

WUNDERLICH SYNDROME - A CASE REPORT

Ankur Sharma¹, Goonj Johr²

¹Assistant Professor, Department of General Surgery, IIMS & R, Integral University, Lucknow.

²Assistant Professor, Department of General Surgery, ELMC, Sarfarazganj, Lucknow.

ABSTRACT: Angiomyolipoma is the most common benign renal neoplasm and with increasing size, the chances of complications also arise. Lesions more than 4 cms require close followup or intervention to minimise the risk of complications. Wunderlich syndrome or Spontaneous Renal Haemorrhage is a potentially life threatening condition, which usually presents as an acute emergency. We present a case of a subacute presentation, in which there was spontaneous haemorrhage in the angiomyolipoma. Also, we would like to highlight the failure to adequately diagnose pre-operatively the correct nature of the tumour.

KEYWORDS: Angiomyolipoma, Bleeding, Spontaneous, Subacute.

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INTRODUCTION: Wunderlich syndrome or spontaneous renal haemorrhage is a rare syndrome with around 300 cases described in the literature. It sometimes presents a diagnostic dilemma to the treating surgeons and radiologists alike due to unawareness of the entity and a therapeutic challenge due to acute presentation. The most common causes of these spontaneous haemorrhages are represented by neoplasm.¹ Angiomyolipoma (31.5%) is the most common benign tumour implicated and renal cell carcinoma is the most common malignant tumour.²

CASE REPORT: A 35 years old man presented with a history of dull aching pain and heaviness in left flank of 2 weeks' duration. There was no history of radiation of pain to groin, dysuria or haematuria or fever or any bowel symptoms. There was no history of trauma. He was evaluated with routine blood investigations which did not show any significant abnormality other than low haemoglobin of 9 gm%. Ultrasonography of abdomen revealed a large cystic lesion with internal echoes in left paraspinal area about 9 x 8 cms in size. Left kidney was displaced laterally. Contrast enhanced CT Abdomen was further done to characterize the lesion. CECT was suggestive of a large heterogeneous predominantly cystic lesion in relation with left kidney.

Since the diagnosis could not be made with certainty, patient was prepared for surgery and on exploration a large tumour was seen in relation to left kidney, around 10x7 cms in antero-inferior location to the kidney and on aspiration around 400 ml altered blood was aspirated and the lesion significantly decreased in size and nephrectomy was done. The biopsy was later reported as Angiomyolipoma.

Retrospectively, a diagnosis of Wunderlich syndrome was made.

DISCUSSION: Angiomyolipoma is a benign mixed mesenchymal tumour that occurs predominantly in the kidney. Angiomyolipomas may also occur, however, in extrarenal sites such as the liver, spleen, abdominal wall, retroperitoneum, uterus, oral cavity, penis, spermatic cord, fallopian tube, vagina, skin, and lung.³ The incidence in the general population is between 0.07 and 0.3%.⁴

Approximately 80% of renal angiomyolipomas occur sporadically and 20% are associated with tuberous sclerosis.⁵ In the sporadic cases, these lesions are to be solitary, larger and unilateral with a female preponderance in the fourth to sixth decade of life. Approximately 64-77% of tumours <4 cms in diameter are asymptomatic, and around 82-90% of angiomyolipoma >4 cms produce symptoms.⁶

Histopathologically, it consists of a collection of thick-walled blood vessels, smooth muscle and mature adipose tissue in varying proportions.

Spontaneous non-traumatic bleeding confined to the subcapsular and/or perinephric space in patients with no known underlying cause was first described as spontaneous renal capsule apoplexy by Carl Reinhold August Wunderlich in 1856.⁷ Presentation of this clinical picture may vary greatly depending on the degree and duration of the bleeding. Wunderlich syndrome is one of the most feared complications of renal angiomyolipoma and can be fatal if not treated promptly and aggressively.

In symptomatic patients of angiomyolipoma, the classic presentation includes flank or abdominal pain, a palpable tender mass and gross haematuria (Lenk's triad). Other less common symptoms include nausea, vomiting, fever, anaemia, renal failure and hypotension. Three types of haemorrhagic aetiology exists including Wunderlich syndrome, bleeding after trauma and rupture during pregnancy. It occurs in up to 50% of patients with tumours larger than 4 cms because of the association with an increased risk of intralesional aneurysm formation and

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Corresponding Author:

Dr. Ankur Sharma, Department of General Surgery, IIMS & R, Integral University, Lucknow.

E-mail: ankurvashishth@gmail.com

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therefore a greater possibility of rupture.⁸ In fact, having abnormal elastin-poor vascular structures, angiomyolipomas are likely to form aneurysms as they grow and as the blood flow entering them increases.

Wunderlich syndrome classically presents with acute flank pain, symptoms of internal bleeding (such as bruising, pallor and signs of shock) and tenderness. Nausea, vomiting, and low-grade fever with decreasing haemoglobin are also seen.^{9,10} It may also be associated with hypotension from a hypovolemic state or a paradoxical hypertension from a condition known as Page kidney. In Page kidney, compression of the kidney by haemorrhage or a mass may result in ischemia, renin release, and renin-dependent hypertension.¹¹

In 1986, Oesterling et al. proposed a treatment protocol based on size and symptoms: Asymptomatic tumours <4 cms should be observed regularly with computerized tomography or ultrasound whereas those >4 cms should be studied by angiography and considered for arterial embolisation or surgery.⁶

Usually, the ultrasonography is the first modality of investigation in nonurgent stable cases. For a patient

suspected to have Wunderlich syndrome, CT scan is preferable. CT scan can give the opportunity to know the entity of the haematoma and the presence of an active bleeding, but however lacks the ability for therapeutic intervention. Selective angiography with embolisation is useful in the acute phase of the haemorrhage in order to control bleeding, contribute to diagnosis and reduce the need for surgery. The management is dictated by the clinical condition of the patient and by the underlying aetiology. Sometimes subjects with unstable haemodynamic condition or when facility for embolisation is not available, an emergency nephrectomy may be required.

CONCLUSION: Wunderlich syndrome can be potentially life threatening and angiomyolipoma which are more than 4 cms in size should either be kept under close followup or advised treatment due to high chances of bleeding. It should be kept as a differential diagnosis in patients presenting with signs and symptoms of internal bleeding without an obvious cause.

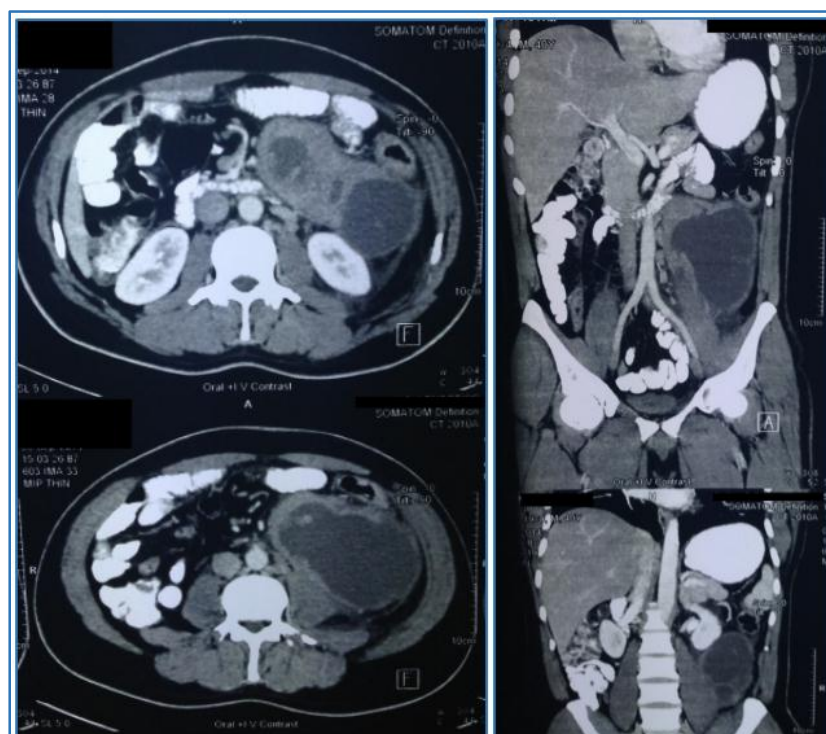


Fig. 1 & 2: CT scan showing the location, relationship of the tumour to the left kidney and internal bleeding

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