CASE REPORT

Wunderlich Syndrome in a Gravid 31-Year-Old with Tuberous Sclerosis Complex and Bilateral Angiomyolipoma: A Case Report

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Wunderlich Syndrome is a rare potentially life-threatening phenomenon that involves spontaneous non-traumatic retroperitoneal hemorrhage. At present, identifying the course of conservative management in these patients, especially in pregnancy, has not been clinically established.

Presented here is a known case of Tuberous Sclerosis Complex with a Bilateral, 10cm Angiomyolipoma in a 31-year-old female, initially managed with active surveillance. At 27 weeks of pregnancy, she presented with a sudden onset of left flank pain with a hematocrit of 22%. Anemia was corrected with blood transfusions. A contrast-enhanced MRI of the abdomen showed a large subcapsular perirenal hematoma of the left kidney. Renal angioembolization of the bleeding segmental renal artery was done. The patient was conservatized until 37 weeks of pregnancy and underwent cesarean section delivery.

Four months after angioembolization, she had a recurrence of the left flank pain associated with gross hematuria and hypovolemic shock. The patient underwent emergency renal exploration of the left kidney via a transabdominal approach. Three liters of hemoperitoneum and a large expanding left retroperitoneal hematoma were noted intraoperatively. Early vascular control before nephrectomy of the left kidney was done. The postoperative course was unremarkable and the patient was discharged with improved condition.

This case displays a unique course in the management of a bleeding angiomyolipoma especially during pregnancy. Renal angioembolization can aid in achieving the age of viability in pregnancy. However, close monitoring for rebleeding should be kept in mind. A lower threshold for conservative management should be utilized when patients have a previous history of bleeding.

Key words: Wunderlich syndrome, angiomyolipoma, tuberous sclerosis complex

Introduction

Angiomyolipoma (AML) is a benign renal tumor composed of dysmorphic blood vessels, smooth muscles and adipose tissue. It is estimated that the prevalence of AML is at 0.13% in the general population and has a strong female preponderance.[1-2] These tumors arise sporadically but are also part of genetic mutation syndromes such as Tuberous Sclerosis Complex (TSC) and Lymphangioleiomyomatosis (LAM).[2]

Wunderlich Syndrome is a rare potentially life-threatening phenomenon that involves spontaneous non-traumatic retroperitoneal hemorrhage. At present, identifying the course of conservative management in these patients, especially in pregnancy, has not been clinically established.[3-4] Several management options depend on patient

stability and other risk factors.[5] There have been a few reported cases discussing the management of these patients, however, these were all solitary in nature.[6-7]

This paper discusses the course of management of a gravid 31-year-old female diagnosed with Tuberous Sclerosis Complex with Bilateral Angiomyolipoma in Wunderlich Syndrome who underwent an emergency nephrectomy of the left kidney.

The Case

The authors a case of a 31-year-old female diagnosed with Tuberous Sclerosis Complex. The patient has no known comorbidities. She is a non-smoker and a non-alcoholic beverage drinker. Heredofamilial diseases were unremarkable.

On physical examination, there were noted facial angiofibromas on the cheeks (Figure 1). A Contrast-Enhanced MRI of the Brain showed multiple cortical tubers consistent with Tuberous Sclerosis (Figure 2).

Figure 1. Facial angiofibromas

On further workup, an incidental finding of a stable bilateral 10cm Angiomyolipoma was seen on a contrast-enhanced abdominal CT scan. Her attending urologist initially managed her with annual abdominal imaging as a means of active surveillance.

After four years of active surveillance, the patient was 27 weeks pregnant with her first baby and presented at the emergency room for sudden onset of severe left flank pain. She was a G2P0(0010) wherein she previously underwent dilatation and curettage for an incomplete abortion. The current pregnancy had regular prenatal checkups. The congenital anomaly scan was also unremarkable.

On current physical examination, she presented with pallor but with no episodes of hypotension or hematuria. The abdominal examination noted a gravid uterus with left flank tenderness. All other findings were unremarkable. The patient was admitted under OB-GYN service and was referred to Urology. Further workup showed anemia (Hematocrit = 22%) which was corrected with multiple blood transfusions. A contrast-enhanced MRI of the abdomen showed bilaterally enlarged kidneys with multiple varisized angiomyolipomas. The largest was in the medial aspect of the left kidney measuring 11.8cm x 5.6cm x 10.6cm (Figure 3). A large subcapsular perirenal hematoma of the left kidney was also seen (Figure 4). The right angiomyolipoma remained stable. The patient was maintained on complete bed rest and serial hematocrit determinations to maximize conservative management and prevent rebleeding of the angiomyolipoma. The status of the fetus was unremarkable and remained stable.

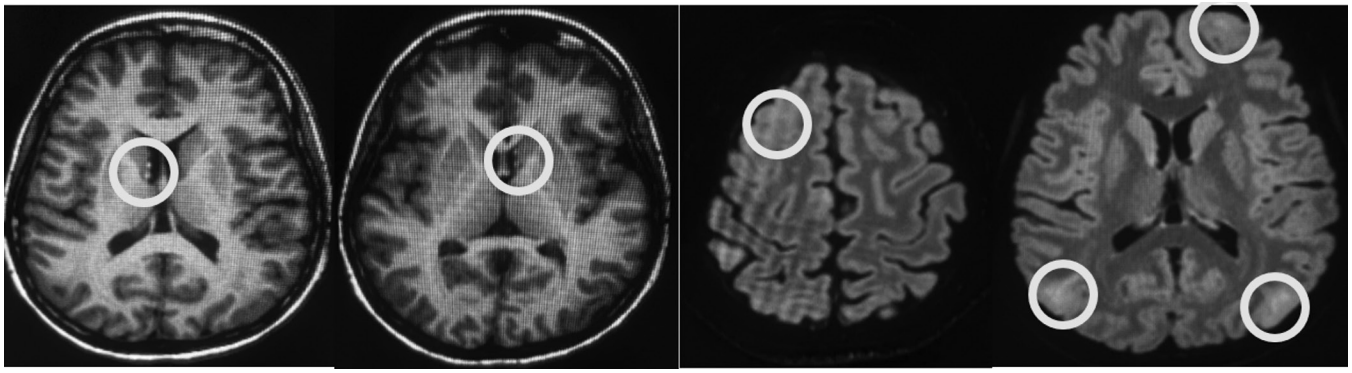

Figure 2. Contrast-enhanced magnetic resonance imaging of the brain showing multiple cortical tubers consistent with tuberous sclerosis.

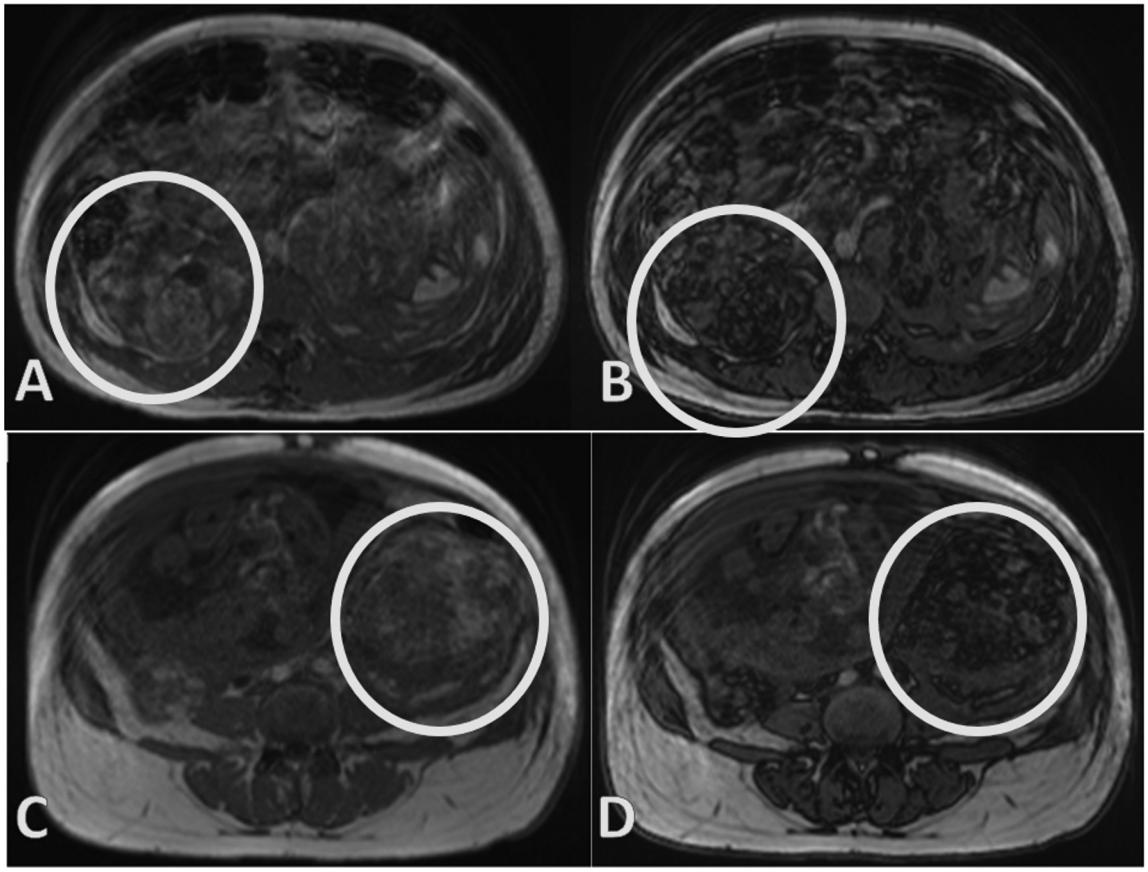

Figure 3. Contrast-enhanced magnetic resonance imaging of the abdomen (Dual Gradient Echo Sequence).
A. Right Angiomyolipoma (In Phase); B. Right Angiomyolipoma; C. Left Angiomyolipoma (In Phase);
D. Left Angiomyolipoma (Out Phase).
Note the Fat Signal Drop on the Out Phase.

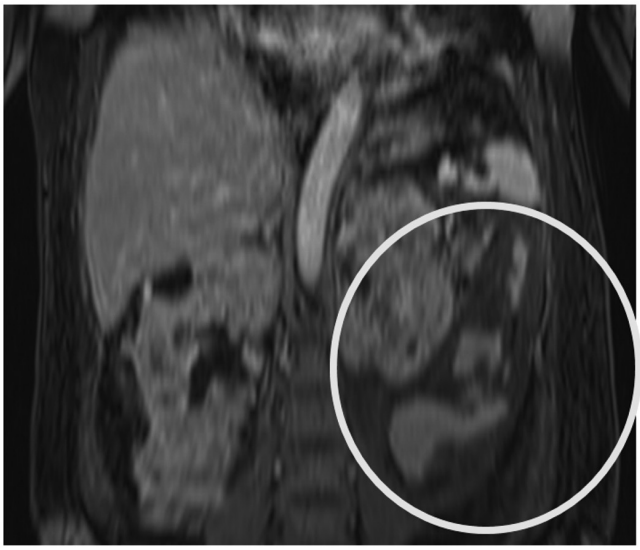

Figure 4. Contrast-enhanced magnetic resonance imaging of the abdomen (Coronal View).
Subcapsular perirenal hematoma.

The patient was scheduled for a renal artery angioembolization to stop retroperitoneal hemorrhage and try to preserve both kidneys. Upon renal angiography, there was contrast extravasation of the superior pole of the left kidney, and the affected superior segmental artery was subsequently embolized (Figure 5). She was then discharged with improved condition and with plans to conservatize the pregnancy until the age of viability.

The patient underwent a scheduled cesarean delivery at 37 weeks of pregnancy after reaching fetal viability. Delivery was unremarkable. She had an unremarkable post-partum course with no hematuria or recurrence of left flank pain.

Four months after angioembolization, the patient presented back to the emergency room due to recurrence of the left flank pain associated with gross hematuria. A contrast-enhanced

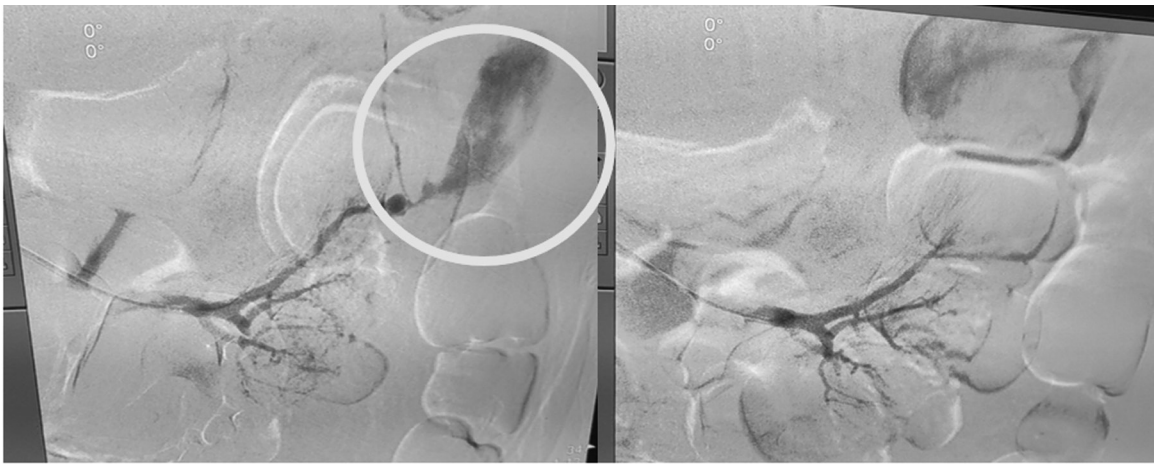

Figure 5. Fluoroscopic images of renal angiography.
A. Pre renal angioembolization showing contrast extravasation of the superior pole of the left kidney;
B. Post renal angioembolization showing normal vascularization of the kidney

CT scan of the abdomen was done showing massive hemoperitoneum, and a large, left suprarenal hematoma extending down to the left retroperitoneal space (Figure 6). She now had episodes of hypotension and was scheduled for emergency renal exploration of the left kidney via a transabdominal approach.

Figure 6. Contrast-enhanced CT scan of the abdomen. Large left retroperitoneal hematoma.

Intraoperatively, three liters of hemoperitoneum was evacuated and a large expanding left retroperitoneal hematoma was noted (Figure 7). Midline early vascular control (Figure 8) was done and urologists proceeded with nephrectomy after ligating the renal hilum. The postoperative course was unremarkable and she was then discharged with improved condition after 1 week.

Figure 7. Large expanding retroperitoneal hematoma.

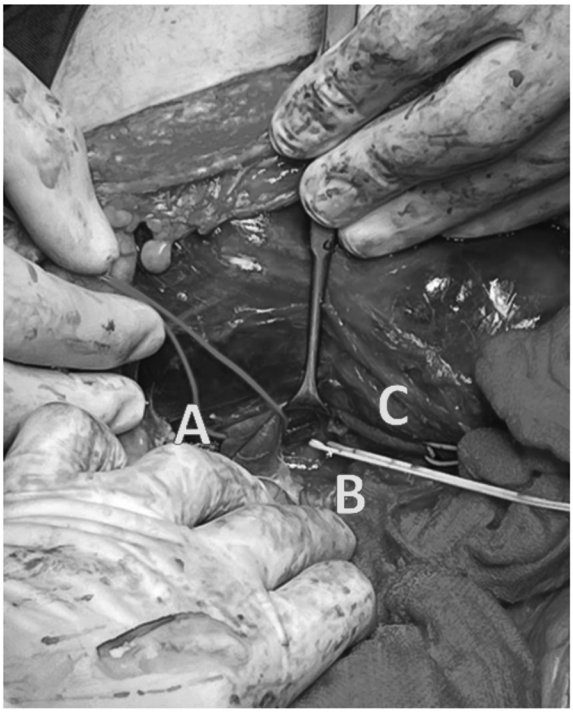

Figure 8. Isolation of the renal hilum via midline early vascular control
A. Renal Vein; B. Renal Artery; C. Ureter

Currently, the right angiomyolipoma, on active surveillance, has remained stable with no intralesional aneurysm or hemorrhage. The patient has been started with everolimus and has been on regular follow-ups every month. A contrast-enhanced CT scan of the abdomen will be done annually for active surveillance of the right kidney.

Discussion

The presented case highlights the different definitive therapeutic interventions of patients with angiomyolipoma. It does not, however, discuss further on the medical and genetic aspect of patients with tuberous sclerosis complex. This case presents a therapeutic dilemma in the management of a bilateral angiomyolipoma in wunderlich syndrome in a pregnant patient.

Angiomyolipomas (AML) are benign renal entities composed of dysmorphic perivascular tissues. Sporadically, they arise as solitary entities. However, when a part of genetic syndromes, such as your Tuberous Sclerosis Complex (TSC), they can affect both kidneys. AML is diagnosed mainly through CT scans or MRIs of the abdomen, in which the mass is predominantly high in fat content.[8]

Wunderlich syndrome is a rare clinical condition of nontraumatic renal hemorrhage which can arise in 15% of patients with Angiomyolipoma.[9] These patients may present with acute hypovolemic shock, sudden onset flank or abdominal pain, and a palpable abdominal mass characterized as your "Lenk's Triad."[10] Pregnancy has been an identified risk factor for spontaneous hemorrhage due to hormonal changes in these tumors.[11]

Current trends in the management of angiomyolipoma have include active surveillance, renal ablation, nephron-sparing surgery and mTOR inhibitors. Angiomyolipomas of less than 4cm have been managed conservatively with annual imaging. Larger tumors have a greater propensity for bleeding leading to nephron-sparing surgery or nephrectomy.[2] Indications to end the active surveillance and proceed to surgical intervention would include persistent pain, hemorrhage, and consideration of probable malignancy. These treatments should be tailored to each individual based on hemodynamic stability and symptom-based therapy.[12]

Steiner et al. suggested that patients with angiomyolipomas must have planned pregnancy and can be managed with pre-pregnancy angioembolization and/or nephron-sparing surgery.[11] However, in the present case, the risk of retroperitoneal hemorrhage and recurrent hemorrhage is high due to pregnancy despite being managed conservatively.

Tanaka et al. reported a case of a ruptured renal angiomyolipoma in pregnancy wherein renal angioembolization was done after successful vaginal delivery.[13] Another case was reported by Shah et al. which discussed angioembolization prior to delivery as a means to conservatize the pregnancy and achieve fetal viability.[14] However, all of these documented cases were solitary in pathology.

This case highlights the management thresholds from active surveillance to active intervention when the patient becomes symptomatic. Although nephron-sparing surgery is the gold standard for angiomyolipomas,[15] this case highlights the difficulty in preserving the kidney when presented

with a large bleeding angiomyolipoma which can lead to emergency nephrectomy. Another dilemma would be to balance out fetal viability as well as achieve adequate conservative management in the ruptured angiomyolipoma.

Conclusion

This case displays a unique management course in a patient with tuberous sclerosis complex and bilateral angiomyolipoma presenting with frank retroperitoneal hemorrhage during pregnancy. Renal angioembolization can aid in achieving the age of viability in pregnancy. However, close monitoring for rebleeding should be kept in mind. A lower threshold for conservative management should be utilized when patients have a previous history of retroperitoneal hemorrhage.

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