

Intestinal complications of Behçet's Disease

3-26% of people with Behçet's have intestinal (gut) problems. The prevalence of intestinal problems may be even higher in Japan and Korea (50-60%) but is much lower in Turkey and the Middle East.

Behçet's can affect any part of the bowel, from the mouth to the anus. Apart from mouth ulcers which occur in almost everyone with Behçet's, the commonest part of the bowel to be affected is the ileocaecal junction; the area where the small and large bowels join, and (incidentally) the location of the appendix. The colon (large bowel) is affected less commonly and involvement of the rectum, oesophagus (gullet) or stomach is quite rare.

Behçet's causes ulcers in the bowel, similar to those seen elsewhere. They may be large or small, single or multiple. They are usually 'punched out' or 'undermining' and may penetrate through the whole bowel wall, causing a perforation and peritonitis.

The mucosa (lining of the intestine) around the ulcers is inflamed and may become thickened forming an inflammatory mass which may mimic the appearance of a polyp, or in extreme cases, may be mistaken for a tumour. The intestine is usually normal in between the areas of inflammation and ulceration. Examination of biopsies of affected intestine under the microscope shows ulcers and inflammation of the whole of the bowel wall. The ulcers are surrounded by areas of neutrophilic phlebitis (inflammation of vein) with a perivascular neutrophilic inflammatory infiltrate, with some predominantly CD4+ lymphocytes (inflammation surrounding the blood vessels). There is typically little fibrosis (scarring) and no granulomata (inflammatory masses typical of another intestinal condition, Crohn's disease).

The following drugs used in Behçet's disease can sometimes affect the intestine:

- Corticosteroids (prednisolone)
May cause dyspepsia (acid indigestion) and stomach or duodenal ulceration.
- Azathioprine
May cause diarrhoea with vomiting or fever- should be stopped if this occurs.
- Tacrolimus
May cause dyspepsia and gut ulceration or inflammation.
- Colchicine
May cause diarrhoea. A reduction in the dose is often effective if this occurs.

Please note; this is not a comprehensive list of drugs which may affect the intestine. Sometimes drugs with a risk of intestinal side effects may nevertheless be the best choice to treat intestinal disease.

Symptoms of Behçet's disease in the intestine

Symptoms of Behçet's in the intestine include abdominal pain, distension, diarrhoea and nausea. Ulcers may occasionally bleed, causing frank blood to appear in the stool if the ulcer is in the lower colon, rectum or anus, or if the ulcer is higher up in the bowel, may cause anaemia of black tarry stool ('melaena') due to blood which has passed around the bowel. Passage of any melaena or of very large amounts of fresh blood is a medical emergency.

If an ulcer perforates the bowel, the patient is extremely ill, with severe pain, and abdominal tenderness, fever, vomiting and cessation of all bowel movements, including flatus.

The bowel symptoms of Behçet's are similar to those of several other inflammatory bowel conditions; Crohn's disease, ulcerative colitis and bowel-related seronegative arthritis. The presence or absence of non-bowel symptoms, the site of involvement of the bowel and the histological (microscopic) appearance of biopsy samples will help differentiate Behçet's from these other disorders.

Investigation of intestinal problems

Tests may include (depending on symptoms):

- colonoscopy (camera passed through the back passage to look around the bowel as far as the ileocaecal junction) with biopsies (small samples of tissue for examination under the microscope).
- Gastroduodenoscopy (camera passed through the mouth to examine the oesophagus, (gullet) stomach and duodenum).
- Barium enema (barium contrast and air are pumped into the colon via the back passage and X-rays are taken. Enables views of colon and rectum)
- CT scanning with contrast. Enables views of whole bowel. May help to identify severely involved areas.
- Barium "follow through" (barium swallowed and Xrays taken as barium travels through small intestine- an area which is not examined by colonoscopy or gastroduodenoscopy).
- Ultrasound (enables examination of major structures in the abdomen)

Treatment of Intestinal Complications of Behçet's Disease

Treatment with bowel anti-inflammatory drugs such as melsalazine, or non-absorbable steroids may be helpful. For more severe problems, or where treatment is needed for other parts of the body, corticosteroids (prednisolone) or immunosuppressive drugs may be required. Azathioprine or infliximab, although unproven, may be particularly helpful for intestinal disease.

If perforation or uncontrolled bleeding occurs, or if there is a large inflammatory mass, surgery may be necessary. Behçet's intestinal disease may recur after surgery. Since the chance of recurrence is not reduced by removing a larger area of intestine, the smallest amount of intestine necessary for control of the problem should be removed.

Dr Hilary Longhurst, April 2008