

Wunderlich's syndrome in a patient of tuberous sclerosis

Abstract

Chondrosarcomas are malignant cartilaginous tumours, commonly found in long bones in this case report a rare presentation of this tumor in spinal cord was presented. The patient was a 45 year old male with history of Swelling & dull aching pain in back for 10 months. His routine Labs were unremarkable.

MRI of Dorsal spine showed a large serpentine multilobulated cauliflower shaped mass lesion of 11.5 X 7.5 X 5.5cm emanating from right side neural foramina at D1-D2 intervertebral disc level. Mass is extending from D1 to D6 vertebrae. It has small intra foraminal & large extra foraminal components. The intra foraminal component is intradural & extramedullary in location. It is causing widening of intervertebral foramina, erosion & scalloping of posterior cortex of vertebral body at D1 transverse process, lamina & spinous process of D1 & D2 vertebrae. There is lateral displacement of spinal cord resulting in moderate compression of spinal cord at D1 & D2 levels. The Large extra foramen component is in right paravertebral region. It is abutting right mediastinal pleura adjacent to apical segment of right upper lung anterosuperiorly & extending caudally along right paravertebral region involving right paravertebral & postvertebral muscles of both sides & pressure erosion is also identified in right posterior 1st & 2nd ribs. The mass is hyperintense with central linear hypointensities on T2WI, uniformly hypointense on T1WI & showing peripheral rim enhancement on post contrast study. It was not suppressed on STIR and fat sat images. Abnormal enhancement also seen in adjacent paravertebral muscles, right posterior 1st & 2nd ribs & lamina & spinous process of D1 & D2 vertebrae representing infiltration.

Complementary focused CT scan done which confirmed pressure erosions in above describe levels & also showed shell like calcification in mass adjacent to spinous process of D2 vertebra. On the basis of CT and MRI findings the diagnosis of plexiform neuro fibroma was suspected. Excision of the mass done. Histopathology report revealed Chondroid neoplasm, features favoring Chondrosarcoma grade II.

Keywords: chondroid neoplasm, chondrosarcoma, plexiform neuro fibroma, lung anterosuperiorly, paravertebral muscles

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Introduction

Tuberous Sclerosis also known as (Bourneville Disease/Epiploia). It is Autosomal dominant neuroectodermal disorder characterized by multifocal systemic hamartomas and malformations that may affect CNS (90%), kidney, lung 4%, skin, heart. Classic triad is seen in 29%; is, facial Angiofibromas, Epileptic Seizures and Mental Retardation.

Wunderlich syndrome is a rare condition in which spontaneous non-traumatic renal hemorrhage occurs into subcapsular and perirenal space. It is clinically characterized by Lenk's triad i.e Acute flank pain, flank mass and hypovolemic shock. It's aetiology is divided into neoplastic and non-neoplastic causes. In neoplastic lesions angiomyolipoma is the most common cause. Classic angiomyolipoma is considered to be benign mixed mesenchymal tumours in kidney. These tumours consist of collection of thick walled blood vessels, smooth muscle and mature adipose tissues in varying proportion.

Case report

We present a patient of tuberous sclerosis complex with left sided renal angiomyolipoma with spontaneous retroperitoneal hemorrhage from right angiomyolipoma (Wunderlich's syndrome). The patient is a diagnosed case of Tuberous sclerosis with epilepsy and a history of right sided nephrectomy due to ruptured angiomyolipomas. now came in emergency with presenting complain of left flank pain and

hematuria. On physical Examination abdominal tenderness, low blood pressure, tachycardia, weak feeble pulse and tachypnea. Lab Investigations showed Hb: 7.1mg/dl, Serum Creatinine: 3.41mg/dl, Urea: 45mg/dl, K: 3.8mg/dl, TLC: 10.5mg/dl. Multiple blood transfusions were done. Patient was dialyzed multiple times. Ultrasound was done in the ER (Figure 1) that showed, enlarged left renal shadow with multiple hyperechoic lesions, located in the cortex representing multiple angiomyolipomas; foci of mixed echogenicity also noted in left kidney and in perirenal space representing bleed in angiomyolipoma. The right renal bed was empty consistent with prior nephrectomy.

CT KUB (Figure 1 & Figure 2) showed multiple mixed density lesions in the renal parenchyma of Left kidney, all containing fat density, representing multiple angiomyolipomas along with a large hyper dense foci in the left kidney and perinephric space suggestive of intra and perirenal hematomas. HRCT Chest (Figure 3 & Figure 4) showed multiple bilateral lung cysts, with bilateral pleural effusion.

Patient was planned for MRI (Figure 5) that showed multiple abnormal signal intensity areas in cortical, subcortical deep white matter of bilateral frontal and right parietal lobes which appear iso intense on T1WI and hyper intense on T2WI and FLAIR. Few subependymal nodules were also noted along the body of lateral ventricle which were isointense on T1W and FLAIR and hypo intense on T2W, enhancing on post contrast, representing multiple tubers.



Figure 1 Ultrasound Left kidney showing peri and intra renal hematomas.

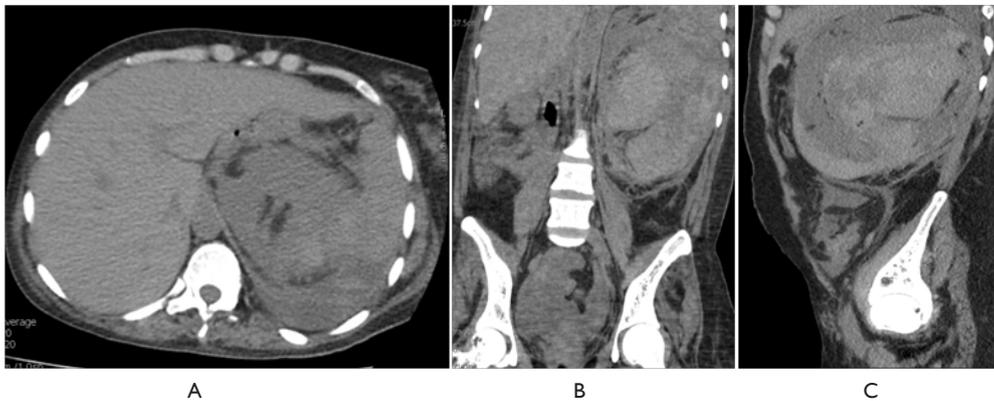


Figure 2 Axial (A) Coronal (B) and Sagittal (C) sections of plain CT showing large mixed density mass in left lumbar region, the hyper dense foci represents bleed. These foci are also seen in retroperitoneum. Note absence of right renal shadow due to nephrectomy.

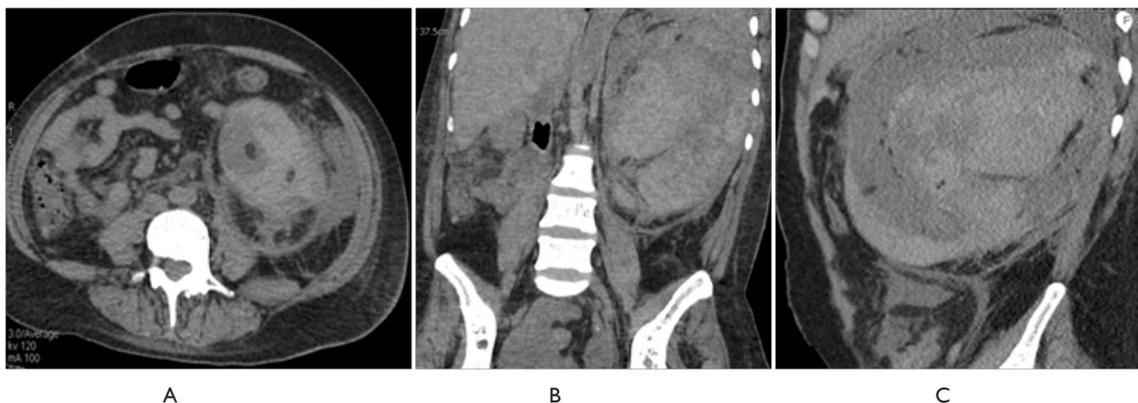


Figure 3 Axial (A) Coronal (B) and Sagittal, (C) sections of plain CT at more distal level, showing hypodense foci representing Angiomyolipomas.

Discussion

Tuberous sclerosis (TS) or Bournville-Pringle disease is a relatively rare autosomal dominant disorder classically described as a clinical triad of adenoma sebaceous, mental retardation and seizures. Multiple body organs can be involved by this condition.

Three types of renal involvement have been described in TS: a) Renal AML (40-80%); b) Cystic disease (occasionally); c) Renal cell carcinoma, with the most common renal lesion being AML.¹

Wunderlich syndrome (WS) is a rare condition characterized by acute onset of spontaneous, non traumatic renal hemorrhage into the sub capsular and perirenal spaces.

The syndrome is classically characterized by the Lenk's triad: acute flank pain, flank mass, and hypovolemic shock. Approximately 60% to 65% of all cases of WS are due to renal neoplasms; the Angiomyolipoma is the most common renal neoplasm responsible for WS.²



Figure 4 Multiple bilateral diffusely distributed lung cysts. Bilateral pleural effusion.

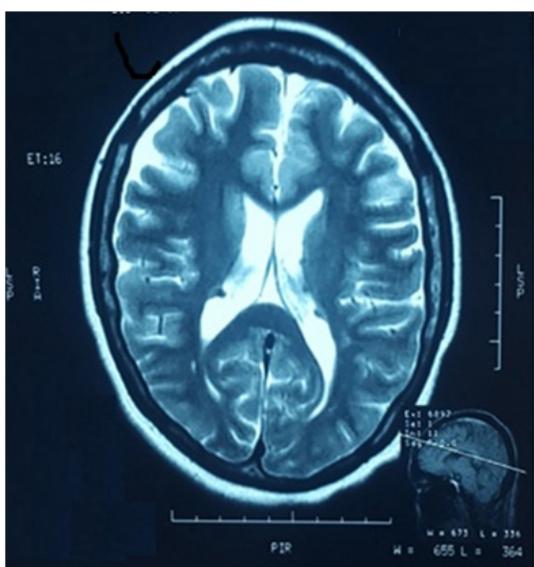


Figure 5 Subependymal nodules on T2W of MRI.

On CT, the classic AMLs appear as heterogeneous masses with varying proportion of macroscopic fat, hyper vascular soft tissue components and intralesional aneurysms, this appearance is often altered by hemorrhage within the lesion and perirenal hemorrhage in patients with WS. The renal AML on MRI shows high signal on T1W images and signal loss on fat-saturated sequences, indicating the presence of macroscopic fat.³

The role of Radiology in a suspected case of Wunderlich syndrome (WS) is confirmation of the diagnosis by identifying renal hemorrhage in the sub capsular and/or perirenal space and detecting the possible cause for the bleeding eg, Angiomyolipoma, other renal tumors or calculi.⁴

In addition the control of active bleeding by, angioembolization of bleeding vessel by interventional radiologist can avoid unnecessary emergent radical surgery. Ultrasound may be the initial modality to detect perinephric hematoma. On US, acute hematoma appears as an isoechoic to hyperechoic perinephric collection causing compression and displacement of renal parenchyma; Multidetector CT imaging is the initial imaging modality of choice. Acute perinephric and sub

capsular hemorrhage appears as an expansile high-attenuation (40Y70 Hounsfield units) fluid collection on unenhanced CT.⁵

Active contrast extravasation may be seen on contrast-enhanced images if there is ongoing bleeding. Magnetic resonance imaging may be performed in whom the bleeding source is not identified on initial CT examination. MRI demonstrates characteristic high signal intensity on T1W; subacute hemorrhage shows hyperintense signal on T1W and T2W images.⁶

In addition, subacute hemorrhage usually will have an inhomogeneous MRI signal output reflecting varying degrees of hemoglobin degradation.⁷ Gadolinium administration helps in the identification and characterization of the renal masses and vascular pathologies.

Conclusion

Wunderlich's syndrome associated with massive retroperitoneal hemorrhage is reported in 20% of all Tuberous sclerosis cases. It is the one of the most feared complications of renal angiomyolipoma and can be fatal if not treated promptly and aggressively; the condition is associated with 10% of all angiomyolipomas. Appropriate treatment for this condition depends on the accuracy of diagnosis and the determination of its cause. Role of radiology is not only in the diagnosis of this syndrome but also in the management of this condition. Selective renal arterial embolization of bleeding vessel in angiomyolipoma is a procedure of choice that helps in immediate and effective control of bleeding in emergency. This procedure has become a procedure that offers greater efficacy, particularly in life-threatening cases.

Acknowledgments

None.

Conflicts of interest

The author declares that there is no conflicts of interest.

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