Spontaneous Nontraumatic Perirenal Hematoma: A Case Report

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Spontaneous perirenal hematoma is a rare medical emergency and is difficult to diagnose early. It mostly occurs secondary to rupture of a renal tumor (61.5%) and vascular disease (17%). We report a case of 28 year-old man without a history of systemic disease or trauma, who had progressive severe right flank pain without hematuria or hydronephrosis for one day. He was initially diagnosed as having muscle pain, and then a hepatic tumor with intra-tumor bleeding was suspected on follow-up abdominal sonography. Finally, a right renal angiomyolipoma rupture with perirenal hematoma was proved pathologically after surgical exploration. The patient was discharged without complications after enucleation of the angiomyolipoma in the right kidney.

Key words: spontaneous perirenal hematoma, non-traumatic, angiomyolipoma

Introduction

Spontaneous perirenal hematoma (SPH) was reported by Wunderlich in 1856, who defined an apoplexy of the renal capsule as the underlying condition. Many reports indicate it is secondary to rupture in renal parenchymal disease. SPH is a rare emergency condition that requires early diagnosis and results in high mortality if surgical intervention is delayed. It always presents clinically as acute abdomen or flank pain in the emergency department, which makes it a challenging diagnosis for most first-line physicians and therefore early appropriate treatment is difficult. Herein, we present a patient with progressive perirenal hemorrhage in the emergency department.

Case Report

A 28 year-old man presented to our emergency department (ER) complaining of severe right flank pain for one day. He denied previous systemic disease or trauma. His vital signs were stable on arrival as follows: blood pressure, 109/75 mmHg; heart rate, 71 beats/min; and respiratory rate 20/min. Physical examination revealed right flank knocking pain and a soft abdomen without a palpable mass or tenderness. Radiography revealed no abnormalities. The urinalysis and blood test results were all within normal limits except for a serum white blood cell count of 16350/μL. Initial bedside sonographic examination in the ER showed no hydronephrosis or ascites in the Morrison pouch. Therefore, anagelsics were prescribed under the impression of possible renal colic or muscle pain. The patient was kept under observation in the ER. However, the symptoms did not improve and the right flank pain became intolerable. A peritoneal sign on the right side of the abdomen was noted over the following 3 hours, and his blood
pressure dropped to 82/50 mmHg. Therefore, the abdominal sonographic examination was repeated, revealing a 10 cm. hypoechoic mass over the right lobe, with a 6 cm. hyperechoic area inside of it (Fig. 1A), and another 7 cm. heterogeneous mass below the larger tumor (Fig. 1B). Hepatic tumor with intra-tumor hemorrhage was then impressed. A contrast abdominal CT scan was done immediately to confirm the diagnosis. It showed a more than 10×21 cm hyperdense hematoma in the right retroperitoneum, mainly in the anterior pararenal space. There was a prominent enhanced engorged vessel within hematoma (Fig. 2). The liver was displaced superiorly and the right kidney was displaced inferiorly. The pancreas, superior mesenteric artery, and superior mesenteric vein were displaced to the right and the inferior vena cava was also compressed. SPH with hypovolemic shock with possible renal tumor rupture was impressed.

Emergency surgical exploration was done and a fragile yellowish tumor with marked vessels engorgement, especially at the tumor base, was found. The retroperitoneum was full of clots, and a hematoma, which compressed the right renal parenchyma, was noted. The renal tumor was enucleated and the right renal parenchyma was preserved. The pathology showed a picture of an angiomyolipoma with variable amounts of cells with blunt-end nuclei. There were an increased number of blood vessels which were dilated, and displayed continuous transition to the surrounding smooth muscle cells, and a few of lipocytes.

The patient was discharged 12 days later. Renal sonography 2 weeks later revealed only an irregular right renal contour from the right enucleation of the angiomyolipoma.

**Discussion**

The etiologies of spontaneous perirenal hematomata are mainly secondary to the rupture of renal tumors (61.5%), predominantly clear cell carcinoma, followed by angiomyolipoma and renal vascular disease (17%), such as renal infarction. Other causes include infectious diseases (2.4%), such as renal abscess, acute or chronic nephritis, and tuberculosis and miscellaneous (12.7%) causes such as polyarteritis nodosa, polycystic kidney, stricture of the pelvis or ureter, blood dyscrasias and idiopathic causes (6.7%) (2,3,5,12,13).

The SPH in our case was secondary to a ruptured angiomyolipoma, which was proved by
Fig. 2 A prominent enhanced engorged vessel (arrow) within the hematoma

pathological findings. An angiomyolipoma usually appears as part of a tuberous sclerosis complex and sporadically in individuals who have no clinical features of that entity. It is uncommon and the current prevalence is 1 in 5,000 in tuberous sclerosis patients and 1 in 15,000 individuals. Pathologically, classic angiomyolipoma is composed of mature fat cells, with thick walled, low elastin vascular tissue surrounded by a cuff of normal smooth muscle with small and regular nuclei. Occasionally the smooth muscle has atypical nuclei with nucleoli. An angiomyolipoma is usually benign, but extremely rare cases have sarcomatous changes with local and distant metastasis. These metastatic tumors are called epitheloid angiomyolipoma, and are found mostly in the lung (50%) and liver (75%). Histologically, staining with protein HMB-45, which is always expressed in angiomyolipoma, is used to distinguished angiomyolipoma from renal cell carcinoma.

The classic triad of angiomyolipoma in clinical presentations includes flank pain, a palpable mass and gross hematuria. But microscopic hematuria remains the most common finding. SPH usually presents with the “Lenks Triad”, consisting of acute flank pain, tenderness and symptoms of internal bleeding, and represents a clinical emergency with possible hemodynamic instability. Our patient had an atypical clinical manifestation of angiomyolipoma at ER arrival, but the typical SPH presentation was noted 3 hours later.

Acute flank pain is a common presentation in SPH, but it is also a common chief complaint in patients in the ER. It is often diagnosed as renal colic in those with hematuria and muscle sprain in those without hematuria, if vital signs are stable. Therefore, it is often misdiagnosed and it is difficult to make an early diagnosis of SPH when radiographic studies do not provide any clues and the patient is hemodynamically stable.

Preserving renal function is the primary concern in clinical management of SPH and angiomyolipoma. Some authors have proposed conservative treatment with angiographic embolization. Selective angiographic embolization is the most common management and it has the advantages of preservation of functional renal parenchyma, and the ability to embolize bleeding vessels selectively, thus circumventing the need for surgery and anesthesia. But some clinicians have proposed
radical nephrectomy in cases of unexplained spontaneous kidney ruptures due to the high rate of renal cell carcinoma (3,10,11). Others have suggested that surgical intervention is indicated only in those cases with significant hemorrhage, hematuria or other symptoms not responsive to conservative treatment, or in lesions with local tissue or vascular invasion and continued suspicion of malignancy after imaging. A complete nephrectomy is indicated if the whole kidney has been replaced by the angiomyolipoma, if a solitary sporadic angiomyolipoma tumor is near the hilum, if there is suspicion of malignancy that requires complete or radical nephrectomy for adequate margins, or if angioembolization is not available or unable to control the bleeding (6).

In order to avoid unnecessary complications such as nephrectomy, we stress the importance of both identification of those at high risk of SPH and early diagnosis of SPH. Then early appropriate treatment can be given before the patient becomes hemodynamically unstable.

A clear medical history is the first step in identifying high risk patients and diagnosing SPH. Non-trauma patient who present with sudden flank pain with or without hematuria, should always raise suspicion of SPH. If the patient has a history of tuberous sclerosis or clinically demonstrates flank pain, a palpable tender mass and gross or microscopic hematuria, angiomyolipoma should be considered (6). Inappropriate hemoglobin and flank pain, associated a past history of unexplained fever or unexplained gastrointestinal symptoms suggests inflammatory vasculopathy (2).

Bedside abdominal sonography should always be done in patients at high risk for SPH and those with flank pain. Sonography is effective in the identification of renal and perirenal fluid collection, although it may be difficult to differentiate between tumors and or abscesses (12). However, sonography relies operator skill and experience, and ability to detect tiny renal abnormalities. An inexperienced operator was the only explanation for failure to detect abnormality of the kidney on the initial abdominal sonographic examination in our case. In acute diseased kidneys, the renal size is usually measured larger than 10.2 cm in diameter on abdominal sonography. A bulging irregular renal shape may be a renal tumor or cystic lesion, and a depressed irregular edge may be renal infarction. The central echogenic complex may be hypoechoic or nonechogenic in renal abscesses and cystic lesions, and an interrupted echogenic or hypoechoic lesion may be carcinoma (7).

In comparing the echogenicity of the renal tumor and renal parenchyma, it is important to remember that renal adenocarcinoma and malignancy are rarely echogenic. But an angiomyolipoma usually has high attenuation and backscatter coefficients due to acoustic impedance differences between fat and smooth muscle components (8). Some studies have reported angiomyolipomas larger then 3.5-4cm are at high risk of severe hemorrhage (5,9). Focal hemorrhage, and myomatous or vascular areas may be hypoechoic in appearance and cause nonhomogenicity resembling malignancy (8).

In cases in which SPH is already suspected, abdominal sonography as a tool of diagnosis is difficult and challenging and can give the wrong impression with tumors (5). Some reports indicate that SPH may be misdiagnosed as a renal tumor or abscess (1). A possible explanation for the misdiagnosis by sonography in our case is that the hematoma may have been seen as hypoechoic mass lesion large enough to blur the edge of the liver and was then misdiagnosed as part of an abnormal liver parenchyma, and therefore considered a hepatic tumor. The hyperechoic lesion was an active hemorrhage within the hematoma that was seen as another tumor with intra-tumor bleeding. The heterogenic mass lesion may have been a pararenal
hematoma mixed with pararenal fat tissue.

We suggest contrast enhanced CT scanning is the most useful tool to confirm the diagnosis of SPH. Also, patients presenting with unexplained flank pain who are identified as high risk for SPH on abdominal sonography, should have contrast enhanced CT scanning.

In conclusion, patients presenting with incongruent laboratory results and clinical symptom should always raise suspicion of SPH. We emphasize that a clear medical history and serial abdominal sonography follow-up of suspected cases are key steps to the early detection of SPH. CT scanning should be used to confirm the diagnosis.

References

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自發性非外傷腎週邊血腫：一例個案報告

馮卓超 張道平 石福來 胡彼得

自發性腎週邊血腫是一罕見醫療急症且不易早期診斷。它最常是因腎腫瘤破裂(61.5%)和血管疾病(17%)。我們報告一案例無任何過去系統性疾病及外傷病史的28歲男性病患，因發生右側腰部嚴重疼痛一天，無血尿、也無腎水腫的合併症。他被初步診斷為肌肉疼痛；隨後又被追蹤的腹部超音波診斷為疑似肝腫瘤並腫瘤內出血。最後經由外科探索術及病理切片證實後，確認是因右腎血管脂肪肌瘤破裂合併自發性腎週邊血腫所導致。該病患經摘除右腎血管脂肪瘤後，在無併發症情形下出院。

關鍵詞：自發性腎週邊血腫，非外傷，血管脂肪肌瘤