Giant Mesenchymal Hepatic Hamartomas with Adrenal involvement precipitating Respiratory Failure: A Myxomatous Mystery in a 3 month old

Background

- Hamartomas are benign, local proliferations of disorganized tissue. Hepatic mesenchymal hamartomas (HMH) are therefore a proliferation of mesenchymal liver tissue and are associated with elevated alpha-fetoprotein levels.
- Although hamartomas are benign, they can cause fetal demise both in utero and perinatally, due to mass effect on the diaphragm, lungs, or other vital organs. Rarely, HMH may present with Placental mesenchymal dysplasia (PMD).

Case

- A new-born month born at 35 weeks with hepatic cysts found on prenatal ultrasound. She presented to the ED with 4 days of non-bilious emesis.
- History revealed uneventful vaginal delivery and an uneventful pregnancy bedsides placental and hepatic cysts found on ultrasound. Over the next 2 months, the patient was admitted to the hospital multiple times for inability to feed and tachypnea. On serial ultrasounds, she was found to have enlarging cysts. A nasogastric tube was placed to help her with feeds. A CT scan was performed which showed large liver cysts and an adrenal lesions. Both were biopsied, but the pathological evaluation was non-diagnostic.
- The patient was admitted to the hospital and a broad workup was initiated. Pertient labs are explained below. The patient had multiple ultrasound exams and an MRI which are mentioned below. She had a US guided biopsy of a left adrenal lesion.

Labs

- Alpha fetal protein was elevated to 5760 with a local reference range of (0-339). Throughout her hospital stay, the patient remained persistently microcytically anemic. HCG, CEA, Coombs test, GGPT, ACTH, urine VMA and HVA, and Random Cortisol were within normal limits.

Figures

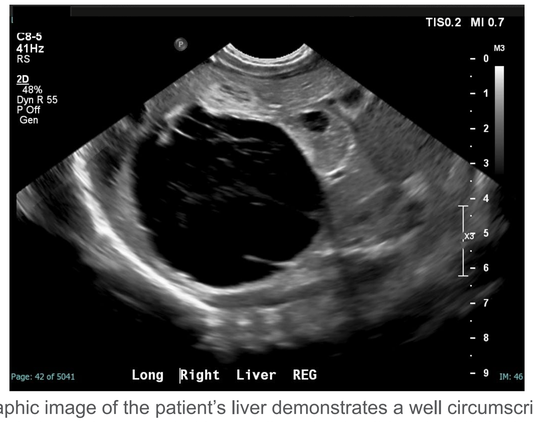

Figure 1: Sonographic image of the patient's liver demonstrates a well circumscribed, multiseptated, anechoic, cyst surrounded by smaller mixed solid and cystic lesions.

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Figures

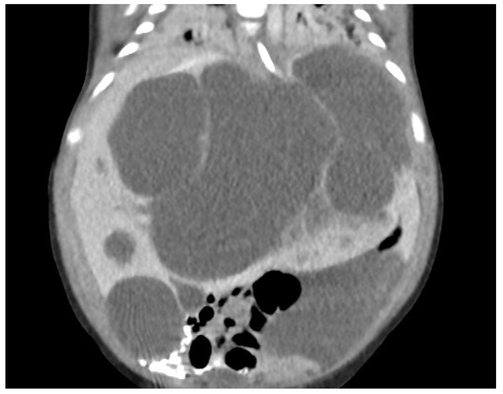
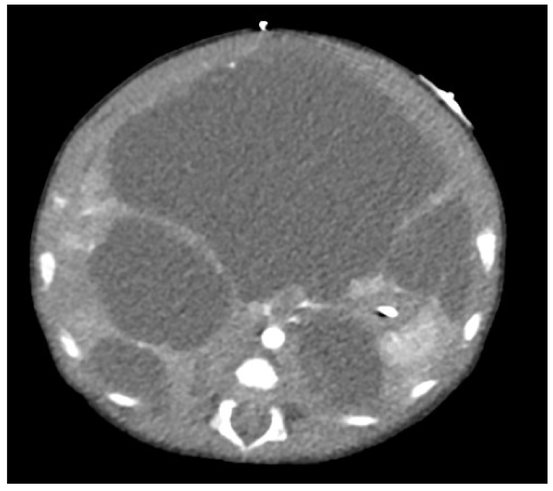


Figure 2: Coronal enhanced CT demonstrates multiple non-enhancing fluid attenuating cysts in the liver causing mass effect.



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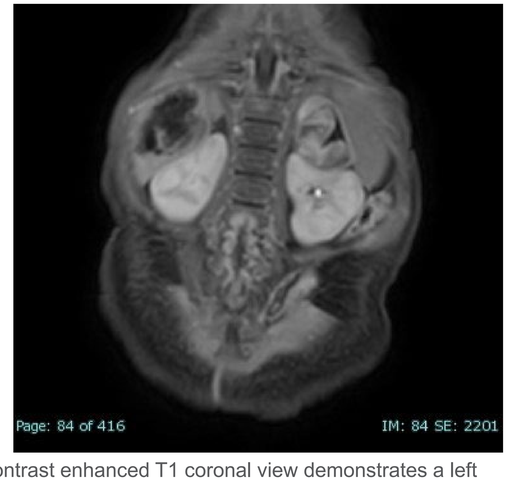

Figure 3: Contrast enhanced T1 coronal view demonstrates a left heterogeneous adrenal lesion that is seen distinct from the left kidney (no claw-sign). The lesion has enhancement of solid and non- enhancement of the cystic portions.

Discussion

- Hepatic Mesenchymal Hamartomas are benign tumors characterized by primitive myxoid stroma. Most commonly these lesions present as painless enlarging hepatic cystic lesions. On ultrasound, HMH presents as both solid and cystic masses of varying sizes. Some experts have theorized that HMH initially presents as solid lesions which then develop cystic change, which ultimately predominate at presentation. MRI is used for further evaluation of the lesions, due to its high resolution of the soft tissues.
- There are 9 reported cases in the literature of PMD and HMH combined and the combination of the two conditions is associated with a poorer prognosis of the fetus. There is no established preferred treatment plan. n most cases, complete resection of the lesions are attempted. For unresectable lesions, liver transplant is the treatment of choice.
- Upon evaluation by a nationally renown outside facility, the interpretation was consistent with a HMH. The report described the following: "Although the fibromyxoid tissue in the liver had similar histological appearance as that in the adrenal gland, immunostaining of the liver did not show any tissue of adrenocortical origin." It is thought that the patient has an adrenal origin of a hepatic mesenchymal hamartoma. To our knowledge, this has not been described in the literature before. Furthermore, this may suggest a malignant potential in an otherwise benign lesion.

Conclusion

- Hepatic mesenchymal hamartomas (HMH) are the second most common benign hepatic tumors behind hemangiomas. They are rare in occurrence and while they have been described in the literature, due to their atypical presentation, they can create diagnostic challenges. As seen in this case, the adrenal cystic lesion presented a distractor for the clinicians. Ultimately expert interpretation of the biopsy showed adrenal involvement of a HMH.

References

1. Harris K, Carreon CK, Vohra N, Williamson A, Dolgin S, Rochelson B. Placental mesenchymal dysplasia with hepatic mesenchymal hamartoma: a case report and literature review. *Fetal Pediatr Pathol*. 2013;32(6):448-453.

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