Case Report

Intestinal Behcet's : A Rare Presentation of Behcet's Disease

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We herein present a case of 48 years old female patient presented with fever, bloody diarrhea followed by palpable purpuric rash over the body along with recurrent oral and genital ulceration. These were associated with history of symmetric polyarthralgia. On examination moderate anemia, signs of anterior uveitis were found. In blood parameters thrombocytopenia along with elevated Erythrocyte Sedimentation Rate, C-Reactive Protein were noted. On further investigations the serological tests were found to be negative for Dengue, Chikungunya, HIV, HBV, HCV. Complement C3 found to be low. Colonoscopic biopsy is diagnostic of Indeterminate Crohn's Disease with IgA, G, M, C3, Fibrinogen immunostaining in skin biopsy. ANA, P-ANCA, C-ANCA were found to be nonreactive for the patient. All of the above mentioned points were pointing towards Behcet's disease. For confirmation, Anti Saccharomyces Cerevisae Antibody was found to be positive . Skin pathergy test was positive. So, we diagnosed this case as behcet's disease.

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Key words **: Intestinal behcet's disease, Skin pathergy test, HLA-B51.**

Behcet disease, an autoimmune disease, affects each and every systems of our body. The entity was first described by a Turkish dermatologist Hulusi Behçet in 1937 as a syndrome having genital and oral ulcerations and inflammation of eye[1,2]. It is more prevalent in Turkey (80-370 cases per 100,000 inhabitants) which is followed by Asia and Middle Eastern countries[3]. Intestinal Behcet disease shares several common characteristics with Crohn's disease and it seems difficult to differentiate between the two. There are many factors in pathogenesis of these two conditions among which genetic, immunological and environmental factors are most important. In this article, we are reporting a challenging case of intestinal Behcet's (Figs 1-3).

Editor's Comment :

- Intestinal manifestation of behcet's disease is rare.
- It is a differial diagnosis of inflammatory bowel disease and iliocaecal tuberculosis.
- Our case had a favourable outcome because of appropriate and timely diagnosis.

Chief Complaints :

A 48 years old female patient presented with chief complains of

(1) Fever for last 45 days.

(2) Rash for last 35 days with oral and genital ulceration.

(3) Pain in eye, blurring of vision for last 25 days.

History of Presenting Complaints :

(1) Fever was for last 45 days which was insidious in onset and gradually progressive , intermittent , high grade and without any chill and rigor.

(2) Rash appeared on the 11th day of fever in the lower limb and gluteal region and then progressed to all over the body. Rashes were palpable and pupuric, nonpruritic. There were appearance of painful perioral, perineal and vulval ulcers.

(3) For 25 days she had redness, pain and blurring of vision of both eyes which was Insidious in onset and associated with intolerance to light.

These were associated with Sudden onset, Progressive bloody diarrhea which did not respond to normal medications. She also developed insiduous onset , gradually progressive joint pain involving knee, elbow joint without any swelling, redness and morning stiffness. There was no history of any orificial bleeding, Cough, haemoptysis, breathelessness, weight loss.

Differential Diagnosis :

(1) Infective causes: viral exanthematous fever, disseminated gonococcal infection, ileocecal TB

(2) HIV/AIDS

(3) Autoimmune causes : Systemic lupus erythematosus, vasculitis, other connective tissue disorder,

(4) Malignancy

Examination :

(1) On examination patient was conscious, alert and co-operative.

(2) Vitals :

Pulse Rate - 90 beats per minutes

Blood Pressure - 128/70 mm of Hg.

(3) Pallor – (+)

(4) Rash - Over gluteal region, lower limb, upper limb, back and Trunk.

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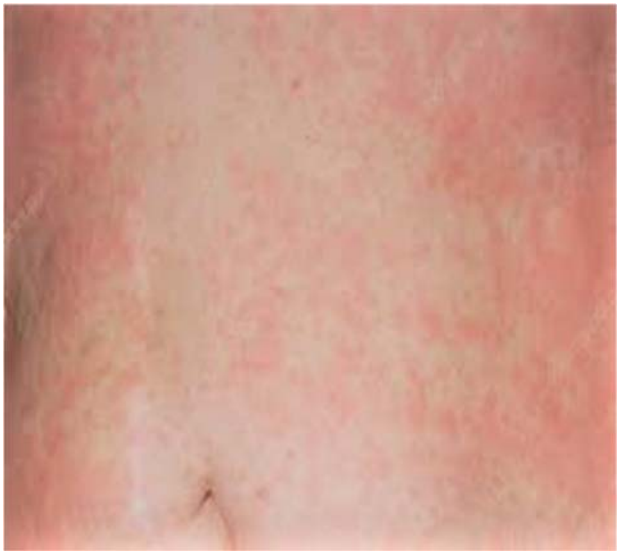

Fig 1 — Purpuric, non pruritic, non blnchable rash

(5) Aphthous ulceration - in buccal mucosa, tongue, inner aspect of lips, genital area specifically over vulval region.

(6) Ocular examination reavealed bilateral mobile hypopyon with circumciliary congestion.

(7) Lymph nodes - Not palpable.

(8) Joint examination- no swelling and no deformity.

(9) Per-rectal examination – no hemmorrhoid, fissure, perianal fistula.

(10) Other Systemic examination - WNL

Differential Diagnosis :

(1) Infective causes: viral exanthematous fever, ileocecal TB

(2) Autoimmune causes : Systemic lupus erythematosus, vasculitis, other connective tissue disorder

Investigation :

- CBC: Hb-9.0 gm% TLC-8800/mm^3 PLT 35000/mm^3 ESR 40mm in 1 hour
- LFT: Bil_T-0.6mg/dl Bil_D-0.2mg/dl Bil_I-0.4mg/dl SGPT/SGOT-321/157U/L Albumin/Globulin – 3.2/3.0 gm/dl
- FBS/PPBS – 76/142 mg/dl
- Urea/Creatinine – 17.5/0.71 mg/dl
- Na^+/K^+-135/3.7 mmol/l
- CRP-98.5 mg/l
- MP,MPDA – negative
- Denhue IgM, IgG- non reactive
- Leptospira IgM – non reactive
- Typhidot IgM- nonreactive
- HBsAg/Anti-HCV/HIV1,2 – non reactive
- Gastric lavage, sputum for AFB, CBNAAT - negative
- On colonoscopy guided biopsy it showed focal area of erosion over colonic mucosa. The glands showed variable distortion and mild mucin depletion. The lamina propria is densely infiltrated by lymphocytes, plasma cells and a few eosinophils. Cryptitis and crypt abscess were present. There was no granuloma. No evidence of dysplasia was observed. Biopsy was suggestive of indeterminate Inflammatory bowel disease.

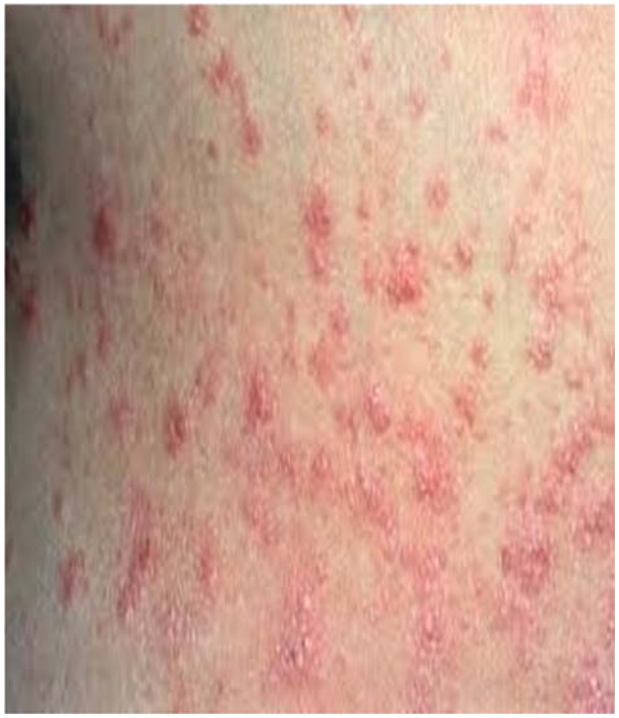

Fig 2 — Purpuric, non pruritic, non blnchable rash

- Skin biopsy findings- superficial and deep perivascular infiltrates of lymphocytes and neutrophils. There were features of vasculitis with endothelial cell swelling.
- On immunofluorescence tyest – IgA, IgG, IgM, C3, Fibrinogen deposits were present in skin biopsy.
- Autoimmune profile- ANA(-) / P-ANCA(-)/C-ANCA(-)
- Serum C3- 52.4 mg/dl which is below normal range

From above these results we were suspecting the case to be a Behcet's disease. To confirm we perform the following tests.

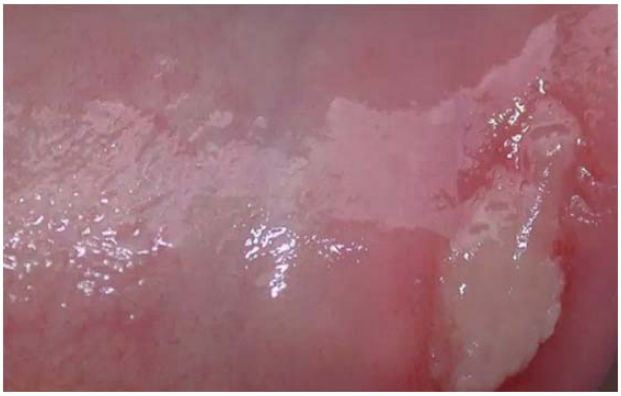

Fig 3 — Painful, aphthous oral ulcer in the inner aspect of mouth

Confirmatory Test :

- Anti Saccharomyces Cerevisiae Antibody – (+)
- Skin pathergy test - reactive.
- HLA-B51 : (+)

Provisional Diagnosis :

This is a case of Intestinal Behcet's Disease.

Treatment and Follow Up :

She was treated with IV Hydrocortisone injection along with Azathioprine and other supportive treatment. After resolution of all symptoms she was discharged. Now she is doing well.

Discussion

Behcet's disease is a type of inflammatory disorder which can affect multiple systems of our body. Most commonly it presents as skin manifestations. Male and female have almost same preponderance towards developing this disease. But males generally develop severe forms of Behcet's. There is a 1990 International Study Group Criteria for clinical diagnosis of Behcet's. It includes presence of recurrent oral ulceration with two of the following[4]

- Recurrent genital ulceration
- Uveitis
- Typical skin lesions
- Skin Pathergy Test. Our case report also fulfilled this criteria.

In Behcet's the oral ulcers are generally characterised by painful, recurrent and aphthous ulcers having necrotic base and they heal without scarring. In contrary the genital ulcers heal with scarring. Apart from theses it involves musculoskeletal system by developing non erosive arthritis of small joints which is collaborating with our case report. Uveitis is most common ophthalmological finding. But gastrointestinal Behcet's like our case report is rare. Only 1-2% of cases having ulcers in GI tract[5] mostly in iliocecal region. Behcet's colitis mimics Inflammatory Bowel Disease specially Crohn's. The exact cause of Behcet's is still unknown. It is believed that there may be immunogenetic and inflammatory cytokines are two components of it's pathogenesis. The autoimmunogenetic association with this disease shows presence of T lymphocytes in lesions. TH1 cells along with Interferon-gamma, IL-12, IL-2 mediates cell mediated autoimmunity. Autoreactive T cells with anti HSP-60 function cause pathogenesis of this. HLA-B51 is more frequently associated with this disease. Due to lack of specific tests, Behcet's is diagnosed clinically. Here elevated ESR, CRP are the nonspecific finding. But ASCA antibody test and skin pathergy tests are the few which can streamline the diagnosis to a certain extent. Apart from them it is associated with HLA-B51, HLA-B5 positivity. Perianal fistula, fissure , etc go in favour of crohn's disease. In our case the mentioned complications were absent. So, the case is more in favour of intestinal behect's rather than crohn's. Steroids are the mainstay of treatment. In serious cases Azathioprine is added for speedy recovery.

Conclusion :

Intestinal BD and inflammatory bowel disease specifically crohn's have significant overlap in terms of sign, symptoms, pathological findings. For these clinicians always face difficulty in distinguishing the two. In both cases Steroids and Immunomodulators are the mainstay of treatment with decreased mortality and morbidity. Many researches are still ongoing to solve this dilemma to streamline the diagnosis. Understanding the mechanism properly will be important for appropriate treatment and prognosis of the disease.

References

1 Baumgart DC, Sandborn WJ — Crohn's disease. Lancet. 2012;380:1590–605. doi:10.1016/S0140-6736(12)60026-9. PMID: 22914295.
2 Behçet H — Rezidivierende aphthose, durch ein virus verusachte geschwure am auge und an den genitalien. *Dermatol Wochenschr* 1937; **105:** 1152-7.
3 Bayraktar Y, Ozaslan E, Van Thiel DH — Gastrointestinal manifestations of Behcet's disease. *J Clin Gastroenterol* 2000; **30:** 144-54.
4 1990 International Study Group Criteria; *Lancet* 1990; **335:** 1078-80.
5 Kasahara Y, Tanaka S, Nishino M, Umemura H, Shiraha S, Kuyama T — Intestinal involvement in Behcet's disease: review of 136 surgical cases in the Japanese literature. *Dis Colon Rectum* 1981; **24:** 103-6.