A child with Behçet’s disease presenting with a spectrum of inflammatory manifestations including epididymoorchitis

Ayhan Pektaş¹, İlker Devrim², Nesrin Beşbaş³, Yelda Bilginer³
Ali Bülent Cengiz², Seza Özen³
¹Department of Pediatrics ²Pediatric Infectious Disease Unit, and ³Pediatric Nephrology and Rheumatology Unit, Hacettepe University Faculty of Medicine, Ankara, Turkey


Behçet’s disease is a systemic vasculitis affecting arterioles and venules and is characterized by recurrent oral ulcers, genital ulcers and ocular inflammation. It can involve any organ but joints, skin, central nervous system and gastrointestinal tract are the most common sites.

Here we report an 11-year-old Behçet’s disease patient presenting with attacks of myositis accompanied by epididymoorchitis and periodic fever attacks as well as a separate attack of thrombophlebitis.

Key words: Behçet’s disease, epididymoorchitis, myositis, thrombophlebitis.

Behçet’s disease (BD) is a systemic vasculitis characterized by presentation with the involvement of joints, central nervous system and gastrointestinal system as well as recurrent oral ulcerations, genital ulcerations and ocular involvement. BD may affect arteries and veins of all sizes in any system. Although the etiology of BD is still unknown, the exaggerated response triggered by infections in genetically susceptible individuals is thought to be responsible for the pathogenesis of the disease.

Behçet’s disease occurs rarely in children. The clinical process of the disease in children is similar to that in adults but periodic fever attacks can be the prominent symptom. We report an 11-year-old BD patient presenting with attacks of myositis accompanied by epididymoorchitis and periodic fever attacks as well as a separate attack of thrombophlebitis.

Case Report

An 11-year-old male patient was admitted to our Children’s Hospital with the complaints of high fever (>39°C) and swelling and sensitivity in the left arm and leg. He refused to walk due to the pain. In his medical history, attacks of fever and similar symptoms of the extremity occurred twice: the first resulted in hospitalization and the second resolved spontaneously. The patient had a history of recurrent aphthous oral ulcerations (Fig. 1) that occurred three-to-four times per year accompanied by high fever, but he had never experienced genital ulcers. Two painful and heat-spot lesions of erythema nodosum (4x6 and 2x3 cm) on the left leg and a similar lesion of 2x2 cm on the right forearm were noticed during the physical examination of the patient (Fig. 2). Hemoglobin, leucocyte count and thrombocyte count were 12.8 g/dl, 12,000/mm³ and 316,000/mm³, respectively. C-reactive protein level and erythrocyte

Fig. 1. Multiple aphthous lesions in the mouth.
sedimentation rate were 7.4 mg/dl (normal
range: 0-0.5) and 62 mm/hour, respectively.
The laboratory examinations of antinuclear
antibodies (ANA), rheumatoid factor (RF)
and anti dsDNA performed for the diagnosis
of autoimmune diseases were all negative. At
his further laboratory examination, he was
found to be heterozygous (M680I/-) for familial
Mediterranean fever (FMF). HLAB5 was not
present in our patient.
Intravenous sulbactam-ampicillin with a
non-steroidal anti-inflammatory agent was
selected as initial therapy and the patient
was hospitalized for further evaluation and
treatment. Pathergy test of the patient was
positive. In his follow-up, epididymoorchitis
developed on the third day of his admission
to hospital (Fig. 3). No uveitis was diagnosed
on ophthalmological examination. Sulbactam-
ampicillin therapy was discontinued on the
fourth day. The patient’s high fever (>38.5°C)
present throughout the first three days of his
hospitalization resolved spontaneously after the
initiation of the therapy with colchicine and
prednisone (1 mg/kg/day). Both the extremity
complaints and epididymoorchitis responded to
this treatment. The patient was discharged with
prednisone and colchicine treatment. Steroid
was discontinued after a month by tapering
while colchicine therapy continued. Two
months after discharge the patient presented
with acute calf pain. Doppler ultrasonography
confirmed a thrombophlebitis. He was started
on low-dose heparin along with prednisone. He
was discharged with coumadin and low-dose
prednisone and was in excellent condition at
his first-month visit.

Discussion
Behçet’s disease characterized by the triad
of recurrent aphthous stomatitis, genital
ulcerations and recurrent uveitis was first
described by Dr. Hulusi Behçet in 1937.
The cases of BD in children were published as series.
The high frequency of erythema nodosum in
Turkish patients was noticed in an international
publication. As there is no specific test for
the definite diagnosis of BD, the diagnosis of
this disease depends on the clinical criteria.
Although several classifications are present, the
criteria of the International Study Group have
been widely accepted since 1990. According to
these criteria, the presence of oral ulcerations
in addition to the presence of two criteria from
among recurrent genital ulcerations, ocular
lesions, skin lesions or positive pathergy test
is sufficient for diagnosis of BD.
In our case, the presence of positive pathergy
test and disseminated erythema nodosum lesions
along with oral ulcers confirmed the diagnosis
of BD. This patient has certain interesting
features, such as the presence of myositis
and epididymoorchitis, as well as the classical
symptoms. The dramatic response of the
patient’s epididymoorchitis to the colchicine and
steroid therapy suggests a probable relationship
between BD and an auto-inflammatory pathology.
Although epididymoorchitis (either recurrent
or non-recurrent) accompanying BD in adult
patients is present in the literature, childhood
cases are very rare.
A peculiar feature of BD is its course characterized
by exacerbations and remissions. In fact, there
is a recent trend to classify this disease as an
auto-inflammatory syndrome. This patient also
displayed a course of recurring inflammation. Initially he had attacks of fever, nodosa, then orchitis and finally thrombophlebitis. He enjoyed good health in between while only on colchicine treatment in the latter period. The clinical features also present similarities with the other monogenic auto-inflammatory syndromes such as fever, painful skin findings and orchitis. FMF should also be considered in the differential diagnosis of such patients. In fact, some authors highlight the association of FMF and BD, whereas others suggest that this is simply because of the increased frequency of both of these diseases in the area. However, our patient fulfilled the criteria for BD and the prominent feature during his attacks was the painful erythema nodosum instead of serositis, which is characteristic of FMF. He did not describe the characteristic serositis attacks; however, these features will also be evaluated in his follow-up. Colchicine is an effective preventive therapy for both of these clinical settings. On the other hand, the M680I mutation in this patient may have worsened his inflammatory disease, as suggested in previous reviews. In conclusion, the presence of oral-genital ulcerations should be investigated in case of epididymoorchitis as well as other inflammatory manifestations with an unknown etiology in order to make a correct diagnosis.

REFERENCES