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Case Report

Low Lying Giant Chorangioma with Myometrial Attachment: A Rare Histopathological Finding

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Abstract

Chorangioma is a placental lesion consisting of capillary proliferation in chorionic villi and typically presents as a nodule or as multiple nodules on the placenta. It is a rare abnormality of placental vascular development and its clinical characteristics depend on size of the lesion. Giant chorangioma is chorangioma with an average diameter greater than 5cm. It is associated with both maternal and foetal complications. This report documents our findings in a hysterectomy specimen removed from a 40-year old female, following emergency hysterectomy for intractable postpartum haemorrhage after an elective caesarean section for three previous sections. A giant chorangioma was discovered in association with both placenta praevia and placenta increta. The outcomes for both mother and child were good.

Keywords: Giant Chorangioma, placental vascular development, haemorrhage, histological examination.

INTRODUCTION

Chorangioma is an expansile nodular lesion composed of capillary vascular channels, intervening stroma cells and surrounding trophoblast [1]. Placental chorangioma is the most common neoplasm of the placenta with an incidence of approximately 1% of all pregnancies [2]. It presents as a solitary nodule or less frequently, as multiple nodules in the foetal surface of the placenta or within the placental parenchyma [3].

The clinical significance of chorangioma is size dependent. Small chorangiomas (<5cm in diameter) are usually asymptomatic and are either not diagnosed or incidentally found at a histological examination or screening obstetric ultrasound examinations [4]. Giant chorangiomas (>5cm in diameter), are rarely seen in obstetric practice, occurring in approximately 1 in 10000 pregnancies [5] and may be associated with hydramnios, haemorrhage, premature delivery, premature placental separation and placenta praevia [6]. Foetal complications arising from giant chorangioma may include non-immune foetal hydrops, heart failure, anaemia, thrombocytopenia, intrauterine growth restriction and death [7].

This report presents an incidental low lying giant chorangioma partly invading the myometrium, in a hysterectomy specimen that followed an elective caesarean section and subsequent hysterectomy for intractable postpartum haemorrhage, with good maternal and neonatal outcomes.

CASE REPORT

Patient is a 40-year old woman, Para 4 (3 Alive) who had an elective repeat caesarean section at a gestational age of 37 weeks, on account of three previous caesarean section deliveries, in May 2021. The antenatal period was uneventful and there were no maternal co-morbidities. A prenatal ultrasound done at 24 weeks showed a posterior, low lying placenta, however there was no warning bleeding or antepartum haemorrhage in the course of the pregnancy and a repeat scan done at 36 weeks showed a fundal posterior placenta. The possibility of a morbidly adherent placenta was entertained due to the three previous caesarean sections. Intraoperatively, Type 2b placenta praevia was identified. A live female baby weighing 2.6kg was delivered, with APGAR scores of 8 at 1 minute and 9 at 5 minutes. The placenta was morbidly adherent to the uterine wall and it was manually removed as much as possible following failed attempts at delivery by controlled cord traction. The portions of placenta removed weighed 350gms.

During placental removal, an intractable postpartum haemorrhage ensued, with a total estimated blood loss of 3 litres and haemodynamic instability (blood pressure dropped to 55/17mmHg and pulse rate went up to 144 bpm intraoperatively). A decision to perform an emergency caesarean hysterectomy was made due to the massive obstetric haemorrhage which could not be controlled. Prior to the surgery, the patient had consented to bilateral tubal ligation for completed family and had given consent for hysterectomy should the need arise. She received 4 units of whole blood intraoperatively. The hysterectomy specimen was subsequently sent for histopathological analysis. Additional 2 units of blood were transfused post operatively. Her post-operative period was uneventful and she was discharged home on the sixth post-operative day in good clinical state. Post-discharge follow-up at 6 weeks indicated that both mother and baby were in good clinical condition.

Histopathology

i. Gross:

A hysterectomy specimen measuring 19cm × 17cm × 10cm and fixed in 10% neutral buffered formalin solution was received. It weighed 1400g. The cervix was deviated to a lateral angulated position, subtending an angle of about 90 degrees to the long axis of the uterus, due to inappropriate containerization and fixation. Other external features of the uterus were unremarkable.

Cut section of the uterus revealed a tan, fleshy mass located in the posterior wall, just above but unattached to the internal os and measuring 7cm × 5cm. It was largely smooth and appeared fairly well circumscribed except for an isolated portion, where it extended 1.5cm into the myometrium, just one millimeter short of the serosal surface. The endometrial cavity above the mass was haemorrhagic. The cervical canal was unremarkable (Figures 1 & 2).

ii. Microscopy

Sections of the circumscribed mass within the hysterectomy tissue showed numerous chorionic villi of varying sizes and shapes. Within the clustered villi were areas of numerous capillary vascular channels, scanty intervening stroma and surrounding trophoblastic epithelia. These capillaries were lined by unremarkable endothelial cells. The trophoblastic epithelial cells were also unremarkable. There were areas of invasion of the myometrial wall by the chorionic villi and free trophoblastic epithelial cells which ranged in depth from 3mm to 5mm (Figures 3 & 4).

Figure 1: Sagittal section of incompletely fixed gross specimen showing the tan mass

Figure 2: Coronal section of completely fixed gross specimen showing myometrial invasion by the mass

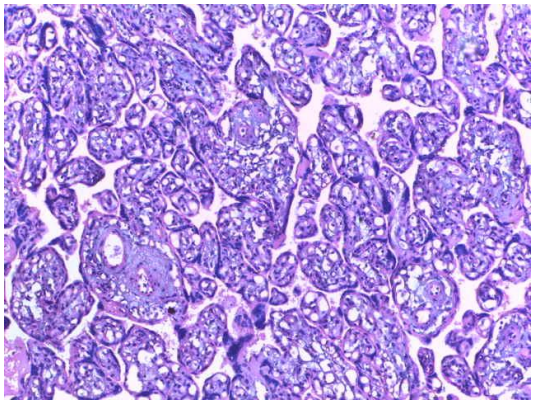

Figure 3: Micrograph showing numerous capillary vessels in the mass (×200)

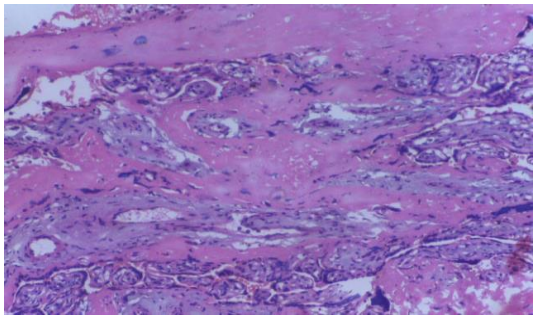

Figure 4: Micrograph showing myometrial invasion by the mass (×200)

DISCUSSION

Chorangioma of the placenta is a benign vascular tumour arising from the primitive chorionic mesenchyme whose aetiology is unknown [2]. It is often associated with metabolic diseases such as hypertension and diabetes mellitus, increased maternal age (above 30 years old), female babies and multiple pregnancies [4]. The index patient was 40 years and carried a female baby. However, there were no documented maternal co-morbidities like systemic hypertension. The facts that she had had three previous caesarean sections and had placenta praevia in the index pregnancy put her at risk of having placenta accreta spectrum.

The pathogenesis of chorangioma is not too clear; some authors attribute its development to pathological placental angiogenesis. Vascular Endothelial Growth Factor (VEGF) as well as Neutral Endopeptidase (NEP) and Receptor Tyrosine Kinase subclass III (KIT) play an important role in placental angiogenesis and the pathogenesis of placental chorangioma. Studies have shown that VEGF, NEP and KIT are expressed in both myofibroblasts of neonatal cutaneous haemangiomas and in placental microcirculation, suggesting a common origin [8].

Chorangiomas are thought to arise in hypoperfused areas, probably due to a hypoxic state, mediated by vascular growth factors, which cause an active proliferation of connective tissue and growth of villous capillaries. The occurrence of chorangioma has also been linked to pre-eclampsia, high altitude pregnancy and foetal anaemia, suggesting that decreased oxygen tension may play an important role in the pathogenesis [1]. These risk factors were absent in our patient and her child.

Antenatal diagnosis is possible if the size of the tumour exceeds 2cm [9]; however, only 50% are detected prenatally [10]. Ultrasound features make it possible to make the diagnosis in utero, but the confirmation of the diagnosis is histological examination of the placenta after birth. Ultrasonographic diagnosis of placental chorangioma can be done by its two-dimensional ultrasound and doppler characteristics. On ultrasound, placental chorangiomas are identified as hypoechoic, rounded mass, located near the chorionic plate, at times close to the umbilical cord insertion [11]. It contains anechoic 'cystic' lesion distinctly separate from the normal placental tissue [11]. Degenerative processes within the mass may appear as heterogenous areas with internal haemorrhage [12]. A large tumour is usually attached to the chorion which is a less perfused area. A few cases could occur in the maternal surface, replacing the whole or some part of the placental lobe [13]. Doppler velocimetry often shows a low resistance pulsatile flow within the anechoic 'cystic' areas representing enlarged vascular channels [12]. In this case, ultrasound only identified presence of a low-lying placenta but missed the lesion. Also, from the location of the mass and insertion of a part of it into the myometrium, this case is likely one of the few where chorangioma is found on the maternal surface of the placenta.

On gross examination, chorangioma is well-circumscribed, devoid of a fibrous capsule, sharply demarcated from the surrounding placental parenchyma by a single or double layer of chorionic epithelium and tends to have fleshy, congested, red to tan cut surface [3]. The lesion in this case is unique because a part of it was attached to the myometrium and there was a rough surface which represents its point of detachment from the other parts of the placenta. However, the free parts of the mass conformed to the classical circumscribed description of chorangioma.

On microscopy, typical chorangioma is composed of proliferation of foetal blood vessels, usually supported by scant connective tissue and covered by trophoblast. Villous expansion is caused by proliferation of blood vessels which is diagnostic. Capillary, cavernous, endotheliomatous, fibrosing and fibromatous variants of chorangioma have been differentiated, but such precision is unwarranted as the clinical outcome depends more on the size of the tumour than on its composition [14]. The morphology of the tumour under consideration is of capillary type. Occasionally, chorangiomas are associated with infarction and degenerative changes such as hyalinization, necrosis, myxoid stromal changes or calcifications. Neither infarction nor degenerative changes were seen in this case.

Immunohistochemically, tumour cells usually show focal staining of Cytokeratin [15]. They also express other vascular markers such as CD31, CD34, factor VIII and GLUT 1 [2]. Immunohistochemistry was not carried out in the index case because of non-availability of appropriate reagents.

The complications of giant chorangiomas are well documented [6, 7]. Our patient had placenta increta, placenta praevia and intractable postpartum

haemorrhage which led to shock and ultimately, emergency caesarean hysterectomy. However, no foetal complications were noted.

On ultrasound, placental chorangioma may pose a differential diagnoses problem with chorionic thrombosis, plaque cyst, deciduous haematoma, placental teratoma, aneurysmal dilatation of a vessel under chorion, placental infarction, partial hydatiform mole, leiomyoma and metastasis. Colour doppler sonography is helpful in differentiating chorangioma from the other entities. It is possible to detect placenta accreta spectrum antenatally using colour flow Doppler ultrasound scan or Magnetic Resonance Imaging if the ultrasound scan is inconclusive. If the diagnosis of placenta accreta spectrum was made prenatally in the index patient, an attempt to deliver the placenta would not have been made as an outright hysterectomy would have been done after the delivery of the baby.

On histology, the differential diagnosis of chorangioma are atypical chorangioma, Chorangiosis and Chorangiomatosis. Atypical chorangioma has similar features as chorangioma, plus increased cellularity, mitotic activity and areas of necrosis. Chorangiosis is capillary hyperplasia with more than 10 capillaries in at least 10 terminal villi (normal villi usually only have 5 capillaries/ villous) [2]. However, there are no distinct gross abnormalities in chorangiosis as in chorangioma that appear as distinct circumscribed mass. Chorangiomatosis is a multiple lesion with capillary proliferation permeating the normal villus structure [2]. Pericytes are normal cellular constituents of vascular tumours such as chorangiomas and haemangiomas that have pro-angiogenic nature.

CONCLUSION

Giant chorangioma is a very rare occurrence and may be associated with a number of complications. The foetus may be normal even when there are maternal complications. Prenatal diagnosis of giant chorangiomas is possible, but can be missed as happened in this case. A thorough prenatal ultrasound examination of the placenta can help identify potential life-threatening complications of placental pathologies which can be prepared for pre-operatively, and thus reduce the risk of incidental findings as happened in the case presented.

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