OMCR

Oxford Medical Case Reports, 2015; 12, 376–377

doi: 10.1093/omcr/omv068
Case Report

CASE REPORT

Sarcoidosis: a diagnostic challenge in atypical radiologic findings of unilateral lymphadenopathy

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Abstract

Sarcoidosis is a chronic systemic disease with a wide array of clinical findings. Given that the clinical symptoms are not pathognomonic, chest radiographs have become essential to the initial diagnosis and choice of treatment modality. Diagnosis hinges on ruling out alternative diagnoses; sometimes, advanced radiologic techniques and histopathology are required. On this occasion, we present a case of a patient with generalized symptoms, no significant chest radiograph findings and lymphadenopathy where advanced imaging and pathology assisted in the diagnosis.

INTRODUCTION

Sarcoidosis is a chronic systemic disease that was initially described in 1869 by Jonathan Hutchinson [1]. Yet, to date, the specific cause and etiology remain unclear [2]. While erythema nodosum was thought to be a key feature at the time of its discovery, sarcoidosis is now much more commonly recognized via the trio of hilar lymphadenopathy, pulmonary and ocular involvement [1]. Symptoms are often generalized and can be markedly diverse [3]. The diagnosis is based on the clinical presentation and radiological findings. If the presenting symptoms do not specifically point to Löfgren's syndrome or Heerfordt's syndrome ***or*** there is asymptomatic bilateral hilar adenopathy, then the diagnosis is reliant on a biopsy [2]. Atypical presentations often confound the diagnostic effort. Here, we present an atypical presentation of sarcoidosis.

CASE REPORT

Our patient is a 38-year-old Caucasian female with a past medical history of papillary thyroid cancer; she had been treated via thyroidectomy 2 years prior to presentation. She first presented to our outpatient clinic with a 20-pound weight loss over 2 months and a persistent, dry cough.

She had initially presented to the emergency department with a cough, tactile fever and a decreased appetite. She was discharged from the emergency department and given a 6-day course of the antibiotic azithromycin. The following day, the patient developed a Bell's palsy, and she finally returned to the emergency department 1 week later with persistent symptoms and new right-sided neck pain. A chest radiograph was unremarkable. A computed tomography (CT) of the thorax was performed and demonstrated an enlarged lymph node in the aorto-pulmonic window, which had not been seen on prior imaging.

On her initial outpatient visit, the patient had been experiencing a chronic cough, fatigue and a loss of appetite for ~6 weeks. The only noteworthy physical exam finding was a right facial droop. Initial chemistries showed a thyroid stimulating hormone (TSH) of 0.22 μIU/ml (0.450–4.500 μIU/ml), a total triiodothyronine (T3) of 72 ng/dl (71–180 ng/dl), a free thyroxine (T4) of 2.63 ng/dl (0.82–1.77 ng/dl), a thyroglobulin antibody of <1.0 IU/ml (0.0–0.9 IU/ml) and a thyroglobulin of <0.1 ng/ml (1.5–38.5 ng/ml). A positron emission tomography (PET) scan showed

Received: October 27, 2015. **Revised:** November 13, 2015. **Accepted:** November 17, 2015

multiple active lymph nodes in the mediastinum with increased activity in the liver at the junction of the left and right lobes.

An endobronchial ultrasound and video-assisted thoracoscopic surgery were performed. A mediastinal mass (4.0 × 2.5 × 2.4 cm in dimension), a large aorto-pulmonary lymph node and two sub-carinal lymph nodes were removed for permanent pathology analysis.

The cytology showed polymorphous lymphocytes, benign bronchial epithelial cells and alveolar macrophages. The mediastinal mass and lymph node biopsy findings indicated granulomas predominantly non-caseating lymphadenopathy with focal necrosis. Pathology was negative for malignancy.

The remaining diagnostic puzzle was the etiology of the lesion in the liver. A magnetic resonance imaging (MRI) scan of the abdomen with and without contrast was performed illustrating two non-enhancing, 1.3-cm lesions straddling the falciform ligament. The enhancement pattern was atypical for metastasis.

This combination of clinical and pathology findings led to a diagnosis of sarcoidosis. The patient has since followed up with evaluation for pulmonary function studies. No systemic treatment has been initiated at this time. The patient has been followed clinically with mild symptomatic improvement. A repeat MRI will be performed again in 3 months.

DISCUSSION

Sarcoidosis is a chronic systemic disease that continues to have an uncertain etiology. Diagnosis is based on clinical, radiologic and histopathologic findings [1, 2]. Broadly described, it is a multisystem inflammatory disease that can affect almost any organ [4]. Sarcoidosis has a strong genetic component and is more prevalent in African Americans with rates 3.6–8 times higher than Caucasians of European descent [4].

The initial presentation typically has pulmonary findings; these are detected in 90% of suspected patients who received chest radiographs [3]. Half of sufferers are typically asymptomatic at the time of diagnosis [3]. The most common symptoms are cough and dyspnea. The most common extra-pulmonary findings include peripheral lymphadenopathy, skin and eye manifestations [2]. Being a systemic disease, thyroid dysfunction has been caused by sarcoidosis with some cases that mimic metastatic thyroid cancer [5, 6]. The chest radiograph can be sufficient for diagnosis in the proper clinical contexts [7]. These include asymptomatic hilar lymphadenopathy, Löfgren's syndrome (bihilar lymphadenopathy, arthralgia and erythema nodosum) or Heerfordt's syndrome (parotitis, Bell's palsy, anterior uveitis and fever). When these pathognomonic sarcoidosis syndromes are identified, a biopsy for diagnostic confirmation is not required [3, 7].

The current radiological staging is based on bilateral hilar lymphadenopathy, parenchymal disease and indicators of lung fibrosis. CT scans should be considered with atypical clinical or radiographic findings and evaluation of suspected complications of other concurrent pulmonary diseases [7, 8]. PET scans are not recommended for standard workup, but may be necessary in rare circumstances to clarify a difficulty diagnosis. This includes explanation of extrathoracic symptoms and determination of activity of inflammation. In addition, MRI has most often been utilized for evaluation of myocardial sarcoidosis [7].

A typical biopsy classically consists of non-necrotizing granulomas surrounded by lamellar hyaline collagen containing macrophages, epithelioid cells and $CD4^+$ lymphocytes [1, 9]. However, these findings by themselves are not specific for sarcoidosis and require correlation with other clinical and radiological findings. Treatment is reserved for symptomatic pulmonary sarcoidosis with parenchymal infiltrates. Additional indications for treatment include, but are not limited to, ocular pain or loss of vision, cardiac arrhythmias, heart failure and other types of end-organ failure [10]. Initial treatment is typically with systemic corticosteroids (usually prednisone) until amelioration of symptoms allows an opportunity to wean the corticosteroids. Anti-metabolite and biologic agents can be considered in cases where there is insufficient clinical improvement or an inability to wean the corticosteroids [10].

The patient presented had a history of thyroid cancer with no epidemiologic or obvious genetic risk factors for sarcoidosis. An initial chest radiograph had only subtle abnormalities. It was fortunate that a PET scan was performed, revealing the multiple active mediastinal lymph nodes, which were later biopsied. Due to the biopsy result, sarcoidosis became the likely diagnosis. Sarcoidosis remains at times a difficult diagnosis, particularly where there are no chest radiograph findings. Careful evaluation and maintaining a wide differential and a high clinical suspicion are needed when other signs are fleeting.

ETHICAL APPROVAL

Conforms to standards currently applied in the country of origin.

CONSENT

Retrieved.

CONFLICT OF INTEREST STATEMENT

None declared.

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