

## Addison's Disease Presenting as Acute Chest Syndrome: Case Report and Review of Literature

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### ABSTRACT

**Background:** Addison's disease is due to primary adrenal failure. It is an uncommon condition with equal prevalence in both males and females. The onset of symptoms is gradual and manifestation is non specific, hence diagnosis is easily missed without a high index of suspicion.

**Methods:** The medical records of a patient who presented with acute chest pain to the cardiac unit of the University of Port Harcourt Teaching Hospital were reviewed. A review of the literature using manual library and Medline search on Addison's disease was also done.

**Result:** A 48years old male presented in our medical outpatient department with a three day history of sudden onset of severe precordial chest pain that started while playing football which was associated with nausea, vomiting and difficulty in breathing. After initial clinical evaluation a diagnosis of acute myocardial infarction and cardiac failure with a suspicion of background Addison's disease was made. Serial electrocardiography done over a two week period did not show evidence of myocardial infarction, but the patient had elevated serum ACTH and very low serum cortisol levels. An abdominal CT scan done two weeks after admission showed absence of the Adrenal glands bilaterally, confirming Addison's disease. He received treatment for cardiac failure, analgesics, prednisolone and a mineralocorticoid to which he responded satisfactorily and has remained healthy. He also received a six months course of antituberculous treatment empirically.

**Conclusion:** Addison's disease is an uncommon endocrine disorder which can present insidiously in a non specific manner. Diagnosis requires a high index of suspicion.

**KEYWORDS:** Addison's disease; Acute chest syndrome.

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### INTRODUCTION

Addison's disease is the clinical syndrome arising from insufficiency of adrenal hormones due primarily to disease of the adrenal glands. It affects all ages, both sexes and all races. The incidence is rare and is commoner in underdeveloped countries<sup>1</sup>. The commonest cause of Addison's disease is idiopathic

autoimmune adrenal atrophy in developed countries but in underdeveloped countries like Nigeria infections such as tuberculosis, histoplasmosis, cryptococcosis, and coccidioidomycosis are the common causes. Less common causes include sarcoidosis, amyloidosis, adrenoleukodystrophy, metastatic tumors to the adrenals, bilateral hemorrhage and infections such as HIV and Cytomegalovirus.<sup>1</sup>

Addison's disease presents insidiously with non specific symptoms such as weakness, anorexia, weight loss, and increased pigmentation of skin, palms, sole and the buccal mucosa. Sometimes presentation may be as a life threatening Acute Addisonian crisis and except with careful clinical assessment and a high index of suspicion the diagnosis may be missed.

### CASE REPORT

A 48year old male presented to the medical outpatient department with a three day history of severe precordial chest pain, difficulty in breathing, nausea and vomiting which started while he was playing football. Pain has been persistent and did not radiate. There was no fever or urinary symptoms, bowel habit and appetite were unchanged. He had noticed an increase in his dark complexion, easy fatigue and weight loss in the last six months. He had a very strong alcohol history but did not smoke. Physical examination revealed an acutely ill asthenic man in obvious pain, not dyspnoeic at rest, moderate pallor, low grade fever, pigmentation of the buccal mucosa, the palms and its creases, knuckles of the fingers, soles, knees, elbows and around the navel. Pulse was 110/min, small volume, regular, blood pressure was 60/40mmHg, and cardiac apex was not displaced. Heart sounds were normal in character, with a gallop rhythm but faint. There were fine bibasilar rales in the chest and a palpable tender liver 5cm below the right subcoastal margin as well as epigastric tenderness. The spleen and kidneys were not palpable. A diagnosis of acute myocardial infarction and heart failure with a suspicion of background Addison's disease was made.

The ECG done on presentation showed generally low voltages and ST segment elevation of 1.5mm in all the chest leads, Lactate Dehydrogenase (LDH) showed only slight elevation 5,1mmol/L (Normal is < 3.38mmol/L) while creatine phosphokinase (CPK) was within normal limits and retroviral screening was

negative. The liver function tests were within normal limits, lipid profile showed HDL cholesterol of 0.2mmol/L, Triglyceride of 2.0mmol/L, LDL cholesterol of 4.8mmol/L and total cholesterol of 5.4mmol/L. Full blood count was within normal limits except for raised ESR (149mm/Hr Westergreen) and anaemia (Haemoglobin of 9.7gm/dl). Kidney function tests were within normal limits. Chest X-Ray showed mild cardiomegaly with right basal pneumonic changes but no effusion.

He received analgesics, digoxin, low dose aspirin, nitroglycerine, heparin injection, and anxiolytics. Streptokinase injection could not be provided. His response to these measures was slow with a persistent intermittent fever. Five days after admission the result of 8.00am serum cortisol and adrenocorticotrophic hormone (ACTH) assay showed very low serum cortisol level, 85nmol/L (Normal=190 to 760nmol/L) and elevated ACTH level. He was commenced on oral steroids, intravenous fluids and antibiotics therapy to which he responded very well and was discharged two week later. Abdominal CT scan done after discharge showed absence of the Adrenal glands. A six month course of antituberculous therapy was commenced empirically on epidemiological basis as Tuberculosis is the most common cause of Addison's disease after autoimmune type in this environment in addition to this the patient also had a strong alcohol history. The patient commenced steroid replacement consisting of both glucocorticoid and mineralocorticoids after counseling. He has remained well three months after discharge.

## DISCUSSION

Addison's disease is an uncommon condition with prevalence of 1:100,000 in the general population and is commoner in the poorer countries and in type I diabetics<sup>2</sup>. It is potentially fatal, develops insidiously and symptoms are non specific and can be easily missed.

The presentation of Addison's disease may be classical and characterized by chronic fatigue, mucosal and cutaneous hyperpigmentation, hypotension made worse by posture and progress to nausea, vomiting, diarrhoea, salt wasting and shock. The presentation may be as acute adrenocortical failure with fever, hypotension, dehydration, and shock.

This case represents one of the latter with an acute Addisonian crisis resulting from the excess energy demand of exercise and possibly chest infection on a background of chronic adrenal destruction. The features of Addison's disease such as progressive fatigue and weakness, increasing pigmentation over the soles, palms and body generally has been present in this patient for several years undiagnosed. Tuberculosis

as a cause of Addison's disease is not common in the developed countries<sup>3</sup>, where most are due to autoimmune adrenal destruction as evidenced by coexistence of other autoimmune endocrine diseases in patients with Addison's disease. The problem of tuberculosis endemicity in Nigeria is well known and is therefore believed to be responsible in this case in the absence of other endocrine abnormalities though adrenal antibodies could not be assayed in this patient. Acute cardiac dysfunction associated with rhabdomyolysis<sup>4</sup> and acute abdomen complicated by cardiomyopathy<sup>5</sup> has been reported as presenting features in children.

Acute Addisonian crisis due to malignant lymphoma infiltrating the two adrenal glands has also been reported<sup>6</sup>. Sometimes an acute illness precipitates an acute Addisonian crisis in a patient with undiagnosed Addison's disease. A case of histoplasmosis capsulatum infection of the liver and adrenal glands was only diagnosed following an Addisonian crisis during which granulomatous destruction of both adrenal glands was found in a Japanese patient<sup>7</sup>.

## CONCLUSION

The rarity of Addison's disease and its unusual presentation as demonstrated by this case indicate the need for careful and thorough evaluation of every patient during an emergency and further re evaluation when the patient is stable.

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