An unusual presentation of Wunderlich syndrome

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Abstract

Spontaneous renal haemorrhage secondary to renal angiomyolipoma, or Wunderlich syndrome, is rare. Renal angiomyolipomata are composed of abnormal vasculature, smooth muscle and adipose tissue. They are more likely to be symptomatic if they are larger than 4 cm, presenting with a history of flank pain, a palpable mass and rarely gross haematuria. Our case illustrates a rare but catastrophic presentation of Wunderlich syndrome. A 29-year-old woman, with no significant past medical history, presented with sudden onset right-sided abdominal pain. The patient appeared pale and revealed localised peritonitis in the right iliac fossa. A diagnosis of acute appendicitis was made and expeditious laparoscopy was performed. At surgery a large right-sided retroperitoneal haematoma extending to the perinephric area was identified. Due to continuing significant haemorrhage an emergency radical nephrectomy was necessary. The patient made an excellent recovery and the histology confirmed large renal angiomyolipoma as the underlying pathology. This is the first case report of Wunderlich syndrome in a previously asymptomatic fit young patient. This novel presentation of Wunderlich syndrome highlights the diagnostic difficulties when faced with the acute abdomen. This report aims to equip clinicians with knowledge and heightened awareness of this rare condition to optimise patient care.

Keywords

Angiomyolipoma; Wunderlich syndrome; retroperitoneal haemorrhage.

Introduction

Spontaneous renal haemorrhage, or Wunderlich syndrome, is a rare cause of acute abdominal pain. This case report describes Wunderlich syndrome secondary to renal angiomyolipoma in a previously fit, young and healthy patient. Angiomyolipomata are uncommon benign lesions that consist of fat, blood vessels and smooth muscle with a prevalence of 0.3–3%. Significant retroperitoneal haemorrhage may occur in approximately 10% of patients with angiomyolipoma, and may be catastrophic[1].

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Case history

A normally healthy 29-year-old woman, with no significant past medical history, presented acutely with right-sided abdominal pain. She described being awoken from sleep with right-sided abdominal pain that radiated to her back and groin. The pain was colicky in nature, and was associated with nausea and anorexia. She reported no urinary or bowel symptoms and denied any injury prior to the pain.

On examination, she appeared pale but was apyrexial with normal vital signs: pulse 80, blood pressure 120/80 and oxygen saturations 97% on room air. Abdominal examination revealed localised peritonism in the lower right quadrant. Urine dipstick was negative. Blood investigations revealed a leucocytosis of 16.5 and a normocytic anaemia 11.8 g/dl. Her C-reactive protein, renal function, liver function and clotting tests were all within normal limits and a β-human chorionic gonadotropin was negative.

A diagnosis of acute appendicitis was made and no further imaging was felt to be necessary. At laparoscopy a large retroperitoneal haematoma, extending from the right pelvis to the perinephric area was identified. It was soft in the pelvis but much firmer in the perinephric area. The pelvic organs were normal and no obvious cause could be identified. The patient became haemodynamically unstable and a laparotomy was crucial. The tense perinephric haematoma, with associated free haemorrhage, was confirmed. The left kidney was normal on palpation and the rest of the exploration revealed no other abnormalities. The whole of the right kidney was macroscopically abnormal and distorted with haematoma, resulting in radical nephrectomy. The patient was admitted to the high dependency unit but overall made an uneventful post-operative recovery.

Macroscopically, the resected right kidney revealed a large haematoma within the perinephric space and a ruptured tumour at the lower pole. Microscopically, the tumour was shown to contain proportions of thick walled blood vessels intermixed with mature adipose tissue and smooth muscle fibres. Immunohistological HMB-45 confirmed the neoplasm to be a benign angiomyolipoma (Fig. 1).

Discussion

Two subtypes of angiomyolipoma have been described in the literature: isolated angiomyolipoma and angiomyolipomas, which is associated with tuberous sclerosis. Isolated angiomyolipoma occurs sporadically. It is often solitary and accounts for 80% of angiomyolipomas. The mean age at presentation of patients with isolated angiomyolipoma is 43 years; this neoplasm is about four times more common in women and 80% of cases involve the right kidney.

Most (60%) of angiomyolipomas are asymptomatic and are commonly found incidentally during routine imaging\[2\]. In a case series that included patients with tuberous sclerosis, 82% of patients with a tumour >4 cm were symptomatic, whereas only 23% of patients with tumours <4 cm were symptomatic. Symptomatic patients may present with a combination of flank pain (53%), a palpable tender mass (47%) and gross haematuria (23%); this is known as Lenk’s triad\[3\]. Spontaneous retroperitoneal haemorrhage is rare and usually presents with pain, haematuria and shock\[4\].
To the best of our knowledge, this is the first case report of Wunderlich syndrome in a young, fit patient with no significant past medical history or classic symptoms, resulting in an emergency nephrectomy. Cases of Wunderlich syndrome associated with severe hypertension, anticoagulants, underlying malignancy and tuberous sclerosis have been described.

In an elective situation, computed tomography (CT) scanning would be helpful in confirming the diagnosis and may allow selective arterial embolisation to control haemorrhage, avoiding radical nephrectomy[5]. In our case, the patient presented as an emergency with an acute abdomen, and rapid onset of haemodynamic instability. This unusual presentation necessitated expeditious resuscitation and emergency life-saving surgery.

We hope that our case report helps to raise clinicians’ awareness of Wunderlich syndrome. Early use of CT in appropriate circumstances and a multidisciplinary team working with radiological colleagues may allow less invasive methods of treatment.

**Teaching point**

This case report illustrates the importance of considering all the differential diagnoses when managing any patient. Although common diseases are the most likely cause of most presentations, there are rarer conditions that can be life threatening and need emergency intervention. Knowledge and heightened awareness of these conditions will expedite the appropriate adjustments required to patient treatment and therefore improve the likely outcome.

This case provides an excellent example in which diagnostic laparoscopy is useful in guiding the surgeon to the correct pathology. If the appendix had been removed as an open procedure there would have been a critical time delay in revealing the retroperitoneal haemorrhage. Open mindedness, flexibility and a multidisciplinary approach facilitates tailoring of medical management to the specific individual to optimise patient care.

**References**