Acute retroperitoneal hematoma mimicking intraperitoneal hemorrhage caused by renal angiomyolipomas associated with tuberous sclerosis

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Abstract
Tuberous sclerosis complex (TSC) is known as a neurological and dermatological disorder, but its renal manifestations are frequent and may cause death. We report a patient with TSC who showed bilateral multiple renal angiomyolipomas. Spontaneous retroperitoneal hematoma caused by renal angiomyolipoma that mimicking intraperitoneal hemorrhage was diagnosed in her clinical follow-up. We performed right total nephrectomy for controlling hemorrhage, but she died due to poor respiratory function, in fifteenth day postoperatively.

Keywords: Angiomyolipoma; hemorrhage; tuberous sclerosis

Introduction
Tuberous sclerosis complex (TSC) is an autosomal dominant disorder that affects approximately 1/6000 individuals. This disease is characterized by the widespread development of benign tumors termed hamartomas, frequently leading to skin rashes, seizures and mental retardation (1). The commonest manifestation in the kidney is angiomyolipoma. The angiomyolipomas associated with TSC usually show multiple, bilateral and symptomatic natures compared with sporadic angiomyolipomas (2).

We report a patient with TSC who showed spontaneous retroperitoneal hematoma caused by renal angiomyolipoma that mimicking intraperitoneal hemorrhage.

Case
A 40-year-old woman presented to our emergency department with right spontaneous pneumothorax. Her past medical history included epilepsy, and she was taking carbamazepine. Diffused bullous emphysema had been observed, and pleurodesis was carried out. In her clinical follow-up, two episodes of spontaneous pneumothorax was observed in the following twenty days.

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Discussion

This patient showed spontaneous retroperitoneal hematoma caused by angiomyolipoma that mimicking intraperitoneal hemorrhage. While bleeding and retroperitoneal hematoma are not rare conditions, intraperitoneal rupture of retroperitoneal hematoma is the first condition according to our knowledge.

Oesterling and co-workers (3) reviewed more than 600 cases of renal angiomyolipomas and concluded that lesions less than 4 cm had a low potential for hemorrhage or growth and recommended conservative management.

Van Baal et al. (2) reexamined 20 patients after 5 years and found that 20% had enlarged tumors, 35% had renal hemorrhage requiring hospitalization, 10% required nephrectomy and 5% died.

Harabayashi et al. (4) were retrospectively analyzed 12 patients with TSC, and they concluded that immediate arterial embolisation is mandatory for symptomatic tumors and that annual follow-up is sufficient for asymptomatic tumors, especially for those showing slow growth or in patients after decade three.

In a case, Kushwaha et al. (5) were reported that multifocal renal angiomyolipoma presenting as massive intraabdominal hemorrhage. During surgery large right-sided renal mass was found measuring 15x15x10 cm and four hundred milliliter of blood was present in peritoneum. In another case, Granata et al. (6) were reported that spontaneous retroperitoneal hemorrhage due to massive rupture of renal angiomyolipoma. It was an unusual onset of TSC, and was treated with nephrectomy (6). Tan and Toh (7) were reported life threatening intraperitoneal hypertension, the nephrectomy was carried out after 48 hour of optimization in the Surgical Intensive Care Unit (7).

Although traditionally recognized as a neurological and dermatological disorder, renal disease is a frequent manifestation of TSC and the leading cause of death in adults with TSC (8). This patient’s cause of death was not directly renal, but it may be accelerated by acute hemorrhage, massive transfusion and operative distress. TSC patients with large renal angiomyolipomas should be evaluated in their routine follow-up with cautious for potential risks of hemorrhage.

References