Fetal myopericytoma: a rare tumour with good prognosis

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Abstract

Reported here is a case of fetal myopericytoma, investigated by prenatal ultrasound and confirmed by autopsy and histopathological examination after termination of pregnancy. On antenatal ultrasonography, there was a large, multicystic neck mass with enhanced Doppler flow in the fetus at 20 weeks gestation. Parents terminated the pregnancy and an autopsy was conducted on the abortus. The tumor was located in the neck and had numerous, large calibre vessels within. Histopathological examination with immunohistochemistry revealed the tumor to be a benign myopericytoma. This is the earliest gestation at which fetal myopericytoma has been reported.

Introduction

Myopericytomas are benign pericytic tumours usually occurring in adults and involving the distal extremities. There are only few case reports of antenatally detected myopericytomas described at 32-33 weeks of gestation. We are reporting a case of fetal myopericytoma involving the neck detected at an early gestation of 20 weeks.

Case report

A twenty five year old primigravida presented for evaluation of a fetal neck mass detected on ultrasound at 20 weeks of gestation. There was a large multicystic mass of size 8 cm X 6 cm on the right side of the fetal neck. There were multiple hypo-echoic spaces within the mass which, on color Doppler, showed increased vascular signals and large feeding vessels arising from the aorta (Figure 1. A-B). Intracranial anatomy was normal and there were no other associated malformations. Amniocentesis was done and the fetal karyotype was reported to be normal. Differential diagnoses of arterio-venous malformations, haemangioma, vascular goiter and teratoma were considered. The extreme vascularity of the mass suggested increased risk of cardiac failure in the fetus. The family opted to terminate the pregnancy in view of the uncertain diagnosis and prognosis.

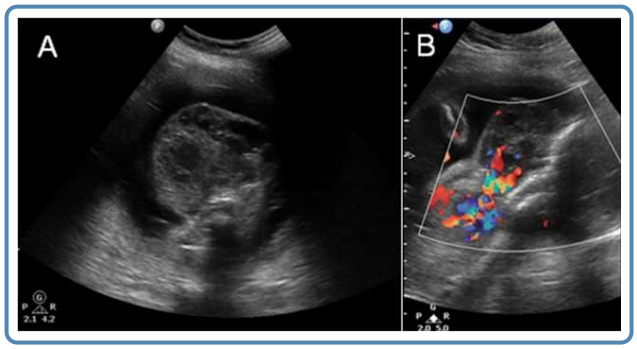

Figure 1 Ultrasound of the fetus showing, (A) the neck mass, (B) extreme vascularity on color doppler.

On postmortem examination of the abortus, there was a mass on the right side of neck, measuring 8 cm X 6.5 cm, extending from the ear lobule upto the clavicle (Figure 2). On dissection, numerous branching vessels were found within the mass. These vessels extended into the thorax and were connected by a major feeding vessel to the aorta. The tumor could be enucleated easily and there were no intracranial extensions or involvement of underlying bones. Histopathological examination,

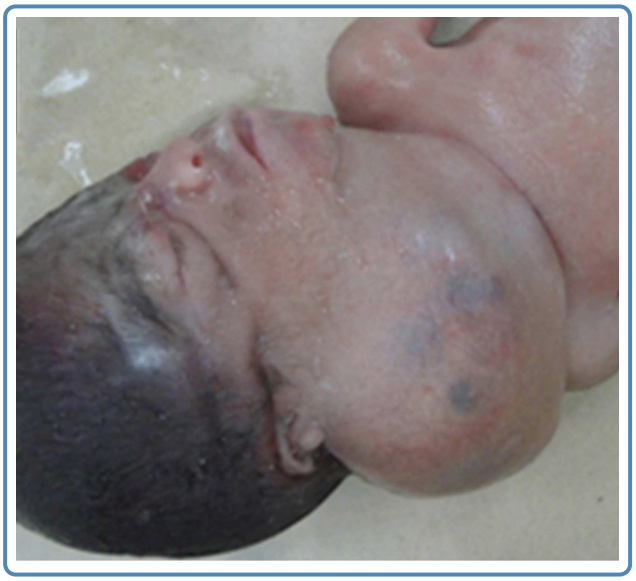

Figure 2 Autopsy of the fetus showing the neck mass.

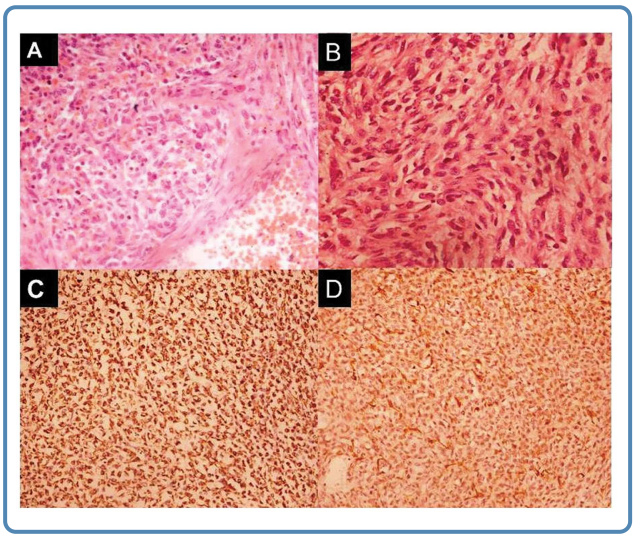

Figure 3 Hematoxylin and Eosin staining of the tumor tissue. (A) Tumor cells seen in sheets with intratumoral blood vessels. (B) Tumor cells displaying nuclear pleomorphism, coarse chromatin, conspicuous nucleoli and eosinophilic cytoplasm with spindling. (C) Positive immunohistochemistry (IHC) staining for vimentin. (D) Positive immunohistochemistry (IHC) staining for smooth muscle actin.

with the addition of immunohistochemistry, revealed the tumor to be a benign myopericytoma (Figure 3. A-D).

Discussion

This is a report of a 20 weeks fetus diagnosed to have a myopericytoma by prenatal ultrasound and confirmed by autopsy and histopathological examination. There are only few reports of prenatally detected myopericytomas involving the lip, nose, brain and fetal neck, all being diagnosed in the third trimester, after 32 weeks of gestation. In our case, the diagnosis was made at an earlier gestation of 20 weeks.

Stout and Murray (1942) were the first to describe hemangiopericytoma as a tumor composed of Zimmerman's pericytes showing the characteristic 'staghorn' branching vascular space. However, the histological features and the branching vascular pattern which were once considered to be its characteristic feature are nonspecific and are seen in many tumors such as benign fibrous histiocytoma, synovial sarcoma, leiomyosarcoma and others (Kempson et al., 2001).

The term myopericytoma was used in 1998 to describe a spectrum of tumours with clinicopathological features that overlap with hemangiopericytoma. WHO describes myopericytomas as pericytic lesions showing differentiation towards myoid/contractile perivascular cells with a characteristic tendency to grow in a circumferential perivascular fashion (Fletcher et al., 2002). Currently, this class of tumours encompasses infantile myofibromatosis-like lesions such as glomangiopericytomas of adults, infantile myofibromatosis and infantile haemangiopericytoma (Mc Menamin et al., 2002).

Myopericytoma usually presents in adults as painless, solitary, slowly growing, subcutaneous nodules involving the extremities, retroperitoneum and rarely the neck and head. Only 5-10% of hemangiopericytomas affect children and the infantile variety is very rare, usually occurring within the first year of life (Ferrari et al., 2001). Ultrasonographic findings are nonspecific and biopsy with immunohistochemical and ultra-structural studies are necessary for the diagnosis. Macroscopically, the tumors are uncapsulated, well circumscribed nodules. There are numerous vessels with characteristic multilayered concentric arrangement of oval-spindle shaped cells around (Fletcher et al.,

2002). Myopericytomas can be distinguished from other tumors with overlapping features by the fact that cells stain positively for alpha-smooth muscle actin (Dray et al., 2006).

There are few published cases of myopericytomas detected prenatally. The first case described by Hornoy et al. (2005), had an associated cerebral extensions of the neck mass on ultrasonography and magnetic resonance imaging at 33 weeks of gestation. In this prenatally detected case, there was a tumor near the temporal region and though there was no connection between the tumor and brain, the brain in the region showed abnormal gyration on fetal MRI. Another case was reported by Chung et al. (2010), where the fetus had a hemangiopericytoma of the forehead on ultrasonography at 32 weeks of gestation. This case was followed up postnatally for 18 months after complete excision at 7 days of birth. The baby had normal growth and development without any recurrence during the period of observation.

Malignant myopericytoma is very rare and shows high mitotic rate, high cellularity, pleomorphism and necrosis on histopathology (Mc Menamin et al., 2002). Malignant tumors may have an aggressive clinical behavior, with recurrence or metastasis, and complete local excision with continued observation is recommended as treatment (Dray et al., 2006).

To conclude, myopericytoma should be considered in the differential diagnosis of any vascular tumor detected antenatally in the fetus. It is especially so in earlier gestation, when a decision to either terminate or continue the pregnancy has to be made. It is important to rule out associated malformations by fetal MRI and utilize color Doppler to look for extension of tumor into the brain. Regular fetal echocardiography is useful for monitoring the fetus for heart failure. This benign tumor is reported to have good prognosis after postnatal surgical excision.

Key messages

Prenatally detected vascular tumors of the neck could be benign tumors. Further management plan needs to be carefully devised to optimize the outcome for the fetus.

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