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Original Research Paper

Radiodiagnosis

A RARE COMPLICATION –"WUNDERLICH'S SYNDROME";SPONTANEOUS RUPTURE OF RENAL ANGIOMYOLIPOMA IN A PATIENT OF TUBEROUS SCLEROSIS COMPLEX – A CASE REPORT

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ABSTRACT We present an extremely rare case of a young female, having spectrum of tuberous sclerosis complex, exhibiting life threatening Wunderlich's syndrome after non traumatic rupture of renal angiomyolipoma. The early ultrasound and Computed tomography (CT) aids in the prompt diagnosis of tuberous sclerosis complex, in patients with retroperitoneal hemorrhage of large right renal angiomyolipoma. Patient also have B/L renal angiomyolipomas, hepatic lipomas, calcified subependymal nodules, lung cysts and facial adenoma sebaceum. Patient underwent immediate laparotomy and thereafter was stable and discharged. All the patients of tuberous sclerosis should be screened radiologically for giant renal angiomyolipoma for prompt treatment of life threatening condition of Wunderlich syndrome.

KEYWORDS : Renal angiomyolipoma, Tuberous sclerosis, Wunderlich's syndrome, Lenk's Triad, Page kidney.

INTRODUCTION

Non traumatic rupture of renal angiomyolipoma (AML) in patients of tuberous sclerosis; Wunderlich's syndrome is an extremely rare complication. A large but contained perinephric hematoma in renal AML usually presents as commonest emergency in 80 % of asymptomatic patients due to abnormal vasculature[(1)].

In patient with tuberous sclerosis (TS), AML accounts for more than 50 % cases and are usually bilateral and multifocal. These are usually small and incidental at times. Our case however presented as symptomatic rupture of renal AML which needed prompt diagnosis and immediate intervention. Hence, as radiologists, our role to recognise its imaging features leads to early action plan for surgeons to act.

CASE STUDY

A young 25 yr old female, visited Surgery O.P.D. for acute pain abdomen and dyspnea. She complains of increased respiratory rate and hematuria. She was on medications for epilepsy. On clinical examination she was pale, had tender and distended abdomen with multiple adenoma sebaceum on her face[Figure 1]. In the initial investigations her haemoglobin was 5gm %.

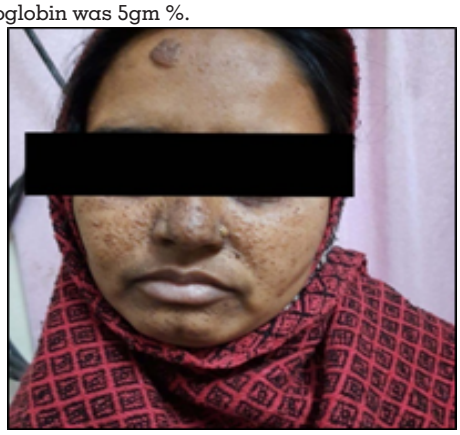

Figure1: Photograph Demonstrates Multiple Facial Adenoma Sebaceum.

On abdominal ultrasound a large heterogenous, well defined space occupying lesion is seen in right lumbar region, extending to right iliac fossa with hyperechoic and hypoechoic areas associated with few spaces showing color flow signal in YIN -YANG pattern [Figure2].

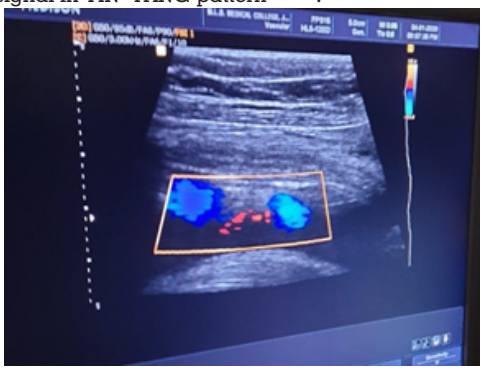

Figure 2: Colour Doppler Usg Showing A Heterogenous Mass In Right Lumber Region With Colour Flow Signal

CECT abdomen imaging suggests a large but contained perinephric heterogenous lesion with hypodense and isodense areas; lesion have 'claw sign' with right kidney exhibiting its origin from the right kidney. The lesion extends cranio caudally from sub hepatic space to the right iliac fossa. Anteroposteriorly the lesion extends from the anterior abdominal wall to the right kidney posteriorly. The lesion appears to cross midline [Figure 3]

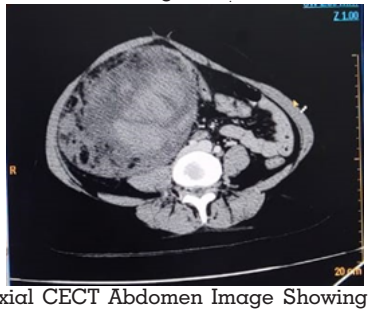

Figure3: Axial CECT Abdomen Image Showing A Massive Right Renal AML With Large Perinephric Hematoma.

On CT angiography the lesion takes up heterogenous enhancement, with opacification of renal arteries. Ongoing bleeding was demonstrated by contrast blush in the hematoma on arterial phase, which appears hyperdense(> 120 H.U.)[Figure4,5].

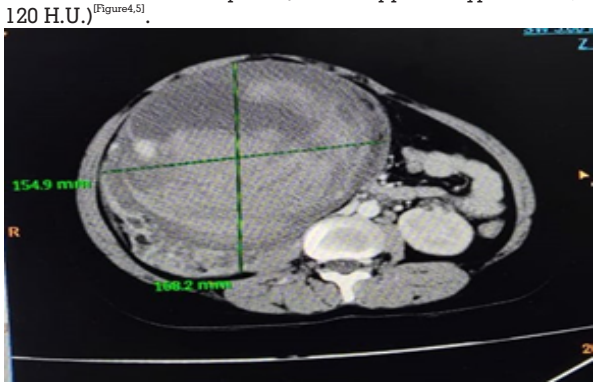

Figure 4: Axial CECT abdomen image revealed a huge fat containing lesion with hematoma formation and active contrast extravasation over right perirenal space in the form of hyperdense area (*asterisk)

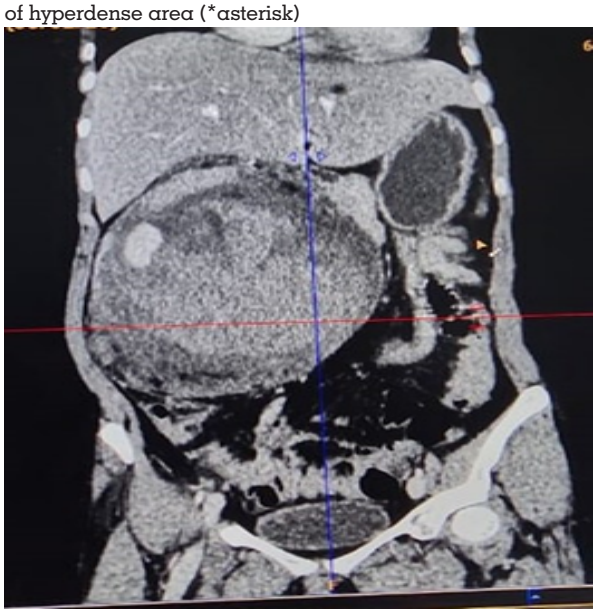

Figure 5: Coronal CECT Abdomen Image Showing A Giant Right Renal Ruptured AML With Heterogenous Density And Ongoing Bleed (*asterisk)

Other findings includes bilateral small renal lipomas (H.U.<-20) and multiple hepatic lipomas[Figure6].

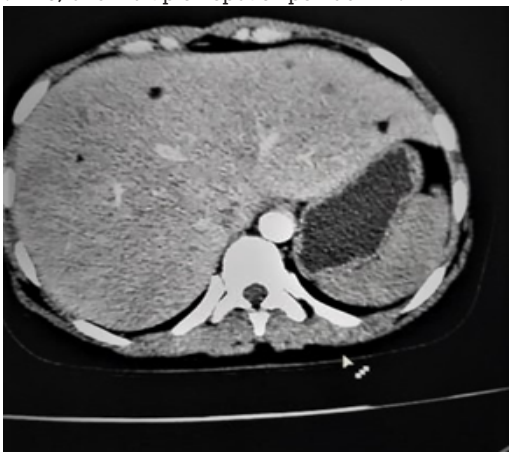

Figure 6: Axial CECT abdomen image showing multiple hepatic lipomas.

Brain CT shows multiple calcified subependymal nodules[Figure7].HRCT Chest demonstrates multiple thin walled small lung cysts with uniform distribution corresponding to Lymphangioleiomyomatosis (LAM)[Figure8].

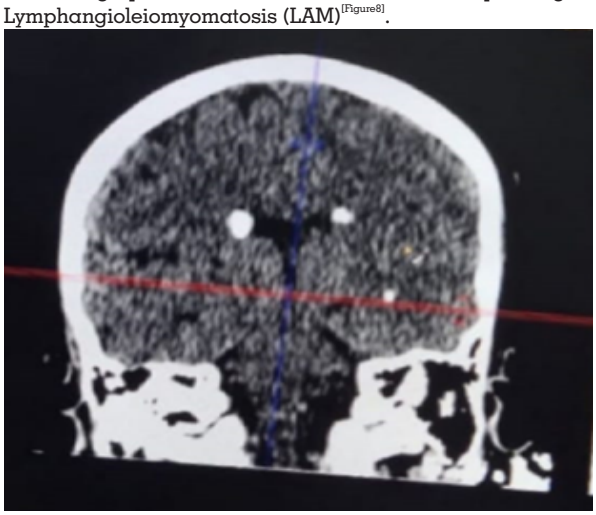

Figure7: NCCT Head Coronal Image Showing Multiple Calcified Subependymal Nodules.

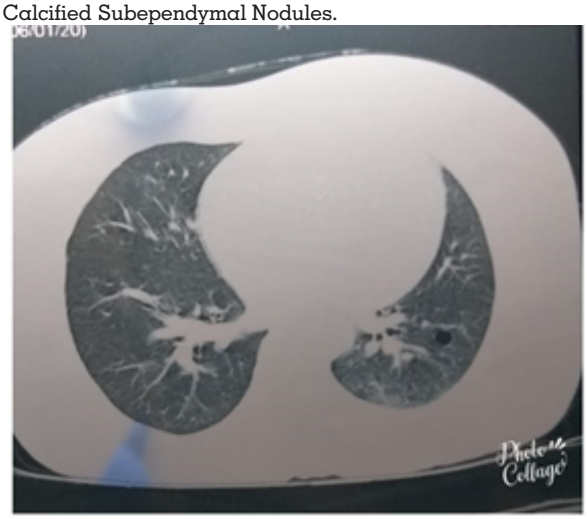

Figure 8: Axial HRCT chest image showing multiple thin walled small lung cysts corresponding to Lymphangioleio myomatosis (LAM).

So glancing all together, rupture of renal AML, bilateral renal and hepatic lipomas or hamartomas, associated with subependymal calcified nodules, lung cysts and adenoma sebaceum on face, points towards diagnosis of tuberous sclerosis complex with Wunderlich's syndrome.

DISCUSSION

Hamartomas in patients of tuberous sclerosis (TS) usually showcases AML, lipomas and fibromas. TS is an autosomal dominant disorder presents with triad of mental retardation, epilepsy and adenoma sebaceum[(2)]. AML in TS is often multicentric, B/L and common in 80% of patients.Rupture of renal AML accounts for a rare urological emergency leading to subcapsular and perinephric hematoma. Other renal manifestations are benign and malignant renal neoplasms, renal vascular disease, nephritis and other space occupying lesions[(3)].

Classic presentation of Wunderlich's syndrome is Lenk's triad including acute flank pain, palpable abdominal mass and hypovolemic shock[(3)]. It can be proved fatal if delay occurs in recognizing its clinical symptoms and imaging features. Neglected long standing perinephric hematomas

can cause compression of renal parenchyma which can further lead to hypertension, deterioration of renal function, termed as Page kidney. Variable complications of AML can be easily diagnosed on CT and hence it is the modality of choice. Ruptured renal AML can be managed by angioembolization as renal sparing surgery, but when bleeding is massive, renal sparing surgery is not possible.

Spectrum of Tuberous Sclerosis Complex includes subependymal calcific nodules, subependymal giant cell astrocytoma (SEGA), white matter lesions and cortical tubers in the brain. The pathognomic cutaneous manifestations include ash leaf spots, angiofibromas, ungual fibromas, subungual red comets and splinter hemorrhages.

AML that occurs in association with tuberous sclerosis on other hand manifests at younger age, they are usually larger, B/L and are prone to grow[4]. We as radiologists have a key role in addressing these issues. CT is the method of choice for the demonstration of perirenal hemorrhage with 100 % sensitivity. A confident diagnosis of AML can be made with CT by demonstrating fat content. If the initial CT shows no mass responsible for hemorrhage, it has been suggested that angiography should be done to reveal vascular lesion and ongoing hemorrhage and it also facilitates embolization[5] as in our case.

CONCLUSION

To conclude, we have witnessed and so presented a case of Wunderlich's syndrome secondary to rupture of right renal AML, which is the commonest cause reported in most series in patient of tuberous sclersosis complex. CT and CT angiography scans plays a key role in making diagnosis and aids in prompt treatment.

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