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Prenatal diagnosis of scalp congenital hemangiopericytoma

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Hemangiopericytoma (HPC) is a rare form of vascular tumour. It consists of extensive proliferation of pericapillary cells (pericytes). The most common locations are the head and neck, lower extremities and retroperitoneum. HPC can be benign or malignant. Malignant HPC can metastasize to other areas of the body, usually to the lungs. Up to 5-10% HPCs present in childhood and 5-40% occur in the 1st year of life. We report a fetal case of HPC presenting as an ultrasound finding of a forehead mass. CASE: The proband was a 23y P1 Caucasian woman and her husband was 22y and of same descent. The couple was healthy and non-consanguineous. Early fetal ultrasounds were normal. IPS was negative. Ultrasound at 19w showed bilateral pelviectasis. A follow-up ultrasound at 32w confirmed the pelviectasis, and also showed a midline mass over the forehead just above the nasal bridge, with slightly amorphous content measuring 4.1 x 4.0 x 3.1 cm. The mass did not appear to penetrate the scalp or skull and no gross abnormalities in the brain were seen. The nose and eyes appeared normal. The findings were confirmed by fetal MRI. The couple decided to continue the pregnancy. Delivery was at 38.8w via CS and was uncomplicated. The birth growth parameters were all normal. The APGAR scores were 9(1) and 10(5). A pedunculated tumor with a large base which measured 4 cm, originating in the right side of the forehead was noted. There were areas of necrosis and excoriation over the skin and no thrill over the tumor. The baby was otherwise not dysmorphic. Complete surgical resection was performed and histopathology was consistent with HPC. DISCUSSION: Congenital HPC behaves in a distinct manner and has excellent prognosis. Etiologically, congenital HPC may arise from a pluripotent cell capable of differentiating into smooth muscle cells, pericytes and retains its capabilities to differentiate into more mature cells, resembling infantile myofibromatosis. A single entity called composite myofibromatosis has been proposed. From our knowledge, this case is the 7th case of congenital/ infantile HPC reported in the literature. Previous cases affect the lip, nose, temporal bone, meninges, the cerebrum and the neck region.

Complete surgical resection is the ideal treatment of choice. Since excellent response to chemotherapy has been seen, extensive surgical excision and radiotherapy are to be avoided. Long-term follow-up is recommended as relapse may occur late.