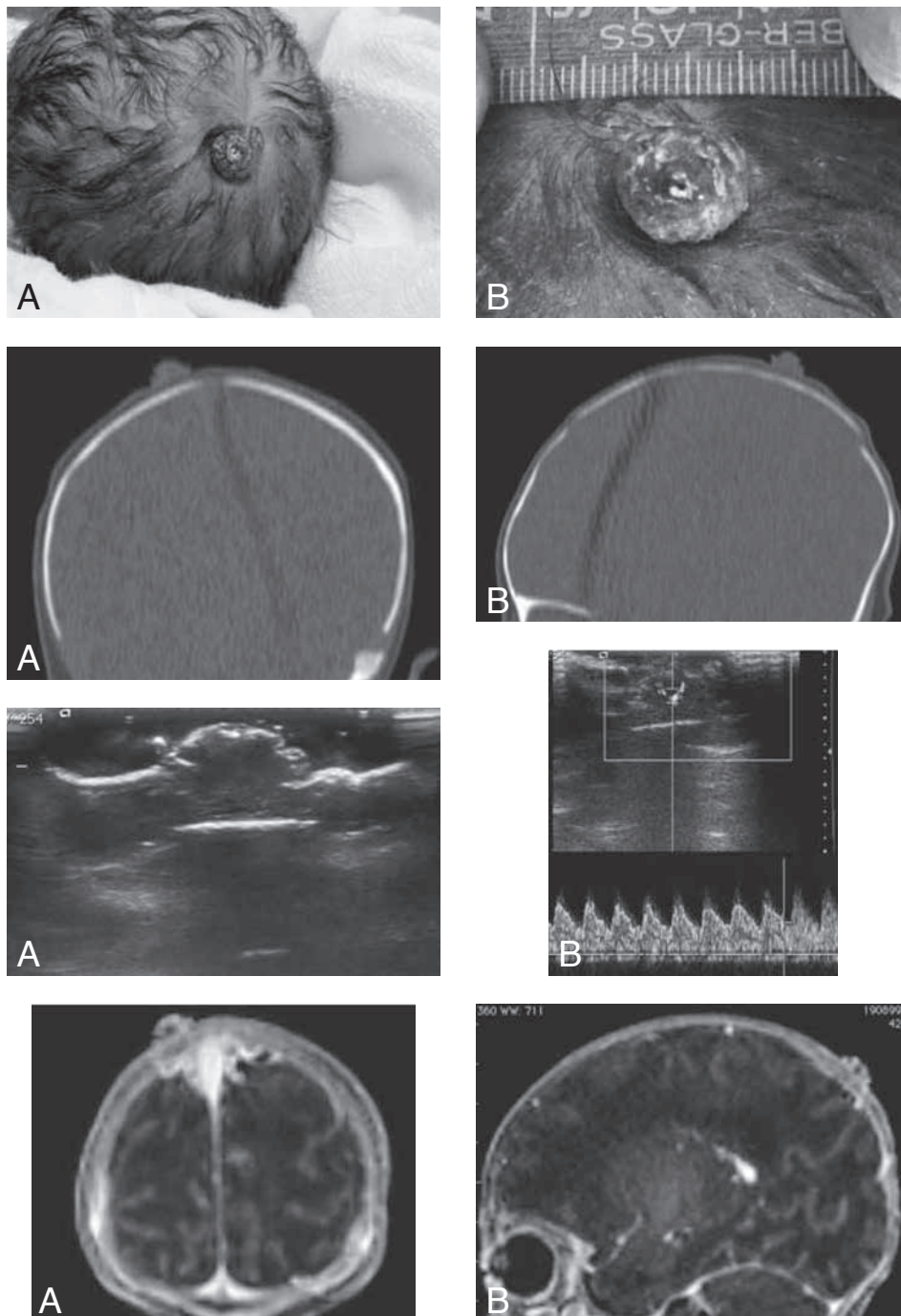


## CONGENITAL HEMANGIOMA OF THE SKULL

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**Key-word:** Angioma

**Background:** A newborn (male, 6 hour-old) was referred to the radiology department for imaging of a solid tumor-like nodular lesion on the vertex of the skull. The treating pediatrician wanted to exclude a (meningo)-encephalocele or other dysraphism-like lesions.



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Fig. 

1A	1B
2A	2B
3A	3B
4A	4B

## Work-up

At clinical presentation a solid raised red lesion is seen on the vertex of the skull (Fig. 1 A,B).

On CT scan of the skull (Fig. 2, A: coronal reconstruction in bone window and B: sagittal reconstruction in bone window), no bone defects nor bony abnormalities are found. A cutaneous, sharply demarcated soft-tissue mass is seen.

On ultrasonography (Fig. 3A) the cutaneous lesion measures 11 x 11 x 4 mm and has a lobulated surface without keratinisation of the skin. The findings were compatible with a cyst or mass. Doppler-ultrasonography of the lesion (Fig. 3B) shows a vascularised lesion with fast flow, suggestive for hemangioma or arteriovenous malformation.

MRI of the brain (Fig. 4, A: contrast-enhanced MPRAGE coronal image and B: contrast-enhanced MPRAGE sagittal image), after gadolinium administration, an intense enhancement of the lesion is seen. Communication with intracranial vessels is formed by small vessels in the diploë. On MRI, the lesion is mildly hyperintense on T2-WI with small intern flow voids.

There are no signs of encephalocele.

## Radiological diagnosis

Based on the clinical presentation and imaging findings, a rapidly involuting *congenital hemangioma* was suspected. Resolution of the lesion during the first weeks of life confirmed the diagnosis.

## Discussion

While infantile hemangiomas are the most common soft-tissue tumors of childhood, congenital hemangiomas are much more rare. Unlike infantile hemangiomas, they are mature at birth and don't have accelerated postnatal growth. Two major types of congenital hemangiomas have been described: rapidly involuting congenital hemangiomas (RICH) and the less common noninvoluting congenital hemangiomas (NICH). Both can appear as bossed plaques or tumors.

Most common locations for RICH are the limb, head, or neck. Three variants of RICH have been described: (1) a lesion with a characteristic red-purple color, often with telangiectasias on its surface or at the periphery, (2) a flat infiltrative tumor with violaceous color, or (3) a raised grayish tumor with tiny telangiectasias surrounded by a pale halo. RICH may involute rapidly resulting in difficult to heal central fissuring and ulceration. In the nuchal area, RICH lesions may be seen on second-trimester ultrasonography and be suggestive for lymphatic malformation, encephalocele, or other forms of cranial dysraphism.

RICH exhibit fast flow at ultrasonography and may show flow voids on MRI. Angiography of RICH shows large and irregular feeding arteries in a disorganized pattern with arterial aneurysms, arteriovenous shunts, and intravascular thrombi. Rapidly involuting congenital hemangioma lesions stain negative with GLUT1, a marker which is positive in infantile hemangiomas. In the majority, involution starts within the first weeks of life and is complete after 6 to 14 months of life. In a patient with typical clinical presentation and features on ultrasound or MRI, observation is usually the first line of treatment.

Firm lesions that show no tendency to involute are difficult to differentiate, even after imaging, and require biopsy to exclude more serious tumors such as fibrosarcoma or rhabdomyosarcoma. Some cases of apparent RICH cease to involute, persisting with clinical characteristics identical to NICH.

NICH are much less common and will never disappear or involute, they undergo proportional growth.

NICH are more plaque-like with a pink or purple color and prominent overlying coarse telangiectasias.

Doppler-ultrasonography shows fast flow, and magnetic resonance imaging shows hyperintensity on T2-weighted sequences with flow voids similar to infantile hemangioma. Arteriography shows arterial-like feeders and a tumorlike capillary blush with small arterial channels. Early venous draining is not seen, which differentiates these lesions from arteriovenous malformations or arteriovenous fistulas. NICH lesions stain also negative with GLUT1 and excision is the recommended treatment.

Most hemangiomas are diagnosed clinically and require no imaging. Ultrasonography and contrast-enhanced MRI are the modalities of choice for assessment of extension, relationship with adjacent structures and atypical features.

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