Miliary Sarcoidosis with Secondary Sjogren’s Syndrome

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Abstract

We report a case of miliary sarcoidosis with secondary Sjogren’s in a 45-year-old male who presented with symptoms of sicca syndrome in the form of dryness of eyes and mouth with parotid swelling. Computed tomography thorax showed mediastinal and hilar lymphadenopathy, bilateral miliary opacities in lung parenchyma. Whole body FDG PET/CT showed involvement of both parotids, liver, diffuse uptake in lungs, mediastinal and retroperitoneal lymph nodes. Patient is on treatment with prednisolone and has responded well.

Introduction

Sarcoidosis is a multisystem granulomatous disorder. Miliary presentation of sarcoidosis is rare and can be very difficult to distinguish from tuberculosis. It can mimic many of the manifestations of tuberculosis and delay the correct diagnosis. Also, it is rare for sarcoidosis to involve liver and retroperitoneal nodes. In the case presented, FDG PET/CT could help in detecting the extent of disease including involvement of liver and retroperitoneal nodes. There is no reported PET/CT study in miliary sarcoidosis.

Case Report

A 45-year-old male was referred to Army Hospital R and R with clinical features of Sjogren’s syndrome. Presenting complaints were dry eyes, dry mouth and swelling around jaws of three months duration. He had an episode of dry cough, coryza, sore throat and fatigability which lasted for few days. There was sand like sensation in both eyes, decreased salivation, difficulty in swallowing of solid food associated with altered taste sensation and dryness of mouth. Swelling around jaws was insidious in onset, gradually progressive, symmetrical and painless. He also complained of photophobia and blurring of vision. There was no history of weight loss, rash, myalgia, muscle weakness, joint pain, chest pain, exertional breathlessness, hemoptysis, abdominal pain, dyspepsia, numbness or paraesthesia or Raynauld’s phenomena. He denied any history of high risk behavior. Local examination showed crusting of eyelids, dry nose (crusting and epistaxis), dry skin, pruritus and excoriation. Both parotid glands were enlarged and non tender. Mouth showed decreased pool of saliva and loss of papillae from the dorsal surface of the tongue. General examination revealed a well built individual who was afebrile with normal vital parameters. Cardiovascular, respiratory, abdominal and musculoskeletal examinations were normal. Investigations showed Hb : 12.3 g/dl, TLC : 19300/cmm, DLC : P 81 L14 E03 M02 B0 (%). Urinalysis was normal. Blood Glucose (F/PP): 95/105 mg/dl, S. Bilirubin: 1.4 mg/dl, AST /ALT : 27/16 U/L, BUN/ S.Creatinine: 08/0.6 mg/dl, S Iron: 25 mg/dl, TIBC : 955 mg/dl, SAP :124 IU/L, LDH : 723 IU/L, Na/K : 134/4.2 mEq/L, CPK : 12 U/L, Lipids (mg/dl): TC/TG/HDL/LDL :162/148/39/93, CRP:5.50 mg/l, S. Calcium : 13.9 mg/dl. ESR : 86 mm in 1st hr. ACE level was elevated: 113.0 Iu/l. HIV, HBsAg, Anti-HCV were negative. Anti Ro and Anti La were negative. 24 hr urinary Calcium was 620 mg. Anitnuclear antibody and rheumatoid factor were negative. Mantoux test was negative. IgG and IgM for MTB were negative. BAL: Turbid, RBC: 30, WBC: 500/cmm, predominant cell lymphocyte. Lung diffusion study was normal. Computed tomography (CT) of the thorax showed mediastinal and hilar lymphadenopathy with bilateral miliary opacities. Based on these clinical and investigation findings, diagnosis of sarcoidosis was made. To know the extent of disease, PET/CT was requested. PET/CT dated 13.05.11 showed FDG avid enlarged parotid and submandibular glands, diffuse FDG uptake in bilateral lungs fields, FDG avid mediastinal and retroperitoneal lymph nodes and FDG avid lesion in left lobe of liver (Figures 1-4). Patient underwent transbronchial lung biopsy and lip biopsy, both of them showed non caseating granulomas with multinucleate giant cells (Figures 5). Bronchoscopy AFB cultures were negative. Based on findings of whole body PET/CT, diagnosis of miliary sarcoidosis with involvement of liver and retroperitoneal nodes with features of Sjogren’s syndrome was made. Patient is now taking prednisolone 60 mg OD and is responding well.

Discussion

This case was initially thought of as a case of Sjogren’s syndrome and subsequently probable case of miliary tuberculosis, but turned out to be case of sarcoidosis with secondary Sjogren’s syndrome based on clinical findings and detailed investigation.
Patient was referred for PET/CT to see the extent of disease. PET/CT scan revealed metabolically active disease involving the parotids, mediastinal and retroperitoneal nodes, bilateral lung parenchyma and liver.

Sarcoidosis is a multisystem granulomatous disorder characterized pathologically by the presence of non-caseating granulomas in involved organs, occurring in patients between 10 and 40 years of age in 70 to 90 percent of cases. It typically presents with bilateral hilar adenopathy, pulmonary reticular opacities, and skin, joint or eye lesions. Lung is the most common organ involved, which is 95% followed by skin and lymph nodes, 15.9% and 15.2% respectively. X-ray chest and Chest CT can demonstrate a variety of abnormalities in patients with sarcoidosis which includes hilar and mediastinal lymphadenopathy, beaded or irregular thickening of the bronchovascular bundles, nodules along bronchi, vessels and subpleural regions, bronchial wall thickening, ground glass opacification, parenchymal masses or consolidation, parenchymal bands, cysts, traction bronchiectasis, and fibrosis with distortion of the lung architecture. Literature search revealed one CT scan proved case of miliary involvement of sarcoidosis in a patient who was treated for miliary tuberculosis. In our case, detailed investigations could not detect presence of tuberculosis. Patient responded well to treatment for sarcoidosis. Hence the miliary involvement of lung is considered as due to sarcoidosis itself. PET/CT image of miliary sarcoidosis of lung has not been reported to our knowledge.

Reticuloendothelial system disease is common in sarcoidosis, manifest as peripheral lymphadenopathy (40%), hepatomegaly (20%) non-caseating granulomas on liver biopsy with or without hepatomegaly (75%), and splenic enlargement (25%). Hypersplenism can lead to anemia, leukopenia, and thrombocytopenia. Calcium metabolism abnormalities are the most common renal and electrolyte abnormalities observed...
among patients with sarcoidosis. The defect in calcium metabolism is due to extrarenal production of calcitriol by activated macrophages. Patients may present with hypercalciuria (occurs in up to 50 %), hypercalcemia (which occurs in 10 to 20 %), and nephrocalcinosis. ACE level is elevated in 75 percent of untreated patients with sarcoidosis.6

Conclusion

PET/CT scan could delineate the correct extent of disease in this case. Miliary sarcoidosis is rare manifestation of sarcoidosis. To our knowledge, this is the first PET/CT image of miliary sarcoidosis. PET/CT helped in detecting the extent of involvement and the same modality may be useful in response evaluation to treatment.

References

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