Rare presentations of sarcoidosis

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Abstract

Sarcoidosis affects multiple organs, forming non-caseating granulomas whose cause is unknown and can present with nonspecific features. The diagnosis requires involvement of two or more organs and the demonstration of non-caseating granulomas on biopsy. Non-specific clinical features make delay in diagnosis. It is for this reason that we report herein two cases, one who presented with abdominal pain due to pancreatitis-hypercalcaemia related and cause of which was sarcoidosis, second patient presented with generalised weakness and shortness of breath. Cases: First case: Thirty-two-year-old man with past history of H. pylori infection, significant weight loss, on and off fever for 1 year, seizures with recurrent bell’s palsy admitted with complaints of pain in abdomen, nausea and vomiting for 3 days. On evaluation his blood calcium level was raised causing pancreatitis which is very rare in sarcoidosis. Diagnosis of sarcoidosis was very challenging in this situation where chest radiograph was perfectly normal and HRCT thorax revealed multiple small nodules, lymphadenopathy and ILD. Lung biopsy revealed chronic granulomatous inflammation. After ruling out infectious and malignancy causes of hypercalcaemia, the patient was diagnosed as sarcoidosis, treated with steroids, is still in follow up and is doing fine. Second case: Fifty-six-year-old female with past history of H- pylori infection, raised blood pressure, weight loss and underlying heart condition presented with complaints of itching all over body, generalised weakness and shortness of breath on exertion. Patient was wandering for treatment for past 1 year for her non-specific symptoms and was investigated in line of lymphoma in the past. On investigation she was found to have raised creatinine, calcium, calcitriol and anterior uveitis, probable diagnosis of sarcoidosis was made trial of steroids was given which showed slight improvement but she died due to cardiogenic shock. Conclusion: Sarcoidosis is usually underdiagnosed or misdiagnosed or lately diagnosed leading to loss of life which can be prevented. Pancreatitis in sarcoidosis is very rare and careful history, knowledge and investigation may be helpful in diagnosis of it.

Keywords: Rare clinical features, Pancreatitits, Sarcoidosis.

INTRODUCTION

Sarcoidosis – a multisystem disorder, characterised by non-caseating granuloma formation at various organs most common of which is lung and lymph nodes. Its presentation is varied from generalised malaise to renal and lung abnormality. It invariably affects almost every organ. Its clinical features are non-specific for this reason we herein discussed 2 cases who presented with non-specific sign and symptoms and turned out to be sarcoidosis and improved on steroids.

CASE REPORT

Case 1: A 32-year-old male known hypertension with history of H- pylori infection, unintentional weight loss (20kg in a year), fever for 1 year on and off, seizures with recurrent bell’s palsy, history of hepatitis admitted with complaints of pain in abdomen, nausea and vomiting for 3 days. On physical examination blood pressure was raised, there were subcentimetric multiple lymph nodes in bilateral submandibular area which were not suitable for biopsy, eye examination suggestive of anterior uveitis. Blood investigation revealed raised serum creatinine: 3.04 mg/dl, serum calcium :14.2 mg/dl which was normalised on treatment, raised amylase :194 U/L, raised lipase level – 405U/L, raised ACE level :86 U/L, raised serum calcitriol (Vitamin D 1,25 di hyroxy) - 262 (pmol/l) (47.76- 190.32), low iPTH level - 3.75 (pg/ml), Vit-D level :67.7 nmol/L, LFT, lipid profile and complete hemogram was normal. Urine routine and microscopy suggestive of calcium oxalate crystals. X ray skull and chest were normal, MRI Abdomen: bulky pancreas with mild peripancreatic fat stranding with mild splenomegaly. Bone marrow suggestive of normocellular marrow with hemophagocytosis and no evidence of hematolymphoid malignamcy. Viral markers: non-reactive. HRCT thorax revealed multiple small nodules and lymphadenopathy.
BAL was negative for tuberculosis, HPE of lung biopsy suggestive of non-caseating granulomatous lesion. Patient was started on oral glucocorticoids for 1 year, responded well and still in follow up.

**Case 2:** A 56-year-old female with history of H-pylori infection, hypertension, weight loss and underlying heart condition presented with complaints of itching over all body, generalised weakness and shortness of breath on exertion. On physical examination vitals were stable, right submamular lymph node was palpable, hepatomegaly present, eye examination revealed anterior uveitis. Blood investigations revealed high serum calcium level: 15.2 mg/dl, iPTH: 5.5 pg/ml, serum creatinine : 3.35 mg/dl, raised calcitriol levels: 313 pmol/l, raised ACE level: 99.4 U/l, Vit D: 186.2 nmol/l in sufficient range, Serum calcitriol level 313 pmol/l high, CA19.9 <3 U/ML, deranged transaminases level, complete hemogram was within normal limit. CECT Abdomen: multifocal discrete masses in spleen and splenunculi. Few subcentrimetric retroperitoneal lymph nodes. Small homogenously enhancing focus in segment 7- ? hemangioma, tiny calcific foci in liver? calcified granuloma, bone marrow examination: Normocellular marrow with no evidence of hematological malignancy, 2D ECHO: LVEF: 30% dilated all cardiac chambers, global hypokinesia. Montoux test was negative. After ruling out malignancy, tuberculosis her probable diagnosis of sarcoidosis was made on the basis of raised calcium, calcitriol, ACE level. Anterior uveitis, renal and heart involvement, trial of short course of steroids was given and showed some improvement. Before we could prove it by biopsy, she died due to cardiogenic shock.

**DISCUSSION**

Sarcoidosis – non caseating granuloma whose cause is unknown and affects many organs [1], it primarily affects the respiratory system – lung and hilar lymph nodes. Nearly all system of body gets affected [2].

In India exact load of sarcoidosis is unknown. In India it has been calculated that ten to twelve per thousand cases reported annually at a Respiratory Unit at Kolkata where as in Vallabhnbhai Patel Chest Institute (VPCI), Delhi, it is 61.2/100,000 new cases annually. As many cases were not reported or gets undiagnosed, we believe that the data reflected here is not exact and very low from actual burden of disease [3].

Among all other causes of hypercalcaemia like primary hyperparathyroidism, hypervitaminosis D, malignancy, sarcoidosis is the only entity which is difficult to diagnose and whose treatment is cheap and is effective but due to its varied presentations and overlapping features with tuberculosis, it is mostly diagnosed lately.

Our both cases were affected with H. pylori one at other time before the diagnoses of sarcoidosis it has postulated that H pylori infection has gradually replaced the parietal cell with autoimmune process [1].

Our patients had all possible complication of sarcoidosis like H Pylori infection, seizures, recurrent bells paly, uveitis, significant weight loss, renal, cardiac injury and pancreatitis (very rare symptom) at variable interval of time. Sarcoidosis mainly affects lung in 90% of patients and is categorized into 4 stages. Stage 1 is hilar adenopathy alone often with right parastracheal involvement, stage 2 is a combination of adenopathy plus infiltrates, whereas stage 3 reveals infiltrates alone. Stage 4 consists of fibrosis. In early disease chest x ray may be normal and may require CT chest to confirm the same if strong suspicion of sarcoidosis is there. Lung biopsy revealed granulomatous lesions and he was started on glucocorticoids and all his symptoms were resolved.

Only five case reports have been reported earlier, that sarcoidosis resulted into acute pancreatitis which may be due to active granulomatous pancreatitis or secondary to hypercalcemia [4-6].

In developing countries like India where Tuberculosis is more prevalent and mimic like sarcoidosis. Diagnosis is more difficult as it will be very unpredictable the response of is either due to antitubercular drugs or by corticosteroids started in combination. Sarcoidosis incidence in India is increasing and cause of which is still not known.

**CONCLUSION**

Due to its varied presentations, sarcoidosis is often diagnosed late and nearly all complications occurs before actual diagnosis as we saw in our cases. Therefore, keeping in mind, sarcoidosis as differentials will benefit patient as treatment is cheap and effective.

**Conflicts of interest**

The authors declare no conflict of interest.

**Funding**

None.

**Data availability statement**

Data can be found from affiliated centre.

**Author contributions**

Dr. Anita Sharma involved in interpretation and conception of study, Dr. Richa Aswal responsible for providing critical and intellectual input, Dr. Arpit Punetha involved in generation of data, whereas Dr. Shilpa responsible for design of study and drafting manuscript.

**REFERENCES**