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A CASE OF INSTESTINAL BEHCET'S DISEASE PRESENTING AS NUTCRACKER SYNDROME: A RARE MANIFESTATION

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ABSTRACT Behçet's syndrome is a systemic vasculitis. It can present with skin and mucosal lesions, uveitis, arthritis, major arterial and venous vessel disease, and gastrointestinal and neurologic manifestations. These manifestations can be present in various combinations and sequences over time. Although the gastrointestinal and systemic features of Behcet's disease and inflammatory bowel disease overlap to a considerable extent, they are generally viewed as two distinct diseases. Ours was a case of a 36 year old Hindu male patient with complaints of abdominal pain, constipation, painful oral and scrotal ulcers and anorexia. CECT abdomen showed colitis with nutcracker syndrome. Colonoscopy showed features of chronic colitis initially thought to be Crohn's disease however biopsy revealed perivasculitis and multiple lymph follicles without granuloma. A diagnosis of Behcet's Disease with GI involvement was made and he was started on oral steroids. On 2 months of follow up he was symptomatically better and in remission. One year later, he was doing well without recurrence of symptoms.

KEYWORDS : Behcet's Disease, Crohn Disease, Colonoscopy, Gastrointestinal tract

INTRODUCTION

Behcet's Disease is a chronic relapsing autoimmune multisystem disease with variable vessel involvement. It is referred to as Oculo-Oro-Genital syndrome with rare involvement of CNS, GIT, lungs, heart. It is a neutrophilic vascular reaction with equal prevalence in both males and females, however males are associated with a more severe form of the disease.

Most commonly affected age is 20-40 years. It is most prevalent in Turkey, with a prevalence of 1 in 250 adults. It is known to show association with HLA B51 however this is not used as a diagnostic test because it is also found in up to 20% of the normal population. The most widely accepted criteria were published by the International Study Group (ISG) for Behcet's Disease in 1990. Diagnosis requires the observation of recurrent oral ulceration (three episodes within any 12 month period) plus any two of the following: recurrent genital ulceration, eye lesions, skin lesions or a positive pathergy test. Gastrointestinal involvement is rare and is commonly associated with Ileocaecal ulcers which are indistinguishable from Crohn's disease however this distinction is important as it has therapeutic and prognostic implications. Below we discuss a rare case of intestinal Behcet's disease presenting with Nutcracker syndrome.

Case Report

36 year old Hindu male patient came with complaint of abdominal pain since 2 months, constipation on and off since 2 months, painful ulcers in mouth and anorexia since 4 months. He was asymptomatic before 4 months when he started to develop painful oral ulcers that recovered spontaneously with topical treatment as reported by him. Then he started to have abdominal pain: periumbilical region, mild to moderate intensity, associated with anorexia since 2-3 months. He also reports weight loss in form of loosening of clothes since 3 months. On admission he was vitally stable. On examination a single discrete superficial ulcer was seen on hard palate. No other significant findings were noted. Hemogram showed Hemoglobin: 12, Total leucocyte count: 9100/cmm, Platelet count 3.15 lakhs. Renal and Liver function tests were normal. ESR: 60mm, CRP: 29 . Urine routine examination was normal. Abdominal Ultrasound showed a normal report. Based on history and reports he was started on symptomatic treatment with PPIs and antispasmodics and topical treatment for oral ulcers with multivitamin supplementation, however he showed no improvement in symptoms. Hence a Contrast CT abdomen was done that showed Irregular diffuse circumferential bowel wall thickening involving caecum, ascending colon and transverse colon suggesting possibility of infective/inflammatory etiology with reduced Aortic-SMA angle: 37degrees (Normal: 38-65 degrees), proximal part of left renal vein compressed between SMA and Aorta with post stenotic dilation suggestive of Nutcracker syndrome. Based on these findings he was started on antibiotics for colitis. For Nutcracker syndrome he was to be kept under conservative management since he had no significant hematuria or occlusion of renal vein. It was concluded that the nutcracker syndrome was due to the significant weight loss as reported in the history however the cause of weight loss was not yet identified. Over the course of hospital stay his oral ulcers improved however he had persistent abdominal pain. He also developed genital ulcers during stay, which on further probing he reported to have had history of previously 2 months back as well. He was then planned for a colonoscopy that showed multiple, round ulcers varying in size from 0.5–1 cm^2 throughout the ileum, caecum, ascending colon, transverse colon, descending colon, and rectosigmoid suggesting possibility of Chronic Colitis due to Crohn's disease. Biopsy taken showed ulcerations with inflammatory infiltration consisting of lymphocytes and plasma cells. Tzanck smear was taken from the genital ulcer that showed normal report and Pathergy test done that turned out to be negative. On corroborating all findings of recurrent oral ulcers, genital ulcers and colitis a possibility of Behcet's syndrome was kept. He was started on oral steroids- Tab Prednisolone 30mg 1-0-0 and discharged. On follow up after a month, he was better and was able to take orally. The dose of prednisolone was tapered and planned to be discontinued. One year later, he was doing well without recurrence of symptoms.

DISCUSSION

Behçet syndrome is a systemic vasculitis, first described by Hulusi Behçet, a Turkish dermatologist in 1937. It can present with skin and mucosal lesions, uveitis, arthritis, major arterial and venous vessel disease, and gastrointestinal and neurologic manifestations. These manifestations can be present in various combinations and sequences over time. Behçet syndrome is diagnosed clinically. Although many diagnostic criteria have been established, there is no universally accepted definition.

The pathogenesis and etiology of Behçet syndrome are unknown. Unlike other autoimmune diseases, however, Behçet syndrome is not typically associated with autoantibodies. In patients with genetic susceptibility (HLA B.51, single nucleotide polymorphism in IL.10, IL.23R/IL.12RB2 genes etc.), trigger factors (bacteria, virus, or protein particle exhibiting molecular mimicry) precipitates perturbation in T.cell homeostasis with predominant Th.1.mediated inflammatory response, unlike Crohn's, where the response is predominantly Th.17.mediated. The most commonly used and best performing diagnostic criteria are the International Study Group (ISG) criteria (sensitivity ~95%, specificity ~96%);

Table: 1 International Study Group Diagnostic Criteria For Behçet Disease (1)

Criterion	Description
Recurrent oral ulcerations	Minor or major aphthous ulcerations or herpetiform ulcerations observed by a physician or patient that have recurred at least 3 times in a 12-month period
Plus 2 of the following criteria in the absence of other clinical explanations:	
Recurrent genital ulcerations	Aphthous ulcerations or scarring observed by a physician or patient
Eye lesions	Anterior or posterior uveitis or vitreous cells seen on slit-lamp examination; retinal vasculitis observed by an ophthalmologist
Skin lesions	Erythema nodosum observed by a physician or patient; pseudo folliculitis; papulopustular lesions; acneiform nodules observed by a physician in post adolescent patients who are not taking corticosteroids
Positive results from a pathergy test	Oblique insertion of a 20—22-gauge needle 5 mm into the skin, causing a papule 2 mm or larger. The test is generally performed on the forearm, and results are read by a physician after 24–48 hours.

Additional clinical manifestations may involve various organ systems, including the gastrointestinal, vascular, pulmonary, and central nervous system.

Table: 2 Common Clinical Manifestations Of Behcet's Disease(2)

Skin, Mucocutaneous	Papulopustular lesions (Behçet's pustulosis), erythema nodosum, superficial thrombophlebitis, minor aphthous ulcers
Eyes	Anterior and posterior uveitis, retinal vasculitis
Vascular	Deep venous thrombosis, large-vein thrombosis, pulmonary artery aneurysm
Musculoskeletal	Arthralgia, arthritis (monoarticular, oligoarticular)
Gastrointestinal	Ileocecal ulcers
Genitourinary	Genital ulcers, epididymitis
Central nervous system	Meningoencephalitis, parenchymal disease (pyramidal signs, hemiparesis, behavioural changes, sphincter disturbance), intracranial hypertension secondary to dural sinus thrombosis

Gastrointestinal (GI) manifestations of Behçet's disease are of particular importance as they are associated with significant morbidity and mortality. GI manifestations usually occur 4.5-6 years after the onset of oral ulcers. Symptoms include abdominal pain, diarrhea, nausea, anorexia, and abdominal distension Intestinal BD occurs in 10–15% of BD patients and shares many clinical characteristics with inflammatory bowel disease (IBD), making differentiation of the 2 diseases very difficult and occasionally impossible. However it is very important to differentiate the two. Despite considerable overlap in the presentation of both diseases, the management approach, complications, and disease outcomes are different. The presence of genital ulcers, especially scarring penile and scrotal ulcers and papulopustular skin lesions, favours BD. Intestinal BD ulcers are mainly located in the ileocecal region, are ovoid or geographic in shape, and may be described as a collar button. Diffuse colonic involvement is rare in intestinal BD. In CD, classic colonoscopic findings include longitudinal ulcers with a cobblestone appearance and discontinuous involvement of various portions of the GI tract. The development of complications such as strictures, fistulae, and abscesses is common in CD. Caecal deformity with incompetence of the ileocecal valveis seen in 95% of cases and a single ulcer averaging 2.7 cm in diameter in 75% of patients based on a study.(5)

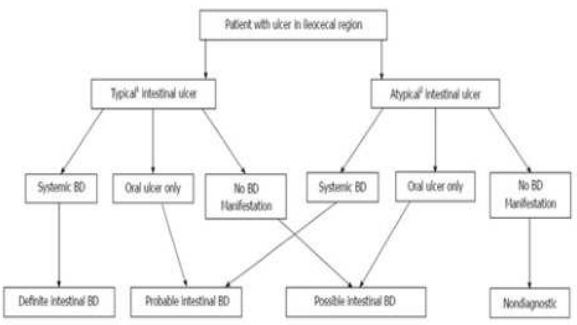

Figure:1 Algorithm to differentiate Crohn's ulcer from Behcet's based on clinical symptoms.(3)

Treatment of Behcet's disease depends on the type and extent of lesions with simple mucocutaneous lesions treated with Topical steroids and PGE2 gel/ Phosphodiesterase 4 inhibitor-Apremilast; Severe relapsing disease with steroids and methotrexate/ thalidomide/ INF A; Systemic disease with oral steroids and azathioprine/ mycophenolate mofetil; Ocular disease with azathioprine (2nd line: Anti TNF A)

Death from intestinal BD is uncommon. Disease-specific mortality in BD is mainly due to major vessel disease (eg. arterial aneurysm) or neurologic involvement.

CONCLUSION

Gastrointestinal manifestations in Behcet's disease are very rare. These are also difficult to differentiate from Crohn's disease. Our patient was diagnosed with Intestinal Behcet's disease based on clinical, radiological, endoscopic and histopathologic findings and was treated accordingly and is currently symptom free and in remission.

Abbreviations: BD- Behcet's Disease, CD- Crohn's Disease, GI- Gastrointestinal

Conflicts Of Interest: None

REFERENCES

1. Criteria for diagnosis of Behçet's disease. International Study Group for Behçet's Disease. Lancet. 1990 May 5;335(8697):1078-80. PMID: 1970380.
2. Skef W, Hamilton MJ, Arayssi T. Gastrointestinal Behçet's disease: a review. World J Gastroenterol. 2015 Apr 7;21(13):3801-12. doi: 10.3748/wjg.v21.i13.3801. PMID: 25852265; PMCID: PMC4385527.
3. Grigg EL, Kane S, Katz S. Mimicry and deception in inflammatory bowel disease and intestinal behçet disease. Gastroenterol Hepatol (N Y). 2012 Feb;8(2):103-12. PMID: 22485077; PMCID: PMC3317507.
4. Yadav AI, Chattopadhyay A, Ahamed R, Muktesh G, Narang T, Nada R, Sharma A, Mandavdhare H, Sharma V, Dutta U. An unusual cause of granulomatous colitis: Behcet's disease. JGH Open. 2019 Aug 15;4(2):303-305. doi: 10.1002/jgh3.12215. PMID: 32280787; PMCID: PMC7144775.
5. Kim ES, Chung WC, Lee KM, Lee BI, Choi H, Han SW, Choi KY, Chung IS. A case of intestinal Behcet's disease similar to Crohn's colitis. J Korean Med Sci. 2007 Oct;22(5):918-22. doi: 10.3346/jkms.2007.22.5.918. PMID: 17982247; PMCID: PMC2693865.
6. Hatemi G, Christensen R, Bang D, Bodaghi B, Celik AF, Fortune F, Gaudric J, Gul A, Kötter I, Leccese P, Mahr A, Moots R, Ozguler Y, Richter J, Saadoun D, Salvarani C, Scuderi F, Sfikakis PP, Siva A, Stanford M, Tugal-Tutkun I, West R, Yurdakul S, Olivieri I, Yazici H. 2018 update of the EULAR recommendations for the management of Behçet's syndrome. Ann Rheum Dis. 2018 Jun;77(6):808-818. doi: 10.1136/annrheumdis-2018-213225. Epub 2018 Apr 6. PMID: 29625968.