

Correspondence:

Giant angiomyolipoma with Wunderlich's syndrome in a patient with tuberous sclerosis

A 25-year-old male presented with right loin pain of two weeks duration. On examination, he had adenoma sebaceum over face (Figure 1), gross pallor, severe tenderness in right lumbar region and a right loin lump extending into hypochondriac and iliac regions. Contrast enhanced computed tomography (CT) of the abdomen revealed right side enlarged kidney with massive subcapsular right renal haematoma (Figure 2) and multicentric nodules with of fat densities (< 20 Hounsfield units) within both the renal parenchyma, suggestive of bilateral renal angiomyolipomas. CT of the brain showed subependymal nodules (Figure



Figure 1: Clinical photograph showing adenoma sebaceum over the face

Received: 22 July, 2013.

3). This pattern of cerebral ependymal nodules together with adenoma sebaceum was diagnostic of tuberous sclerosis. Patient was resuscitated and planned right sided renal artery embolization but the procedure failed due to altered renal hilar anatomy due to displacement by haematoma.

In view of expanding retroperitoneal haematoma, emergency laparotomy and right partial nephrectomy was planned. Bilateral subcostal (Chevron) incision was made and right kidney was exposed transperitoneally by reflecting the right colon and hepatic flexure by Centrallization. Duodenum was kocherized to expose hilum of right kidney. Owing to huge haematoma crossing midline, technical difficulty of approaching the hilum was faced. As there was only small amount of normal right renal parenchyma noted in lower pole right total nephrectomy was planned. After ligating the renal artery and renal vein, kidney along with the haematoma was removed which measured 23 cm in maximum diameter (Figure 4). Histopathology it was confirmed as angiomyolipoma (Figure 5). Postoperatively patient has normal serum creatinine and is on regular follow-up.

Spontaneous non-traumatic renal haemorrhage (Wunderlich's syndrome) is known to occur in renal angiomyolipoma and may be the first manifestation of the disease.

Classic angiomyolipoma is considered to be benign mixed mesenchymal tumour that occurs

Tyagi A, Reddy CS, Anil Kumar N, Reddy AR, Ganesh A, Radhika K. Giant angiomyolipoma with wunderlich's syndrome in a patient with tuberous sclerosis. *J Clin Sci Res* 2014;3:210-13.



Online access

http://svimstpt.ap.nic.in/jcsr/jul-sep14_files/corr314.pdf

DOI: <http://dx.doi.org/10.15380/2277-5706.JCSR.13.039>

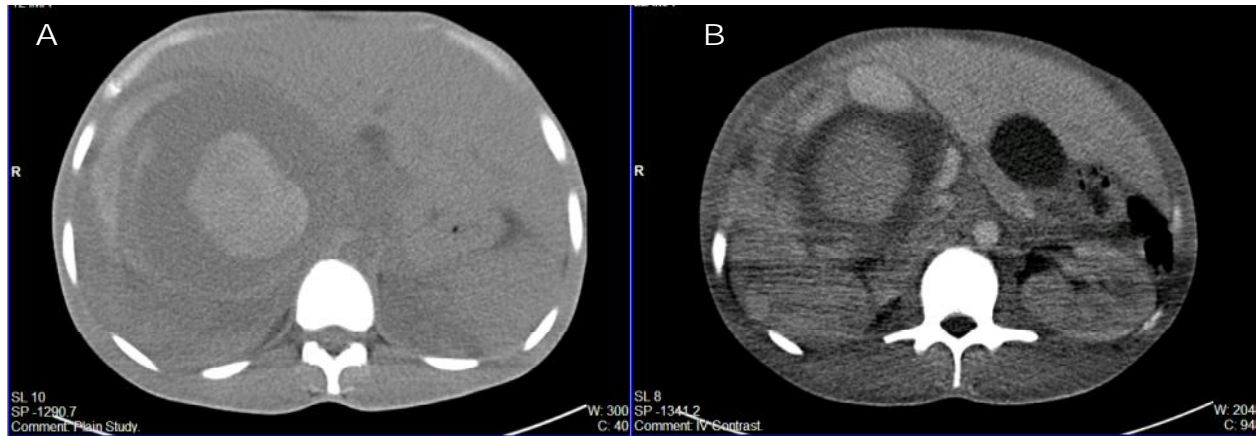


Figure 2: CECT abdomen showing right-sided angiomyolipoma of kidney with haemorrhage. Pre (A) and post contrast (B) CT axial images of abdomen showing large heterogeneously enhancing mixed density lesion with hyperdense components suggestive of hemorrhage in the right kidney

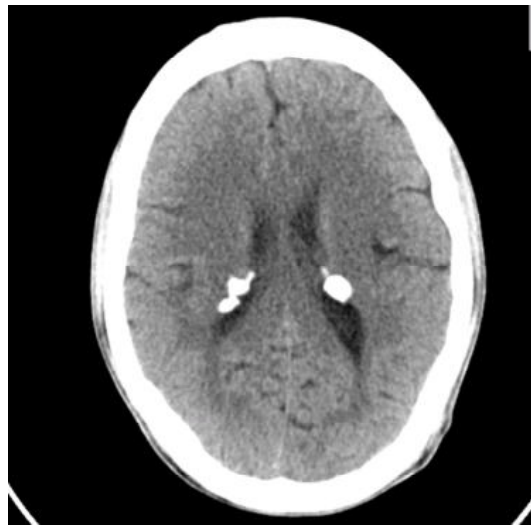


Figure 3: CECT brain showing calcified sub-ependymal nodules abutting body of lateral ventricles

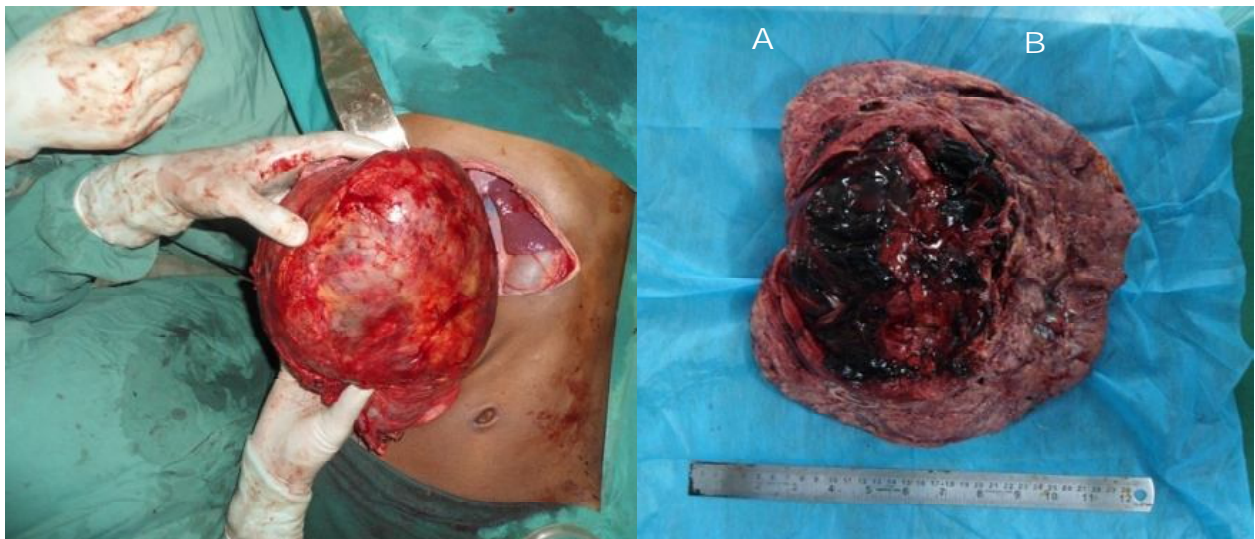


Figure 4: Intra-operative photographs showing a large, ovoid, encapsulated lesional mass (A). The cut-surface shows brilliant yellowish and central haemorrhagic areas (B)

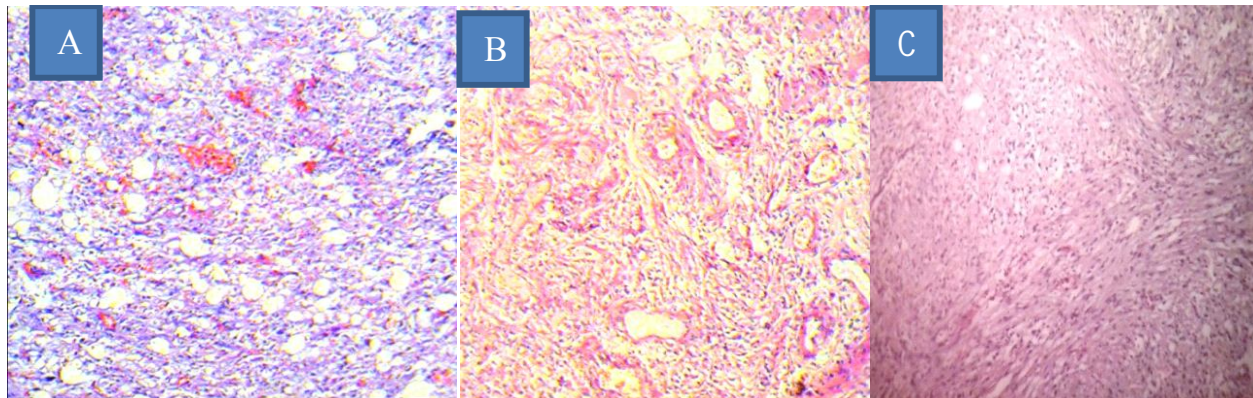


Figure 5: Photomicrograph showing (A) proliferation of adipose tissue, blood vessels (B) and smooth muscle (C) Photomicrograph showing the presence of mature adipose tissue (A), thin walled tortuous blood vessels (B) in the midst of spindle shaped smooth muscle cells, another area exhibiting proliferating smooth muscle cells (C), (Haematoxylin and eosin, $\times 40$)

predominantly in the kidney. These tumours consist of a collection of thick-walled blood vessels, smooth muscle and mature adipose tissue in varying proportions. A second type of angiomyolipoma is described containing a large fourth component, the perivascular epitheloid cells, making it more aggressive than the classical form. The incidence in the general population is between 0.07% and 0.3%.¹ Approximately 80% of renal angiomyolipomas occur sporadically and 20% are associated with tuberous sclerosis. In the sporadic cases, these lesions are found usually larger, single and unilateral, with a female preponderance (approximately 4:1) and occur in the fourth to sixth decade of life.

The frequency of symptoms and the risk of bleeding increase with the size of the lesion. Approximately 64%-77% of tumours less than 40 mm in diameter are asymptomatic, although 82%-90% of angiomyolipomas larger than 40 mm produce symptoms.²

Wunderlich's syndrome, a urological emergency, refers to spontaneous nontraumatic renal bleeding confined to the subcapsular and/or perinephric space. Various aetiological causes for Wunderlich's syndrome include benign and malignant renal neoplasms, vascular

disease (vasculitis, renal artery arteriosclerosis, and renal artery aneurysm rupture), nephritis, infections, undiagnosed haematological disorders, and anatomical lesions. Of all these aetiologies, the most common cause is renal angiomyolipoma.

Angiomyolipomas are known to occur in Tuberous sclerosis, an autosomal dominant disorder characterized by mental retardation, epilepsy, and adenoma sebaceum, a distinctive skin lesions.³ Angiomyolipoma in tuberous sclerosis is usually bilateral and multicentric.

Clinical presentation of Wunderlich's syndrome depends on the degree and the duration of the bleeding. Classically, these patients present with flank or abdominal pain, a palpable tender mass, and gross hematuria (Lenk's triad).⁴ Other symptoms and/or signs are nausea, vomiting, fever, anaemia, renal failure, and hypotension. The syndrome can be fatal if not identified and promptly treated. CT is the investigative modality of choice in diagnosing angiomyolipoma and its related complications. Presence of fat in a noncalcified renal mass lesion on computed tomography confirms the diagnosis of angiomyolipoma in most cases.⁵

The management of Wunderlich's syndrome depends on the general condition of the patient. In patients with solitary tumour or tumours

confined to one part of the kidney, renal embolization can arrest the bleeding and these patients can later be considered for a nephron sparing surgery in the form of partial nephrectomy. Selective embolization and nephron sparing surgery should be considered especially in patients with bilateral disease and pre-existing renal insufficiency. However, coalescent renal angiomyolipoma replacing entire renal parenchyma is a much more vexing clinical problem. When bleeding occurs in this circumstance, it can be impossible to identify which lesion is the source. Nephron sparing surgery is not feasible when entire renal parenchyma is replaced by angiomyolipoma.

The role of embolization in the management of existing or impending haemorrhage is expanding.⁶ Both embolization and nephron-sparing surgery have been reported with excellent success and minimal complications. Embolization can temporize and in many cases, arrest bleeding. A nephron-sparing approach in the form of partial nephrectomy should be considered when possible particularly in patients where preservation of renal function is at issue.⁷ Patients with life-threatening hemorrhage, failed embolization require urgent exploration and in majority of the cases require total nephrectomy.⁸ We considered nephrectomy as our patient was having dropping haematocrit levels and increasing size of hematoma.

As the renal angiomyolipoma were bilateral in our patient, he was prone for Wunderlich syndrome on the contralateral side also. The patient and her relatives are well educated about

the nature of pathology. He has been informed to report immediately in case of sudden abdominal pain and/or haematuria. He was also educated as to the possible need of renal transplantation should he require contralateral nephrectomy owing to failed embolization or torrential bleed.

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