A Case of Sarcoidosis: Multiple Osseous Nodule and Thyroid Involvement

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Abstract

Sarcoidosis is a chronic granulomatous inflammatory disease of unknown etiology. It affects predominantly lungs, thoracic lymph nodes, skin and eyes. Thyroid gland, skull and vertebrae are rarely involved. The diagnosis is suggested on the basis of clinical and radiologic manifestations and is supported by the histological demonstration of noncaseating granulomas in the affecting tissues. In recent years FDG positron emission tomography (PET) has proved to play an important role in the diagnosis of sarcoidosis with systemic distribution. We reported a unique case of sarcoidosis with Horner’s syndrome, thyroid gland involvement and multiple osseous nodules in vertebrae and skull detected by PET scan, in response to corticosteroid treatment.

INTRODUCTION

Sarcoidosis is a multifocal, immune mediated, chronic inflammatory systemic disease of unknown etiology [1,2]. The overall prevalence of sarcoidosis is 10-40 cases per 100,000 populations. Females affected more often than males. Age of onset is usually during early adulthood [3]. Accurate diagnosis often requires pathologic examination. However, in recent years FDG positron emission tomography (PET) has proved to play an important role [4]. We report a female case of sarcoidosis patient with previous biopsy proven thyroid involvement, who presented with recent Horner’s Syndrome and multiple osseous nodules detected on PET scan.

CASE PRESENTATION

A sixty one year old female has been followed up in our clinic for sarcoidosis since 2003. She had bilateral subtotal thyroidectomy due to multinodular goiter 25 years ago. In 2004, an enlarged remnant thyroid gland has found on the CT scans. A thyroid biopsy was performed to exclude malignancies and a noncaseating granulomatous process was determined. After ruling out other causes of granulomatous diseases, sarcoidosis involvement was diagnosed (Figure 1A). In 2006, she had a Lumber MRI because of low back pain. Widespread nodular vertebral implants, compression fracture and a narrowed spinal canal at L4-L5 were observed (Figure 1B). By the bone marrow aspiration biopsy, any malignancies or metastatic diseases were excluded. The patient underwent cyphoplasty operation. After the operation, she noticed that her left eyelid dropped and left side of her face remained dry when she sweated. She started coughing two months after the surgery. On physical examination ptosis and miosis detected on her left eye and anhidrosis on the left side of her face. Crackles were present at the left lung base on auscultation. Other systemic findings were normal. A positive phenylephrine test supported the diagnosis of Horner’s syndrome. Laboratory tests were normal except for hypercalciuria (438 mg/24 hr urine) and she had a minimal peripheral airway obstruction. On CT scan of the thorax; the thyroid gland was enlarged, paratracheal, prevascular, parasternal, bilateral hilar lymphadenopathies and multiple widespread parenchymal, sub pleural and pleural nodules were also observed (Figure 1C). A cranial CT was performed with normal findings. Cranial MRI scans showed 1 cm diameter hyper intense lesions on the sphenoid bone on T2 weighted sequences and 1.2x1 cm diameter hypo intense lesions on the clivus on T1 weighted sequences (Figure 1D). The brain tissue was normal. PET scan showed increased activity on the clivus, sphenoidal bone and multiple locations on the skin. In addition; cervical, bilateral supraclavicular, bilateral axillary, mediastinal, prevertebral, bilateral inguinal and iliac lymph nodes; lungs, sternum, costae, vertebrae, pelvic bones, bilateral femur and humerus also showed increased activity (Figure 2A). Bone scintigraphy was also done with normal findings. With all these findings, we considered the nodular bone lesions as sarcoid involvement and Horner’s Syndrome was assessed to be secondary to the nodular involvement in the cranial bones. Since she had multiple osseous involvements and calciurea, we started chloroquine (2x200mg/day) and prednisolone (60 mg/day) as a treatment. Two months later her control PET scan was found normal, Figure 2B but Horner’s syndrome persisted. The patient is doing well on oral steroid treatment.

DISCUSSION

A wide array of organs and tissues are involved in sarcoidosis. The most prominent sites were lungs. Spleen, liver, bone, heart, kidney or central nervous system was affected in 2% to 6% of the patients. Asymptomatic involvement of these organs is far more common. Thyroid involvement has been often described but rarely confirmed histologically. In the review of the literature, there are less than 50 documented cases with sarcoid involvement of the thyroid gland [5]. Patients with mediastino-pulmonary sarcoidosis with a thyroid nodule like our case; should be investigated for sarcoidosis thyroid involvement [6,7]. The incidence of skeletal involvement ranges from 3% to 36% [1,8]. Bone lesions occur most frequently in the phalanges of the fingers and toes. Skull, vertebrae, and pelvis are less commonly affected areas [7]. Clinically, skeletal sarcoidosis is usually well tolerated and causes few symptoms. Radiographic evaluation of the skeleton often shows normal findings. For diagnosis, bone scintigraphy is more sensitive to locate involvements than radiography. MRI is the currently preferred technique in
detection and specific evaluation of the lytic and sclerotic lesions [8]. FDG PET showed to be a sensitive tool for evaluating the bone involvement [1]. Moreover, it was reported that FDG PET appear to be more accurate and contributes to a better evaluation of extra pulmonary involvement in sarcoidosis patients but it should be kept in mind that it can mimic widespread skeletal metastases [9]. In our case, widespread nodules on vertebrae and the skull were detected incidentally on lumber and cranial MRI. After excluding any malignancies by bone biopsy and with the increased FDG uptake shown on the PET, these nodules were assessed as sarcoid lesions.

As known, integrating PET with the glucose analogue 2-18F-fluoro-2-deoxy-d-glucose (FDG) has made a significant impact on the management of many types of solid-organ malignancy [8]. Furthermore, it has also been recognized that FDG accumulates not just in tumor tissue, but also in areas of active infection and inflammation. The uptake of FDG increases when the metabolic activity is increased by inflammatory cells. In sarcoidosis, granulomatous lesions in which the activated macrophages play important role, shown a strong FDG uptake. The degree of FDG uptake has been related both to the activity of disease and to the response of the treatment [10]. Lewis et al., showed that FDG was taken up at sites of intra- and extra thoracic involvement with sarcoidosis and several larger studies have confirmed the utility of FDG PET and PET/CT in the evaluation of these patients [11]. In the present case, all intrathoracic and intra abdominal enlarged lymph nodes and nodular osseous lesions captured FDG. After two months of steroid and chloroquine treatment, a control PET scan documented the disappearance of the osseous nodules supporting our diagnosis of osseous involvement of sarcoidosis.

In sarcoidosis, Horner’s syndrome is not a common finding. Lower brainstem, eye, middle ear and hypophysis lesions can also cause Horner’s syndrome [12]. In our case, sarcoid nodules on the cranial bones neighbouring the course of the sympathetic chain are thought to be the most probably cause of the Horner’s syndrome. Although the osseous lesions disappeared after treatment, Horner’s syndrome still ensued probably owing to the slow remodeling of the nervous tissue. This patient is a unique case of sarcoidosis presenting with Horner’s syndrome, very rare site involvements, namely the thyroid gland and osseous tissue. It should be emphasized that PET scan can be a useful tool in the diagnosis and follow-up of sarcoidosis patients.

REFERENCES