

Placental mesenchymal dysplasia and associated neonatal hepatic mesenchymal hamartomas

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Patient Presentation

27 year-old G3P1011 at 22 weeks' referred to Maternal-Fetal Medicine for evaluation of cystic placenta

- Ultrasound findings: Thickened placenta, 6.7 cm, cystic area adjacent to normal appearing placenta, with some vascular flow within (Fig. 1)
- Differential diagnosis: Placental mesenchymal dysplasia (PMD), chorioangioma, partial molar pregnancy
- Cell-free DNA low risk for triploidy

Prenatal Course

- Serial ultrasounds for biometry, MCA Dopplers & hydrops surveillance
- 27 weeks' – Normal MCA Doppler, no evidence of hydrops
- 28 weeks' – Membrane separation with dilated vessels on fetal surface, concern for placental hemorrhage, MCA Doppler PSV 1.5 MoM
- Patient sent to labor & delivery for consideration for percutaneous umbilical blood sampling & transfusion
- External fetal monitoring showed recurrent deep & prolonged decelerations necessitating cesarean delivery
- Placenta pathology: Features of PMD & fetal thrombotic vasculopathy/fetal vascular malperfusion, thrombi involving umbilical cord, fetal chorionic vessels & fetal stem vessels (Fig 2)

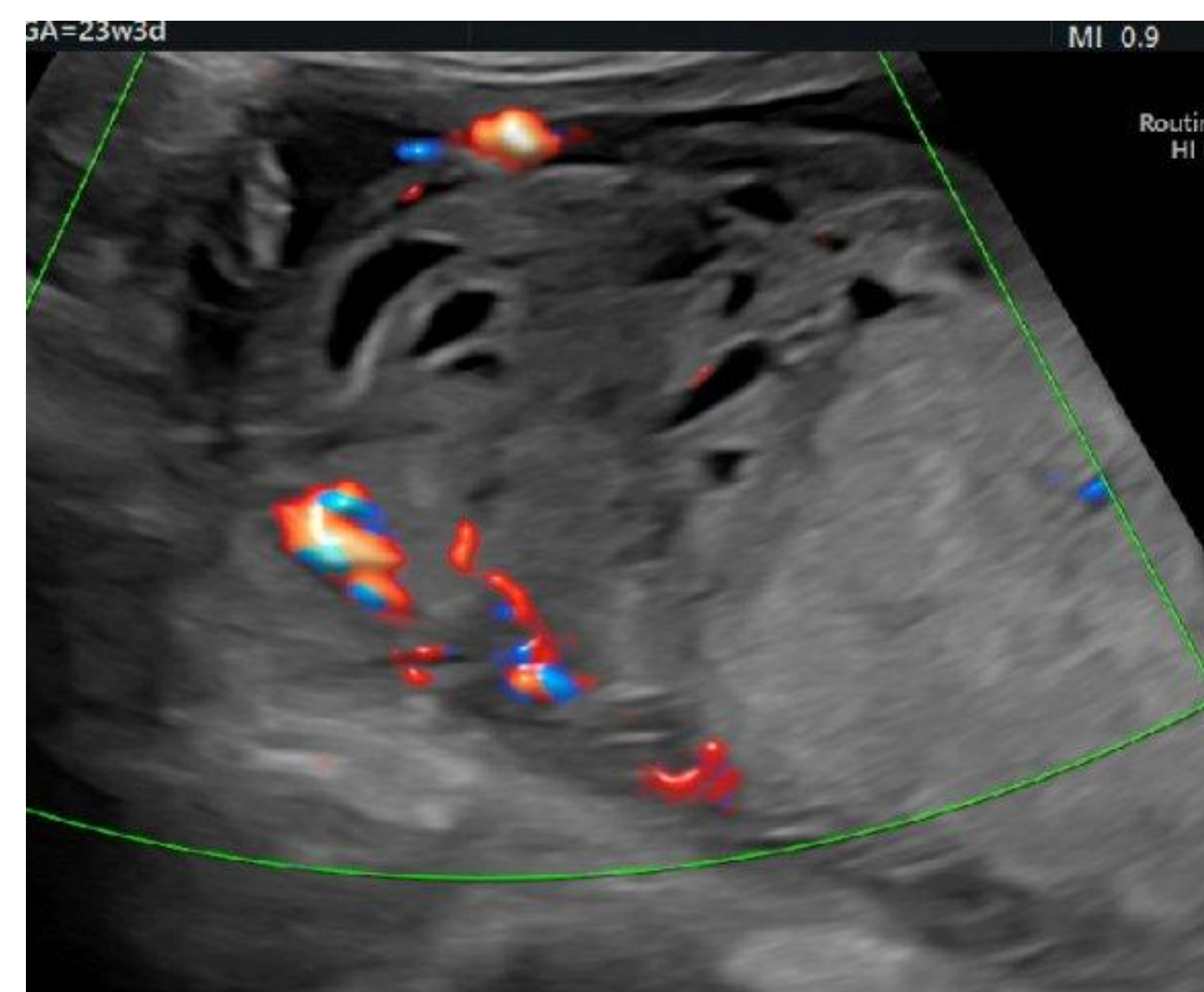
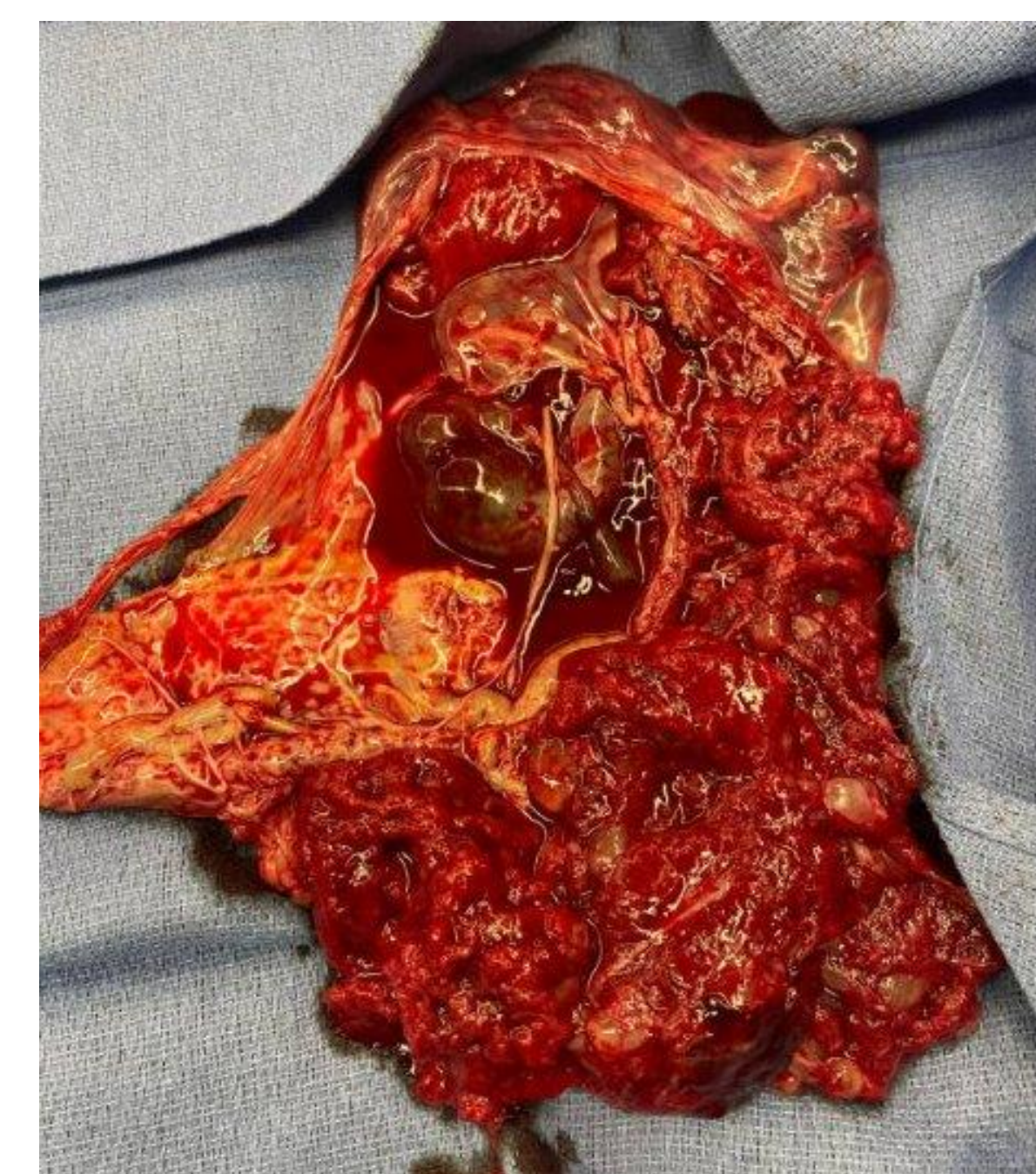


Figure 1: Ultrasound image, cystic placenta adjacent to normal placenta

Figure 2: Placenta



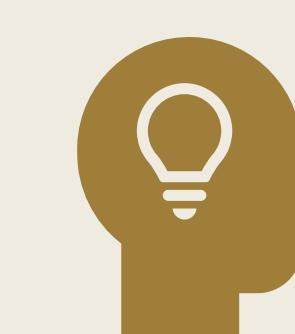
Neonatal Course

- Neonatal weight: 830 grams, appropriate for gestational age
- Hemoglobin 8.9, transfused PRBCs
- Genetics consult: Exam without features of Beckwith-Wiedemann Syndrome (BWS)
- 62 days: During TTE, noted liver cysts
- MRI: innumerable variable sized septated liver cysts consistent with hepatic mesenchymal hamartomas (HMH)
- 141 days: CT – marked hepatomegaly with cysts, displacement & distortion of abdominal organs, nonocclusive thrombus in proximal intrahepatic IVC, more inferior IVC & portal venous branches very small due to mass effect
- 240 days: Confirmed liver transplant candidate
- 13 months – Liver transplanted (explanted liver pathology consistent with HMH)

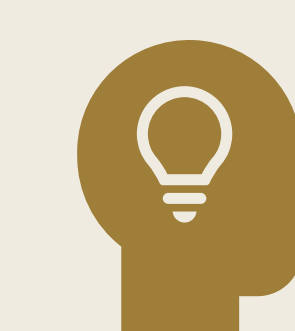
Teaching Points

- PMD is rare, true incidence is unknown
- Literature on associated HMH limited to case reports
 - Harris et al. reviewed 9 cases resulting in 3 fetal deaths, 1 neonatal death, 4 postnatal surgical resections, 1 neonatal resolution after serial percutaneous decompressions
- PMD is also associated with fetal anemia, growth restriction, BWS, maternal hypertension

CONCLUSION



PMD is associated with fetal anemia & serial ultrasound surveillance including MCA Doppler studies is recommended



In cases of PMD, thorough evaluations of the fetus & neonate, including liver and genetics, are warranted

REFERENCES

- 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 Dec;32(6):448-53. doi: 10.3109/15513815.2013.835893. Epub 2013 Sep 17. PMID: 24044629.