Aseptic Abscess Syndrome with Severe Skin Involvement: Case Report

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Sir,

Aseptic abscess syndrome is an autoinflammatory disease that was first reported in 1995.[1] It manifests as round and sterile abscesses composed of neutrophils, and it involves the liver, abdominal lymph nodes, lungs, skin, and especially the spleen. The symptoms include fever, elevated erythrocyte sedimentation rate, and elevated C-reactive protein; however, it does not respond to antibiotic therapy. Its response to steroids is excellent.[2] Aseptic abscess syndrome needs a quick diagnosis, and a differential diagnosis should be done as well.[1-3] We report a case of multiple abscesses involving the spleen, liver, and skin, who was treated with steroids and colchicine.

A 43-year-old male patient presented with complaints which included fever, fatigue, weight loss, and multiple wounds with discharge on the extremities. His history included abdominal pain, nausea, and fatigue that started 4 years ago, and an abscess was found in the liver with magnetic resonance imaging. The patient was followed up for malignancy, and he was managed with symptomatic treatment. He had severe abdominal pain 1 month back, and then multiple splenic abscesses were identified; hence, he underwent a splenectomy. He developed abscesses on the distal parts of the upper and lower limbs, with purulent discharge that rapidly increased 2 weeks after splenectomy; therefore, he used ceftriaxone for 14 days but did not benefit from the therapy. The dermatologic examination of the patient on admission revealed that the distal upper and lower extremities had a number of subcutaneous nodules, each limb had 6–7 nodules, especially centered around the wrists and ankles [Figure 1]. These nodules then deepened to form abscesses, and finally, well-circumscribed necrotic ulcers with detached edges appeared, which were formed by the dehiscence of the abscesses [Figure 2]. The ulcers extended to the tendon over time, and there were lesions in different phases that progressed in the same way.

The laboratory workup was as follows: C-reactive protein level was 213 mg/L (0–5); erythrocyte sedimentation rate was 110 mm/h (0–30); white blood cell count was 18,000 (3.98–10.2); aspartate aminotransferase level was 82 U/L (<50); alanine transaminase level was 142 U/L (<50); gamma-glutamyl transferase level was 235 U/L (<55); alkaline phosphatase level was 265 U/L (30–120); and procalcitonin level was 0.02 ng/mL. No growth was observed in blood, urine, or throat cultures. The patient received ertapenem, meropenem, and daptomycin for 14 days; however, no clinical or laboratory response was achieved. The workup results for human immunodeficiency virus, the venereal diseases research laboratory test, the treponema pallidum hemagglutination assay, hepatitis B virus, hepatitis C virus, cytomegalovirus, Brucella, toxoplasma, and the antineutrophil cytoplasmic autoantibody test were all negative. Tissue samples from ulcerous lesions were collected for the examination of nonspecific bacteria, aerobes, anaerobes, and tuberculosis, as well as a mycotic examination. No growth was seen in cultures, and the tuberculosis complex polymerase chain reaction was negative.

Considering the differential diagnosis, polyarteritis nodosa, disseminated tuberculous abscess, ecthyma gangrenosum, ecthyma, pyoderma gangrenosum, deep mycosis, atypical mycosis, and syphilitic gumma, multiple biopsies were carried out. The histopathology of the epidermis revealed spongiosis, slight acanthosis, and exocytosis abscesses in the epidermis, focal abscesses in the epiderm and mesoderm, and more abscesses in the deep dermis, with increased leukocytes accompanied by a few lymphocytes and multinuclear giant cells [Figure 3]. The patient was examined with a chest X-ray, thoracic tomography, sacral and lumbar magnetic resonance imaging, and computed tomography for pulmonary tuberculosis and extrapulmonary tuberculosis, and no tuberculosis was identified. The Quantiferon test was negative, and chronic granulomatous disease was excluded with neutrophil function test.

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Infective endocarditis was excluded by echocardiography; polyarteritis nodosa was excluded by magnetic resonance imaging; and the absence of stenosis and aneurysm was found with angiography. Abdominal tomography revealed dispersedly localized hypodense lesions in the liver parenchyma, with the largest one being 1.5 cm [Figure 4], and a biopsy was conducted.

The sections of the liver biopsy showed mild lymphocytic inflammatory cell infiltration on the portal area, focal interphase hepatitis, intralobular sinusoidal dilatation, and several focal lytic necrosis areas.

The patient was diagnosed with aseptic abscess syndrome using the available clinical and histopathological findings.

Methylprednisolone therapy was started with a dose of 1 mg/kg/day, and the clinical and laboratory conditions quickly improved [Figure 5].

Abdominal dynamic contrast-enhanced magnetic resonance imaging performed on day 45 of treatment showed that the hypodense areas in the liver had completely disappeared [Figure 4].

The dose of steroids was tapered in 2- to 4-week intervals and ceased after 4 months of treatment and 1 year of colchicine therapy. The patient had no recurrence for 6 months without therapy.

Aseptic abscess syndrome is a disorder included in the group of neutrophilic diseases.[1,4,5] The diagnosis is established by excluding other diseases in a differential diagnosis.[3] Our case had a liver abscess for 4 years, and his general condition rapidly deteriorated when he also developed an abscess in the spleen, and then he developed cutaneous abscesses. The skin lesions mimicked the process of a gummatous lesion. The initial ulcers had deepened to the tendon until a diagnosis was established, and the treatment was started. After treatment, the subcutaneous lesions immediately became superficial and quickly improved.

Aseptic abscess syndrome may present with other neutrophilic diseases. In general, aseptic abscess syndrome has been reported to be accompanied by inflammatory intestinal diseases.[2,4,5] The literature reports that visceral abscesses are accompanied by pyoderma gangrenosum, Sweet’s syndrome, and leukocytoclastic vasculitis.[1-5] Most of the reported cases of aseptic abscess syndrome are from continental Europe, with France in the lead with a series of 30 cases. It is believed that the reason for this limited number of reports is that the disease is not well-known, not that it is rare.[6] It took more than 4 years to diagnose our patient. The patient received many treatments for visceral abscesses and finally underwent a splenectomy. The severe skin involvement allowed us to observe the patient, thus making it easy to diagnose and treat him. One case reported from France had necrotic cutaneous abscesses in the distal extremities, which is clinically
similar to our case, and the patient developed Crohn’s disease 6 months later. In our case, ileocolonoscopy was performed with a biopsy, and no inflammatory bowel disease was found. The literature includes cases that developed inflammatory intestinal disease after 3–4 years.

Cutaneous abscesses evolve into deep ulcers. Large ulcers appear similar to pyoderma gangrenosum at the end, but all ulcers begin as subcutaneous nodules, and there is no type of pyoderma gangrenosum following such a path. Almost all the cases reported in the accessible literature used systemic steroids for the treatment of aseptic abscess syndrome; however, dapsone, colchicine, tumor necrosis factor-alpha blocker, and interleukin-1β blocker have been included in the treatment for treatment-resistant and recurrent cases.

Aseptic abscess syndrome should be considered if any tissues have abscesses and there is fatigue, arthralgia, and myalgia or if a microbial factor is not identified, and steroids and colchicine should be included in the treatment.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**
There are no conflicts of interest.

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