E-ISSN: 2616-3470
P-ISSN: 2616-3462

www.surgeryscience.com
2020; 4(1): 185-187
Received: 01-11-2019
Accepted: 03-12-2019

Sushant Deole
Department of Urology, JN Medical College, KLE Academy of Higher Education & Research (Deemed-to-be-University), JNMC Campus, Belagavi, Karnataka, India

Vikram Prabha
Department of Urology, JN Medical College, KLE Academy of Higher Education & Research (Deemed-to-be-University), JNMC Campus, Belagavi, Karnataka, India

Nerli RB
Department of Urology, JN Medical College, KLE Academy of Higher Education & Research (Deemed-to-be-University), JNMC Campus, Belagavi, Karnataka, India

Priyeshkumar Patel
Department of Urology, JN Medical College, KLE Academy of Higher Education & Research (Deemed-to-be-University), JNMC Campus, Belagavi, Karnataka, India

Shridhar C Ghagane
Department of Urology, KLES Kidney Foundation, KLES Dr. Prabhakar Kore Hospital & Medical Research Centre, Belagavi, Karnataka, India

Corresponding Author:
Sushant Deole
Department of Urology, JN Medical College, KLE Academy of Higher Education & Research (Deemed-to-be-University), JNMC Campus, Belagavi, Karnataka, India

Emergent angio-embolization with elective nephrectomy in ruptured Angiomyolipoma: A case report

Sushant Deole, Vikram Prabha, Nerli RB, Priyeshkumar Patel and Shridhar C Ghagane

DOI: https://doi.org/10.33545/surgery.2020.v4.i1d.330

Abstract
Renal Angiomyolipoma is a common tumour in clinical practice and mostly follows a benign course. Angiomyolipoma is the most common renal neoplasm associated with spontaneous retroperitoneal haemorrhage due to rupture of micro and macro-aneurysms. Urgent trans-catheter angio-embolization of the bleeding vessel is very effective in patients suffering from life threatening hemorrhage secondary to rupture of Angiomyolipoma. Elective surgery is usually indicated to avoid re-bleeding or abscess formation. We report a case of Spontaneous Rupture of Angiomyolipoma of Right kidney in 63 years old female. The female underwent emergent Trans-arterial Embolization followed by elective nephrectomy.

Keywords: Angiomyolipoma, nephrectomy, Wunderlich's syndrome and Lenk's triad

Introduction
Renal Angiomyolipoma was originally described in 1900 by Grawitz. It is a benign neoplasm with definite radiological and histological characteristics [1, 2]. It consists of thick-walled poorly organized blood vessels, smooth muscle, and varying levels of mature adipose tissue [3]. Angiomyolipoma mostly occurs in kidneys but can be found in uterus, fallopian tubes, spleen and liver [4, 5]. Renal AML are usually sporadic and are found in association with Tuberous Sclerosis and lymphangioleiomyomatosis like genetic diseases [6]. Renal AML has male to female ratio of 1:2 and has hormonal component to tumor growth [5, 7]. Retroperitoneal hemorrhage and compression of the tumor on kidney and other organs are the major complications of Renal AML [5]. Most patients present in 5th or 6th decade of life. Most of the patients are asymptomatic and diagnosed incidentally [5]. Lenk's triad is classical presentation of Renal AML – Palpable tender mass, flank pain and gross hematuria [8]. Ten percent of cases present with hypovolemic shock due to retro-peritoneal hemorrhage known as Wunderlich's syndrome [8, 9]. Presence of fat confirmed on CT scanning by -20 Hounsfield units or less in a renal lesion is diagnostic of AML [10]. Most patients are asymptomatic and smaller lesions are managed conservatively with regular Ultrasonography follow up. Lesions >8cm are usually symptomatic and are prone for spontaneous or traumatic rupture. We present a case of Spontaneous Rupture of Angiomyolipoma of Right kidney in 63 years old female who underwent emergent Trans-arterial Embolization followed by elective nephrectomy

Case Report
A 63-year-old female was brought to the urological services of the hospital with complaints of sudden onset severe right flank pain with distension of abdomen. She had severe abdominal tenderness with guarding. She had tachycardia, BP was normal. A lump approximately 10*10 cm was palpable in right lumbar, right hypochondriac region. She was anemic (Hb – 5.6 gm%). Serum creatinine was 0.82 mg%. Computed tomography (CT) showed Right kidney replaced by heterogeneously enhancing ill defined mixed density area of fat and blood with perinephric edema and ill-defined hypo and hyper dense collections. The extension of collection was into peri-nephric region, retro peritoneum, right para-colic gutter, supra-renal and hepato-renal pouch. (Figure 1 a and b) A diagnosis of Ruptured Angiomyolipoma of Right Kidney with retroperitoneal hematoma and hemoperitoneum was made. She underwent emergent Trans-arterial Embolization in view with 3 Packed Cell Transfusions (Figure 2 a and b).

Post TAE and blood transfusions, her Hb improved to 10.4 gm%. Pain did not subside. She was taken for Elective Nephrectomy 3 days post TAE. The entire kidney was replaced by tumor with extensive organized hematoma (Figure 3 a & b). The female had an uneventful post-operative period and was discharged 7 days post-operatively. The histopathology examination revealed Angiomyolipoma.

Discussion

The management of AML is multimodal and the type of treatment is individualized. Most patients with tumor < 4cm are usually asymptomatic and are managed conservatively. Radio-frequency ablation can be done for small tumors in patients with solitary kidney, risk of renal dysfunction following partial nephrectomy or in elderly patients. Tumors >8 cm are symptomatic and at risk for spontaneous / traumatic rupture. Renal artery embolization is the first line of management for bleeding AML. This is nowadays being used as preventive treatment for patients with risk of bleeding [11]. Advantages of embolization over surgery include low complication rate, less trauma [12], acceptable short term outcome [13, 14] and preservation of renal function [15].

Ewalt *et al* found that transcatheter embolization of large AML is minimally invasive, prevents hemorrhage and preserves renal function [16]. Wang *et al*, respectively, reviewed 46 patients who underwent super-selective renal artery embolization (SRAE) for renal hemorrhage, and indicated that SRAE is an effective and minimally invasive method for the control of renal hemorrhage [17]. However, embolization also has its limitations.

Tumor shrinkage which occurs in most patients after embolization is not a reliable exclusion criterion for recurrent hemorrhage [18, 19]. Boorjian *et al*. pointed out that embolization had higher risk of relapse and recurrent bleeding compared with partial nephrectomy [20]. Kara *et al*. recommended Partial nephrectomy in for patients with high risk of rupture [21]. The complication rate of embolization is 10% and the most common complication is abscess formation (5%). Post embolization re-bleeding, need for re-embolization or need for surgical intervention are noted after embolization. Post-embolization need for surgical intervention is usually due to persistent of symptoms and recurrent bleeding [22]. Adhesions usually form within 72 hours after injury and are more extensive from 10 days to 2 weeks [23]. So surgery within 1 week after embolization seems to be a better option. The advantages of early surgery include less surgical and anaesthetic risk. Patients are hemodynamically stable after embolization, less tissue adhesion, no need for renal pedicle control and decreased bleeding during procedure. The female patient underwent emergency renal artery embolization with Blood transfusions. After hemodynamic stability, the patient underwent elective total nephrectomy.

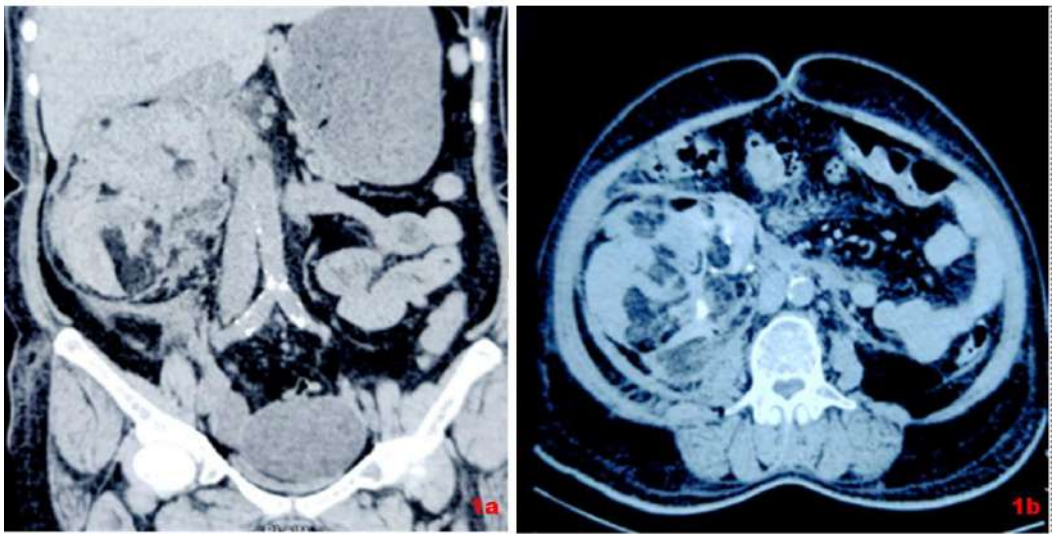

Fig 1a & b: Computed tomography (CT) showing a Right kidney replaced by heterogeneously enhancing ill defined mixed density area of fat and blood with perinephric oedema and ill-defined hypo and hyperdense collections. The extension of collection was into peri-nephric region, retroperitoneum, right para-colic gutter, supra-renal and hepato-renal pouch

Fig 2a: Angiography of right renal artery showing contrast extravasation of dye due to rupture of AML.

Fig 2b: Post embolisation Angioography showing no extravasation of dye.

Fig 3a: Intra-operative photograph showing sub-capsular organized hematoma and tumor.

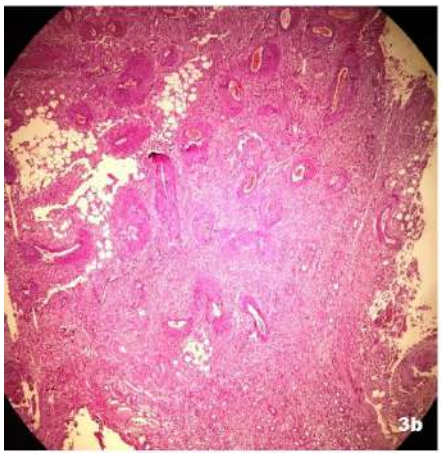

Fig 3b: Histomicrograph (H & E) showing adipose tissue, blood vessels and muscle fibres normal kidney tissue confirming diagnosis of Angiomyolipoma.

Conflict of Interest: The authors declare as None.

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