Adrenal Angiomyolipoma with Gastric Outlet Obstruction

ABSTRACT

Angiomyolipoma is a neoplasm that derives from perivascular epithelioid cells. It is a rare mesenchymal tumor, usually found in the kidney. Extrarenal angiomyolipoma is uncommon and the most common extrarenal site is the liver. Angiomyolipoma of adrenal is extremely rare, with only four cases reported in the literature. It usually presents as incidentaloma. We report a case of a 45-year-old female patient presenting with pain abdomen, nausea, and repeated episodes of vomiting. Contrast-enhanced computed tomography (CECT) abdomen revealed heterogeneous nonenhancing hypodense fatty lesion in right adrenal gland. Biochemical investigations were negative for functioning adrenal tumor. Right adrenalectomy was done with good outcome. Diagnosis was confirmed by histopathology. The patient recovered without any complications following surgery.

Keywords: Adrenal, Angiomyolipoma, Extrarenal, Gastric outlet obstruction.

INTRODUCTION

Angiomyolipoma is a benign mesenchymal tumor consisting of varying amounts of mature adipose tissue, smooth muscle, and thick-walled blood vessels. It arises from perivascular epithelioid cells and is commonly seen in the kidney. The extrarenal sites reported include the bone, colon, heart, lung, parotid gland, skin, spermatic cord, gynecologic organs, and retroperitoneum, with the most common extrarenal site being the liver. Only four cases of angiomyolipoma of the adrenals have been reported in the world literature.

CASE REPORT

A 45-year-old female presented with off and on epigastric pain with repeated episode of nonbilious vomiting. There was no history of fever, jaundice, or weight loss. Ultrasound abdomen revealed a well-defined 16 × 14 cm mass in the right retroperitoneum. The CECT abdomen defined the mass as of right adrenal origin pushing the duodenum anteriorly with inferior vena cava anteromedially. The diagnosis was made as possibility of angiomyolipoma (Fig. 1). Laboratory investigations, serum catecholamine, cortisol, and urinary vanillylmandelic acid were within normal limits. Exploratory laparotomy by right transverse incision revealed a 16×14×10 cm mass, firm in consistency, quite separate from the right kidney with no definable right adrenal gland. The whole mass was removed without rupture (Figs 2 and 3). Histopathological examination revealed mature fat cells, smooth muscle fibers, and thin-walled blood vessels with peripherally...
compressed adrenal cortical tissue suggestive of angio-
myolipoma of adrenal. Patient had an uneventful recovery
and was normal at follow-up.

DISCUSSION

Angiomyolipomas are very rare lesions, often arising
in the kidney and are a part of a group of tumors with
a diverse appearance known as PEComas (tumors
of perivascular epithelioid cell origin). Angiomyoli-
poma most commonly occurs in the kidney. The next
common site is the liver. Extrarenal angiomyolipomas
are extremely rare and have been reported in the liver,
colon, suprasellar region, small intestine, skin, intranodal,
omentum, breast, and adrenal. About half of the angio-
myolipomas are associated with tuberous sclerosis and
in these cases, they are usually multiple and bilateral.4 It
has been estimated that approximately 80% of the severe
and complete forms of tuberous sclerosis have angio-
myolipoma.5,6 On CT scan, the presence of even a small
amount of fat suggests the diagnosis of angiomyolipoma.
On magnetic resonance imaging, the typical features
of the fatty component include bright signal intensity
on nonfat suppressed images, with dropout of signal
on fat-suppressed images.1 Grossly, angiomyolipomas
appear well-circumscribed and depending on the relative
amount of adipose tissue, they range from a glistening
yellow (“fatty”) appearance to a more white-tan and firm
appearance depending on the relative amount of adipose
tissue.2 Histology typically shows a variable mixture
of mature adipocytes, thick-walled blood vessels, and
spindled and epithelioid stromal cells often radiating
out from blood vessel walls. Management should be
the same as that for any adrenal mass. Assessment of
functional status of the tumor should be done, although
all the five adrenal angiomyolipomas reported so far
were nonfunctional. Surgery is indicated if the patient
is symptomatic or the tumor is more than 6 cm since the
risk of malignancy increases with size.7 Also, the risk of
spontaneous rupture increases with size, owing to the
presence of abundant and abnormal elastin and poor
vascularity in the tumor.4 Laparoscopic adrenalectomy
is an option and had been successfully done for a 6 cm
adrenal angiomyolipoma.8

Angiomyolipoma of adrenal is extremely rare, with
only four cases reported in literature. It usually presents
as incidentaloma. We report the case of a 45-year-old
female patient presenting with pain abdomen, nausea,
and repeated episodes of vomiting. Ultrasonography
abdomen showed features of retroperitoneal tumor.
The CECT abdomen revealed heterogeneous nonen-
hancing hypodense fatty lesion in right adrenal gland.
Biochemical investigations were negative for functioning
adrenal tumor. Right adrenalectomy was done with good
outcome. Diagnosis was confirmed by histopathology.
The unusual association of adrenal angiomyolipoma with
gastric outlet obstruction has not been reported till date.

REFERENCES

Gupta A. Adrenal angiomyolipoma: a rare entity. Indian
2. Elsayes KM, Narra VR, Lewis JS Jr, Brown JJ. Magnetic reso-
nance imaging of adrenal angiomyolipoma. J Comput Assist
Tomogr 2005 Jan-Feb;29(1):80-82.
3. Lam KY, Lo CY. Adrenal lipomatous tumours: a 30 year
clinicopathological experience at a single institution. J Clin
Pathol 2001 Sep;54(9):707-712.
4. Eble JN. Angiomyolipoma of the kidney. Semin Diagn Pathol
5. Bernstein J, Robbins TO, Kissane JM. The renal lesions of
in the tuberous sclerosis complex. Pathology 2003 Dec;35(6):
505-512.
7. Grumbach MM, Biller BM, Braunstein GD, Campbell KK,
Carney JA, Godley PA, Harris EL, Lee JK, Oertel YC,
Posner MC, et al. Management of the clinically inappar-
ent adrenal mass (“incidentaloma”). Ann Intern Med 2003
8. Sutter R, Boheler A, Willmann JK. Adrenal angiomyolipoma
in lymphangioleiomyomatosis. Eur Radiol 2007 Feb;17(2):
565-566.