

A Rare Case with Fatal Disseminated Coccidioidomycosis and Poorly Differentiated Carcinoma of Adrenal Gland in a Young Male

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ABSTRACT

Coccidioidomycosis, the cause of valley fever, is acquired by inhaling fungal spores, present in the soil in the deserts of southwestern USA, central and South America. Most people recover but less than 1% acquires fatal disseminated Coccidioidomycosis (DC). We reported a rare autopsy case of DC in a young male with an incidental finding of a poorly differentiated carcinoma of adrenal gland.

The decedent was a 26-year-old incarcerated Hispanic male who presented with fatigue, nausea, and vomiting. Imaging showed a liver mass, miliary lung nodules and a cavitary lung lesion. The autopsy grossly showed widespread lesions in the lungs, spleen and liver. A 13 x 13 x 4 cm mass, replacing the right adrenal gland and infiltrating into the right posterior aspect of liver was identified. Microscopically, DC with granulomas and necrosis was seen in heart, spleen, liver, lungs and kidneys. The sections of the mass demonstrated a poorly differentiated neoplasm comprised of large pleomorphic cells with rhabdoid features. The tumor showed weak and patchy positivity for pancytokeratin, MART-1, and inhibin but was negative for desmin, myogenin, SMA, arginase-1 and HMB45. INI-1 was intact. This immunoprofile is most compatible with an adrenal cortical primary tumor/carcinoma.

Risk factors for DC include diabetes, HIV, and immunosuppressed state. The descendant's post mortem HIV test was negative however DC is an AIDS defining disease. Younger patients with severe or relapsed DC should be considered for HIV and genetic screening for primary immune defects.

Key words: Coccidioidomycosis; Immunosuppressed state; Pancytokeratin

INTRODUCTION

The decedent was a 26-year-old single Hispanic male with a past medical history of hypertension, alcohol abuse and marijuana use. He was incarcerated at prison. He presented with fatigue, nausea, vomiting, and loss of appetite. On evaluation, he was found to have a liver mass with cavitary lung lesion in the right upper lobe with military nodule diffusely in the lung along with pancytopenia and coagulopathy. He shortly became hypotensive and developed acute renal failure. He coded the next day and passed away shortly afterwards. Medical Examiner's autopsy was limited due to potential exposure to an infectious agent.

The autopsy was remarkable for diffused papular rash over the chest, abdomen, back, flanks and lateral thigh. There was military distribution of small nodules throughout both lungs

and the spleen and a large granuloma in the right upper lobe of the lung. Another significant gross finding was a large, cancerous nodule (13 x 13 x 4 cm) replacing the right adrenal gland and infiltrating the right side of the liver. Cultures of the lung and blood reveal coccidioidomycosis infection. Microscopically, many organs include the heart, spleen; bilateral lungs, pancreas and liver (Figure 1) et al were extensively involved by diffuse coccidioidomycosis infection with associated acute and chronic inflammation and necrosis. The sections of the tumor nodule demonstrate poorly differentiated carcinoma consisting of nests of tumor cells with pink granular eosinophilic cytoplasm, hyperchromatic large nucleus and prominent nucleoli (Figure 2). Necrosis is evident in the tumor. Pancytokeratin, MART-1 and inhibin are weak and patchy positive (Figure 3 and 4). Desmin, myogenin, Arginase, HMB45, and SMA are negative. Overall the

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tumor is a poorly differentiated carcinoma. The immunoprofile is most compatible with an adrenal cortical primary tumor/ adrenal cortical carcinoma.

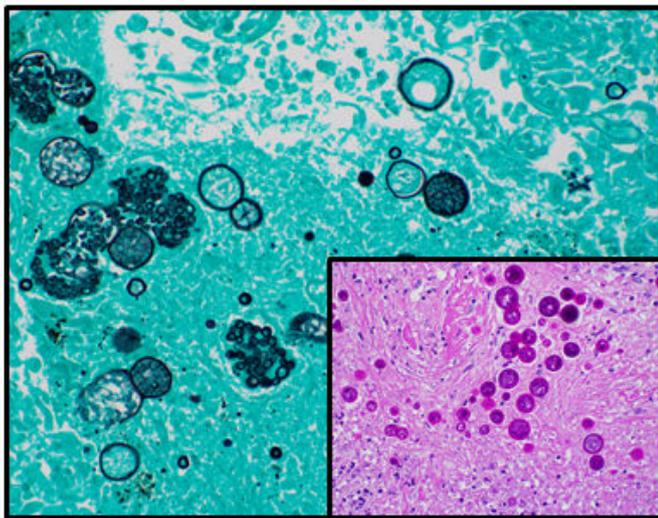


Figure1: GMS and PAS special stain demonstrate different stages of coccidiomycosis forms including sperules and the ones actively releasing endospores (PAS [inset] 20 x total magnification).

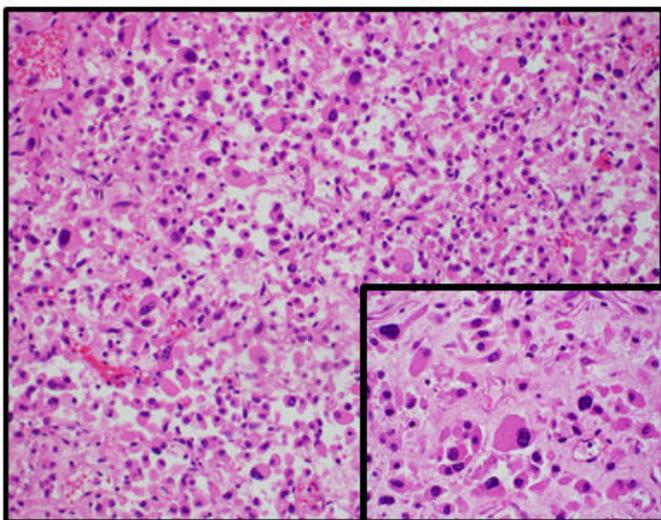


Figure2: Tumor nodule infiltrating the liver demonstrates nests of tumor cells with abundant eosinophilic cytoplasm and bizarre hyperchromatic large nucleus. (hematoxylin & eosin stain, 20x and 60x [inset] total magnification).

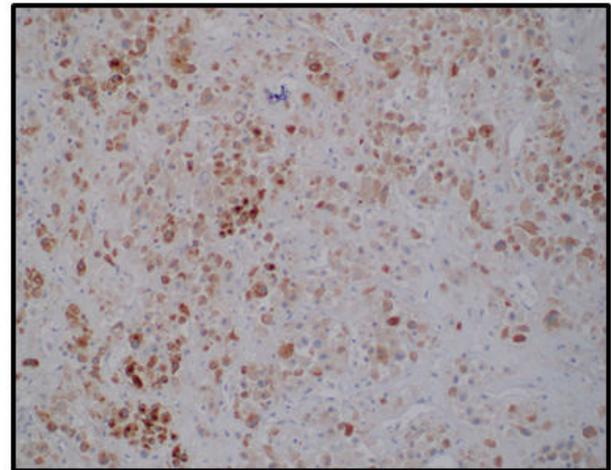


Figure3: Pancytokeratin immunohistochemistry stain of the tumor nodule shows weak and patchy positivity (20x total magnification).

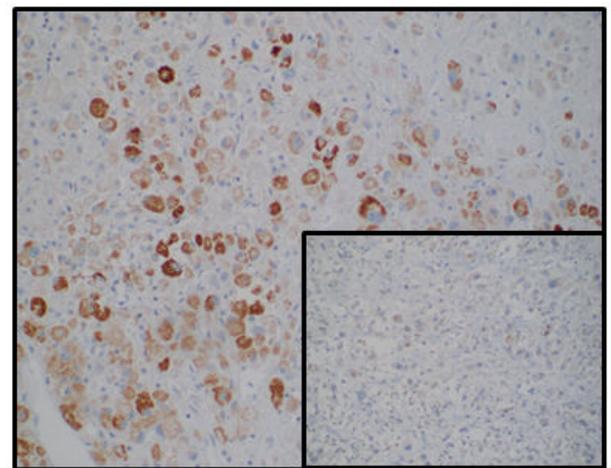


Figure4: MART-1 and inhibin [inset] immunohistochemistry stain of the tumor nodule shows weak positivity (20x total magnification).

DISCUSSION

Coccidiomycosis, also known as Valley fever, is acquired through inhalation of *Coccidioides* species.

It is caused by a dimorphic fungus of which there are 2 subspecies, *Coccidioides immitis* and *Coccidioides posadasii*. It is a fungus endemic to the southwestern United States, the Central Valley of California, Mexico and parts of South America. There are approximately 150,000 new cases occurred in the United States every year [1]. In 2016, a large increase in coccidiomycosis incidence was observed in California compared with previous years, with 5,372 reported cases [2]. It is also a leading cause of community-acquired pneumonia in some endemic zones [3]. The infection causes a broad spectrum of illness from mild flu-like syndrome to uncomplicated pneumonia to severe life-threatening, disseminated disease. Interestingly, African-Americans, Hispanics are more susceptible to severe forms of disease [4]. Immunocompromised population

is particularly vulnerable to developing more severe or disseminated disease.

Hematogenous spread of coccidiomycosis is rare. Of all the infected population, there are approximately 1% will become disseminated [5]. A study done among Prison inmates in California showed that inmates of African American race/ethnicity who were older than 40 years were at significantly higher risk for primary coccidiomycosis than their white counterparts [6].

CONCLUSION

We present a case of a previously healthy 26-year-old young HIV negative Hispanic male who was deceased shortly after presenting with nausea, fatigue and nonspecific symptoms. Imaging suggests multiple lung nodules and remarkable skin lesions. Autopsy examination demonstrated disseminated coccidiomycosis infection involving multiple organs. Characteristics fungal organisms were identified and culture of postmortem blood and tissue confirmed coccidiomycosis infection. A large tumor nodule in the region of unidentifiable right adrenal gland with an immunoprofile most consistent with poorly differentiated carcinoma most compatible with an adrenal cortical primary. This rare case with previously young healthy incarcerated male with unidentified high grade malignancy and DC. His Hispanic ethnicity and probable

immunocompromised status due to the high grade malignancy may lead to the DC infection. Prison is another possible environmental contributor. This case highlights the importance of recognizing the DC in the younger, potentially immunocompromised (HIV et al) population.

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