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## Case Report

# Congenital hemangioma of the face—Value of fetal MRI with prenatal ultrasound

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## ABSTRACT

We report the presentation, workup, and pre/perinatal management of a fetus with a large congenital hemangioma of the face. Hemangiomas are benign vascular neoplasms frequently encountered in neonates and infants. The less common congenital variant develops in utero and can present on prenatal ultrasound with diagnostic uncertainty as well as clinical implications for delivery. The differential diagnosis for these solid vascular masses when located in the head and neck includes myofibroma, kaposiform hemangioendothelioma, teratoma, and encephalocele. Potential clinical issues relate to size and location of the mass and include airway obstruction, disruption in development or compression of the globe, invasion into the cranial vault, bleeding and ulceration, and high output heart failure. Prenatal ultrasound and MRI play an important role in the diagnosis of congenital hemangiomas and planning for delivery and immediate postnatal cares.

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## Introduction

Hemangiomas are benign vascular neoplasms frequently encountered in neonates and infants. The more common infantile type usually develops shortly after birth and follows a characteristic clinical course of rapid growth with subsequent stabilization and gradual involution. The typical cutaneous appearance of superficial infantile hemangiomas usually enables diagnosis without tissue sampling. The less common

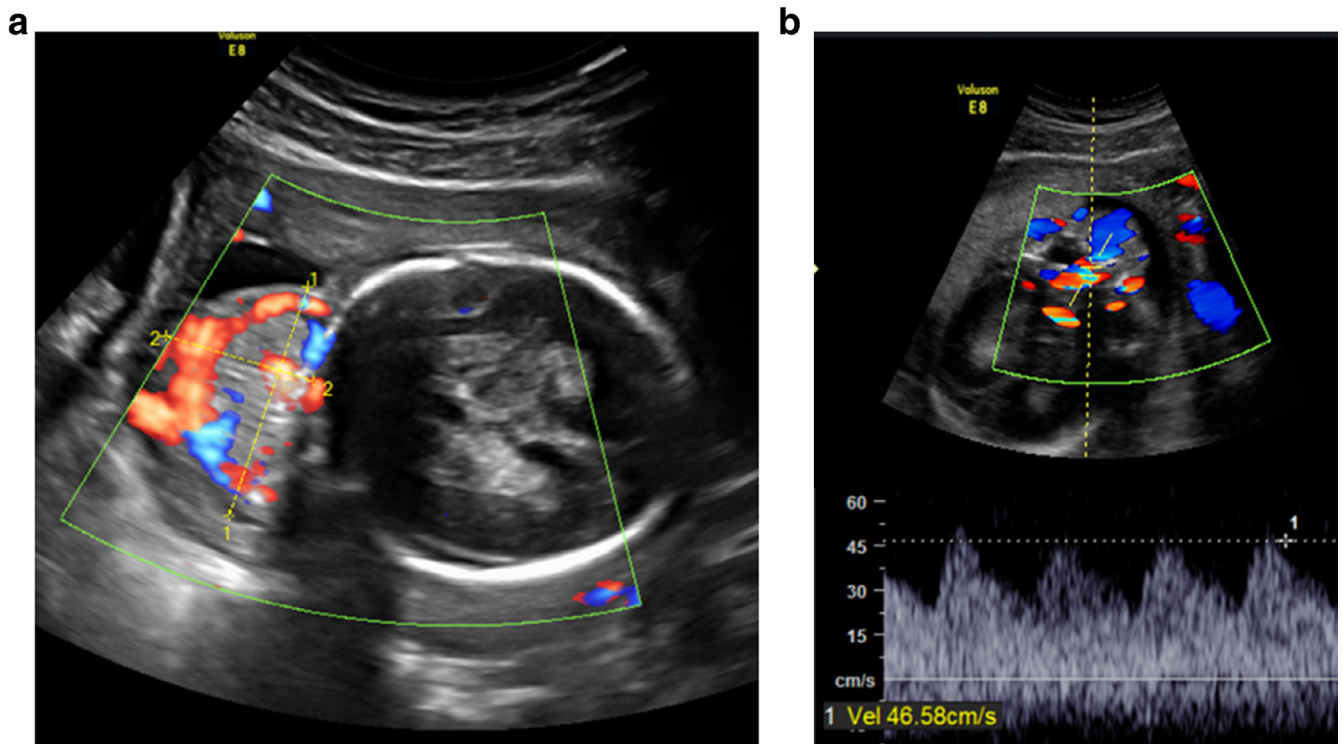
congenital hemangioma is pathologically and clinically distinct, having completed its proliferative phase in utero with variable patterns of involution observed during the first year of life (including rapidly involuting, partially involuting, and noninvoluting subtypes). When congenital hemangiomas are detected in utero, the gross appearance cannot be assessed, and a differential diagnosis must be considered. Furthermore, their highly vascular nature and predilection for the head and neck can have serious clinical implications before, during, and after delivery. This case illustrates the utility of

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**Fig. 1 – Prenatal ultrasound at 20 weeks. Homogeneous solid soft tissue mass over the fetal forehead with increased vascularity (A) and low resistance arterial waveforms (B). Shadowing of the cranium obscures evaluation of intracranial contents.**

prenatal ultrasound and MRI in diagnosing and managing these lesions in utero.

### Case report

A 17-year-old G1 POAOL0 female was referred for a fetal facial mass noted on a transabdominal ultrasound performed at 18 weeks gestation at another institution. Maternal and paternal histories were noncontributory, and the fetus had been naturally conceived.

Repeat ultrasound at our center at 20 weeks revealed a well circumscribed, homogeneous soft tissue mass arising from the left forehead with prominent internal vascularity demonstrating low resistance arterial waveforms (Figs. 1A and B). Shadowing from the cranial vault prevented a detailed evaluation of the brain and limited assessment for intracranial extension.

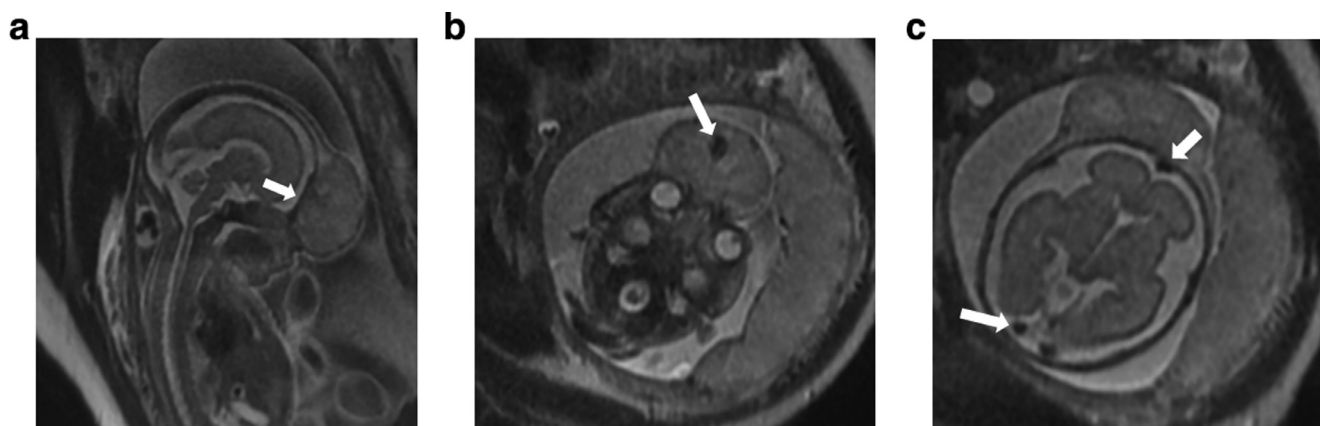
A fetal MRI was performed at 26 weeks for further characterization of the mass and brain. The mass was centered in the soft tissues of the forehead and extended over the left globe and nose, with slight flattening of the underlying frontal bones (Fig. 2A). The mass measured 5 × 5 cm and was mildly T2 hyperintense with hypointense flow voids from prominent vessels (Fig. 2B). There was no intracranial extension, cystic change, calcification, or hemorrhage. The dural venous sinuses were enlarged due to arteriovenous shunting in the mass (Fig. 2C), but the appearance of the brain was normal.

No airway extension or obstruction was seen, and there was no evidence of fetal hydrops.

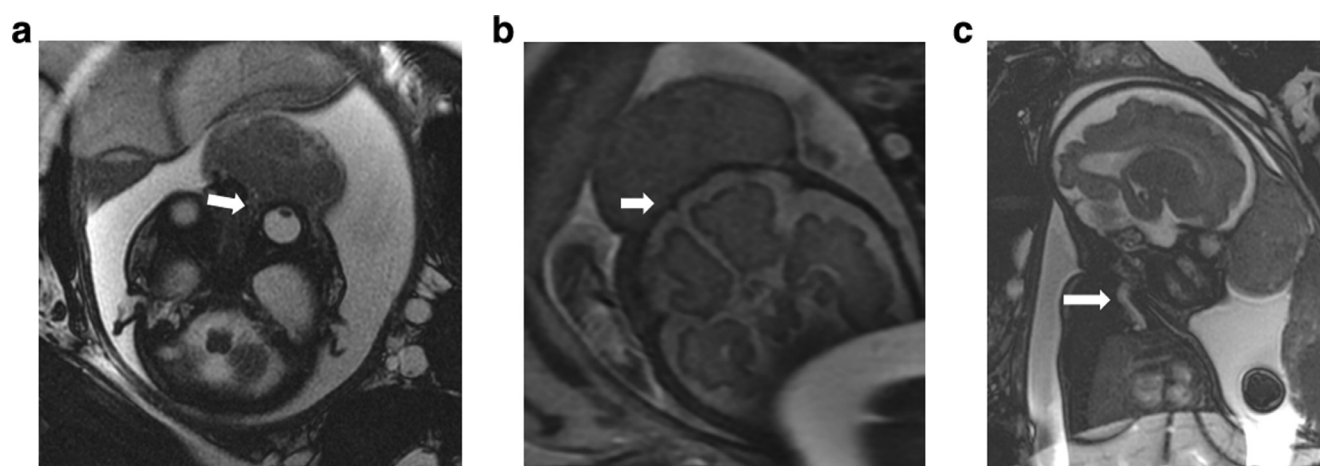
Given the solid and highly vascular appearance of the mass by ultrasound and MRI, a congenital hemangioma was the favored diagnosis. Other solid congenital soft tissue masses, including myofibroma, kaposiform hemangioendothelioma, and sarcomas were considered less likely, though increased vascularity can be seen in these lesions. Teratoma was considered unlikely given the location and homogeneous, solid appearance. The lack of intracranial extension excluded encephalocele.

MRI was repeated at 33 weeks to evaluate growth of the mass and detect any new intracranial or airway extension that would impact delivery and immediate postnatal care. The mass had grown in size proportionally to the fetus and measured 5 cm by 7 cm. The possibility of extension into the left orbit was raised (which could affect long-term vision), and there was increasing mass effect on the nares (Fig. 3A). The dural venous sinus enlargement appeared resolved (Fig. 3B), but prominent vessels extended over the surface of the mass with persistent jugular vein enlargement, suggesting shunting through extracranial veins (Fig. 3C). The heart was mildly enlarged, but there was no evidence of fetal hydrops.

Given these findings, a cesarean section was planned to minimize trauma to the mass with potential hemorrhage, and ENT was to be present at the birth for advanced airway management if necessary. Weekly ultrasounds over the remainder of the pregnancy showed no development of hydrops.



**Fig. 2 – Prenatal MRI at 28 weeks. Sag SSFSE (A) and axial SSFSE (B and C) images show the soft tissue mass abutting the eye and nasal structures with mild flattening of the underlying frontal bone (A, arrow), enlarged intratumoral vessels (B, arrow), and engorgement of the dural venous sinus system from shunting of blood flow through the tumor (C, arrow). There was no intracranial extension or evidence of brain abnormality.**



**Fig. 3 – Prenatal MRI at 33 weeks. Axial bSSFP (A) shows that the mass has increased in size in proportion to the fetus with possible intraorbital and nasal extension (A, arrow). Axial SSFSE (B, arrow) shows resolution of the dural venous sinus engorgement, though the jugular vein remained enlarged on the sagittal bSSFP image (C, arrow).**

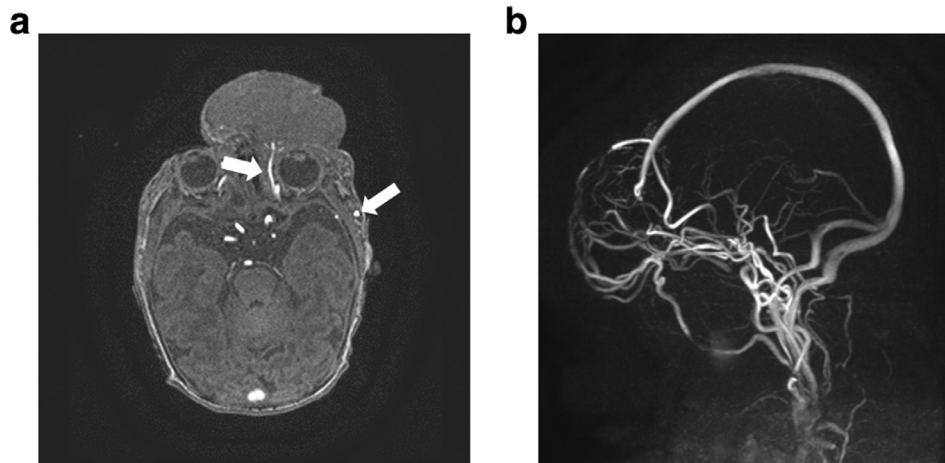
The mother presented at 38 weeks, 5 days gestation with spontaneous rupture of membranes and underwent uneventful C-section. The neonate took spontaneous breaths with Apgar scores of 8 at 1 minute and 8 at 5 minutes and required nasal oxygen for mild hypoxia immediately after birth. The large protuberant 7 cm by 9 cm frontal mass extending over (but not involving) the globe showed a purplish-red vascular discoloration of the overlying skin with foci of erosion or exudate. Flexible endoscopy showed no extension into either nasal passage. The infant was eventually transitioned to room air with no further respiratory issues.

Confirmatory postnatal MRI/MRA showed prominent feeding vessels arising from the ophthalmic and superficial temporal arteries (Fig. 4A) with drainage through engorged scalp veins (Fig. 4B). Surgical excision was not pursued due to concern for ophthalmic artery compromise. Limited biopsy confirmed a hemangioma with negative Glut-1 staining, consistent with a congenital hemangioma. Treatment with

propranolol in a dose of 1 mg/kg/day was instituted, and the mass showed partial regression over 9 months, consistent with an involuting subtype of congenital hemangioma.

## Discussion

Hemangiomas are benign vascular tumors comprised of endothelial cells [1,2]. These lesions undergo a defined natural progression with initial rapid proliferation followed by stabilization and, in most cases, gradual involution and regression. Infantile hemangiomas, which are not present at birth, are common and typically proliferate during the first year of life. They then slowly regress over the first decade of life. Congenital hemangiomas are less common, undergo proliferation only in utero, and present with maximal size at birth (in the absence of subsequent hemorrhage). After birth, they either



**Fig. 4 – Postnatal MRA. Time-of-flight MR angiography (left) shows enlargement of the left ophthalmic artery (ICA supply) and left superficial temporal artery (ECA supply) (A, arrows). 3-D phase contrast MR venography (B) shows drainage through engorged scalp vessels with relatively less intracranial drainage.**

rapidly or partially involute (RICH or PICH, respectively) or show no regression (NICH, or noninvoluting congenital hemangioma). Histologically, infantile hemangiomas stain positive for Glut-1 while congenital hemangiomas do not.

Clinical issues encountered with hemangiomas relate to their size and location [2]. Facial hemangiomas can obstruct the airway, compress or cover the globe (potentially disrupting vision long term), invade the cranial vault, and bleed or ulcerate. Of sufficient size, congenital hemangiomas may also lead to shunting with high output heart failure.

Prenatal and postnatal imaging cannot reliably differentiate the subtypes of hemangiomas. On ultrasound, hemangiomas are very vascular solid masses which demonstrate abundant venous and low resistance arterial waveforms. On MRI, they are typically T1 hypointense, T2 hyperintense, and show avid post contrast enhancement, though contrast is not administered in fetal cases. The congenital types are more likely to be poorly defined and heterogeneous [3,4] with calcifications, venous thrombi, and larger vessels, including possible aneurysms [5-7]. As hemangiomas regress postnatally, they demonstrate decreasing vascularity with increasing resistance of arterial waveforms plus gradual fatty infiltration of the mass.

While several medical treatments (primarily propranolol and steroids) are highly efficacious for infantile hemangiomas, therapy for congenital hemangiomas is limited. The rapidly involuting subtype (RICH) is usually observed as it shows regression in size of 50% by 7 months and nearly 100% by 14 months [8]. As drug treatment has not been shown to accelerate involution in RICHs, it was likely unnecessary in our case. Symptomatic RICHs (as well as PICHs and NICHs) may be embolized or resected, and pulsed dye laser or sclerotherapy may improve a superficial lesion's color [2,8].

This case illustrates the utility of fetal MRI, and its complementary role with ultrasound, in evaluating congenital soft

tissue lesions. For facial masses, MRI can demonstrate critical organ invasion that may alter perinatal management.

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