ORIGINAL RESEARCH

Unilateral Adrenal Myelolipoma and Minimal Autonomous Cortisol Excess (MACE) with Disseminated Histoplasmosis: A Case Report

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Abstract

Background and aim: Adrenal myelolipoma is a benign tumor composed of both lipomatous and myeloid components. It is usually asymptomatic in presentation. Another spectrum may present with rupture and hemorrhage. Usually, myelolipoma are non-functional but an endocrine abnormality is seen in 7% of cases. Radiologically diagnosed and asymptomatic adrenal myelolipomas can be kept on follow-up. Symptomatic cases need adrenalectomy. Adrenal Histoplasmosis usually occurs as a part of disseminated histoplasmosis. This is a fungal infection which seen in both immunosuppressed and immunocompetent individuals. Here by reporting a patient presented with Unilateral adrenal myelolipoma with minimal autonomous cortisol excess (MACE) and disseminated histoplasmosis.

Case description: This is a case report of a 64-year-old gentleman presented with vague abdominal discomfort and evaluation found to have large unilateral Adrenal myelolipoma with MACE. The patient underwent Laparoscopic adrenalectomy. Histopathological examination showed histoplasmosis, and was managed by antifungals.

Conclusion: Symptomatic adrenal myelolipoma is managed by adrenalectomy. Prompt antifungal treatment has avoided complications of disseminated histoplasmosis.

Clinical significance: This type of disseminated histoplasmosis in unilateral myelolipoma with MACE is a rare presentation and worth reporting.

Keywords: Computed tomography, Disseminated histoplasmosis, Minimal autonomous cortisol excess, Unilateral adrenal myelolipoma.

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Introduction

Arnold was the first person who described Adrenal myelolipoma as "adrenal lipoma" in 1866. The myeloid component was described by Gierke in 1905. Oberling, in 1929 coined the term "myelolipoma".[1] Adrenal myelolipomas are neoplasms with a mixture of adipose tissue and hematopoietic (myeloid) elements.[2] The detection rate of myelolipoma has become more nowadays because of the widespread use of radiological investigations like ultrasonograms, CT, and MRI, constituting up to 10–15% of adrenal incidentaloma.[1] Histoplasmosis, also known as Darling Disease was first described by Samuel Darling, from America a century ago.[3] *H. capsulatum* var. *capsulatum* and *H. capsulatum* var. *Duboisii* are two species of *H. capsulatum* that cause illness in humans.[4] Adrenal histoplasmosis is usually due to a previous infection or is seen bilaterally with disseminated Histoplasmosis.[5] It can be present in both immunocompetent and immune-suppressed patients. It may also present as an Addisonian crisis. This is a case report of disseminated histoplasmosis presenting in unilateral adrenal myelolipoma with minimal autonomous cortisol excess (MACE), which is not reported in the literature.

Case Description

A 64-year-old man came in with 6 months of upper abdomen burning sensation and discomfort. There was no history suggesting recent cortisol or catecholamine excess, except known diabetic for 20 years, with sugar level controlled with medication. His vitals were normal. On abdominal examination, right renal angle fullness was noted, and a large mass was palpable below the right costal margin in the right hypochondrium and right lumbar region with clinical features of a retroperitoneal site of origin. On radiological evaluation, USG of the abdomen showed a mass measuring 11 × 14.7 cm above the kidney abutting the right lobe of the liver superiorly with heterogeneous, solid predominantly hyperechoic and a few hypoechoic areas with no demonstrable internal vascularity (Fig. 1). Contrast-enhanced computed tomography (CECT) abdomen showed a well-defined heterogeneous lesion measuring 12.7 × 9.2 × 11.7 cm with predominantly fat density and enhancing soft tissue component noted in the right supra renal region superiorly abutting the liver, medially abutting the right renal vessels. Inferiorly abutting the upper pole of the right kidney, suggestive of right adrenal myelolipoma; the left adrenal gland appeared normal (Fig. 2).

The patient underwent Laparoscopic Transperitoneal Adrenalectomy under the cover of perioperative steroids in

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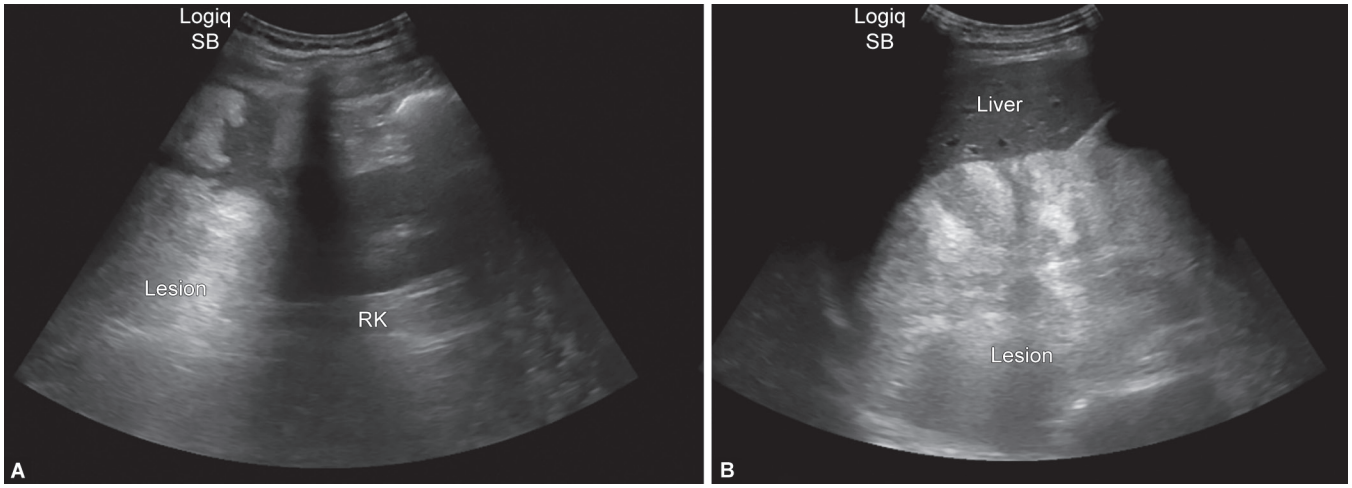
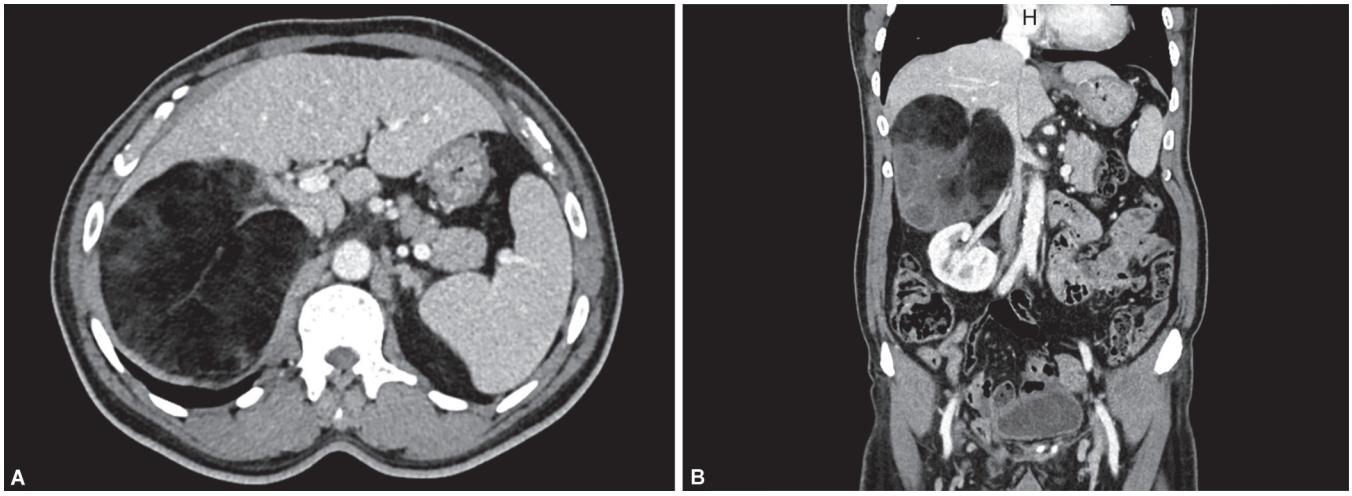


Figs 1A and B: USG image of right adrenal mass



Figs 2A and B: CT image of the right adrenal mass

Table 1: His relevant preoperative biochemical evaluation

Investigation	*Results*
Serum cortisol 8 a.m.	17.3 µg%
Serum cortisol post 1 mg dexa	2.2 µg%
Urinary free cortisol	160 µg/24 hours
Urine 24 hours metanephrine	67 µg/24 hours
Urine 24 hours normetanephrine	270 µg/24 hours
DHEAS	67.1 µg%
Direct renin	5.0 µIU/mL
Aldosterone	24 pg/mL
Sodium	137 mmol/L
Potassium	3.8 mmol/L

view of MACE shown in Table 1. The postoperative period was normal. Histopathological examination of the specimen reported Adrenal myelolipoma with coexistent necrotizing granulomas with numerous fungal yeast forms, possibly histoplasmosis, and the possibility of mixed cryptococcal infection to be ruled out. Histoplasma enzyme-linked immunosorbent assay (ELISA) was positive, and cryptococcal antigen was negative. The patient was managed with conventional amphotericin B for 10 days and discharged on oral Itraconazole 200 mg twice daily to be continued for 1 year. The patient was given Prednisolone postoperatively, tapered, and stopped after 3 months. He was found to be asymptomatic on follow-up after 3 months and his histoplasma ELISA test was reported negative.

Discussion

The most frequent adrenal lipomatous tumor and the second-most common adrenocortical tumor are both adrenal myelolipoma. It is a benign neoplasm in the adrenal cortex composed of mainly mature adipose tissue and intermixed myeloid tissue.[6] Around 3–4% of primary adrenal neoplasms are adrenal myelolipoma. Adrenal myelolipomas are usually non-functional.[3] Around 7% of adrenal myelolipoma endocrine dysfunction was noted.[1] Adrenal myelolipoma with coexistent fungal infection is rare. Abdominal pain or discomfort was the most common symptom reported. Adrenal myelolipoma may present as acute abdomen due to hemorrhage, rupture, or abscess.[1,7] Myelolipomas present as elliptical or spherical tumors with varying diameters reported from less than a centimeter and up to 43 cm, with an average diameter of 10.2 cm. Giant myelolipomas are the tumors exceeding 10 cm in diameter.[7] Investigations commonly done are USG, CT, and MRI.

In CT scans, myelolipomas are well-circumscribed, elliptical or round, hypodense, and heterogenous masses. The attenuation values characteristic of the adipose tissue are from −120 to −90 HU, and the presence of fat density is the hallmark in making the diagnosis of myelolipoma. Management decisions for adrenal myelolipomas should be individualized due to the lack of general guidelines. Most myelolipomas are found incidentally. Imaging confirms the myelolipoma diagnosis and since there are no symptoms associated with the condition, there is no need for treatment. The treatment of choice is adrenalectomy if needed. If there is a considerable increase in size, symptoms (such as stomach pain), or endocrine disruption, surgery is recommended.[1,2,7,8] Large tumors may need laparotomy though even tumors up to 14 cm have reportedly been removed by the laparoscopic method. The patient who presents as an emergency with myelolipoma rupture, endovascular embolization with gelatine sponge particles can arrest retroperitoneal hemorrhage from the tumor, and plan adrenalectomy as an elective operation later.[7]

Histoplasmosis is an infective condition caused by a dimorphic fungus, *Histoplasma capsulatum*. It is contracted by breathing in its spores. It can be found in soil that has had bird and bat droppings decompose, and it also occurs in the atmosphere as a mycelium. This disease is endemic in the United States, Africa, and Asia.[3,9,10] Usually asymptomatic, but when a substantial amount of aerosol is inhaled, it can cause self-limiting acute pneumonitis and hilar lymphadenopathy. Immunosuppressed patients may exhibit a disseminated form that affects the liver, spleen, lymph nodes, bone marrow, and adrenal glands. Primary cutaneous, progressive disseminated, and pulmonary histoplasmosis are the typical clinical manifestations. In immunocompetent hosts, a progressive disease may present as a chronic illness, while in immunocompromised hosts, it may present as an acute illness.[3,4,11] Involvement of the adrenal is seen in disseminated disease. Adrenals may be the only organ affected.[12] Adrenal involvement, overall, has been reported in 30–40% of patients with Histoplasmosis in clinical studies performed globally. The investigation of choice is a CT scan, which shows bilateral enlargement with maintaining normal contour or central hypodense areas with peripheral enhancement. The definite diagnosis of Histoplasmosis requires image guided or laparoscopic biopsy of the adrenal for visualization of the fungus within the invaded tissue, growth of Histoplasma on tissue culture; alternatively, antigen detection in urine or serum and serological test for antibodies can aid the diagnosis.[5] Demonstration of the organism in culture remains the gold standard, but it can take up to four weeks to isolate the organism. A dose of 0.7–1 mg/kg per day of conventional amphotericin B or a dose of 3–5 mg/kg per day of liposomal amphotericin B should be used to treat patients with severe disseminated illness.[3] Patients with adrenal insufficiency should be given steroids. Currently lipid formulations of amphotericin B are used due to reduced toxicity compared to the conventional type. Asymptomatic patients can be treated with oral itraconazole, 200 mg twice daily at least for 1 year.[3]

Conclusion

Adrenal myelolipoma with MACE presenting with disseminated histoplasmosis is extremely rare. Timely management of the condition avoided deadly complications of disseminated histoplasmosis, reinforcing the need for detailed assessment.

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