44526 - EMERGENCY NEPHRECTOMY DUE TO RUPTURED RENAL ANGIOMYOLIPOMA IN A PATIENT WITH PULMONARY LYMPHANGIOLIOMYOMATOSIS

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PURPOSE:

Pulmonary lymphangioliomyomatosis (PLAM) is a rare disease often associated with renal angiomyolipoma. We report a patient presented for emergency laparotomy due to ruptured renal angiomyolipoma seven weeks after uneventful LSCS.

CLINICAL FEATURES:

Patient consent was obtained for publication.

A 32-year-old woman was admitted to the A&E department with sudden and severe abdominal pain. On examination, she was in severe hemorrhagic shock with distended abdomen. The patient was able to inform that she was seven weeks post LSCS with a history of PLAM and bilateral RAML. She underwent an emergency laparotomy, which revealed a large left sided retroperitoneal hematoma in association with an actively bleeding left RAML. Nephrectomy was performed.

She received multiple blood and blood component therapy. She was transferred to the ICU. Her trachea was successfully extubated after few hours and all biochemical investigations normalized within the day. She was transferred to the surgical ward on the second postoperative day and discharged from the hospital on 9th day. CT scan of chest, performed during postoperative period demonstrated numerous small cystic air filled spaces scattered throughout both lungs (figure). Retrospectively her past medical history includes embolization of the left sided renal AML in early 2001. Pulmonary function tests and ultrasonography of abdomen to evaluate RAML size were frequently performed. Her PFTs and renal AML were stable at 28 weeks during pregnancy although there is no record of any further follow up. She attended chest clinic 6 weeks post delivery (1 week before this admission) where her PFT's remained stable although her renal AML were not evaluated at this point.

CONCLUSION:

PLAM is a rare debilitating disease of unknown etiology and is characterized by marked hamartomatous smooth muscle proliferation along the lymphatic system and in both the peribronchial and perivascular regions. In the UK the estimated incidence is one per 1.1 million populations (1). PLAM is predominantly a disease of women in their reproductive years. PLAM may present with haemo or chyloptysis (2) and as obstructive or restrictive airway diseases. A chest CT is essential in the diagnosis of PLAM, the results typically demonstrating the presence of thin-walled cysts spread in relative uniformity throughout the lungs (figure). Up to 47% of PLAM patients may have a

RAML. Regular radiological evaluation of RAML during the third trimester and postpartum period is recommended to determine increase in size. It can increase in size during this period and rupture can cause life threatening hemorrhage. In the elective setting (a) PFT in the preoperative period (b) prevention of pneumothorax with mechanical ventilation and during extubation and (c) measures to prevent chest infection are important implications for anesthesiologist.

1.Thorax. 2000 55:1052-7.

2.Am J of Med Sci. 2001 321:17-25.