



## A rare presentation of right adrenal angiomyolipoma with accelerated hypertension: A case report

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### ABSTRACT

Right adrenal angiomyolipoma with accelerated hypertension is usually an incidentaloma and infrequent finding, and only adrenal angiomyolipomas are reported but not with tumor-associated accelerated hypertension. In this case, a 45-year-old male patient presented with the typical symptoms of a headache, neck pain and also giddiness in the last four days. Right adrenal angiomyolipoma diagnosed by using CT and USG scans of the abdomen, and majorly by histopathology. Mostly the management of this case requires surgery, but the preoperative and postoperative management and monitoring of high blood pressure is the utmost importance due to accelerated hypertension. Laparoscopic excision of adrenal tumor and adrenalectomy was done under general anesthesia. Postoperative recovery was uneventful, and patient blood pressure became normal.

**Key words:** Adrenal angiomyolipoma, incidentaloma, adrenalectomy, accelerated hypertension

### Introduction

Angiomyolipomas of the adrenal glands are rare benign mesenchymal neoplasms with an incidence rate of 0.3-3%. This tumor poses a diagnostic difficulty to pathologists as it can resemble a variety of the other tumors found in the adrenal gland and also extra-renal [1,2]. Adrenal tumors can occur separately or as a part of systemic syndromes. Asymptomatic and incidentally found adrenal angiomyolipomas are isolated tumors, most of them (about 80% of the lesions) are sporadic, and they are often solitary. Compared to males, the average frequency of appearance in 40 years of age, is four times higher in females [3].

To best of our knowledge, only 16 patients diag-

nosed with adrenal angiomyolipoma have been reported in the English literature [1,2,4–15] (Table 1). In this case, we reported a rare case of right adrenal angiomyolipoma with accelerated hypertension. The adrenal angiomyolipoma is not an unusual tumor, however, as far as we know this is the first case presented with tumor-associated accelerated hypertension.

### Case Report

A 45-years-old male patient presented with chief complaints of a headache, neck pain, and burning micturition in the last four days. He was fainted due to giddiness on the day of admission with the past six years history of hypertension on irregular medication and regular alcoholic. On general examination, he was afe-

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**Table 1.** The summary of the sixteen patients diagnosed with adrenal angiomyolipoma from the English literature (modified from Ghimire et al. 2017 [15] and updated by authors).

Pt. No.	References	Age/Sex (yrs.)	Presentation	Side	Size (cm)/Imaging procedures
1	Lam et al., 2001[1]	46/F	Incidental finding	L	8 cm diameter (CT)
2*	Lam et al., 2001 [1]	20/M	Incidental finding at nephrectomy	L	0.2 cm diameter on nephrectomy specimen (Smallest)
3	Li et al., 2015 [4]	53/M	L upper abdominal pain	L	9×6 (US)
4	Godara et al., 2007 [2]	45/F	Epigastric discomfort	L	15×12 (US)
5	Hu et al., 2012 [5]	55/F	R upper abdominal pain	R	15×16 (US) (Largest)
6**	Sutter et al., 2007 [6]	32/F	Diffuse abdominal pain	R	6 cm in maximum transverse diameter (CT)
7	Gupta et al., 2011 [7]	42/M	Upper abdominal pain	R	8×5.5×4.5 (CT)
8	Goswami et al., 2014 [8]	43/F	R loin pain	R	9.5×8×2 (MRI)
9	Yener et al., 2011 [9]	45/F	R subcostal pain	R	5×6 (US)
10	Monowarul et al., 2012 [10]	37/M	Generalized jerking discomfort and weakness	R	5.4×4.5 (US)
11*	Elsayes et al., 2005 [11]	49/F	Referred for MRI	R	12.2×9.8×6.8 (MRI)
12	Chee et al., 2010 [12]	61/M	L loin pain	L	10×10×10.1 (CT)
13	Hafeez et al., 2013 [13]	72/F	R upper quadrant pain	R	9×8.9 (CT)
14	Kwazneski et al., 2016 [14]	65/F	R upper abdominal pain	R	11.3×9.4 (US)
15	Obin Ghimire et al., 2017 [15]	36/M	Incidental finding	R	5.2×4.2×3.1 (CT)
16	Obin Ghimire et al., 2017 [15]	61/F	Incidental finding	R	8.6×9.5×8.1 (CT)
17***	Present Case	45/M	Incidental finding and associated with Accelerated Hypertension	R	6.67×6.29 cm (CT), 6.17×6.72 cm (USG)

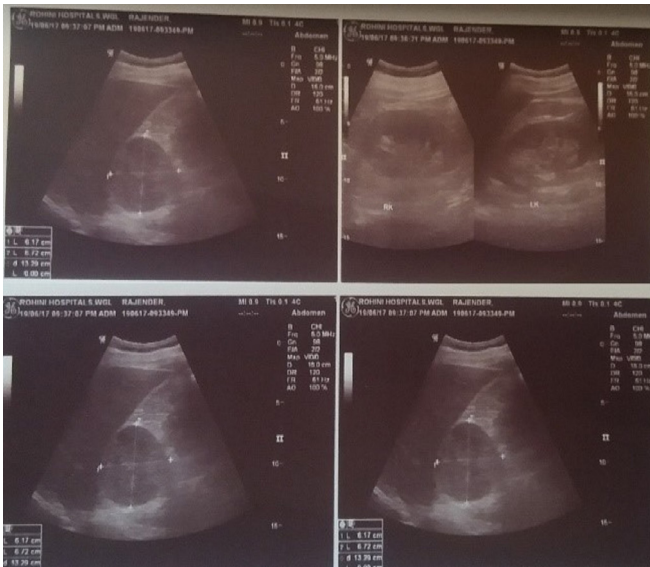
\*Associated with tuberous sclerosis; \*\*Associated with lymphangioliomyomatosis; \*\*\*Associated with Accelerated Hypertension; F – female; M – male; L – left; R – right; US – ultrasound; CT – computed tomography; MRI – magnetic resonance imaging.

brile, hypertensive (170/100mmHg), pulse rate was found to be normal. The patient was advised for complete blood count (CBC), random blood glucose, blood urea, serum creatinine, liver function tests (LFTs) and serum electrolytes which are found to be normal (Table 2). 2D echo Doppler showed concentric left ventricular hypertrophy and Grade 1 diastolic dysfunction. Ultrasound (USG) (Figure 1) and Computed tomography (CT) (Figure 2) scan of the abdomen showed a mass on the right adrenal gland. On admission patient was treated with Olmesartan 20 mg once a day, Clonidine 100 mg twice a day, Rosuvastatin 10 mg at bedtime, Nifedipine 5mg administer when required, Aspirin 150mg once a day and Clopidogrel 75mg once a day.

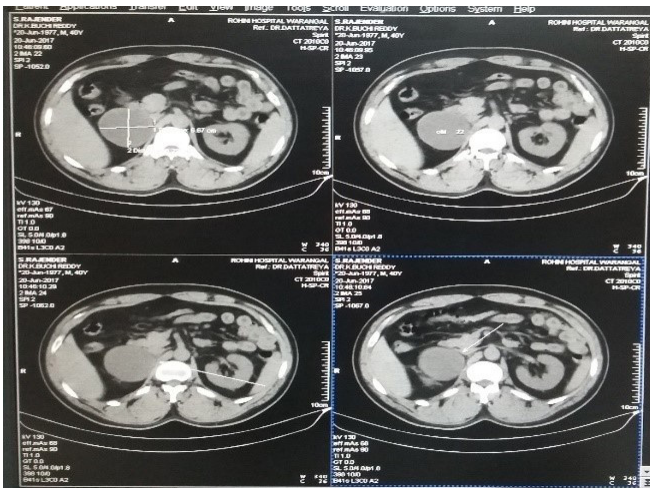
**Table 2.** Abnormal Serum electrolyte levels on Day-3.

Serum electrolyte levels	Day-1	Day-3
Sodium (mEq/L)	133	122 ↓
Potassium (mEq/L)	3.4	3.6
Chloride (mEq/L)	103	88 ↓

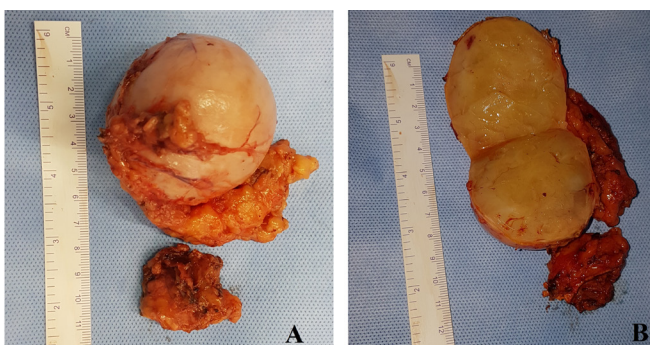
On day-2, accelerated hypertension (170/100 mmHg) was found and advised with plasma metanephrine (metabolites of epinephrine), and it was found in normal, i.e., 47.6 pg/ml (normal range <65 pg/ml) and decided to continue the same medications along with Cilnidipine 10 mg once a day. On day-3, blood pressure was 150/110mmHg, pulse rate was 82/min, and serum sodium and chloride levels were reduced (Table 2). Thus, the diagnosis was made to be accelerated hypertension, hyponatremia and pheochromocytoma. Intravenous infusion of 3% Sodium chloride in normal saline (3mg/100ml) and Metoprolol 50 mg once a day was added and continued the same medications. On day-4, vitals were found to be normal, blood pressure was 130/90 mmHg and advised with Cortisol estimation to evaluate the Cushing's syndrome. However, it was found to normal, i.e., 11.15 µg/dl (normal range: 4.8-19.5 µg/dl) and same medications were continued. On day-5, blood pressure was elevated again, i.e., 170/100 mmHg and metoprolol was stopped, and other medications were continued.



**Figure 1.** Ultrasound scan of the abdomen (USG) showed well defined isoechoic lesion (6.17x6.72 cm) near the upper pole of the right kidney.



**Figure 2.** Computed tomography (CT) of the abdomen showed heterogeneously enhancing solid cystic lesion arising from the right adrenal gland could be a degenerative adenoma (6.67x6.29 cm).



**Figure 3.** (A) Gross examination of right adrenalectomy specimen with attached peritoneum with grey, white globular mass of tissue measuring 5.5 x 5 x 3 cm. in dimension. (B) Cut section of right adrenalectomy.

The patient was referred to the higher center for surgery with the diagnosis of pheochromocytoma and accelerated hypertension. The prescription drugs during his discharge were Olmesartan 20 mg once a day,

Clonidine 100 mg twice a day, Rosuvastatin 10 mg at bedtime, Nifedipine 5 mg (if required), Aspirin 150 mg once a day, Clopidogrel 75 mg once a day, Cilnidipine 10mg once a day, Metoprolol 50mg once a day and infusion of 3% Sodium chloride in normal saline (3mg/100ml, IV) three times a day.

### Operation Procedure

The patient was admitted one day before surgery for preparation and assessment. Adrenalectomy was done under general anesthesia; all precautions and preparations were taken for managing the preoperative hypertensive crisis.

We used a transperitoneal lateral approach by lying the patient at his right side. After the patient was rotated, four ports were placed at a distance of 5-10 cm from the costal arch. The first 10/12-mm trocar, used for videoendoscopic control, was routinely inserted through a mini-laparotomy incision between the midclavicular and anterior axillary line, depending on the patient's habitus and shape of the thorax, always laterally into the edge of the rectus abdominis muscle. The remaining three trocars were typically inserted under direct vision after establishing a CO<sub>2</sub> pneumoperitoneum with the abdomen inflated to the pressure of 12-14 mm Hg. Two 10-mm trocars were used to insert a laparoscopic hook, a clip applicator, and other surgical instruments, and one 5-mm trocar was used for auxiliary instruments.

After routine diagnostic inspection of the peritoneal cavity, the immediate operative area was exposed by elevating the right hepatic lobe and transection of the triangular ligament. Through the translucent parietal peritoneum, the adrenal gland was easily identified because of its characteristic color, shape, and location. Next, the retroperitoneal space was opened, and dissection of the adrenal gland with the involving tumor was begun at a low coagulation power, which allowed the maintenance of hemostasis. Exposure of the operative area was maintained by copious irrigation with saline and laparoscopic suction. The adrenal gland and its tumor were totally excised (Figure 3) and hemostasis maintained. Finally, the gland and its tumor placed in a plastic bag with morcellation. The laparoscopic surgery was completed uneventfully, and patient blood pressure became normal.

### Surgical Pathology

The gross examination of right adrenalectomy specimen with attached peritoneum was a grey, white globular mass of tissue measuring 5.5 x 5 x 3 cm in greatest dimensions (Figure 3). The external surface of the specimen was smooth and attached peritoneum measuring was 6.5 cm in length. The microscopic examination showed a well-encapsulated tumor, and the tumor cells were arranged in sheets with perivascular distribution. Perivascular epithelioid cells, hypocellular and myxoid areas were seen. Adipose tissue was scattered in peripheral areas. Peripheral compressed adrenal tissue was seen. Mitosis, nuclear atypia, and necrosis were not present. (Immunohistochemistry (IHC) with Human Melanoma Black-45 (HMB)).

### Discussion

In the general population, the incidence rate of angiomyolipoma is about 0.13% and twice in females compared to males [16]. Typical symptoms of angiomyolipoma are back/flank pain and hematuria. Retroperitoneal hemorrhage of the tumor due to friable blood vessels is the more drastic presentation of the angiomyolipoma. When the tumor is symptomatic, to achieve hemostasis and prevent the further complications, the treatment options include tumor excision, angioembolization, or cryotherapy. The patients who have small or asymptomatic lesions need active surveillance with the typical recommendation of serial ultrasounds [13]. The retrospective studies show that at the time of autopsy, about 91% of the pheochromocytomas had a history of "typical" symptoms include headaches, palpitations, and sweating. Atypical symptoms include abdominal pain, nausea, vomiting, dyspnea and 61% of the patients had a history of hypertension [17]. In this case, the patient admitted due to malignant or accelerated hypertension (170/100 mmHg) with the typical symptoms of a headache, neck pain and also giddiness presented for last four days. Two mechanisms are proposed when pheochromocytoma resulting in neurological injury; hypertension and vasospasm. First proposed mechanism suggest that high blood pressure may overwhelm cerebrovascular autoregulation leading to hypertensive encephalopathy during the excess release of catecholamines. The second mechanism suggests that spasm of the cerebral arteries leading to the infarction or transient

impairment of circulation during the excess release of catecholamines [18,19]. The malignant/accelerated hypertension is the main sign for pheochromocytoma. Although accelerated hypertension is present in our case, this is not a pheochromocytoma because of plasma metanephrine metabolites found in normal. Symptomatic patients with larger than 4 cm angiomyolipomas have an increased risk of bleeding and spontaneous rupture. Additionally, in large angiomyolipomas, malignancy increases with the tumor size, and the best treatment options are surgery, i.e., excision of the tumor or selective arterial embolization [8,11]. It can be challenging to distinguish angiomyolipomas from other tumors sharing fat densities. In most cases, angiomyolipomas are an incidental finding on imaging studies [20].

In the presented case, all precautions were taken, and preparations were made for managing the preoperative hypertensive crisis. Laparoscopic excision of adrenal tumor and adrenalectomy was done under general anesthesia. Postoperative recovery was uneventful, and patient blood pressure became normal.

### Conclusion

We report a rare case of right adrenal angiomyolipoma with accelerated hypertension. Majorly accelerated hypertension was seen in patients with pheochromocytoma. To best of our knowledge, a total of 16 cases of adrenal angiomyolipomas have been documented up to date. In that, 11 cases were right side tumors, and this is the 12th case diagnosed by using histopathology, CT and USG scan of the abdomen. However, as far as we know this is the first case of right adrenal angiomyolipoma in the literature presented with accelerated hypertension. The patient recovered uneventfully with stabilized blood pressure.

### Conflict of interest statement

The authors have no conflicts of interest to declare.

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