

A Rare Presentation of Sarcoidosis with SIADH: A Case Report

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Abstract

We report a case of 60 year old male, who presented with acute change in mental status characterized by lethargy and decreased level of consciousness since one day. Sarcoidosis is a granulomatous disorder of unknown etiology which involves multiple systems. The central nervous system is affected in only 5-15% of patients with sarcoidosis. Neurosarcoidosis is a rare disorder with variable clinical course and prognosis. Treatment consists mainly of high-dose corticosteroids, which usually have to be taken long-term.

Keywords: Lymphadenopathy; Hyponatremia; Sarcoidosis; Inappropriate ADH Syndrome.

Introduction

Sarcoidosis is a female-predominant multisystem granulomatous illness.¹ A majority of patients develop respiratory system involvement,¹ however only 5-15% patients with sarcoidosis have neurological involvement.² Most common neuroendocrine manifestation (accounting for half) is diabetes insipidus, followed by amenorrhoea or galactorrhoea. SIADH is very rare and only few cases have been reported.³ The syndrome of inappropriate antidiuretic hormone secretion (SIADH) is caused due to inappropriate continued secretion or action of the hormone despite normal or increased plasma volume which results in hyponatremia and hypo-osmolality.⁴ In this case we describe a rare presentation of sarcoidosis with SIADH.

Case report:

We report a case of 60 year old male, who presented to our hospital in December 2013, with acute change in mental status characterized by lethargy and decreased level of consciousness since one day. Patient also had history of cough, dyspnea and hoarseness of voice for past 6 months history. His general physical examination was normal except for axillary lymphadenopathy and vitals were stable. On evaluation of nervous system, patient was in altered sensorium, not oriented to time, place and person. Cranial nerves examination was normal. Reflexes were 1+. Plantars were bilaterally flexors. No signs of meningeal irritation. Respiratory system- diffuse rhonchi present.

On investigations; CBC was normal. Urea, creatinine, RBS, LFT was normal. Electrolytes- sodium-112meq/l, potassium

5.0 meq/l, chloride was 76 meq/l. Patient's sodium deficit was calculated and treated with 3% Normal saline infusion. It was noticed that patient had recurrent hyponatremia inspite of repeated corrections with 3% NS. Urine sodium was 127meq/l, urine osmolality was 314.5mosm/kg, serum osmolality-254.32 mosm/kg urine glucose was 20mg/dl. Thyroid profile and cortisol were within normal limits. Chest X ray showed hilar lymphadenopathy and reticulonodular pattern.

CT thorax showed bilateral hilar and paratracheal lymphadenopathy with hazy or egg shell calcification and small well defined subpleural nodules as shown in (Fig1).

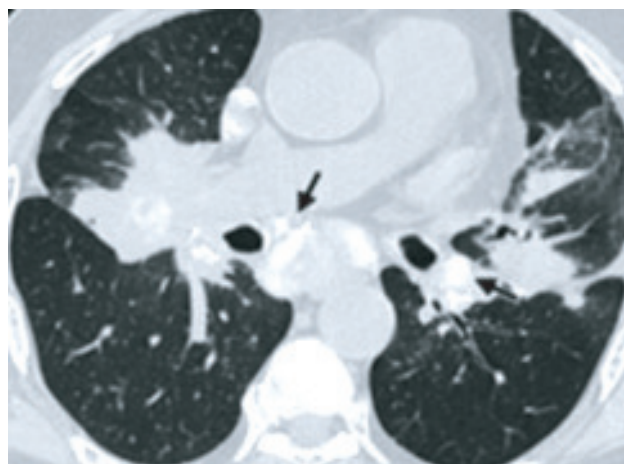


Fig 1: CECT axial view showing bilateral hilar & paratracheal lymphadenopathy with hazy or egg shell calcification.

Differential diagnosis of sarcoidosis and interstitial lung disease was given. Serum ACE levels were 73 and serum calcium was 11meq/l. Patient underwent biopsy of axillary lymph node. It showed non caseating granuloma (Fig. 2).

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So a diagnosis of sarcoidosis with SIADH was made and patient was started on tab prednisolone 1 mg/kg body weight and tablet tolvaptan 15mg/day. Patient's sodium levels improved from 119 to 133 meq/l over a period of two weeks with improvement in clinical symptoms.

Discussion:

Sarcoidosis is ubiquitous disease of unknown etiology, seen worldwide with a multisystem involvement.⁵The central pathologic hallmark of Sarcoidosis is the granuloma formation, consisting of macrophages, epithelioid cells and multinucleated giant cells which secrete cytokines. Sarcoidosis commonly involves lungs, skin and eyes. Lung involvement occurs in >90% of sarcoidosis patients.⁶ Lymphadenopathy is seen in about 15% cases. Intrathoracic nodes (hilar nodes and paratracheal nodes) are enlarged in 75 to 90%. Peripheral lymphadenopathy is also very common, particularly involving the cervical axillary, epitrochlear, and inguinal nodes.⁷ Diagnosis of sarcoidosis is a matter of exclusion, as there is no specific test for the condition. Important differential diagnoses of granulomatous disease like Crohn's disease, lymphoma and vacuities should be excluded. Pathological findings of sarcoidosis are usually non necrotizing granuloma, but occasionally necrosis can be observed. Serum ACE levels in neurosarcoidosis have low sensitivity rates.⁸ Abnormal CXR is seen only in 30% of neurosarcoidosis.³ In our case axillary lymph node biopsy showed not caseating granuloma. CT thorax showed bilateral hilar and paratracheal lymphadenopathy with hazy or egg shell calcification and small well defined sub pleural nodules suggestive of sarcoidosis.

Neurosarcoidosis is a rare but potentially devastating manifestation of sarcoidosis, with a prevalence of approximately 5%.⁹ However asymptomatic central nervous system (CNS) involvement is found in a much higher proportion.¹⁰ In half of the patients with NS Neurological symptoms are the presenting manifestation and in the remaining half symptoms may appear within 2 years of being diagnosed with systemic sarcoidosis.¹⁰ The most classic clinical patterns of nervous system involvement in sarcoidosis include cranial neuropathy, meningeal diseases or hypothalamic-pituitary axis involvement. Facial nerve palsy is most frequently involved nerve in sarcoidosis seen, followed by optic nerve.¹¹ Possible endocrine manifestations of sarcoidosis includes obesity, body temperature dysregulations, personality change, SIADH, diabetes insipidus (50%), hyperprolactinemia, hypoadrenalism, growth hormone deficiency and impairment in counter-regulatory response to hypoglycaemia.³ Thyroid dysfunction

is seen in 4.2–4.6% of cases. SIADH has been very rarely reported in literature. It is mainly caused in sarcoidosis due to deposition of granulomas in the posterior pituitary.^{12,13}

Conclusion:

As neurosarcoidosis is a serious devastating disease which poses a variety clinical challenges, this paper tries to highlight that SIADH presenting with severe hyponatremia can be primary presentation in sarcoidosis. Also as per our pubmed search, neurosarcoidosis with primary presentation as SIADH has not described in literature previously. This paper also tries emphasizes on the need to consider it as differential diagnosis in unexplained causes of hyponatremia.

Conflict of Interest : None

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