Case Report

DOI: 10.7860/NJLM/2017/24793:2223

Pathology Section

A Rare Case of Giant Placental Chorangioma with Adverse Fetal Outcome : A Case Report with Review of Literature

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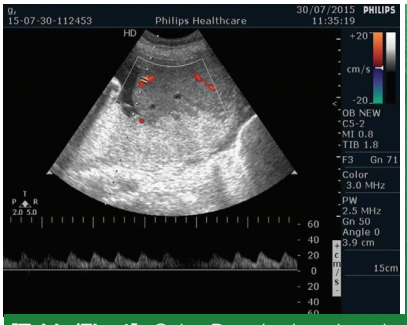
ABSTRACT

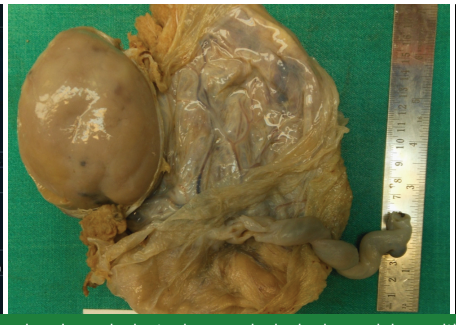
Chorangiomas are a rare benign non-trophoblastic neoplasms of the placenta. Small sized chorangiomas often go undetected with uneventful pregnancy. However large sized chorangiomas, which if undiagnosed antenatally can cause adverse fetomaternal effects. We present a case of giant chorangioma which was undetected antenatally resulting in acute hydramnios and fetal hydrops leading to preterm labour and causing neonatal death. A review of literature is done with emphasis on use of colour Doppler ultrasound as a diagnostic modality and a conclusion that timely diagnosis, active surgical intervention could have prevented this fatal outcome.

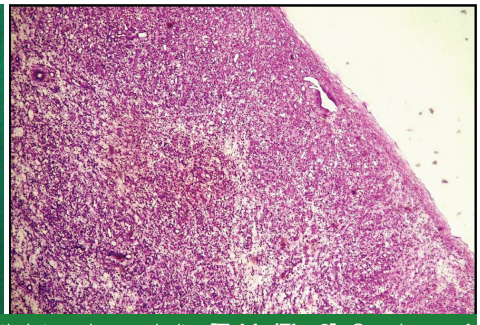
Keywords: Antenatal, Benign, Non-trophoblastic

CASE REPORT

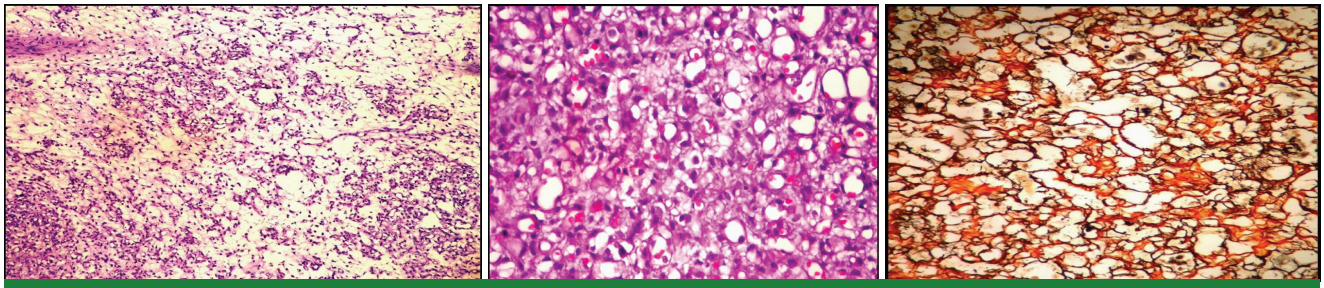
A 34-year-old 2nd gravida with one living child presented with breathlessness and pain abdomen in her 26th week of gestation. On examination, her blood pressure was 110/70 mmHg and abdomen distention was slightly above the umbilicus. Fetal heart sounds were present on auscultation. PV examination revealed a closed Os. Her hemoglobin was 6 gm%. Ultrasound revealed a single intrauterine live fetus of approximately 29 weeks growth with enlarged cardia and minimal pericardial effusion. Rest of her organs were normal. Placenta was posteriorly implanted with an iso to hypoechoic lesion measuring 8x8.6 cm. Color Doppler ultrasound showed internal vascularity with arterial waveforms indicating a vascular lesion [Table/Fig-1]. Amniotic fluid index was 25 cm indicating moderate to gross polyhydramnios (Normal AFI ranges from 8-20 cm). Clinically, a diagnosis of acute hydramnios with placental chorangioma was made. A differential diagnosis of placental hematoma was also considered. An ultrasound guided amniocentesis was performed and 100 ml of amniotic fluid was drained. However, the patient's condition deteriorated. After obtaining consent for surgery, an emergency LSCS was done by which the patient delivered a live baby. The baby did not cry immediately after birth and was transferred to the NICU where the rescusitation efforts failed and the baby died. We recieved the placenta for histopathological study. It weighed 280 gm with a trivascular 20 cm long cord. Fetal surface revealed a lobular well circumscribed fleshy mass measuring 8x7x7 cm [Table/Fig-2]. Microscopy revealed a complex network of proliferating capillaries lined by plump endothelial cells [Table/Fig-3,4]. Reticulin stain showed a loose lattice of reticulin fibres [Table/Fig-5,6]. A final diagnosis of giant







[Table/Fig-1]: Color Doppler imaging showing Isoechoic to hypoechoic lesion arising with internal vascularity. **[Table/Fig-2]:** Specimen of placenta with attached umbilical cord weighing 280 gm. Attached umbilical cord measured 20 cm, inserted slightly eccentrically and was trivascular. A large globular soft tissue mass is attached to the placenta. **[Table/Fig-3]:** A well circumscribed vascular tumour (H&E 40X).



[Table/Fig-4]: Focal areas with myxoid change (H&E 100X). **[Table/Fig-5]:** Tumour composed of capillary sized blood vessels (H&E 400X). **[Table/Fig-6]:** Reticulin stain highlighting the blood vessels.

chorangioma was given on basis of positivity for reticulin stain and radiological correlation.

The patient recovered well and was free of symptoms on post surgery follow-up.

DISCUSSION

Placental chorangiomas are benign, primary non-trophoblastic placental vascular tumours which originally was described by Clarke in 1798 .These lesions were previously classified as hamartomas rather than true tumours resulting from abnormal angiogenesis from primary stem villi. Specific genetic alterations have been detected in these lesions indicating a neoplastic origin [1]. True incidence of this tumour is unknown as most of them are small, clinically insignificant and may be missed during obstetric screening procedures.

Fox accounted for 344 published cases and gave incidence figures of 1 in 9000 to 1 in 50000 placentas [2]. However, the prevalence is as high as 1 in 100 pregnancies. Chorangiomas are common in pregnancies associated with female foetus and in primigravida. They are associated with increased maternal age, diabetes mellitus and hypertension. Many studies have shown a higher incidence of chorangiomas in pregnant women living in higher altitudes [3,4] and that it is the prevailing partial pressure of oxygen rather than hypoxia per se that plays a role in regulating capillary proliferation. Hyper expressivity of various angiogenesis factors like angiopoietin 1 and 2 with their receptors have been demonstrated by Guschmann M et al., [4].

Chorangiomas present as solitary, nodular expansile growth bulging on the fetal surface at the placental margin, 55% of them are located in subchorionic regions. Several studies have revealed the origin of the chorangiomas from the primitive angioblastic tissue of the placenta within a single primary stem villous stem [5].

Macroscopically size may vary from few millimetres to several centimetres. In one of the case series, a size range between 0.5 to 23 cm was reported [6]. Tumours larger than 5 cm are known as giant chorangiomas. In our case the placenta measured 8x7x7 cm. Although, voluminous chorangiomas with successful outcome of pregnancy have been previously reported [7] most commonly these tumours are associated with fetomaternal complications like placentomegaly, hydramnios, pre-eclampsia, preterm labour, fetal hydrops, fetal cardiomegaly, fetal high output cardiac failure, thrombocytopenia ,fetal growth restriction, anaemia and non-immune hydrops [2,8-10]. Fetal hydrops may be due to varied causes like compression of umbilical vessels by the tumour, increased secretion of fetal metabolites through the tumour or may be due to transudation of the fluid from the tumour [10].

Microscopically capillary, cavernous, endotheliomatous, fibrosing and fibromatous tumours have been described by Marchetti [11]. However, the clinical outcome depends upon the size, vascularity and location of chorangiomas . Angiomatous pattern is the most common and is composed of numerous small endothelial lined capillaries surrounded by trophoblastic tissue. Fibroblasts, macrophages and collagen make up the perivascular stroma. Mitotic figure is less than 3 mitosis/10 hpf. Mitosis more than 7/10 hpf with association of cytologic atypia and necrosis are termed atypical chorangioma. Mitotically, active lesions with nuclear atypia raise the suspicion of a malignancy. However, invasion and metastasis were never demonstrated in these tumours. Chorangioma admixed with leiomyoma elements has been reported by Miliaras et al., for which the term suggested was chorangiolieomyoma [12]. Secondary changes that can be seen are infarction, hyalinization, myxoid change and calcification. In our case angiomatous pattern with focal myxoid degeneration was seen without any atypia or mitosis.

Immunohistochemistry shows positive staining with vimentin and alpha smooth muscle actin along with focal staining of Desmin and Cytokeratin 18 [6]. Ultrastructural features of chorangiomas are very similar to hemangioma. Endothelial nature of the lesion can be detected by electron microscopy and with positive immunostaining for factor VIII and CD34. Hormonal stimulation may have a role in its growth and clinical behaviour as demonstrated by joint positivity to estrogen and progesterone receptors.

The importance of imaging cannot be refuted. Gray scale findings such as intraplacental subchorionic location, well defined circumscription, complex echogenecity different from the rest of the placenta single or multiple tumours and protrusion into the amniotic cavity near the insertion of the umbilical cord are helpful in the diagnosis of chorangioma

[13]. Ultrasonographically, intraplacental or subchorionic hematoma, partial mole, teratoma or metastasis falls under the differential diagnosis of chorangioma [14].

The 3D doppler ultrasound can accurately diagnose these conditions and can also detect altered hemodynamics. Color Doppler sonography may show prominent flow in numerous small to medium sized vessels with hypo to anechoic spaces, this flow being continuous with that of the umbilical cord. Fibrosis, vascular septa or acute haemorrhage within the lesion is represented by hyperechoic regions.

Chorangioma should be differentiated from chorangiosis and chorangiomatosis. In chorangiosis, terminal villi are involved with involvement of minimum of 10 villi, each with 10 or more vascular channels [15]. In chorangiomatosis the lesion permeates the normal villi instead of an expansile lesion [16].

Treatment options like serial fetal transfusions, fetoscopic lazer coagulation of the vessels supplying the tumour, chemosclerosis with absolute alcohol, antenatal embolization using percutaneous glubran 2 injection and endoscopic surgical devascularisation. Therapeutic amniocentesis and maternal endomethacin therapy with steroid administration for acceleration of fetal lung maturity before 34 weeks is indicated.

CONCLUSION

Although, chorangiomas are benign in nature, large sized lesions can lead to a variety of fetomaternal complications. Inspite of our hospital being a tertiary centre, the lesion was undetected antenatally. The tumour has been detected by ultrasound and Doppler, close monitoring and timely intervention could have averted the fatal fetal outcome. Thus, there is a need for awareness of this condition and this case report also emphasizes on detecting, monitoring and active surgical intervention.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Publishing: **Apr 01, 2017**