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Mesenchymal Tumor of the Liver in an Eight-Year Old Filipino Child

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Background

- Primary hepatic tumors are rare in children
- Less than 2% of neoplasms in the pediatric population
- Embryonal sarcoma of the liver (ESL), or undifferentiated embryonal sarcoma of the liver (UESL), is a rare condition but is the 3rd most common primary liver malignancy among children
- Constitutes about 6-13% of pediatric liver tumors
- Only 30 cases of ESL have been reported in the Philippines over the past 15 years

Methods/Case

- This is a case of an **8-year-old female**
- Two-week history of a non-tender, non-movable mass near the epigastrium
- **Initially managed as a case of parasitic infection**
- Imaging studies showed a **complex cystic mass** in the left hepatic lobe
- Given the age, the clinical considerations included hepatoblastoma, hepatocellular carcinoma, teratoma, and neuroblastoma
- The patient then underwent left lateral hepatic segmentectomy

Methods/Case

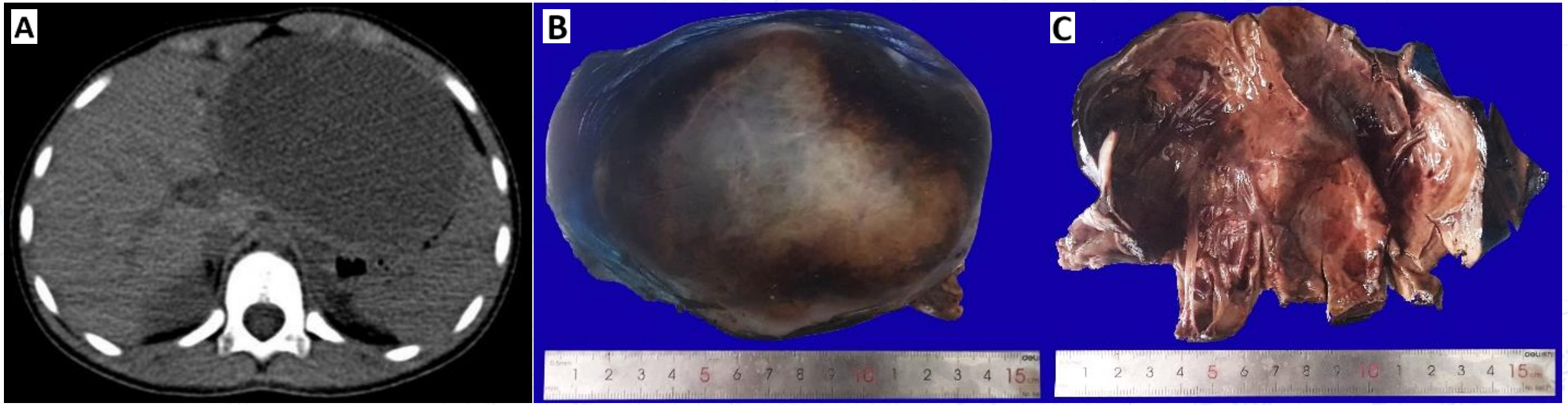

Figure 1. (A) CT scan of the abdomen showed a septated cystic mass in the left hepatic lobe.
(B) Gross examination showed a well-circumscribed, brown to dark gray, wedge-shaped left lobe with a fluctuant cystic structure.
(**C)** Cut surfaces revealed unilocular tan-brown septated cyst containing dark brown, turbid fluid.

Methods/Case

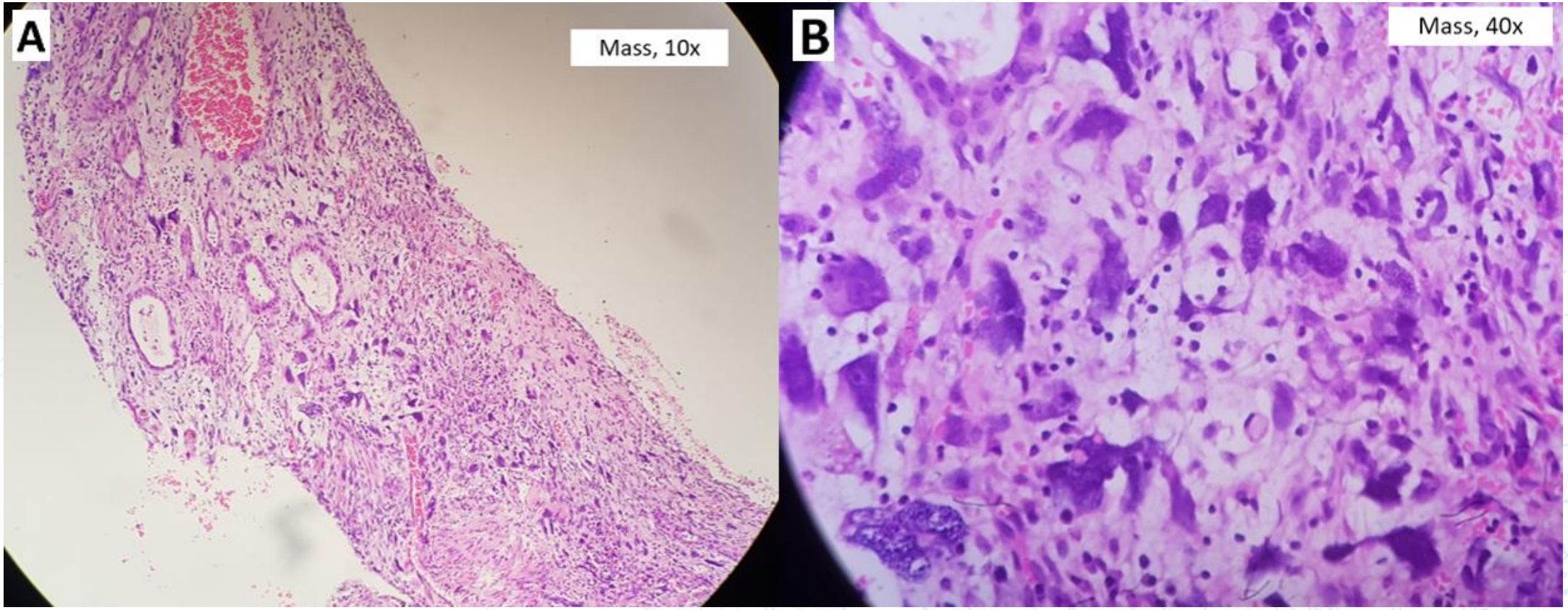

Figure 2. (A-B) Histologic examination of the tumor showed abundant pleomorphic atypical giant cells loosely arranged in a myxoid stroma. Entrapped hepatocytes and bile ducts were also seen (hematoxylin and eosin stain; original magnification: A, x10; B, x40).

Methods/Case

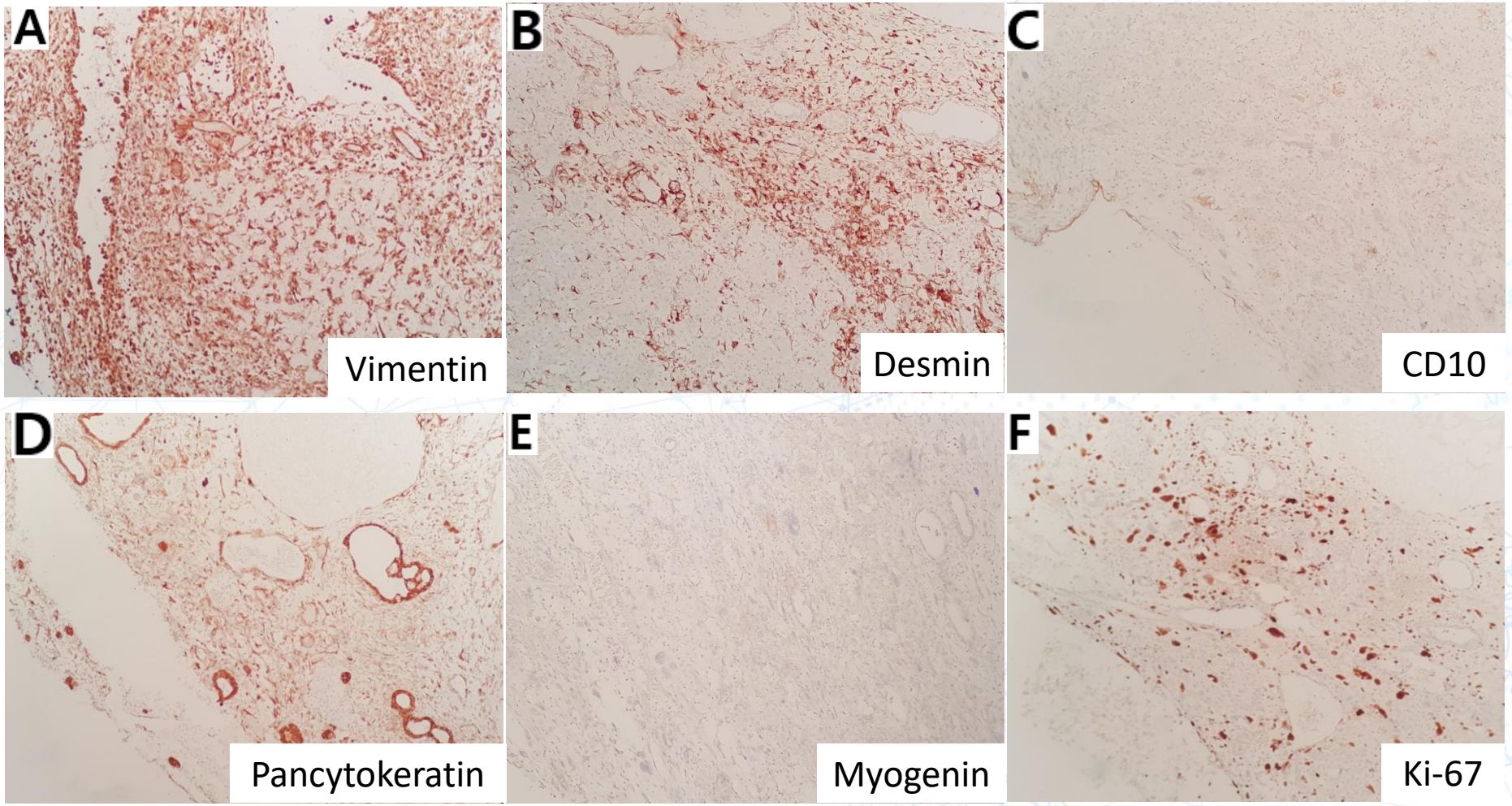

Figure 3. Immuno-histochemical staining showed strong diffuse cytoplasmic positivity for Vimentin **(A)** and Desmin **(B)**,

focal cytoplasmic positivity for CD10 **(C)** and Pancytokeratin **(D)**,

but were negative for Myogenin **(E).**

There was a 90% proliferative index with Ki-67 **(F)**, (immunoperoxidase; original magnification: x10).

Methods/Case

- Diagnosis: **Embryonal Sarcoma of the Liver (ESL)**
- **French Federation of Cancer Centers Sarcoma Group (FNCLCC)** Grade 2 (Differentiation: 3, Mitosis: 1, Necrosis: 0)
- **American Joint Committee on Cancer (AJCC) 8th edition:** pT1 (organ confined)
- Postoperatively, the patient was started on chemotherapy, consisting of Vincristine, Dactinomycin, and Cyclophosphamide

Results/Discussion

- ESL usually arises in children **5-15 years old** without racial or sex predilection but has also been reported in adults, as old as 86 years of age
- Most common symptoms include abdominal distension, pain, nausea, vomiting, anorexia, weight loss, and fever
- Rapid growth of the mass can cause spontaneous rupture, leading to intraperitoneal hemorrhage
- Liver function tests and tumor markers **may often be normal**
- **Laboratory results are nonspecific,** such as leukocytosis, eosinophilia, anemia, hypoalbuminemia; elevated GGT, LDH, CA 19-9.
- **Abdominal ultrasound**: predominantly solid mass with cystic components.
- **CT scan**: cystic to solid, heterogeneous, hypodense, often multiseptated masses

Results/Discussion

- The mainstay of diagnosis is **immunohistomorphologic examination**
- ESL **does not also have a specific immunophenotype**
  - At least 2 positive antibodies is recommended to confirm the diagnosis
- Possibly a malignant evolution of mesenchymal hamartoma
  - Seem to involve the **chromosome 19 microRNA cluster (C19MC)**
  - FISH for **t(11;19)(q13;q13.4)** may confirm this evolution in up to 45% of cases
    - This translocation may represent the first hit, and **TP53 mutations** serve as the second hit
- Treatment shifted from surgery alone to multimodal **with adjuvant chemotherapy**, improving the five-year survival up to 80%
- 15% of cases develop **metastasis** to the lungs, adrenals, peritoneum, pleura, and even the heart

Conclusion

- Clinical manifestations, laboratory and imaging findings for ESL lack specificity
- Radiologic findings of a predominantly solid mass on ultrasound and cystic on CT scan may raise the suspicion for ESL
- **THE DEFINITIVE DIAGNOSIS LIES ON THE PATHOLOGIC EXAMINATION, HENCE, AWARENESS ABOUT THE EXISTENCE OF THIS TUMOR IS IMPORTANT**
- Although it may pursue an aggressive clinical course, complete resection of the tumor is associated with a favorable prognosis
- However, some studies reported that negative resection margins was not a significant prognostic factor
- **Prompt diagnosis and a multimodal treatment are key to improving the outcome**