Extensive Gastrointestinal Involvement in Behcet’s Syndrome — A Case Report

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SUMMARY

Behcet’s syndrome is a relatively rare entity, originally characterized by a diagnostic triad of oral and genital ulceration with ocular abnormalities. Since the syndrome was described in 1937 by Behcet, a Turkish dermatologist, many papers have described multisystemic involvement in the disease. Recognized clinical features include arthritis, arterial thrombosis, encephalitis, cardiomyopathy, erythema nodosum, amyloidosis, glomerulonephritis, and pneumonitis. We report here a case of Behcet’s syndrome in 29 year old male presenting with orogenital as well as colonic ulcers.

INTRODUCTION

Behcet's disease is a chronic, relapsing, inflammatory disease characterized by recurrent oral aphthae and any of several systemic manifestations including genital aphthae, ocular disease, skin lesions, neurologic disease, vascular disease, or arthritis. Disease prevalence and expression vary geographically and it affects people of Middle Eastern or Far Eastern ancestry more often than those from other regions. Even in these populations Behcet’s disease is an uncommon disorder. Behcet's disease is considered to be a type of multisystem vasculitis. The prevalence rates in Turkey and Chinese populations are approximately 100 and 3 per 100,000 persons, respectively.

Severity is generally greater in men. The greatest morbidity and mortality occurs with ocular disease (affecting up to two-thirds of patients), vascular disease (affecting up to one-third of patients), and central nervous system disease (affecting 10 to 20 percent of patients).

CASE REPORT

A 29 year old mentally subnormal male presented in OPD with intermittent high grade fever of 22 days, and loose stools sometimes associated with fresh bleeding per rectum and tenesmus. Patient also developed difficulty in swallowing due to deep oral ulcers. There was past history of skin rash 6 months back which was vesicular in nature with scarring on healing along with bilateral ankle swelling when patient was put on steroids for 4 months which resulted in relief. Patient was also diagnosed as a case of T.B 3 year back and took ATT for 9 months.

On examination, patient was having oral as well as genital ulcers which were of punched out variety with yellow base bluish edges and no discharge , there was no undermining or regional lymph node enlargement. There was right knee swelling without redness or tenderness. Patient was mentally retarded could not talk or clean his bowels but can walk. Bilateral conjuctival redness noticed. Fundoscopic examination was normal except mild AV nipping there was no exudation or hemorrhage.

Systemic review was normal.

On investigations, raised inflammatory markers with hypoalbuminaemia and normal renal profile (Table 1).
Table 1: raised inflammatory markers with hypoalbuminaemia and normal renal profile.

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<td>32/30sec</td>
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<td>APTT</td>
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<td>Anti-HIV</td>
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Pathergy Test was negative.

USG Abdomen—Liver 16cm normal echotexture. No focal lesion or dilated ducts seen. Gall bladder was contracted. CBD 6mm, PV 13mm. Pancreas, spleen and kidneys were unremarkable. Mid and lower abdomen showed gaseous distended bowel loops. No ascites.

Colonoscopy—multiple deep ulcers of various sizes with normal intervening mucosa from rectum onwards.

Rectal biopsy—moderate degree of mildly active chronic colitis with benign ulceration. No granulomas or crypt abscesses seen.

He was diagnosed as a case of Behcet's syndrome with gastrointestinal manifestation based on orogenital ulcers, joint involvement, raised inflammatory markers and rectal biopsy findings. Patient was given high dose Methylprednisolone therapy with calcium supplements and proton pump inhibitors and later discharged on oral steroids and azathioprine. On follow up oral and genital lesions have healed with relief of GI symptoms.

**DISCUSSION**

Behcet's is a multisystem relapsing chronic inflammatory disease with its skin, joint, ocular and nervous system involvement. Acneiform lesions may be more common in those with associated arthritis.

**Fig. 1. Multiple aphthous ulcers in sigmoid colon and rectum.**
Pathergy test is less frequently positive in Northern European and North American patients. Thus, it has been suggested that other features might be substituted for pathergy in these populations, including aseptic meningoencephalitis, cerebral vasculitis, recurrent phlebitis, arteritis, synovitis, or focal bowel ulceration. Behçet's disease is remarkable for its ability to involve blood vessels of all sizes—small, medium, and large—on both the arterial and venous sides of the circulation. Glucocorticoids are the mainstay of Behçet's disease treatment for patients with moderately severe to severe disease.

REFERENCES


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