ORIGINAL RESEARCH PAPER

General Surgery

AN INTERESTING CASE SERIES OF SMA VASCULAR INSUFFICIENCY IN BEHCETS SYNDROME PATIENTS

KEY WORDS: Behcets disease ,silk road disease, vasculities, bowel gangrene

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ABSTRACT

Behcets disease is DEFINED as an auto-inflammatory systemic vasculitis of unknown etiology. It is characterized by mucocutaneous manifestations, including recurrent oral and genital ulcerations, ocular manifestations, especially chronic relapsing uveitis, and systemic vasculitis involving arteries and veins of all sizes. Behcets disease usually affects young adults 20 to 40 years of age and is also seen in children less frequently.Both genders are equally affected by the disease although slight male preponderance is seen in japan[7] .we would like report a rare case series on Behcets aka silk road disease presenting in a 16 year old female complicated with SMA vascular insufficiency.

INTRODUCTION

Behcets aka Silk Road disease is a rare autoimmune disorder of unknown etiology among Indians .usually presents as ocular involvement as non-granulomatous panuveitis with unfavorable visual outcome in third of the cases1.The diagnosis was based on the Japanese diagnostic criteria and patients were classified into complete, incomplete, suspected, and possible Behçet's disease.It was first described by Hulusi Behçet from Istanbul in 1937, who described three patients with oral and genital ulcerations, uveitis, and erythema nodosum[2]

Etiology

Exact etiology not known ,multiple factors including environmental,host ,microbial has been putforward but none proved to be of significant importance. Increased prevalence along the "Silk Route" and a genetic element suspected, but Behcet disease does not follow a mendelian inheritance.

HLAb51/B5 single most important important factor identified and significant association with behcets have been proven recently. Many other genes have been identified, including heat shock proteins, major histocompatibility complex class I chain-related genes and TNF related genes.

Hypersensitivity type reactions have been postulated especially to herpes simplex type 1 antigen,staph aureis ,streptococcus sanguinis ,prevotella antigens.

HLA alleles has been shown to protect against or decrease the development of BD, such as HLA-A*33:03 in Korea , DRB1*15 in China , HLA-B*35 in Spain , and -A*03 and B*52 in Egypt 3-6. Distribution and pathophysiology Age group which are vulnerable are 20-40 years of age,however 1-10% of younger age group have been identifies. Both genders are equally affected however slight male preponderance seen In Asian and Turkish people. It appears that Behçet's disease in India is predominantly 'mucocutaneous' ,while 'arthritic'; 'ocular','vascular' and 'neuro' Behçet's being uncommon.

CASE SERIES

We have encountered 5 patients in our institution

CASE1

Miss jasmine 16 yr old female known case of behcets disease for which was on treatment with colchicine for the past 1 year came to emergency department with acute onset severe abdomen pain starting from epigastric and umblical region radiating to back.within 24 hours she developed generalized guarding and rigidity.pt had severe vomiting .

Vitals:
T:100.6 *F
Bp 100/54 mm of hg
Pr:140/min
Spo2:97% on Room Air
Blood Investigations:
Ph:7.10
Pco2:24
Po2:92
Hco3:14
Basedeficit:-8
Urea:50
Creatnine:1.4
Na:134
K:2.9
Tw:18000
Hb:9g/dl
Mcv:80
Haematocrit:30
Platelets:2lakhs
Radiological investigations:

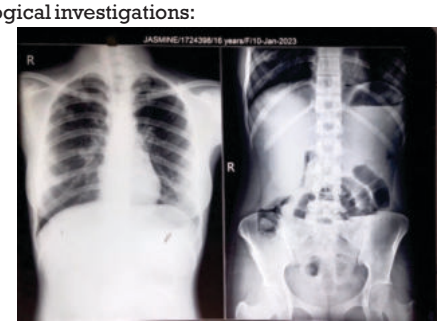

Figure 1 x-ray abdomen showing air under diaphragm and multiple air fluid level

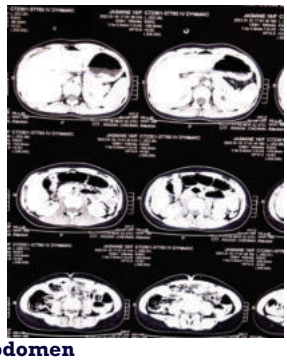

Figure 2 CT Abdomen

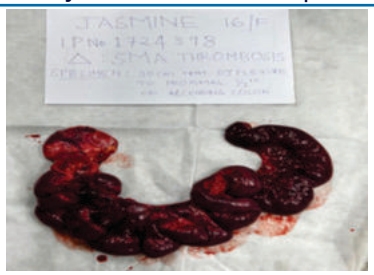

Figure 3 Resected bowel-30 cm from Dj flexure upto proximal 3rd of ascending colon

Progress at hospital

Pt was taken up for emergency laparotomy,findings -30 cm from dj flexure upto proximal 3rd of ascending colon SMA territory was affected .primary resection and proximal jejunostomy and distal mucus fistula was created.

Post op HPE:

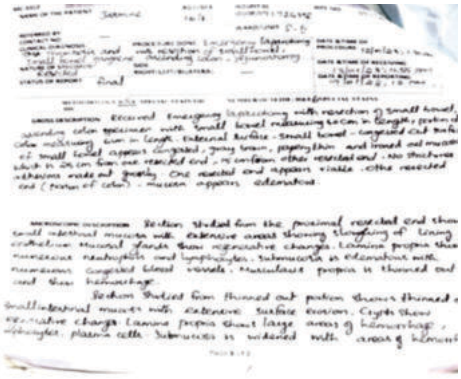

Figure 4 HPE-A

Pt was consulted with rheumatologist and vascular surgeons ,she was started on heparin 5000 units q6 hourly and high dose methyl prednisolone 40mg/kg was used and subsequently tapered to dexamethasone 8mg bd and od dosage.

Post operatively she recovered well and nutritional management was quite challenging owing to jejunostomy which had output of more than 2.5 Litres ,pt was supported with stoma refeeding and TPN .

CASE 2:

Miss jothi 30 yr old female ,presented with abdomen pain for past 1 month ,with increased in severity for the past 1 day.she had no previous comorbids except recurrent oral ulcerations for past 2 years for which no doctor has been consulted. she presented with shock to our emergency department.

Vitals on arrival

T:102*f
Bp 70/40 mm of hg
Pr:122/min
Spo2:90 percent on room air

Blood investigations

Ph:7.25
Pco2:20
Po2:88
Hco3:16
Basedeficit:-7
Urea:75
Creatnine:2
Na:130
K:5.3
Tw:22000
Hb:8.6g/dl
Mcv:81
Haematocrit:27
Platelets:1lakhs

Radiological investigations

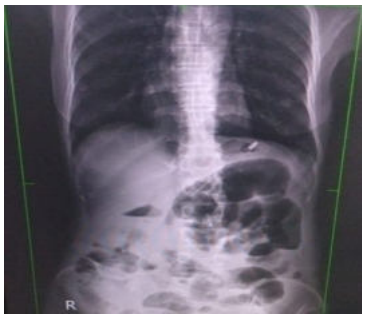

Figure 6 xray abdomen showing multiple air fluid levels

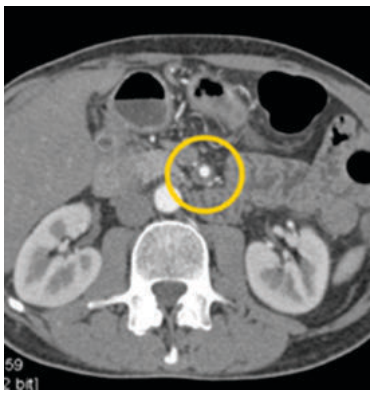

Figure 7 SMA wall thickening

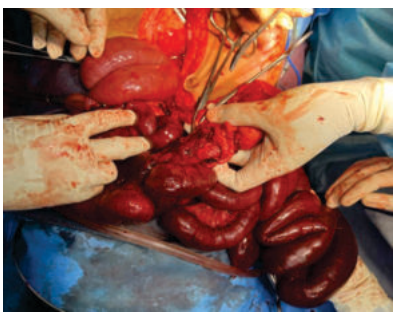

Figure 8 Gangrenous bowel-40 cm from dj flexure upto proximalr 3rd of ascending colon-SMA territory

Pt was taken up for emergency laparotomy after resuscitation and gangrenous bowel was resected 40cm from DJ flexure upto proximal 3rd of ascending colon was resected , proximal jejunostomy and distal mucus fistula was created Post operatively pt was monitored in ICU.pt expired due to refractory acidosis and septic shock with MODS on Post op day 3

HPE:
1)sloughing of epithelial lining mucosa
2)lamina propria showed numerous lymphocytes ,plasma cells,lymphoid aggregates and neutrophils
3)submucosa edematous
4)muscularis propria showed haemorrhage and thinned out
Diagnosis:
SMA vasculitis.

CASE3
Ms senthamil selvi 32 yr old female ,who is a labourer by occupation and known case of behecets disease on treatment .she started to have symptoms 1 year ago with fever ,oral and genital ulcers for 2 weeks with bilateral lower limb petechias c3:1.57 and c4:0.324 started on colchicine 0.5 mg bd and dexamethasone 4mg iv od initially and then tapered out to oral prednisolone 5mg od,for the past 5 days she had vague post prandial abdomen pain ,1-hr after eating and for past one day severity increased with vomiting x3 episodes.

Vitals
T:100*f
Bp:112/70 mm of hg
Pr:112/min
spo2:99% on Room Air

Blood Investigations
TW:16000
Hb:11g/dl
Haematocrit:37
Platelets:80000
Urea:34
Creat:1.2
Na:136
K:4.2
Ph:7.28
Pco2:38
Po2:99
Hco3:17
Basedeficit:-5

Radiological investigations:

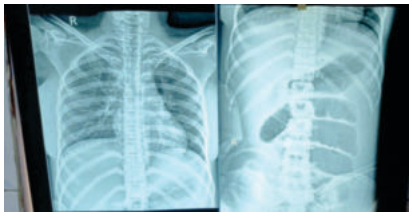

Figure 9 xray abdomen

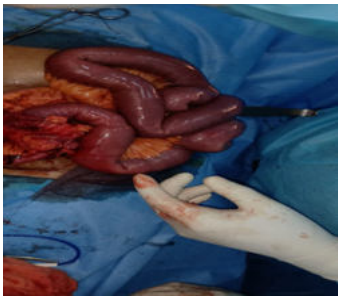

Figure 10 Gangrenous part-70 cm from DJ flexure upto 20 cm from IC junction

Post op HPE:
Showed diffuse infiltrates of lymphocytes and plasma cells in muscularis propria of resected bowel

Post op care :
Pt was started on heparin 5000 units q 6hrly which was bridged to acitrome 2 mg as per vascular surgeons advice and discharged home with a functioning stoma and prednisolone 5mg od.

We have encountered another 2 patients with similar history within age range of (36-42 yrs) ,diagnosis SMA thrombosis with background of Behecets disease–they have not consented to share their history

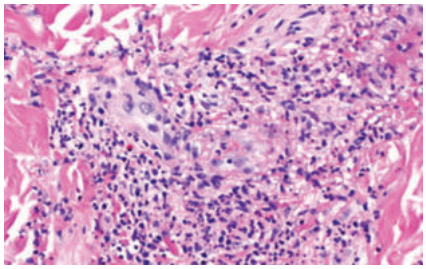

Figure 11 HPE-lymphocytic aggregates

DISCUSSION
Behçet's disease (BD) is a chronic, relapsing inflammatory, multisystem disease of autoimmune etiology with unknown trigger. The disease has a wide clinical spectrum of mucocutaneous lesions,ocular, vascular, articular, neurologic, gastrointestinal and cardiac involvement. Although the number of effective drugs used in the disease's treatment has increased in recent years, BD is still associated with severe morbidity because of mainly mucocutaneous, articular and ocular symptoms and an increased mortality because of large vessel, neurological, gastrointestinal and cardiac involvement[8].

Vascular involvement is one of the most important causes of mortality in BD. Although BD can affect vessels of any size and type [9-10], venous system is the major affected site, and superficial and deep vein thrombosis are the most frequent type of vascular involvements. Thromboses of the inferior and superior vena cava, dural sinuses and Budd-Chiari syndrome can also be seen and are associated with poor prognosis. Although rare, pulmonary artery aneurysm is the most common cause of death

Literature review

No	Author	Age/ sex	Previous diagnosis of Behcets	Time of diagnosis	Mode	Vascular manifestations
1	Hassen et Al[11]	43/Male	yes	9days	Angiography	Sma thrombosis
2	Mercie et al[12]	26/f	no	0	angiography	Sma thrombosis
3	Bayraktar et al[13]	41/male	yes	15	Surgical exploration	Sma thrombosis
4	Wu et al[14]	35/male	no	0	Surgical exploration	SMA aneurysm and thrombosis
5	Chubachi et al[15]	37/male	no	0	cect	Rt renal artery Aneurysm and SMA
6	Men et al[16]	23/male	no	0	Usg & angiography	Aneurysm SMA AND IMA
7	Hafsa et al[17]	32/male	no	0	Usg &cect	SMA aneurysm
8	Yokota et al[18]	24/male	yes	4	angiography	Sma ,rt renal,celiac artery aneurysm

9	Saiki et al[19]	50/male	yes	4	angiography	Sma ,iliac artery aneurysm
10	Ogula et al[20]	40/male	yes	9	angiography	Sma dissection

Some of the literature found ,sma aneurysm >sma thrombosis>sma dissection are the frequent patterns encountered

The literature review revealed that vasuculo-BD in the SMA can occur in both the chronic phase and the acute phase of BD. Multiple vascular lesions were more common than single lesions. CECT seems to be useful for whole- body blood vessel evaluation. Early initiation of steroid ther- apy is indicated if the situation does not require surgical treatmen The literature review revealed that vasuculo-BD in the SMA can occur in both the chronic phase and the acute phase of BD. Multiple vascular lesions were more common than single lesions. CECT seems to be useful for whole- body blood vessel evaluation. Early initiation of steroid ther- apy is indicated if the situation does not require surgical treatmen The literature review revealed that vasuculo-BD in the SMA can occur in both the chronic phase and the acute phase of BD. Multiple vascular lesions were more common than single lesions. CECT seems to be useful for whole- body blood vessel evaluation. Early initiation of steroid ther- apy is indicated if the situation does not require surgical treatmen The literature review revealed that vasuculo-BD in the SMA can occur in both the chronic phase and the acute phase of BD. Multiple vascular lesions were more common than single lesions. CECT seems to be useful for whole- body blood vessel evaluation. Early initiation of steroid ther- apy is indicated if the situation does not require surgical treatmen The literature review revealed that vasuculo-BD in the SMA can occur in both the chronic phase and the acutephase of BD. Multiple vascular lesions were more common than single lesions. CECT seems to be useful for whole-body blood vessel evaluation. Early initiation of steroid ther-apy is indicated if the situation does not require surgical treatment Our case series is of particular importance as it is female dominant(all 5 cases are female) ,age range from and SMA thrombosis is the major finding in all the cases.

Vascular pathology are thought to be of chronic in nature in behcets disease.however from our recent encounter it can present acutely as well. When there is unexplained abdominal pain in patients with BD, vasculo-BD should be considered as a differential diagnosis. When there is unexplained abdominal pain in patients with BD, vasculo-BD should be considered as a differential diagnosis. Di-agnostic modalities used to investigate vascular involvement in patients with BD include ultrasonography, CT, CECT, and angiography. Although abdominal ultrasonography combin- ing color and pulse Doppler can be performed initially, this method is unsuitable for evaluation of the intraperitoneal cavity, and so evaluation of the SMA is difficult. Angiogra- phy seems to be the best method for the diagnosis of aneu- rysms and pseudoaneurysms; Di-agnostic modalities used to investigate vascular involvement in patients with BD include ultrasonography, CT, CECT, and angiography. Although abdominal ultrasonography combining color and pulse Doppler can be performed initially,. Angiography seems to be the best method for the diagnosis of aneurysms and pseudoaneurysms[21]

The main therapeutic aim in vasculo-BD is to avoid arte- rial rupture (28). In the acute phase of aneurysm, high-dose prednisolone (1 mg per kg per day) or intravenous methyl-prednisolone pulse (1 g per day for 3 days) are recom- mended, and furthermore, the combined use of cyclophos- phamide, methotrexate, azathioprine, cyclosporine A is posi- tively considered (29). In addition, effective cases of inflixi- mab, a biological agent, have also been reported (30), so considering combination therapy. Although early prednisone therapy was successful in the present case, prednisone ther- apy alone is more likely to result in recurrence of vasculo- BD than combined treatment with methotrexate or other im- munosuppressive agents.

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CONCLUSION

Behcets is a multispectrum,autoimmune,relapsing inflammatory diseaseof unknown aetiology.although male preponderance has been documented in various case studies,from our institutional followup female were more likely to get affected.also once thought to be of chronic in nature ,more and more acute symptoms with varying clinical presentation such as thrombosis ,aneurysm and dissection are encountered.5 patients that we have encountered sofar in our institution were taken up for upfront surgery without delay (median 8 hrs)from entry into emergency department, isolated sma pathology was so far identified.out of 5 ,1 patient succumbed on POD 3.remainder of pateints recouperated well,however how output stomas was the major post operative problem .Nutritional care,multimodality approach using TPN,stoma refeeding, physiotherapy, incentive spirometry, psychosocial support all played part in the recovery of these patients.they were followed for period of 6 months and 3 of them underwent stoma reversal with good outcome.

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