Tanaffos (2009) 8(2), 69-71

A 49-Year-Old Woman with Refractory Chronic Sinusitis

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WHAT IS YOUR DIAGNOSIS?

A 49-year-old married, non- smoker housewife had purulent rhinorrhea, nasal congestion, post nasal drip (PND), and feeling of sinus pressure following an episode of common cold. She had no complaints of fever, cough, dyspnea, or arthralgia. Physical examination revealed PND and a posterior auricular lymphadenopathy (1cm × 1cm). The patient received amoxicillin for 2 weeks with mild improvement. But again, she experienced the exacerbation of signs and symptoms a week later. CT-scan of the paranasal sinuses was performed (Figure 1). She also had erythematous patches on her face around the right eye and her back, along with splenomegaly.

The patient was treated with amoxicillin/clavulanate (for two weeks), loratadine, and beclomethasone nasal spray as well as normal saline nasal wash. There was a slight improvement in patient's signs but, repeatedly after two weeks signs and symptoms exacerbated. Due to the lack of response to antibiotics, a biopsy of the skin lesion was taken which revealed the following histopathological findings (Figure 2). ***(Tanaffos 2009; 8(2): 69-71)***

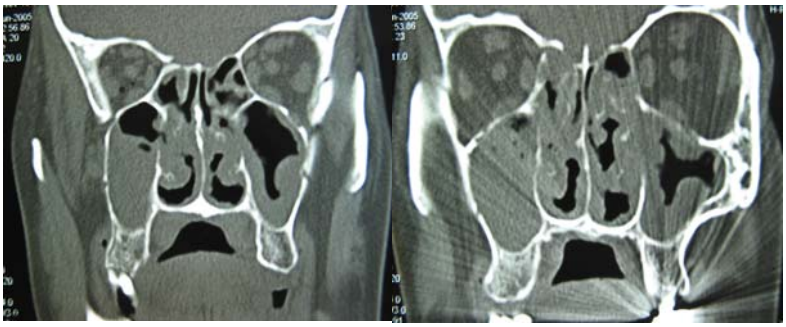

Figure 1. CT-scan of the paranasal sinuses

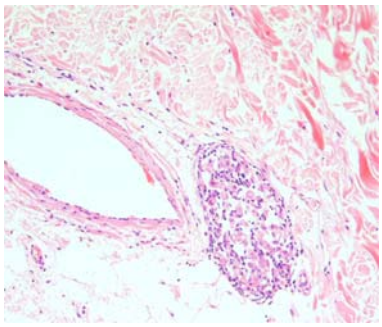

Figure 2. Histopathology of the skin biopsy sample

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Diagnosis: Sarcoidosis with granulomatous sinusitis

Chest x-ray (Figure 3) demonstrated bilateral hilar lymphadenopathy and histopathological examination of the skin lesion, sinus and posterior auricular lymph node revealed non-necrotizing granulomatosis (Figure2). Laboratory examinations showed high ACE (Angiotensin Converting Enzyme) level, normal level of C-ANCA and negative PPD test. According to the constellation of findings on chest x-ray, histopathological examination of the skin, lymph node and sinus associated with elevated ACE level, and lack of response to multiple courses of antibiotic therapy and exclusion of other causes of granulomatous sinusitis, the diagnosis of "sarcoidosis" was confirmed.

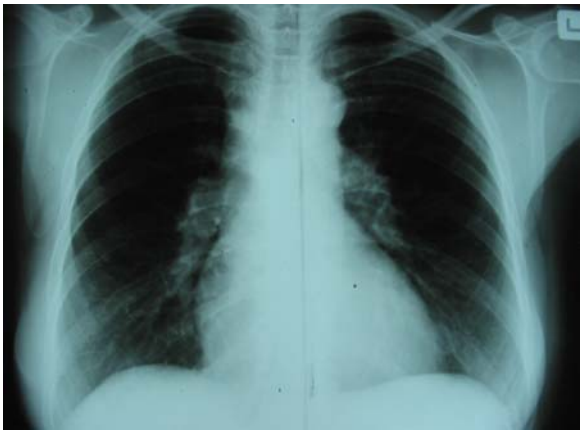

Figure 3. Chest x ray with bilateral hilar lymphadenopathy

Sarcoidosis is a chronic, multisystemic disorder of unknown cause. All body parts can be involved, but the organ most frequently affected is the lungs. Involvement of the skin, eye, liver and lymph nodes is also common (1).

Sarcoidosis is more commonly found in young adults, between the third and fifth decades. There is a female predilection of 2: 1, which is even higher in those with upper respiratory tract manifestations (3). Incidence of sinonasal sarcoidosis is not precisely known but appears to be reasonably low and is estimated that <1% of patients with sarcoidosis ultimately display sinonasal involvement (4).

The first manifestation of systemic sarcoidosis often affects the upper respiratory tract (3). Nasal and paranasal sinus involvement can be most difficult to diagnose and treat properly. Presenting symptoms are usually nonspecific, including nasal congestion, postnasal drip, sinus pressure and headache (2).

Sarcoidosis uncommonly occurs in the sinonasal system and rarely in the absence of pulmonary disease (5). Sinonasal sarcoidosis is unusual and may be misdiagnosed (6). Several other diseases involving the sinonasal region are characterized by granulomatous lesions. Thus, the diagnosis of sinonasal sarcoidosis is often difficult, and is made by correlating clinical, radiological, histopathological, and laboratory findings (7). Diagnostic criteria for sinonasal sarciodosis consist of:

1. Histopathological confirmation of non-necrotizing granuloma.
2. Chronic rhinosinusitis poorly responsive to conventional treatments and radiological evidence of rhinosinusitis, often nodules on the septum and/or the turbinates.
3. Elevated level of angiotensin converting enzyme.
4. Positive gallium scan if performed.
5. Evidence of systemic particularly pulmonary sarcoidosis.
6. No evidence of other granulomatous diseases, such as Wegener's granulomatosis (7).

There are a number of management strategies for sinonasal sarcoidosis, depending on disease severity. Krespi and colleagues have proposed a staging

system that provides some general guidelines for therapy. Stage I is defined as a mild nasal disease without paranasal sinus involvement. These patients can typically be managed successfully with saline nasal spray, nasal irrigation, and topical nasal steroids. Stage II is a moderate disease, with involvement of both nasal and paranasal sinuses; it is typically treated with both stage I therapy and intraregional steroids. Stage III is characterized by severe, often irreversible, nasal and sinus disease that usually requires the therapeutic interventions of stage I and II, as well as systemic therapy. Courses of systemic antibiotics are also required for many patients who develop secondary infections related to mucus stasis or sinus ostium obstruction. Surgical interventions have traditionally been avoided due to concerns about worsening the tissue destruction and inflammation; however, some reports suggest that surgery can be performed safely and successfully in selected cases. Examples of surgical indications include severe nasal obstruction that has failed to respond to medical therapy, sinus obstruction with chronic infection, and biopsy for diagnosis (2).

The therapy of choice for sarcoidosis is glucocorticoids. Methotrexate is usually the second-line medication. Other various drugs have been used in refractory cases, including indomethacin, oxyphenbutazone, chloroquine, hydroxychloroquine, thalidomide, infliximab, etanercept, pentoxifylline, tacrolimus, allopurinol, levamisole, azathioprine, p-aminobenzoate, and cyclophosphamide. Cyclosporine is ineffective for the pulmonary manifestations of the disease; anecdotal reports suggest that it may be useful in extrathoracic sarcoidosis not responding to glucocorticoids (1).

The patient was treated with prednisolone and later addition of methotrexate, then azathioprine and finally cyclosporine with no response. Consequently, a 3-month course of hydroxychloroquine sulfate (200 mg/ daily) led to complete recovery of clinical signs and symptoms which is being continued, though there was no sign of improvement in the control sinus CT-scan obtained 6 months after initiation of chloroquine treatment.

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