Case Report

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Adrenocortical Adenoma with Foci of Myelolipoma in a Patient of Conn's Syndrome

Pathology Section

DEBOSMITA BHATTACHARYYA, ANADI ROY CHOWDHURY, DEBAJYOTI SINGHA ROY, RITA BASU MITRA

ABSTRACT

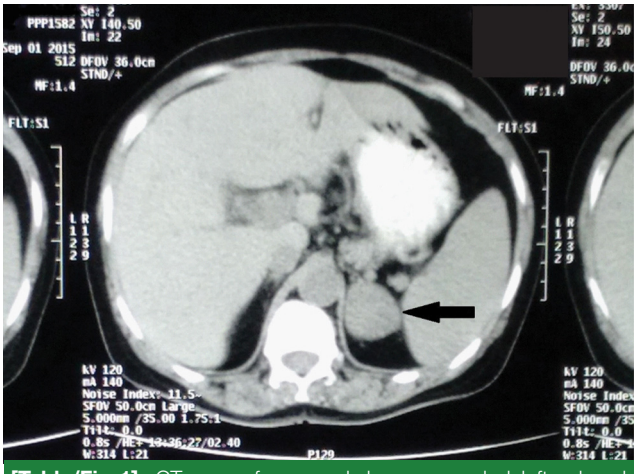
Adrenal myelolipoma is an uncommon benign tumour usually discovered by chance in patients. It is usually inactive hormonally and composed of mature adipose tissue with normal haematopoietic cells. Rarely, adrenocortical adenoma and adrenal myelolipoma can coexist. We report a case of a 40 years old Indian woman with 3 years history of hypertension. CT scan showed a left adrenal tumour measuring 3×2.6 cm. Clinical history and laboratory result suggests a metabolic disorder like Conn's syndrome. The patient underwent left adrenalectomy and the histopathology study revealed adrenocortical adenoma with foci of myelolipoma.

Keywords: Adrenal incidentalomas, Primary hyperaldosteronism, Secondary hypertension

CASE REPORT

A 40-year-old female patient presented with right upper quadrant pain for 3½ months. She was a known hypertensive patient and was on medication for last three years. On clinical examination, her blood pressure was found to be 180/98 mmHg. There was moderate amount of anaemia. Liver and spleen were not palpable. CT-scan of upper abdomen revealed left adrenal tumour measuring 3 x 2.6 cm [Table/Fig-1]. Right adrenal gland was slightly enlarged.

Serum aldosterone level was 403.10 pg/ml (reference range- supine: 10-105 pg/ml, upright: 34-273 pg/ml). Plasma renin activity was 0.27 ng/ml/hr (reference range- supine: 0.50-1.90 ng/ml/hr, upright: 1.90-6.00 ng/ml/hr). Aldosterone/PRA ratio was 149.30 (reference range >30 suggestive of primary hyperaldosteronism, >50 diagnostic of primary aldosteronism). The serum potassium level was found to be 2.80 mEq/L (reference interval 3.50-5.50 mEq/L). The serum cortisol (6 AM) level was 15.71 µg/dl (reference range AM- 6.2 to 19.4 µg/dl, PM- 2.3 to 11.9 µg/dl). Her urine for VMA was found to be 4.0 mg/24hrs (normal range <13 mg/24hrs).



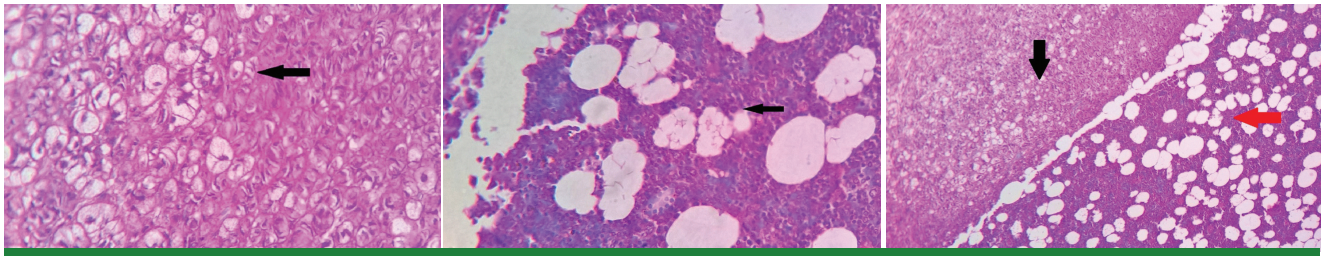
[Table/Fig-1]: CT-scan of upper abdomen revealed left adrenal tumour measuring 3 x 2.6 cm marked with black arrow.

In further investigations her serum CA- 19.9 was 76.31 U/ml (reference interval 0-37 U/ml).

Above biochemical investigation coupled with CT-scan pointed towards primary aldosteronism (Conn's syndrome) due to adrenal tumour. Left adrenalectomy was performed without any complications. The post-operative evaluation was favorable, the patient remained asymptomatic.

On macroscopic examination, left adrenal gland was enlarged measuring 3.5×3.2×3.0 cm and brownish yellow in color with a space occupying lesion in it measuring 3.0x2.8 cm [Table/Fig-2].

[Table/Fig-2]: On macroscopy, left adrenal gland shows a space occupying lesion measuring 3.0 x 2.8 cm marked with black arrow.



[Table/Fig-3]: On histopathology, adrenocortical adenoma showing oval shaped cells with hyperchromatic nuclei are marked with black arrow. (Hematoxylin-Eosin Section; 400X magnification). **[Table/Fig-4]:** On histopathology, focal area of myelolipoma with haematopoietic cells within adrenocortical adenoma marked with black arrow. (Haematoxylin-Eosin Section; 400X magnification). **[Table/Fig-5]:** On histopathology, adrenocortical adenoma marked with black arrow and focal area of myelolipoma marked with red arrow. (Hematoxylin-Eosin Section; 100X magnification).

On histopathological examination- the sections from the tissue showed presence of spindle to oval cells arranged in trabecular, solid and acinar pattern [Table/Fig-3]. Capsule was intact. Focal area showed presence of myelolipoma [Table/Fig-4]. The Weiss score [1] is as follows:-

Eosinophilic cells <75 %

Necrosis- Absent

Fuhrman's Nuclear Grading- 2/4

Mitotic Figure- <5/50 HPF

Vascular/Venous/Capsular Invasion- Absent

So, the diagnosis was made to be adrenocortical adenoma with focal area of myelolipoma [Table/Fig-5].

Her postoperative follow-up was uneventful and her blood pressure was well controlled and her antihypertensive medication was slowly weaned off.

Consent was taken from the patient for the publication of her case report.

DISCUSSION

Adrenal myelolipomas are small, asymptomatic, non-functional tumours [2]. They are uncommon and are composed of mature adipose tissue with normal haematopoietic tissue. Its pathogenesis remains unclear [3]. Adrenal myelolipoma occurring within adrenal cortical adenoma has only been reported in 16 cases so far and only one amongst them had associated Conn's syndrome [4]. Here, we report a myelolipoma that is located within adrenal cortical adenoma. The diagnosis is made by pathological examination of the specimen. Relying on laboratory tests and clinical examination, adenoma's functioning property was proved.

There are different explanations about this matter but according to a widely participated theory, pathogenesis of myelolipomas occurs in response to stimuli, such as necrosis, infection or stress, reticuloendothelial cells of blood capillaries in adrenal gland with metaplasia [5].

Histological examination of myelolipomas prove that haematopoietic elements consisting of myeloid and erythroid precursors as well as megakaryocytes are interspersed within adipocytes [6]. Myelolipomas in fact equally affect both sexes between the ages of 50 to 70 [7]. But our patient is a 40 years old lady presenting with a left sided adrenal myelolipoma.

The adrenal cortical adenoma was well encapsulated. The tumour was composed of spindle to oval cells arranged in trabecular, solid and acinar pattern. In this case, in the tumour well demarcated areas of myelolipoma was found in focal areas. The size of the tumour may vary from few millimeters to centimeters and they constitute about 7-15% of adrenal 'incidentalomas' [7].

In this case the adrenal mass was 3×2.8 cm. The patient underwent left adrenalectomy for this. Three radiological patterns of myelolipoma are noted- isolated myelolipoma, myelolipoma with haemorrhage, and foci of myelolipoma within other adrenal tumours often with extensive calcifications [8]. In our case, histological findings showed foci of myelolipoma within adrenal cortical adenoma. Coexistence of these two entities in literature is rare, which makes the case more interesting [9]. The differential diagnosis of myelolipomas includes retroperitoneal neoplasms like renal angiomyolipoma, lipoma, liposarcoma [10]. These fatty tumours can be differentiated on the basis of CT, sonography imaging techniques major difference between myelolipomas and other adrenal neoplasms is the presence of mature fat. However, if the case contained myeloid material with extensive haemorrhage, the fat content may not be recognised [10].

CONCLUSION

Myelolipoma with adrenal cortical adenoma are usually asymptomatic. But if the tumour presents with complications like Cushing's syndrome and Conn's syndrome, it requires immediate attention and management. The diagnosis and differential diagnosis rely mainly on pathological examination and imaging techniques.

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AUTHOR(S):

1. Dr. Debosmita Bhattacharyya
2. Dr. Anadi Roy Chowdhury
3. Dr. Debajyoti Singha Roy
4. Dr. Rita Basu Mitra

PARTICULARS OF CONTRIBUTORS:

1. Post Graduate Trainee, Department of Pathology, RG Kar Medical College & Hospital, Kolkata, West Bengal, India.
2. Associate Professor, Department of Pathology, RG Kar Medical College & Hospital, Kolkata, West Bengal, India.
3. Demonstrator, Department of Pathology, RG Kar Medical College & Hospital, Kolkata, West Bengal, India.
4. Demonstrator, Department of Pathology, RG Kar Medical College & Hospital, Kolkata, West Bengal, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Debosmita Bhattacharyya,
Som-Lok, EF-6, East Kolkata Township,
1729, Rajdanga Main Road,
Kolkata-700107, West Bengal, India.
E-mail: dr.debosmitabhattacharyya@yahoo.in

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