatric YSTs.
8. AFP is a sensitive serum tumor marker for diagnosis and in follow-up after therapy in YSTs.
9. Differentiation of YST, HCC, and hepatoblastoma is essential for appropriate treatment as HCC are chemoresistant and requires complete surgical resection while YST and hepatoblastoma involve combination treatment which includes chemotherapy and surgery.

Take home message

All cases of elevated AFP and liver mass in young childrens doesn't mean its Hepatoblastoma or HCC. Primary hepatic GCTs should be considered in the differential

diagnosis of hepatic tumors especially in young children and in biopsy specimens to ensure correct diagnosis for definite management and prognostication of the patient as platinum-based therapy and surgery has improved the prognosis of primary hepatic GCTs.