รายงานผู้ป่วย

Spontaneous rupture of a renal angiomyolipoma during pregnancy in patient with tuberous sclerosis: a case report.

Apichai Vasuratna*

Pongsak Wannakrairot**

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A case of 22 -year-old pregnant woman at 34 weeks' gastation with underlying tuberous sclerosis complicated by spontaneous rupture of the left renal angiomyolipoma is presented. The treatment consisted of cesarean section, tubal resections, total left nephrectomy and partial left adrenectomy. The patient was discharged home 8 days after the operation. However the infant died 8 days after delivery.

Key words : *Renal angiomyolipoma, Tuberous sclerosis, Pregnancy.*

Reprint request: Vasuratna A, Department of Obstetrics and Gynecology, Faculty of Medicine, Chulalongkorn University, Bangkok 10330, Thailand.

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*Department of Obstetrics and Gynecology, Faculty of Medicine, Chulalongkorn University.

**Department of Pathology , Faculty of Medicine, Chulalongkorn University.

อภิชัย วสุรัตน์, พงษ์ศักดิ์ วรรณไกรโรจน์. การแตกเองของเนื้องอกที่ไตชนิดแองจิโอมัยโอไลโปมาระหว่างตั้งครรภ์ในผู้ป่วยทูเบอร์รัส สเคอโรซิส : รายงานผู้ป่วย 1 ราย. จุฬาลงกรณ์เวชสาร 2539 มกราคม; 40(1): 49-56

รายงานผู้ป่วย 1 ราย อายุ 22 ปี ตั้งครรภ์ 34 สัปดาห์ เป็นโรคลมชัก เนื่องจาก ทูเบอร์รัส สเคอโรซิส มีการแตกของเนื้องอก แองจิโอมัยโอไลโปมาที่ไต ได้รับการรักษาโดยการผ่าตัดคลอดบุตรทางหน้าท้อง, ทำหมัน, ตัดไตข้างซ้ายออกทั้งหมด และตัดต่อมหมวกไตข้างซ้ายออกบางส่วน หลังผ่าตัดผู้ป่วยสบายดีสามารถกลับบ้านได้ในวันที่ 8 หลังผ่าตัด ทารกเสียชีวิตหลังคลอด 8 วัน

Renal hamartoma (angiomyolipoma) is an uncommon benign tumor of the kidney. Spontaneous rupture of renal angiomyolipoma during pregnancy is exceeding rare; to date ,we can find only 9 such cases in the world literature.[1-8] Intrapartum rupture of kidney presents a dilemma in both diagnosis and treatment. The attending physician has to choose between surgical intervention and conservative management. This report describes a pregnant woman with tuberous sclerosis and bilateral renal angiomyolipoma who suffered from massive retroperitoneal hemorrhage.

Case report

A twenty-two-year-old woman, gravida 1, para 0, at thirty-four weeks gestation came to the delivery room due to severe left costal pain after generalized tonic-clonic convulsion three hours ago. She was diagnosed as atrial septal defect (ASD) and tuberous sclerosis with mild mental retardation since childhood. At about 12 years old, ASD was surgically corrected and her functional class was I since then. She has to take phenobarbital to prevent seizure. The last convulsion was at 12 weeks' gestation. She had a prenatal visit once at about 29 weeks' gestation. On physical examination, the patient was found to be drowsy, pale and acutely ill. The blood pressure was 90/60 mm.Hg. The heart rate was 120 beats per minute. Her face showed butterfly pattern adenoma and the rough thickening skin over the back . The uterine size was normal for her stage of pregnancy. The fetus was in cephalic presentation. Its heart rate was 100-120 beats per minute irregularly. There was no uterine contraction. Below left costal margin, there was an exquisitely tender fixed mass which measured about 10 cm. in diameter. Tenderness on left costovertebral angle palpation was also noted. The pelvic examination revealed firm cervix and closed cervical os without evidence of bleeding or rupture of the membranes. No other abnormalities were detected. Immediate resuscitation with volume expander and blood transfusion was undertaken. Initial laboratory data revealed a hematocrit of 26%, white blood cells 10,500 cells/mm^3, platelet count 300,000/mm^3, BUN 10 mg/dl, creatinine 0.9 mg/dl,prothrombin time 11.7 seconds (control 12.5 seconds), partial thromboplastin time 31.1 seconds (control 32.3 seconds). Urinalysis was within normal limits. After half an hour of management her vital signs were improved (blood pressure 100/60 mm.Hg, pulse rate 92/min). The fetal heart sound could be demonsrated between 136-152/min regularly. She also gained full consciousness without evidence of neurological deficit. We consulted general surgeon, urologic surgeon and neurologist to take care this patient as a team. The presumptive diagnosis at that time was ruptured spleen or left kidney with underlying tuberous sclerosis in postictal state. Because her vital signs,fetal monitorings and hematocrits were stable,we decided to further investigate her by abdominal ultrasonographic study then computerized tomography.

Ultrasonographic study of the abdomen revealed left hyperechoic retroperitoneal mass. Its size was 10 x 14 x 17 cm. Slightly enlarged right kidney and moderate amount of peritoneal fluid were also noted. CT scan of the kidney (Figure 1 to 4) showed a 17 x 10 x 14 cm. fat attenuation mass (measured about was -70 HU) with

inhomogeneous contrast enhancement at upper pole of left kidney. Peripheral to the mass, round and cresentic hyperdensity lesions were observed in the pre-contrast study and became slight hypodensity in post-contrast study suggestive of hematoma. Right kidney also contained multiple small hypodensity masses. Bilateral renal angiomyolipoma with left intrarenal and/or subcapsular hematoma were suggested.

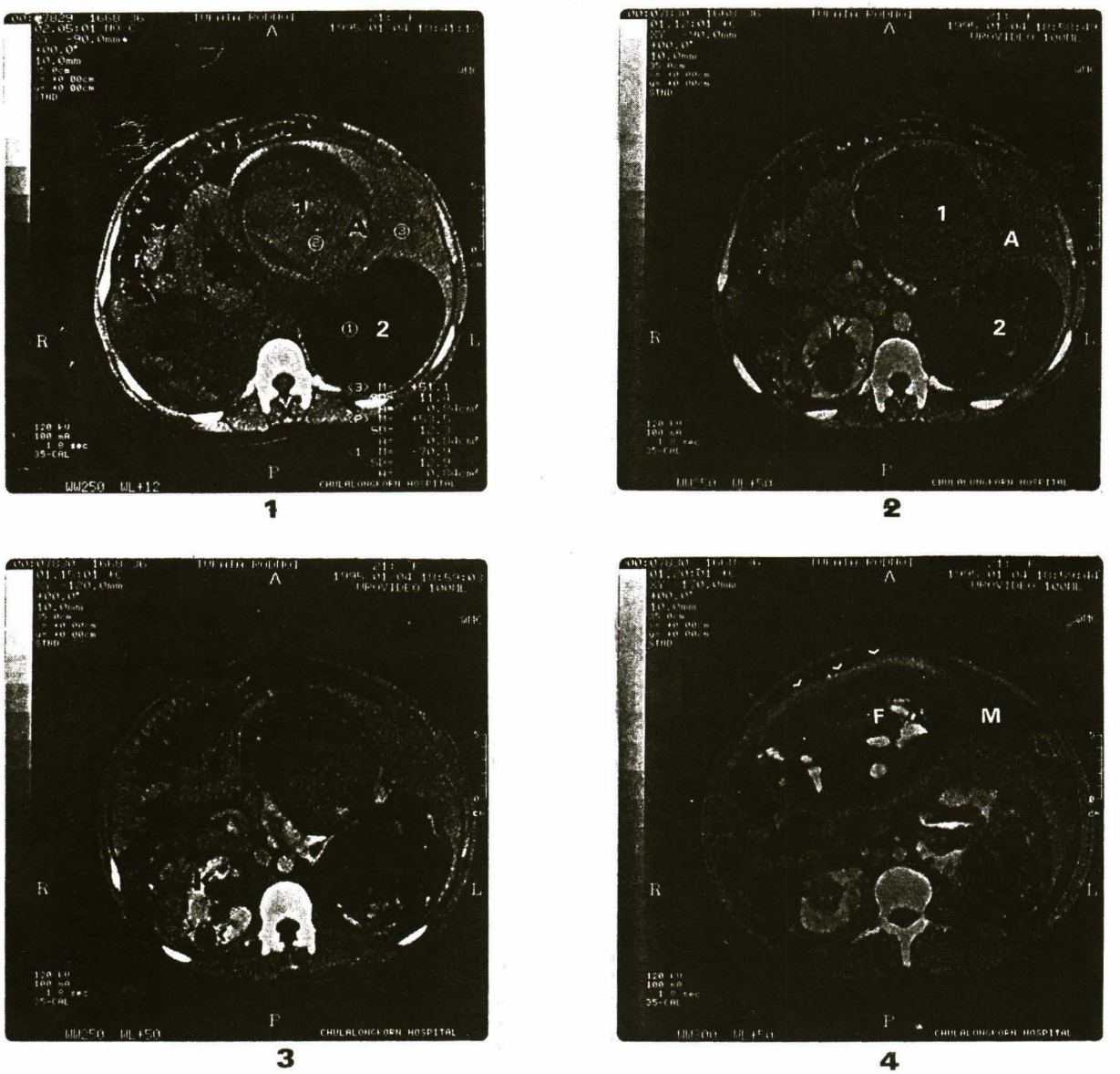

Figure 1-4. CT scans show mixed echogenic mass at left kidney (A) with fat attenuation component(2). The right kidney also contains multiple fat attenuation masses (black arrow heads in figure 1-3). In figure 4,a fetus (F) is demonstrated in the uterus(White arrow head). Moderate amount of fluid is also noted (M).

During investigation procedures her vital signs and fetal heart sound were within normal limits, but after that she developed more severe left upper quadrant pain and the fetal heart sound became irregular again (between 100-120 /min). We decided to perform immediate cesarean section, tubal resections and exploratory laparotomy, after counseling the patient about her diseases and prognosis. The preoperative diagnosis was rebleeding of ruptured left renal angiomyolipoma with fetal distress.

Cesarean section and tubal resections were accomplished first with delivery of a male infant (birth weight 2,500 gm.; Apgar scores were 4 and 9 at 1 minute and 5 minutes respectively), then followed by abdominal exploration which showed serosanguinous fluid approximately 1,000 ml., a large retroperitoneal hematoma with necrotic tumor (15 x 18 x 20 cm.) at the upper pole of left kidney and a tumor measured 3 x 4 x 4 cm. at the lower pole of right kidney. Since left kidney was severely damaged,a left total nephrectomy with partial adrenectomy was performed.

Seven units of blood and blood components were used during operation. The total estimated blood loss was 2,800 ml. The patients postoperative course was uneventful and she was discharged home on the 8th postoperative day.

On the first day of life, the neonate developed hypoglycemia which recovered after treatment. During the 6th to 8th postoperative day, the clinical course of the neonate ran down. His abdomen was distended. Because of severe neonatal sepsis, attempts at resuscitation were unsuccessful and he died 8 days after delivery. Autopsy was not allowed. Blood and cerebrospinal fluid cultures from the neonate revealed Pseudomonas spp.

Histologically, the section of the left kidney showed multiple foci of fatty tissue admixed blood vessels. The vessels revealed hyaline thickening wall and surrounded by plump spindle cells. No mitotic figure was demonstrated. (Figure 5) The final diagnosis was bilateral renal angiomyolipomas with spontaneous rupture at the left one.

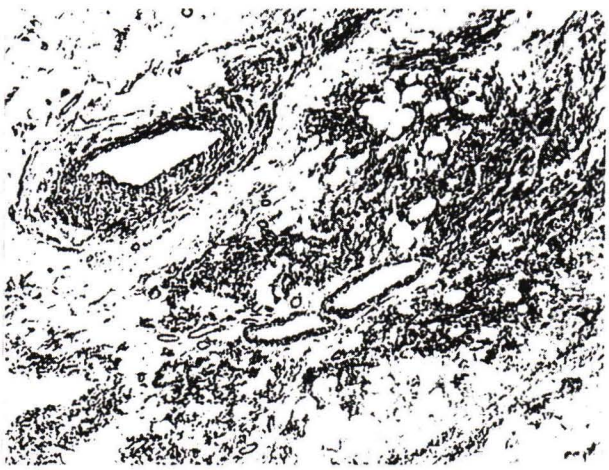

Figure 5. Illustrate mixture of various sizes thick-walled vessels, fat and smooth mucle cells.(H & E stained, x 40)

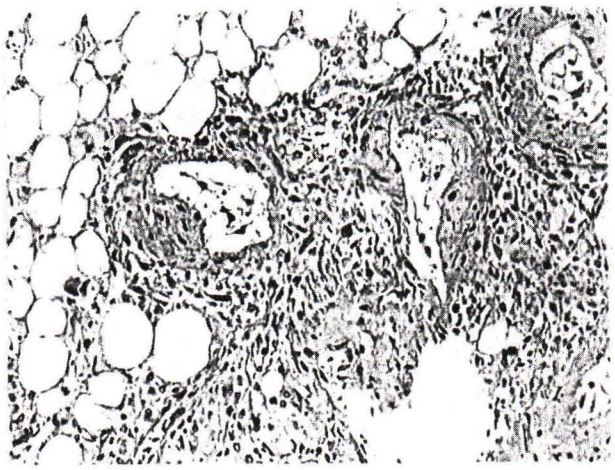

Figure 6. High power menification depicts bundles of spindle cells around irregular thick-walled blood vessels. Note the mature adipocytes on the left upper conner.(H & E stained, x 100)

Discussion

Tuberous sclerosis or Bourneville's disease is a congenital disease inherited in an autosomal dominant fashion.[9] The estimated prevalence of tuberous sclerosis in western society is 1:10,000.[10] Clinical characteristics of tuberous sclerosis includes convulsion, mental retardation, adenoma sebaceum, shagreen patch, rhabdomyoma in heart and angiomyolipomas in several organs.[9,11] About 50% to 80% of tuberous sclerosis patients have renal hamartoma (angiomyolipoma) which have the tendency to be multiple, small, bilateral and asymptomatic.[2,5,6,7,11,12,13]

Renal angiomyolipoma may present in the other clinical features, if it is not associated with tuberous sclerosis. These include single, large, unilateral and symptomatic tumor. Although the incidence of renal angiomyolipoma is about 0.1-2.1% of surgically treated kidney,[12,14] but it is not known about the incidence in pregnancy. We can review only 9 cases of ruptured renal angiomyolipoma during pregnancy in the literatures and three of them occured in tuberous sclerosis patients.[6-8]

The presentation of acute abdominal pain in pregnancy confronts the obstetrician and surgeon with unusual and complicated problems, especially in this patient whom there was the history of convulsion before the abdominal pain started. The differential diagnosis should include the followings:[4,6,7]obstetric caused antepartum hemorrhage (i.e., uterine rupture), ruptured intraabdominal organs (i.e., ruptured spleen), hematologic disturbances, anticoagulation medications, arteriovenous malformations, renal artery aneurysm, tumors (i.e., angiomyolipoma and hypernephroma), parenchymal diseases, renal cystic diseases, renal vein thrombosis and adrenal hemorrhage. About 5-10% of renal angiomyolipomas can be presented with shock and loin pain.[10,12] The speed with which the workup proceeds is highly dependent on careful patient monitoring. CT scan and magnetic resonance imaging have shown to be helpful in diagnosis of renal angiomyolipoma. There also are reported cases of antepartum diagnosis of tuberous sclerosis fetus in utero.[15,16]

Some authors believe that the increased blood volume and mechanical obstruction of enlarged uterus during pregnancy might have some relationship with the spontaneous rupture of these renal tumors.[2,3,5-7] Our case may also be precipitated by shooting blood pressure during convulsion. But there appears to be no relationship between rupture of an angiomyolipoma and the patients age,parity or stage of gestation.[7] Even in the non-pregnant, most of renal ruptures occur in kidneys with pathology.[3]

Once it is recognized that a renal rupture has occurredin a pregnant women , rapid exploration to evaluate and control hemorrhage is mandatory. In literatures reviewed by Middleton,[3] all cases of spontaneous renal ruptures in pregnancy led to nephrectomy. But for nonpregnant one, treatment should depend on size and symptoms of patients. If a renal angiomyolipoma is larger than 3.5 or 4 cm., it should be treated whether conservative or not.[13,17] Almost all literatures say that renal angiomyolipoma should be conservative management, but in the situation that severely damaged kidney is found, like our patient, the best way to do is total nephrectomy. In review litera-

tures of Oesterling et al,[13] they suggested nephrectomy in patients with uncontrollable, life-threatening hemorrhage or when a symptomatic angiomyolipoma consumes an entire renal unit. It also may be necessary in patients with a renal cell carcinoma coexisting in the same kidney ; several such cases have been reported.[18] Because some renal angiomyolipomas show progression and this patient still has renal hamartomas on the right kidney, so we recommend ultrasonographic followup every 6 months.[10]

The neonate was hypoxia due to maternal hypotension and then developed septicemia later. We think that early diagnosis and treatment in both mother and infant may prevent this catathrophic event.

Summary

A case of bilateral renal angiomyolipomas in a 22-year-old,34-weeks pregnant woman is illustrated. Her underlying diseases were tuberous sclerosis and atrial septal defect (which was surgically corrected). Her presenting symptoms were convulsion, left upper abdominal pain with hypotension. Cesarean section, tubal resections, total left nephrectomy and partial left adrenectomy were done. The patients postoperative course was benign but unfortunately, her child died eight days later from sepsis. Earlier diagnosis and treatment may prevent maternal and perinatal morbidity and possibly mortality.

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