Löfgren’s Syndrome with Splenic Involvement: A Case Report

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SUMMARY

Löfgren’s syndrome is an acute form of sarcoidosis characterized by fever, erythema nodosum (EN), bilateral hilar lymphadenopathy (BHL), and polyarthritis. This syndrome is a self-limiting disease. A 45-year-old woman was admitted to our hospital complaining of fever, visual blurring, and arthralgia. A chest radiograph showed bilateral hilar mass lesions. High-resolution computed tomography (HRCT) scans of the chest revealed multiple nodules at different segments of the lung, bilateral hilar and paraesophageal lymphadenopathy. Also, abdominal and pelvic CT scan showed mild splenomegaly, with multiple hypodense nodules in the spleen and an accessory spleen. She was diagnosed with Löfgren’s syndrome.

Key words: Löfgren’s syndrome, sarcoidosis, polyarthritis, erythema nodosum

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INTRODUCTION

Sarcoidosis is a granulomatous disorder of unidentified cause that affects several organs in the body (1). This systemic disease is characterized by non-caseating granulomatous inflammation with some giant cells. Sarcoidosis may affect the lymph nodes, eyes, heart, spleen, and skin (2). This disorder produces enlarged lymph nodes of pulmonary hila. About 90% of patients with sarcoidosis have lung engagement (3). This multi-system disease involving the musculoskeletal system is infrequent. Furthermore, it can involve the synovitis of large joints of lower extremities and proximal muscles (4). Sarcoidosis usually occurs in young and middle-aged people, mainly affecting women (5). Timely diagnosis of this disorder still poses difficulties because of varying presentations. For that reason, clinicians must be aware of the varied range of this disease manifestations, and avoid a delay in exact diagnosis and treatment. Löfgren’s syndrome is an acute form of sarcoidosis and approximately 30% of sarcoidosis cases present with Löfgren’s syndrome (6). It is characterized by several signs and symptoms such as fever, rash, erythema nodosum (EN), polyarthritis, and bilateral hilar lymphadenopathy (BHL) (7). Its prognosis is excellent and almost all of these cases respond to corticosteroid therapy. Herein, we report a middle-aged woman with Löfgren’s syndrome with splenic involvement.

CASE REPORT

A 45-year-old woman presented to the Infectious Disease Clinic with three weeks of intermittent fever, visual blurring, and arthralgia. She had no other complaints but an acute episode of rectorrhagia. On examination, she had frank bilateral ankle arthritis and EN was noted bilaterally over the lower limbs. Ophthalmic examination showed blood-vessel branching suggestive of scleritis. The results of laboratory analysis were as follows: serum glutamic-oxaloacetic transaminase (SGOT), 30 U/L; serum glutamic pyruvic transaminase (SGPT), 23 U/L; alkaline phosphatase (ALP), 154 U/L; lactate dehydrogenase (LDH), 340 U/L; creatine kinase (CK), 1213 U/L; C-reactive protein (CRP), 2+; erythrocyte sedimentation rate (ESR), 58 mm/h; serum angiotensin-converting enzyme (ACE), 121 U/L; serum calcium, 7.3 mg/dL. The serum agglutination test (SAT) and 2-mercaptoethanol (2 ME) agglutination were negative. Stool examination results and complete blood count (CBC) were normal. Her chest x-ray showed bilateral hilar mass lesions (Figure 1). High-resolution computed tomography (HRCT) scans of the chest revealed multiple nodules at different segments of the lung, bilateral hilar and paraesophageal lymphadenopathy. Abdominal and pelvic CT scan showed mild splenomegaly, with multi-
Coronal CT scan disclosed splenomegaly, with multiple hypodense nodules in the spleen and an accessory spleen (Figure 2, 3). Echocardiography found no vegetation. Colonoscopy was performed because of her rectorrhagia that showed decreased vascularity.

After consulting a pulmonologist, an almost definite diagnosis of sarcoidosis was made based on her complaints and imaging findings. Despite the oncologist’s recommendation to perform biopsy of the lymph nodes to rule out lymphomas as the most probable differential diagnosis, according to the pulmonologist opinion, her diagnosis was definite, and our patient was diagnosed with Löfgren’s syndrome based on her symptoms. Therefore, we treated the patient with prednisone 50 mg daily with a slow tapering rate, without the need for biopsy. Her symptoms and radiographic abnormalities resolved progressively and responded well to treatment.

DISCUSSION

Sarcoidosis is a chronic granulomatous and inflammatory disease and Löfgren’s syndrome is an acute form of sarcoidosis (8). What should be taken into account in diagnosing this disorder is that its differential diagnosis with other diseases such as lymphomas, carcinoma, and tuberculosis should be evidence-based. Two of the following three criteria are needed to diagnose the Löfgren’s syndrome: EN, BHL, and arthralgia (7). Our patient had all of these three criteria. Symmetric arthritis in sarcoidosis is very common and ankles are frequently involved joints in this disease; therefore, it is reported in over 80% of patients (9). Along with the wrists, knees, and elbows are engaged but are less visible. Our patient presented with bilateral ankle arthritis.

Ophthalmic involvement in sarcoidosis includes posterior, intermediate, and anterior uveitis (iritis), conjunctival involvement, iris granuloma, posterior synechiae, pupillary abnormalities, episcleritis, scleritis, cataract, vitreous, retinitis, macular edema, and many other manifestations (10). Scleritis was also diagnosed in this case.

The pulmonary involvement is the most usual presentation in Löfgren’s syndrome, so that cough, pain, and heaviness in the chest and breathing difficulty are common (11). Our patient had radiographic signs of lung involvements, but these symptoms were absent.

Spleen involvement is not uncommon during sarcoidosis as approximately 5% of cases with sarcoidosis have splenomegaly on the physical examination and about 30% on imaging (12). Hypodense splenic nodules are seen on CT in about 10% of cases (13). This is similar to what happened to our patient. Moreover, splenic granulomas are found at autopsy in about half of the patients with sarcoidosis and hypersplenism due to sarcoidosis, which can lead to pancytopenia (14). A chest x-ray showed bilateral hilar mass lesions and chest HRCT revealed bilateral hilar and paraesophageal lymphadenopathy. It should be noted that tissue biopsy of the most accessible lesion is needed for an exact diagnosis in the absence of Löfgren’s syndrome. Therefore, to further investigate these pulmonary masses, biopsies of hilar lymph nodes were recommended by our oncologist, but due to the typical manifestation of Löfgren’s syndrome as the classic criteria, we concluded that biopsy was unnecessary and eventually the treatment began.

Erythema nodosum, as a kind of panniculitis, is observed in 90% of Löfgren’s syndrome patients. Sarcoidosis-associated erythema nodosum is usually self-limited and responds to steroids, which indicates good and proper prognosis (15). In our case, a sarcoidosis-related symptom rapidly resolved after the steroid treatment, which was compatible with the characteristics of erythema nodosum in Löfgren’s
syndrome. Laboratory data can help to confirm the diagnosis. Meanwhile, it is important to note that an elevated serum ACE level is not a reliable indicator of disease activity and excludes Löfgren’s syndrome, but it can be used as a marker for follow-up of treatment responses and monitoring disease activity (16).

CONCLUSION

As described, Löfgren’s syndrome comprises a triad of BHL, erythema nodosum, and arthritis. Having two of these criteria confirms the diagnosis of the disease, along with investigating the presence of unusual engagement of Löfgren’s syndrome, such as spleen involvement which should be taken into consideration by clinicians. Also, exclusion of diseases such as lymphomas should also be considered in the exact diagnosis.

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Conflict of interests

All of authors report no conflict of interest.


Lofgrenov sindrom i zahvaćenost slezine: prikaz slučaja

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SAŽETAK


Ključne reči: Lofgrenov sindrom, sarkoidoza, poliartritis, eritema nodozum