Adrenal myelolipoma: a case report

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ABSTRACT

Myelolipoma is a relatively rare benign tumor of adrenal gland. Most cases are non-functioning and asymptomatic and usually discovered during various diagnostic-imaging procedures performed for other diseases of abdomen. Cases associated with excessive hormone secretion have been reported which may constitute adrenocortical adenomas with associated myelolipomatous components. Histopathological findings are very interesting and the tumor is composed of bone marrow elements with various amounts of fatty tissue. The fat component in myelolipoma can facilitate the diagnosis by radiological examination. Here, we report a case of adrenal myelolipoma and its clinical presentation and evaluation. In this case, initially the patient presented with nonspecific abdominal symptoms. Her adrenal tumor was found incidentally and the final diagnosis was proved by histopathology.

Key words: adrenal gland, myelolipoma, haemopoietic element.

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INTRODUCTION

Myelolipoma is a relatively rare benign tumor of adrenal gland and accounts for 2.6% of all primary adrenal tumors. The lesion was first described by Glerke in 1905. Although extra-adrenal myelolipomas are very rare but documented. The tumor is more common in female than in male and patients usually present during the fifth to seventh decades of life. The majority of the cases have been small and asymptomatic and discovered incidentally, either at autopsy or during investigations for other diseases. However, some become symptomatic due to hemorrhage, necrosis or pressure

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on surrounding structures.⁶ Histologically, this tumor is composed of mature adipose tissue and scattered islands of hematopoietic cells including erythroid, myeloid and lymphoid series as well as megakaryocytes representing the most important diagnostic feature.⁶ It is thought to arise from metaplasia of undifferentiated stromal cells.⁷ Diagnosis is done either pre- or post-operatively but the therapeutic strategy is still controversial. Surgical treatment or watchful waiting are the options depending on the size of the tumor and the presence of symptoms.⁸ However, as a consequence of progress in diagnostic imaging techniques, the frequency of diagnosis is expected to rise in the future.⁹

CASE REPORT

A 35-year-old female patient presented with 2-year history of pain in lower abdomen and right lumber region with hypertension. She was admitted to Gynaecology and Obstetrics Department of Combined Military Hospital, Dhaka Cantonment for evaluation. Abdominal ultrasonography was done and a right adnexal mass was detected. Then abdominal computed tomography (CT) scan was performed. It revealed a right adnexal mass with a right suprarenal mass which was about 7.1 cm × 6.6 cm in size with well-defined contour and fat component in density (Figure 1).



Figure 1 Abdominal CT scan revealed a right adnexal mass with a right suprarenal mass

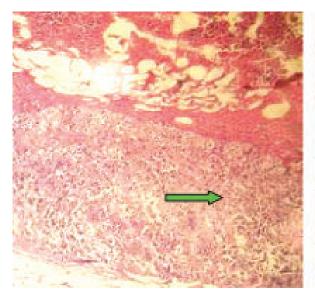
The laboratory investigations revealed that, all blood biochemical profiles including blood glucose, blood urea, serum creatinine, cholesterol, triglycerides and liver function tests were within normal limit. All haematological profiles also revealed normal. Twenty four hours urinary vanyllmendelic acid was 4.28 mg (normal: 1.0 - 7.5 mg/24 hour). Initially, the patient was treated by gynaecologist and the adenxal mass was removed by right sided oophorectomy and the specimen was sent for histopathological examination. Histopathological diagnosis was chocolate cyst.

Then the patient was transferred to the urology department for further management. Based on clinical history of hypertention and radiological findings, the case was clinically leveled as pheochromocytoma. Then suprarenal mass was removed by laparoscopic right adrenalectomy and the tissue was sent for histopathological examination in Armed Forces Institute of Pathology (AFIP), Dhaka Cantonment. Gross examination revealed multiple fragments of redyellowish tissue collectively measuring (3 cm x 2.5 cm x 2 cm) (Figure 2).



Figure 2 Multiple fragments of red-yellowish tissue on gross examination

Microscopic examination revealed the features of myelolipoma of the adrenal gland. The neoplasm was composed of mature adipose tissue mixed with normal bone marrow elements. Thin rim of adrenal cortical tissue was found in the periphery of the tumour (Figure 3).



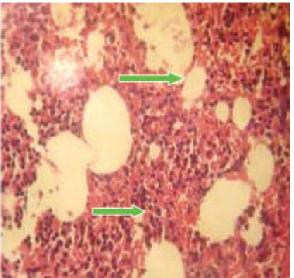


Figure 3 Features of myelolipoma of the adrenal gland with thin rim of adrenal cortical tissue in the periphery of the tumor

DISCUSSION

Adrenal myelolipoma was first reported as "myeloadipose formation" in 1905. Its exact cause is unknown. There are many theories to explain the etiology of myelolipoma to justify the presence of myeloid cells in the adrenal gland. First theory is that the bone marrow emboli probably lodged in the adrenal gland, second theory is probably due to metaplasia of the reticuloendothelial cells within the capillaries of the gland and the third one is due to deviation of the embryonic primitive mesenchymal stem cells. ¹⁰ However, the most widely accepted theory is the metaplasia of the reticuloendothelial cells of blood capillaries.

This tumor is generally unilateral, asymptomatic and functionally inactive. The majority of the cases are discovered incidentally, either at autopsy or during investigations for other diseases.⁴ The incidental findings in autopsy varies from 0.08 to 0.2%.¹¹ Most of the tumors originated from the right adrenal gland and bilateral synchronous tumors may rarely develop. The tumor is more common in female than in male. The lesion has been found in patients from 17 to 93 years of age with mostly diagnosed during 5th to 7th decades of life. The youngest patient reported is a 17-month-old child with Beckwith–Wideman syndrome.¹² In case of symptomatic patients, the most common symptoms observed are nonspecific abdominal pain, hematuria and hypertension.

Interestingly, although myelolipomas are commonly found in adrenal glands but rarely extra-adrenal myelolipomas are also documented. They have been found in various sites, including mediastinum, liver, stomach, lungs, pelvis, spleen, retroperitoneum, presacral region, thoracic spine and mesentery.³ It should be differentiated between extra adrenal myelolipoma with extramedullary hematopoieses which are also composed of hematopoietic elements but may lack adipose tissue and are associated with anemia and marked bone marrow hyperplasia.¹³

Imaging studies such as CT scan and magnetic resonance imaging (MRI) are relatively specific and have made the differentiation of adrenal myelolipomas from other retroperitoneal tumours. Ultrasonographic, fluoroscopic or CT guided needle biopsy have been reported to be useful to establish the definitive diagnosis and select optimal treatment.¹⁴

Therapeutically, the strategy is still controversial. Most authors suggested that myelolipoma, if correctly diagnosed, could be treated conservatively with careful follow-up, while their surgical management is dependent on size or symptoms.

Conclusion

In conclusion, adrenal myelolipomas are rare and always benign adrenal tumours. Radiological imaging, such as CT and MRI are usually used to characterize a lesion and are specific enough to provide a pre-operative diagnosis of adrenal myelolipoma. Definitive diagnosis depends on cytological or histological evaluation. Biopsy performed under ultrasound or CT scan guidance has been suggested as the most direct approach to establish the diagnosis in difficult cases. Its prognosis is excellent with rare recurrence potential or malignant transformation. Although myelolipoma is rare, it should be considered for differential diagnosis of adrenal tumours.

Authors' contribution: MSH was involved in diagnosing the case and preparation of the case report. SMMA, SMJI were involved in diagnosing the case. SY was overall supervisor.

Conflicts of interest: Nothing to declare.

Consent: Informed written consent was taken from the patient.

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