

A case of fetal hepatic haemangioendothelioma

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Objective

To present a case of prenatally diagnosed hepatic haemangioendothelioma, a rare benign vascular neoplasm that can be associated with life-threatening complications and review of literature for prognosis and management.

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Retrospective review of a case of hepatic haemangioendothelioma diagnosed prenatally in an inner city London Hospital.

Results

A 26 year old, nulliparous woman of South Asian descent was booked as low risk pregnancy. She had a normal anomaly scan at 20 weeks and her antenatal period was uneventful until 35+4 weeks gestation, when she perceived reduced fetal movements and was admitted for a suboptimal cardiotocogram (CTG). Growth scan performed at this stage revealed fetal growth around the 5-10th centile with abdominal circumference around 75th centile, normal liquor and fetal dopplers. There was a well defined mass of mixed echogenecities with areas of calcifications and peripheral vascularity in the right lobe of the liver, measuring 63 x 66mm. The heart appeared to be mildly enlarged, but with normal function and there was mild skin edema with bilateral hydroceles. She was reviewed in the tertiary fetal medicine referral centre and a differential diagnosis of hepatic adenoma, hepatoblastoma or haemangioendothelioma was entertained. Maternal alpha feto protein (AFP) was found to be elevated at 146. 74 U/ml. A semi-elective caesarean section was planned in view of the suboptimal CTG and the liver tumour and a baby boy was delivered at 36+2 weeks gestation in good condition. Postnatally the baby was seen by the neonatal / Paediatric hepatologists and a diagnosis of hepatic haemangioendothelioma was confirmed based on the ultrasound appearances aided by the Doppler flow velocities of the portal circulation. Propranolol was commenced postnatally and there was a significant reduction in the tumour size within few weeks.

Conclusion

Hepatic haemangioendothelioma is a rare benign vascular tumour of unknown aetiology, usually characterized by a low grade malignancy and good long-term prognosis. Though majority tend to regress spontaneously, they have the potential for progressive growth and can cause hepatic failure, coagulopathy, extra hepatic metastasis, congestive cardiac failure and death. The mainstay of diagnosis is imaging and histological findings. Around 14% of patients have an elevated AFP, though the rise is not as marked as in hepatoblastomas. Caesarean section is the recommended mode of delivery for prenatally diagnosed liver tumours because of potential tumour rupture and intra-abdominal haemorrhage during labour. Prenatal diagnosis is important as early evaluation and treatment improve prognosis. Oral Propranolol is safe and efficacious treatment with significant tumour regression in majority of cases. Resection, embolization or arterial ligation should be considered in case of complications.

