Case Report. Rupture of Angiomyolipoma of the Kidney Presenting as Puerperal Collapse

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SUMMARY: We report an unusual presentation of angiomyolipoma of the kidney. Whilst a conservative surgical approach is ideal, the greatly increased cardiac output in pregnancy makes haemorrhage more severe and hence more radical surgery is often required. The difficulty in reaching a correct pre-operative diagnosis can lead to an unusual approach to nephrectomy, of which the surgeon should be forewarned.

Case Report

Our patient, a 39 year old para 2, gravida 9, went into spontaneous labour at term after an uneventful pregnancy. After a 10 hour labour augmented with oxytocin she delivered a live male infant. Placental separation did not occur and the placenta was removed manually under general anaesthesia without any problem. The blood loss was estimated as 400 mls.

The haemoglobin had fallen from 10.4g% pre-delivery to 6.3g% on the third day and, as she felt tired and was breast feeding, four units of packed cells were transfused, following which the haemoglobin rose to 10.7g%.

Later, on the fifth day, she suddenly developed severe right loin pain which was marginally relieved by flexing her hips. A presumptive diagnosis of renal colic was made and, after intramuscular pethidine had been given, emergency intravenous urography was performed. The control film showed loss of the right psoas shadow. After injection of contrast material there was a prompt left nephrogram with a normal left ureter and bladder. No nephrogram appeared on the right and therefore an ultrasound examination was performed which confirmed the presence of a right kidney but showed it to have an indistinct outline. No calculi were seen.

Over the next two hours she developed signs of hypovolaemia, with peripheral circulatory shutdown and tachycardia. Her urine output remained over 30mls/hour without haematuria. She also developed hyperaesthesia over the right side of her back at the level of

the lower thoracic vertebrae, right shoulder-tip pain and increasing abdominal pain and distension with absence of bowel sounds.

The diagnosis was revised to retroperitoneal haematoma (from torn uterine vessels) with continuing bleeding and, after resuscitation, laparotomy was performed through a midline incision. Nine hundred millilitres of blood were aspirated from the peritoneal cavity, after which a large right retro-peritoneal haematoma was seen extending from the base of the broad ligament to above the right kidney. The uterus, broad ligaments and ovaries were all normal so the peritoneum overlying the haematoma was opened and the clot evacuated. The source of the bleeding was traced upwards to the mass of clot surrounding the right kidney. As it was impossible to identify the origin precisely, a right nephrectomy was performed. The upper pole of the kidney was found to have ruptured. No evidence of calculus or obstruction was found in the distal ureter.

Following the operation, during which she received 10 units of blood, she made a rapid and uneventful recovery. She was discharged home 10 days later, and has remained well.

Pathology

The kidney weighed 220g. In the upper pole there was a tumour some 3cm in diameter (Fig. 1). The tumour was yellow in colour with areas of haemorrhage. It was sharply demarcated from the renal parenchyma and there was a ragged rupture through the renal capsule.

Microscopic examination revealed normal fat together with numerous atypical blood vessels (some of which were surrounded by recent haemorrhage) and much tissue composed of spindle shaped cells with bizarre nuclei. The blood vessels were thick walled, with thickened intima devoid of elastica, a basophilic background and a few irregularly placed nuclei. The muscle coat often disappeared into the surrounding cellular tissue composed of ill-formed smooth muscle cells. In places these cells formed a continuous, irregularly arranged network; elsewhere a plexiform mass of bundles of muscle fibres was seen. A small nodule in a renal papilla contained fibroblasts without atypical blood vessels or fatty tissue. The appearances were typical of an angiomyolipoma (a hamartoma).

Discussion

Spontaneous rupture of a renal angiomyolipoma during pregnancy is an extremely rare event. It is difficult to diagnose because carrying out the full range of diagnostic procedures may expose the fetus to unacceptable levels of radiation. The majority of reported cases have occurred during pregnancy and only two have been reported in the puerperium.

The term 'hamartoma' was first used by Albrecht (1904) to describe a tumour-like malformation. Since then hamartomata of renal origin reported include benign arterioleiomyoma, angiolipoleiomyoma, benign mesenchymoma, lipomyohaemangioma, myoangiolioma, and, most commonly, angiomyolipoma¹. Such lesions are usually unilateral, and present most frequently in adults and mainly in women^{2,3}. Angiomyolipomata, often when multiple, are associated with tuberous sclerosis, and this may be the sole clinical

manifestation; this patient, however, had no stigmata of tuberous sclerosis.

Ballonius⁴ first reported spontaneous circumrenal haemorrhage in 1616. Post-partum rupture of a renal angiomyolipoma was first reported by Rusche⁵ and only one further case was found in the literature⁶. This contrasts with the much larger number of cases reported during pregnancy⁷.

Parenchymal rupture is more common than rupture into the renal pelvis, and the majority are acute. The final stimulus to rupture is often a sudden increase in intra-abdominal or intra-renal pressure^{8,9} such as that occurring in labour. Minor trauma has also been reported to cause rupture¹⁰⁻¹². In our case, there may well have been a slow initial haemorrhage following delivery, with a later acute bleed precipitating the eventual clinical presentation. It is unlikely, however, that the manual removal of the placenta can be blamed for the rupture.

The three main signs of spontaneous rupture of the kidney are pain (almost invariably present), cardiovascular collapse due to haemorrhage, and the appearance of a palpable tender mass in the loin. Haematuria occurs only when the bleeding site is within the renal pelvis, or if blood tracks into the pelvis from the site of the rupture.

The diagnosis may be confirmed by intravenous urography and sometimes by retrograde pyelography. Ultrasound can now provide a more accurate picture and aid the diagnosis of renal hamartoma.

Prompt exploration is mandatory to control the haemorrhage, which may otherwise prove fatal¹³. The best treatment for significant rupture of the kidney is nephrectomy, although where the rupture is small, the cause known to be non-neoplastic and conditions suitable, an attempt may be made to repair it as advocated by Chung et al¹⁴. The greatly increased renal blood flow in pregnancy¹⁵ and the puerperium will, however, usually preclude this approach.

This rare and life threatening complication of pregnancy may present considerable diagnostic difficulties, but careful clinical observation and appropriate investigations will demonstrate the need for laparotomy. Transabdominal nephrectomy is not an operation for the tyro, and early consultation between obstetricians and surgeons is advised so that operation may be planned as a joint procedure from the outset.

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Fig 1. Tumour erupting through upper pole of kidney.

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