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A 48-Year-Old Woman with Left Subclavian Artery Occlusion and Intrathoracic Lymphadenopathy

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ABSTRACT

Sarcoidosis is a systemic disorder characterized by noncaseating granulomas, involving multiple organs including thyroid and great vessels. We present a 48 year-old women with sarcoidosis, left subclavian artery occlusion and sarcoidal thyroid gland involvement. The patient presented with a 1 week history of progressive left upper limb pain with coldness of left hand and fingers. On examination, radial, ulnar, and brachial artery pulses were not palpable. She had also enlarged thyroid gland with firm consistency. CT angiography of aortic arc demonstrated occlusion of left subclavian artery. Because of progressive ischemic necrosis of left hand and fingers, amputation above elbow was performed. Fine needle aspiration (FNA) was suspicious for thyroid neoplasm and total thyroidectomy was performed. Thoracic CT scan showed mediastinal and bilateral hilar lymphadenopathy. Fiberoptic bronchoscopy with transbronchial needle aspiration (TBNA) from right hilar lymph nodes and endobronchial biopsy showed multiple granulomas with negative acid-fast stain. Pathologic examination of thyroid also revealed fibrosis and granulomatous inflammation. On follow up, the ACE level was 104 u/l. ***(Tanaffos2010; 9(3): 75-79)***

Key words: *Sarcoidosis, Sarcoidal thyroid involvement, Subclavian artery occlusion, Mediastinal lymphadenopathy*

INTRODUCTION

Sarcoidosis is a systemic disorder characterized by noncaseating granulomas, involving multiple organs with highly variable clinical course and outcome (1-4). The diagnosis is based on compatible clinical and/or radiographic manifestations, noncaseating granulomas on histopathological examination and also exclusion of other disorders with similar clinical and pathologic findings (5).

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The most commonly involved organ is the lungs, seen in >90% of patients, sometimes with atypical radiologic manifestations (5,6). Other organs including central nervous system, eyes, heart, joints, kidneys, gastrointestinal tract, thyroid and great vessels may be involved (7-10).

CASE SUMMARIES

A 48-year-old woman was admitted to the hospital with a 1 week history of progressive left upper limb pain. She also complained of coldness and color change of her left hand and fingers. She denied any fever, dyspnea, cough, articular pain and

swelling. Her medical history was significant only for thyroid enlargement for 4 years, for which she was receiving therapy with Levothyroxine.

Physical Examination

On physical examination, the patient had a temperature of 37.3ºC, respiratory rate of 14 breathes/min, heart rate of 100 beats/min, and right arm BP of 125/85 mmHg. She had enlarged thyroid gland with firm consistency without bruits or palpable nodules. Cardiac examination was normal other than sinus tachycardia. Chest auscultation and percussion were normal. Left hand and fingers and forearm were cold with skin mottling. Radial, ulnar, and brachial artery pulses were not palpable. There was tenderness in palpation of thenar and hypothenar muscles. The patient was alert with no neurologic deficits other than sensory deficit in her left upper limb.

Laboratory findings

Laboratory evaluation revealed normal WBC and platelet counts but low hemoglobin level of 8.6 g/dl with MCV of 61.1, MCH of 19.2 and MCHC of 31.4 g/dl. The erythrocyte sedimentation rate was 8 mm/h. Electrolytes, renal parameters, hepatic panels, and coagulation profiles were all normal. A chest radiography revealed bilateral hilar enlargement (Figure 1).

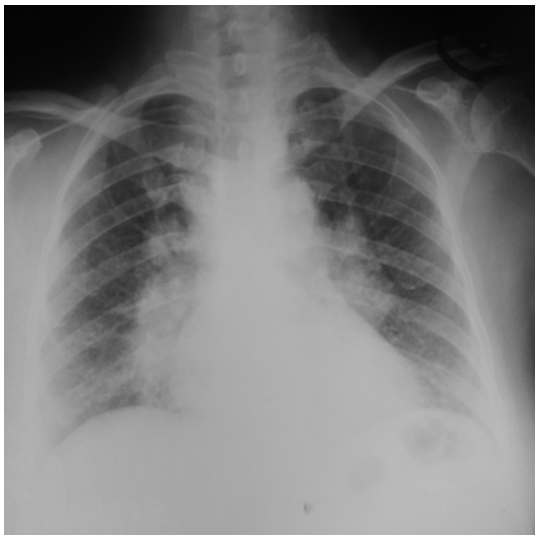

Figure 1. Initial chest radiograph demonstrates bilateral hilar enlargement.

Transthoracic echocardiography revealed normal ejection fraction, normal chamber sizes, and no valvular disease. CT angiography of aortic arc and its branches is shown in figures 2 (A,B,C).

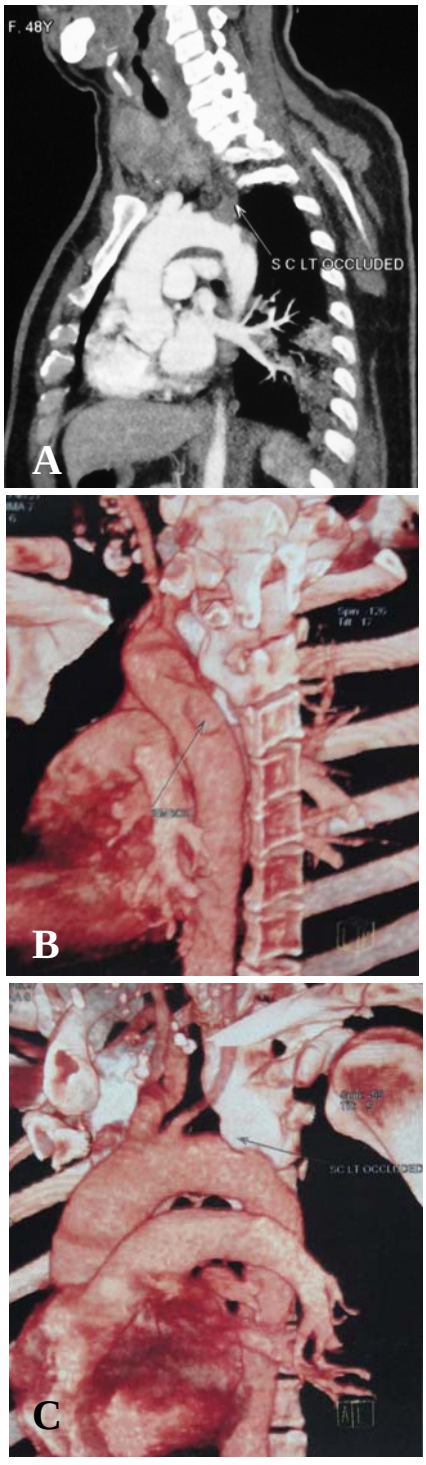

Figure 2. CT-angiography of aortic arc demonstrating occlusion of left subclavian artery.

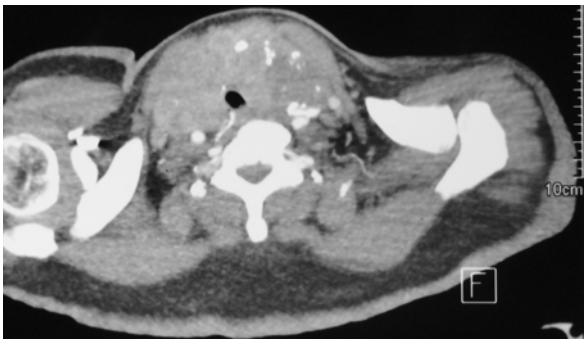

Figure 3. CT scan demonstrating thyroid enlargement with calcification.

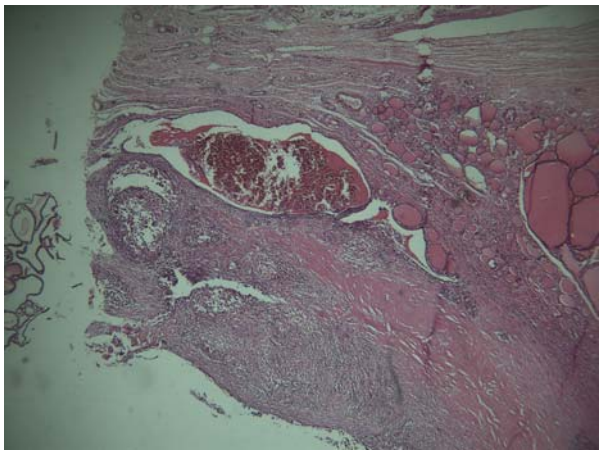

Figure 4. Section of thyroid on high power revealing fibrosis and granulomatous inflammation.

Thoracic CT-scan showed right paratracheal, periaortic, bilateral hilar, and subcarinal lymphadenopathy. Fiberoptic bronchoscopy with TBNA from right hilar lymph nodes and endobronchial biopsy were performed. Multiple granulomas with negative acid-fast stain were reported. On follow up, the ACE level was 104 U/L. The diagnosis of sarcoidosis with left subclavian artery occlusion and sarcoidal thyroid gland involvement was made based on clinical, radiologic, and pathologic findings.

DISCUSSION

Although multiple organs may be involved in sarcoidosis, the constellation of intrathoracic lymph node, thyroid, and aortic involvement, as seen in our patient, has not been reported before as far as we know.

The first description of sarcoidal thyroid involvement was in 1938 (11). Although, thyroid involvement is rare (12), variable clinical presentations have been reported including painful enlargement of the gland, painful nodule, multinodular goiter with hyperthyroidism, diffuse toxic goiter, euthyroid goiter, cold thyroid nodule, graves`disease, and subclinical hypothyroidism (12-19). Thyroid involvement in sarcoidosis most commonly is associated with intrathoracic findings but isolated thyroidal sarcoidosis has also been reported (20, 21).

Involvement of great vessels including aorta, superior vena cava, pulmonary artery and veins has been found in sarcoidosis (22). Pulmonary embolism, diffuse arterial thrombosis, upper extremity venous thrombosis and systemic emboli associated with mural thrombi have also been reported (23-26). Antiphospholipid antibodies were detected in sarcoidosis but they were not associated with arterial or venous thrombosis (27).

Although Takayasu's arteritis, giant cell arteritis and Behçet's disease are the most common causes of inflammatory aortitis, it is rarely associated with sarcoidosis (28).

Association of sarcoidosis and Takayasu’s arteritis was reported in several studies, sometimes with many years lag between detection of the two diseases (29). It suggested that Takayasu’s arteritis or Takayasu’s arteritis –like granulomatous vasculitis probably are a complication of sarcoidosis and to detect asymptomatic underlying inflammatory arteritis, complete vascular clinical examination has been recommended in patients with sarcoidosis (30). On the other hand, in patients with Takayasu’s arteritis, nodular skin lesions which are atypical in this disease should be carefully evaluated by skin biopsy to assess the presence of concomitant sarcoidosis (31).

In conclusion, as shown in the present report great vessels involvement has been found in sarcoidosis,

therefore, complete vascular clinical examination should be considered in patients with sarcoidosis. Thyroid involvement is rare in sarcoidosis, but variable clinical presentations may be seen, most commonly in association with intrathoracic findings.

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