An Unusual Case of Fetal Distress: Spontaneously Ruptured Maternal Renal Angiomyolipoma

SPONTAN RUPTURE OLMUŞ MATERNAL RENAL ANGİOMYOLİPOM NEDENİ İLE OLUŞAN ALIŞAGELMEDİK BİR FETAL DİSTRES VAKASI

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Özet

Olgu sunumu: G3,P1,A0, D&C1, 28 yaşında bir gebede , 6 santimetre açıklığa kadar problemsiz ilerleyen travayda , aniden gelişen fetal distress ve maternal hipovolemi belirtileri nedeni ile sezaryen ile doğum yaptırıldığında; fetal distress ve maternal hipovoleminin nedeni, spontan olarak rüptüre olmuş renal bir angiomyolipoma olarak tespit edildi.

Sonuç: İngilizce literatürde yaptığımız taramada ; gebelik, travay ve erken puerperiumda rüptüre olmuş renal angiomyolipom vakalarının sunulduğu 14 makale ve toplam 24 vaka tespit edebildik. Retroperitona olabilecek bir kanamanın fetal distrese yol açabileceğine dikkat çekmek istedik.

Anahtar Kelimeler: Angiomyolipoma, Gebelik

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Summary

Case report: A labouring patient went into fetal distress at 6 cm. cervical dilatation. An immediate caesarean section was performed, and the cause of the fetal distress and hypovolemia was found to be a retroperitoneal haemorrhage due to a ruptured renal angiomyolipoma.

Conclusion: Ruptured angiomyolipoma during pregnancy, labour and puerperium is extremely rare. In the literature we could have detected 14 papers presenting 24 cases of spontaneous rupture of renal angiomyolipoma in pregnancy, labour and puerperium. We want to draw attention to a very unusual cause of fetal distress during labour.

Key Words: Angiomyolipoma, Pregnancy

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Angiomyolipoma is a rare renal neoplasm, composed of an intimate mixture of fat, blood vessels and smooth muscle. Renal angiomyolipoma may cause massive and sometimes fatal haemorrhage. Acute abdomen and fetal distress during pregnancy, secondary to retroperitoneal haemorrhage due to rupture of renal angiomyolipoma is very rare. In the literature we could have spotted only 24 cases of spontaneous rupture of renal angiomyolipoma during pregnancy, labour or in the immediate puerperium.

Here we present another case of angiomyolipoma, which ruptured in a labouring patient and caused fetal distress, resulting in an emergency caesarean delivery.

Case Report

Our patient, a G3, P1, A0, D&C1, 28 year old, white woman at 39 th gestational week was in labour with a vertex presentation and the labour progressed to a cervical dilatation of 6 centimetres uneventfully. At this stage of labour, the patient complained of a left flank pain and became restless and signs of hypovolemia appeared. Pulse was 120 beats/min and arterial blood pressure was 100/80mm Hg. Simultaneously severe fetal heart rate decelerations were recorded at the external fetal heart rate monitor. Upon pelvic examination there was no sign of bleeding. Fearing a rupture of the uterus, the patient was immediately transferred to the operating theatre for an emergency caesarean section. A 3000 g male infant with an Apgar score of 6 at the first minute was delivered abdominally with a midline skin incision and a lower segment uterine incision. After the delivery of the infant, the uterus was examined and there was no sign of uterine rupture. Examination of pelvic and abdominal contents revealed an unusual retroperitoneal mass on the left side of the patient, above the pelvic brim. The mass was 20 centimetres in diameter and bluish purple in colour and was crossing to the right side of the columna vertebralis for 5 centimetres and was non-pulsating. Immediate consultations were arranged with general surgeons and urologic surgeons. An entry was made to the retroperitoneal space trough the left lateral border of the mass and a haematoma of 1250 cc was aspirated.

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Within the haematoma were tissue fragments of dark yellow colour and of soft consistency with diameters of up to 1,5 cm. After the evacuation of haematoma and those tissue fragments, exploration progressed cephalad and towards midline and the ruptured lower pole of the left kidney was visualised. Urologic surgeon assessed the damage to the left kidney to be unrepairable and performed an immediate left nephrectomy and ureterectomy. Drains were left in the retroperitoneal space and the abdominal incision was closed in the anatomic plane. During the operation 2000 cc of whole blood was transfused and the postoperative course of the patient was uneventful and the patient was discharged on the seventh postoperative day. Pathologic diagnosis of the operative specimen was reported to be an angiomyolipoma

Discussion

Angiomyolipoma of the kidney is a rare neoplasm and these tumours are classified as hamartomas (tumour-like malformations in which there is only abnormal mixing of the normal components of the organ in which they occur). An hamartoma of renal origin has been referred to by several terms in the literature including benign arterioleiomyoma, angiolipoleiomyoma, benign mesenchymoma, lipohemangioma, myoangiolipoma and most commonly angiomyolipoma (1).

Angiomyolipoma share more than a casual relationship with tuberous sclerosis. Some series report about an 80% incidence of angiomyolipoma in patients with tuberous sclerosis (4). When occurring with this entity the tumour is usually bilateral and multiple. When occurring in the absence of the tuberous sclerosis complex, angiomyolipomas are usually unilateral, occur most frequently in adults and predominantly in women (2). Our patient had no sign of tuberous sclerosis.

The gross appearance varies depending on the predominant tissue type (smooth muscle, blood vessels and fat). Microscopically the vascular component is composed of tortuous, thick walled blood vessels that frequently lack an elastic lamina. The adipose tissue is of mature type. The smooth muscle component is troublesome because it may exhibit hypercellularity, marked pleomorphism and moderate mitotic activity. These features may prompt a mistaken diagnosis of leiomyosarcoma (1).

Renal angiomyolipoma resulting in spontaneous rupture during pregnancy and the immediate puerperium is very rare. We could have spotted only 14 papers presenting 24 cases of spontaneous rupture of renal angiomyolipoma during pregnancy, labour or puerperium (1-14). The largest series belongs to E.L. Lewis et al., they published their three cases with tuberous sclerosis in 1985 (7).

In the literature there is a report of hepatic angiomyolipoma diagnosed in the pregnancy(15). And there is a report of successful selective embolization of bleeding renal angiomyolipoma (16).

We believe that our case is unique because all the other cases in the literature were admitted to the hospital for indications other then active labour. The most common indications for hospitalisation were flank pain or shock.

Whether the increased blood volume during pregnancy might have some relationship with the spontaneous rupture of these renal tumours, can only be a matter of conjecture at this point. During pregnancy and especially when the patient is in active labour, the differential diagnosis of an acute abdomen with internal bleeding is a challenge for the physician in charge. In those cases obstetric complications such as placenta previa, abruptio placenta and even uterine rupture must be ruled out first (2). The causes of retroplacental haemorrhage in pregnancy, in addition to angiomyolipoma, are rupture of renal aneurisms, spontaneous rupture of hydronephrotic kidneys, rupture of renal vessels and other less common vascular accidents.

Fortunately, massive retroperitoneal haemorrhage during pregnancy is very rare. The case reported above illustrates the importance of close monitorization of the labouring patients and life saving effect of prompt surgical intervention.

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