Bilateral Giant Adrenal Myelolipoma: A Case Report

Mandeep Phukan¹, Nitin Gupta², Rajeev T.P³, Saumar J. Baruah⁴

¹Assistant Prof., ²M.Ch Trainee, ³Prof. and Head, ⁴Professor, Department of Urology, GMCH, Guwahati, Assam, India.

Corresponding Author: Mandeep Phukan

ABSTRACT

Myelolipomas as the name itself suggests are tumors consisting of myeloid tissue and lipomatous tissue in variable proportion. Though rare, these tumors are usually confined to retroperitoneum with adrenals being the most common site. Adrenal myelolipoma (AML) is a rare benign tumour with reported incidence being less than 0.5%. Usually these tumors are detected incidentally on imaging done for some other reasons and most of the times they are unilateral and small. When small these tumors are mostly asymptomatic and hormonally inactive. Giant and bilateral adrenal myelolipomas are very rare tumors. We here report a rare case of bilateral giant adrenal myelolipoma in a 47 year old male who presented to us with history of lump and pain on left side of the abdomen. On evaluation, he had bilateral giant adrenal myelolipomas, left being 22x15x12 cm and right being 11x8.2x7cms with hypertension. Laboratory examination of adrenal function revealed normal results. Bilateral adrenalectomy was done and HPE revealed adrenal myelolipoma. Post operative period was uneventful and the patient is successfully on hormone replacement therapy since then.

Keywords: Bilateral, Giant Myelolipoma, Adrenalectomy.

INTRODUCTION

Adrenal myelolipoma is a rare benign tumour of the adrenal gland composed of adipose tissue and bone marrow elements in varied proportions. Though these tumors are mostly asymptomatic when small and usually detected incidentally on imaging studies done for some other reasons, occasionally they may present with abdominal pain because of mass effect due to large size or from spontaneous hemorrhage or rupture, more likely when predominantly composed of myeloid tissue. Incidences of adrenal myelolipoma have been reported to be between 0.08 to 0.4% and they constitute 15% of all adrenal incidentalomas. There is no sex preponderance with age predilection for fifth to seventh decade of life. [¹] Though exact etiopathogenesis of these tumors is yet to be established, these tumors are frequently found to be in association with congenital adrenal hyperplasia. [²] Bilateral adrenal myelolipoma is an extremely rare entity and when present they are usually small and do not produce bothersome symptoms to the patient. Here we present a case of bilateral giant adrenal myelolipoma which became bothersome to the patient because of its large size and was successfully managed by bilateral adrenalectomy.

CASE REPORT

A 47 year old hypertensive male presented with history of pain and lump on the left side of the abdomen for duration of
three months. The pain was gradual in onset, constant dull aching and often radiating to the back. Patient also complained of a lump on left side of the abdomen which he could appreciate since last two months and was gradually increasing in size. On clinical evaluation, general examination was insignificant, heart rate was 84 beats/min and blood pressure was 150/94 mmHg. On her abdomen examination there was a palpable lump of approximately 10x8 cm size in left hypochondrium and left lumbar region and a small ill defined lump on right hypochondrium. There were no other significant findings clinically.

Ultrasonography revealed homogenous, hyperechoic, well defined mass lesion in both adrenal fossa (right side = 12.2x7.9 cm and left side = 22.3x12cm). CECT revealed bilateral adrenal mass lesions, left side measuring 22x15x12 cms and right side measuring 11x8.2x7 cms with variable mass effect over kidneys (Left>Right), pancreas, liver and spleen [Figure1]. Routine haematological parameters were within normal limit. Functional evaluation of adrenals was done using 24 hour urinary metanephrines and serum cortisol, which were both within normal range (24 hour urinary metanephrines - 280 micrograms/24 hrs, serum cortisol – 12.0 micrograms/dl).

Considering the patients symptoms and size of adrenal masses patient was explained regarding the need for bilateral adrenalectomy and post-operative lifelong need for hormone supplementation. Patient was started on alpha blockers and his blood pressure was well controlled. The patient underwent bilateral staged adrenalectomy, left side first and then on the right side six weeks later. [Figure 2 and Figure 3] Histologically both the lesion showed haematopoetic element, admixed with mature adipocytes, surrounded by adrenal tissue with foci of hyalinization and hemorrhage suggestive of adrenal myelolipoma [Figure 4]. Post operative period was uneventful. The patient is on regular follow up since then without any symptoms.
DISCUSSION

Adrenal myelolipomas are rare, benign lesions that are usually metabolically inactive, without any sex predilection seen in all the age groups but more frequent in fifth to seventh decade of life. Myelolipoma of adrenal gland were first described in 1905 by Gierke as a lesion composed of mature adipose tissue mixed with myeloid and erythroid cells. [3] In 1929 Oberling described them as formations myelolipomatosis. [4]

Etiopathogenesis of these tumors is not well defined and they are thought to arise from metaplasia of undifferentiated stromal cells and congenital adrenal hyperplasia has been seen to be frequently associated with them. In the past these tumors were usually detected at autopsy, however, due to advanced and frequent use of radiological techniques like ultrasonography, computerized tomography (CT), and magnetic resonance imaging (MRI) they are now mostly detected as incidental findings. Myelolipomas are not only confined to adrenals, but extra adrenal myelolipomas have been reported in presacral, retroperitoneum, thorax, and pelvis, renal and even hepatic regions. [5]

With an incidence of less than 1% myelolipomas are usually noted in late adult life with a mean age at presentation of 62 years in one of the larger series by Lam KY et al. [6] frequent association with obesity, type 2 diabetes mellitus and hypertension is noted, possibly coincidental. [7] Usually unilateral, small and asymptomatic, a limited number of bilateral tumors have been described in the literature. [8] Average size of most adrenal myelolipomas is less than 4 cms and giant adrenal myelolipoma has been defined as one being more then 8 cms of size. [9] These tumors are generally hormonally-inactive, although there are case reports of their association with over production of adrenal hormones. Myelolipomas have been associated with overproduction of dehydroepiandrosterone sulphate (DHEAS), congenital adrenal hyperplasia caused by 21-hydroxylase deficiency, congenital adrenal 17 α-hydroxylase deficiencies, Cushing disease, Conn syndrome, adrenal insufficiency, and pheochromocytoma. [2,10,11]

When small and asymptomatic most myelolipomas are treated conservatively. Follow up guidelines for conservatively managed myelolipoma have not been standardized, but serial reevaluation with ultrasonography or CT for several years has been suggested. [12] Large symptomatic adrenal myelolipoma causing bothersome symptoms because of mass effect warrant surgical intervention in form of adrenalectomy. Rapid increase in size of myelolipoma, spontaneous rupture or hemorrhage with minor trauma during conservative management has been reported. [13] This can sometimes be life threatening and highlights the need for close follows up.

Due to rarity of bilateral giant adrenal myelolipoma, treatment should be individualized. If bilateral adrenalectomy is contemplated, every effort should be done to preserve normal adrenal tissue is possible to save the patient from lifelong adrenal insufficiency another important aspect while considering bilateral adrenalectomy is that it should be
performed in a staged fashion to prevent severe post operative adrenal insufficiency, surgical complications and enhance the post operative recovery. Hence the best option for bilateral adrenal myelolipoma is removal in two different stages of surgery at different times.  

In our case, due to large size of the mass and associated symptoms we decided for resection of the left tumour first. Right adrenalectomy was performed six weeks later. Post operative period was uneventful. Endocrinological consultation was taken and hormone supplementation was started. The patient is on regular follow up without any symptoms.

CONCLUSION

Adrenal myelolipoma is a rare benign tumor and bilateral tumors are further more rare entity. When small they are usually asymptomatic and need only close follow up, but when large and symptomatic, surgery remains the treatment of choice. Staged bilateral adrenalectomy followed by hormone replacement therapy in case of giant adrenal myelolipoma remains the optimal treatment and yields favourable outcome.

REFERENCES

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