

Incidentaloma: An incidentally discovered adrenal mass; a study of two cases

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Abstract

Background: “Incidentaloma” is an adrenal mass, generally 1 cm or more in diameter, discovered serendipitously during radiological imaging performed for indications other than an evaluation for adrenal disease.

Case presentation: We present two cases for their rare presentation as incidentalomas. First case was incidentally discovered adrenal myelolipoma in a 47 year female. Another case was incidentally discovered adrenocortical carcinoma in a 53 year female who initially presented with abdominal pain and deep vein thrombosis. On routine abdominal ultrasonography, adrenal mass was discovered in both the cases and subsequent pathological evaluation revealed diagnosis.

Conclusion: These cases highlights the importance of thorough investigation to rule out the possibility of lethal malignancy. The evaluation of incidentalomas focuses on identifying functional masses & treating or resecting them appropriately; identifying adrenal carcinomas early with the intent of attempting complete surgical extirpation; and also to reassure the patients whose masses are neither functional nor malignant.

Keywords: adrenal, adrenocortical carcinoma, incidentaloma, myelolipoma

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Introduction

“Incidentaloma” is an adrenal mass, generally 1 cm or more in diameter, discovered serendipitously during radiologic examination performed for indications other than an evaluation for adrenal disease.^{[1][6]} Majority are non hypersecreting benign adrenocortical adenomas, cortisol-secreting adrenocortical adenoma, pheochromocytoma, adrenocortical carcinoma and metastatic carcinoma. In autopsy series, prevalence of undiagnosed adrenal masses ranges from 1.4% to 3%–6%.^[4] Abdominal CT yields similar findings; a recent study reported prevalence of adrenal incidentaloma of 4%.^[1] A recent study on prevalence of incidental adrenal mass confirmed histopathologically, shows that adenomas and myelolipomas are most common lesions, whereas adrenocortical carcinoma is about 0.1%.^[4] We report two such cases.

Case 1

A 47 year female, known hypertensive, presented with severe pain in abdomen since 1 week. Patient had one year history of mild dull aching abdominal pain. Her previous abdominal ultrasound findings dated one year back were normal except for the presence of small

right adrenal incidentaloma measuring 2x1.5x1cm. At admission, her general and physical examination was unremarkable except for mildly raised blood pressure 140/90 mmHg. Her abdominal ultrasound revealed right adrenal mass measuring approximately 5x5x4cm. CT scan findings were suggestive of adrenal myelolipoma (Fig.1). Complete blood count and peripheral smear findings were normal. Biochemical investigations revealed only raised serum cortisol levels 14.3 pg/ml, that fell below basal level post dexamethasone administration with normal urinary metanephrine levels. The associated hypertension is explained by the raised serum cortisol levels suggesting its functional nature. She underwent right adrenalectomy as the size of lesion was more than 4cms associated with intense pain. Finally histopathology of resected mass confirmed it as myelolipoma [Fig. 2(A) & 2(B)]. On follow up, post operative recovery was uneventful. Her pain subsided and blood pressure returned to normal level.

Case 2

A 55 year female presented with 2 months history of abdominal pain associated with pedal edema and pain in lower limbs. Her general and physical examination findings were unremarkable except for the presence of skin discolouration with edema of both lower limbs. Routine blood investigation findings were normal. Patient was kept on heparin followed by warfarin for control of deep venous thrombosis (DVT). As she also had history of post menopausal bleeding, she was referred to gynaecology department. Her abdominal ultrasound revealed right suprarenal mass

with mild ascites. CT scan revealed right suprarenal solid lobulated mass (Fig. 3). The urinary metanephrines, biochemical and hormonal assays were normal. Finally, following series of radiological investigations and after ruling out pheochromocytoma, patient underwent image guided FNAC and Tru-cut biopsy of adrenal mass which revealed the diagnosis as adrenocortical carcinoma (ACC) (Fig. 4 & 5). Adrenalectomy is the best method to manage the patients with ACC, as the complete tumor resection is possible even in the presence of tumor thrombus in inferior vena cava. Medical treatment should be preferred in elderly patients, in those unwilling to undergo surgery and in patients with co morbid

conditions that preclude surgery ^[4]. Our patient presented with advanced tumor and her diagnosis was delayed because of the unusual presentation with the symptoms of deep venous thrombosis and abdominal pain. Though the remote history of post menopausal bleeding could have suggested possible hormonal excess as an etiology but unfortunately the search was directed towards discovering possible gynecological cause. In due course of time, the patient developed various complications such as gross ascites, severe DVT, various side effects of warfarin etc, precluding the surgical option. So as a last resort, chemotherapy was started as a palliation. Unfortunately the patient died within 3 months of the final diagnosis.

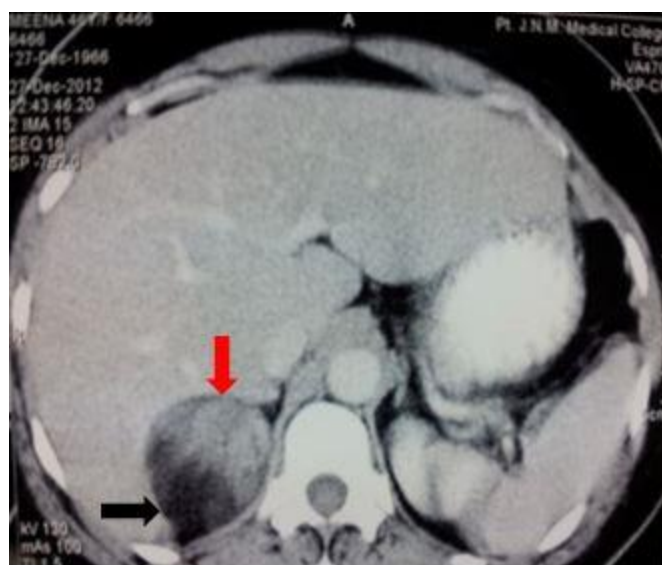


Fig. 1: Computerized tomography scan (abdomen) revealed that the right adrenal is replaced by a well defined large mass lesion which shows soft tissue density and primarily fat ranging from -63 to 53 HU, measuring approximately 5.3 x 4.6 x 5.1 cms, likely suggestive of adrenal myelolipoma. Lipid rich areas- black (black arrow); soft tissue density- white (red arrow).

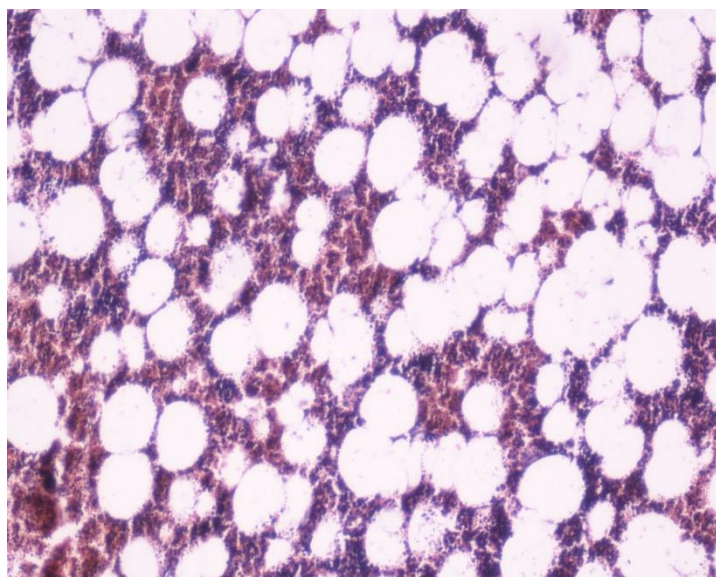


Fig. 2(A): Section of adrenal myelolipoma shows adipocytes with interspersed haematopoietic elements. [H&E; X100]

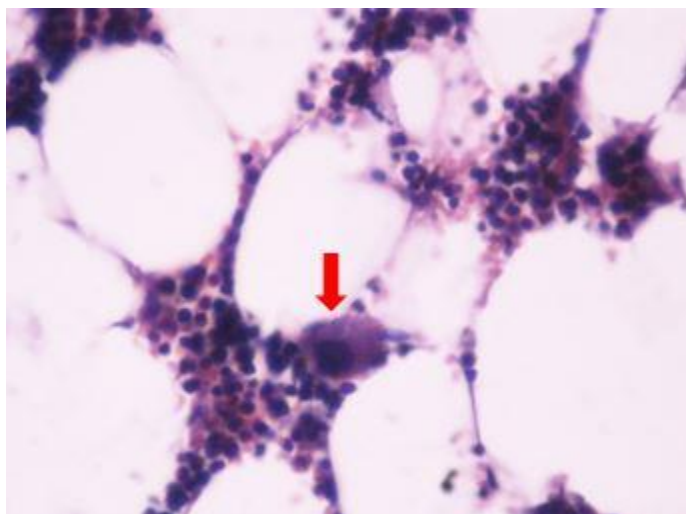


Fig. 2(B): Section of adrenal myelolipoma shows adipocytes with interspersed myeloid and erythroid precursors, as well as megakaryocyte (red arrow). [H&E; X400]

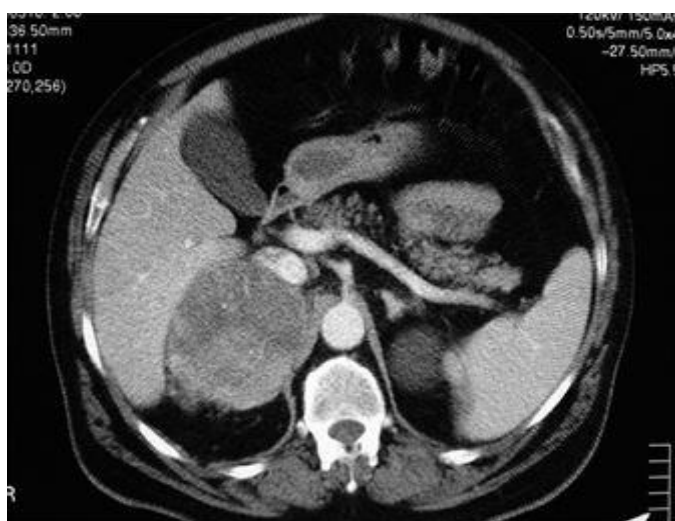


Fig. 3: Computerized tomography scan (abdomen) revealed right suprarenal solid lobulated mass measuring 160 x 90 mm showing areas of internal necrosis and few calcifications with renal metastases. Occlusion of IVC is also evident.

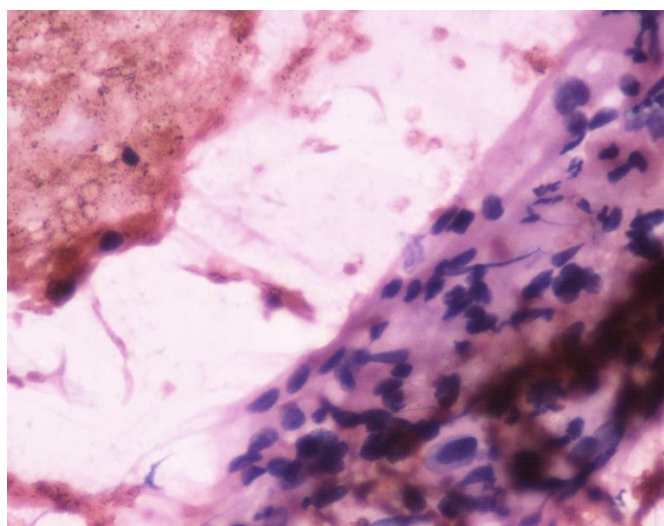


Fig. 4: Fine needle aspiration cytology smear, adrenal mass: Photomicrograph shows cluster of pleomorphic cells with hyperchromatic nuclei and eosinophilic cytoplasm, suggestive of adrenocortical carcinoma. [H&E; X400]

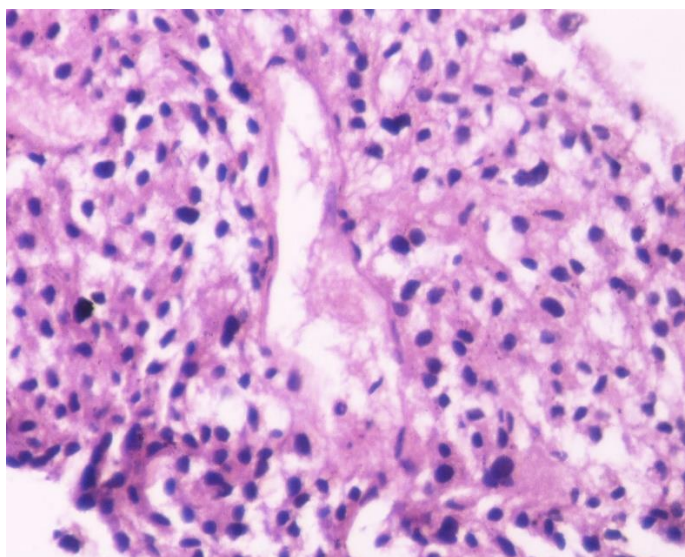


Fig. 5: Adrenocortical carcinoma: trucut biopsy section revealed cells with eosinophilic cytoplasm and round hyperchromatic nuclei showing atypia. [H&E; X400]

Discussion

Adrenal myelolipoma is a rare benign tumour comprised of mature adipose tissue & hematopoietic elements. They are discovered incidentally at autopsy or through CT scan done for other reasons.^[2] Frequency of myelolipomas in adrenal incidental-mas varies between 7-15 %. Incidence at autopsy has been reported from 0.08% to 0.4%.^[2] Mostly, the patients are asymptomatic. Occasionally, they may present with abdominal pain if the tumour is large or due to traumatic rupture leading to hemoperitoneum or from spontaneous hemorrhage. Asymptomatic mass less than 4cms diameter, diagnosed on imaging & or by cytological studies, should be followed up with watchful monitoring. When malignancy is suspected or in case of symptomatic tumors more than 4 cms in diameter, should be extirpated, because of the risk of spontaneous rupture with retroperitoneal bleeding. Prognosis of myelolipoma is excellent. Malignant change is still not documented.^[3]

Adrenocortical carcinoma is a rare malignancy, with an incidence of 1 in 1 million population. Clinically, they present heterogeneously. This neoplasia constitutes 0.02% to 0.2% of all cancer-related deaths, with median survival of 18 months. Women are more prone than men (ratio 1:5).^[4] The prognosis of adrenocortical carcinoma remains poor even after surgery. The overall 5-year survival rate ranges from 16% to 38%.^[6] The neoplasm has been classified according to Weiss histological criteria. Features associated with high probability of malignant clinical behaviour included tumour weight (> 400 g), tumour size (> 10.5 cm), vena caval invasion, severe nuclear atypia, >15 mitotic /20 high power field & the presence of atypical mitotic figures, nuclear grade III or IV, clear cell comprising 25% or less of the tumour, a diffuse architecture, microscopic necrosis & invasion of

venous, sinusoidal & capsular structures. Previously, the presence of 4 or more of these histologic findings was defined as indicative of malignancy, later it was modified to 3 or more.^[5] In our case four criterias were fulfilled; tumour size (>10.5), vena caval invasion, severe nuclear atypia and necrosis. Adrenal cortical carcinoma can cause thrombosis in inferior venacava, portal vein and renal vein.^[7] Our patient had deep venous thrombosis secondary to inferior vena cava and renal vein obstruction. This case highlights the value of correct diagnosis of tumour thrombosis & to distinguish it from venous thrombosis; as it is essential for deciding treatment protocol, especially to decide whether to use anticoagulant or anti cancer therapy. In this case, image guided FNAC and biopsy of adrenal incidentaloma spared the patient from unnecessary and extensive surgical exploration for diagnostic reasons.

Conclusions

The cases described here substantiate, that the term incidentaloma is a misnomer when applied in the widest context to serendipitously discovered lesions in the adrenal. Clearly, a clinically relevant disorder which requires treatment should not be labelled just as an incidentaloma, a term which should be restricted to the detection of a lesion that is associated with no obvious consequence. The pathology associated with incidentalomas represents a broad spectrum of risk for patients and reaffirms the necessity for a meticulous clinical, biochemical, imaging and histopathological evaluation in order to make appropriate decisions.

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