

9. Intestinal Complications of Behçet's disease

How are the intestines affected by Behçet's disease?

Behçet's disease can affect any part of the intestines (gut), from the mouth to the anus. Apart from mouth ulcers, which occur in almost everyone with Behçet's disease, the most common part of the intestines 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 disease 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 (*Perforation is when there is an abnormal opening in the bowel wall which causes the contents of the bowel to leak out. Peritonitis is inflammation of the thin layer of tissue that lines the inside of the abdomen.*)

The mucosa (lining of the intestine) around the ulcers is inflamed and may become thickened, forming an inflammatory mass that 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 (*removal of a small amount of tissue for examination under a microscope*) 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; it 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 that may affect the intestine. Sometimes drugs with a risk of intestinal side effects may nevertheless be the best choice to treat intestinal disease.)

Symptoms

3–26% of people with Behçet's disease have intestinal problems. The prevalence (how many people are affected by the illness) of intestinal problems may be even higher in Japan and Korea (50–60%) but is much lower in Turkey and the Middle East. Symptoms of Behçet's disease in the intestine include abdominal pain, distension (stretching or swelling), diarrhoea and nausea. Ulcers may occasionally bleed, causing frank (obvious) blood to appear in the stool if the ulcer is in the lower colon, rectum or anus. If the ulcer is higher up in the bowel, bleeding may cause anaemia of black tarry stool ('melaena') due to blood that 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 (breaks through) the bowel, the patient is extremely ill, with severe pain, and abdominal tenderness, fever, vomiting and cessation of all bowel movements, including flatus. (gas from the stomach or bowels more commonly known as "wind ")

The bowel symptoms of Behçet's disease are similar to those of several other inflammatory bowel conditions, such as 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 to differentiate Behçet's disease from these other disorders.

Investigations

Tests may include (depending on symptoms):

- Colonoscopy (a camera is 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 (a camera is 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; this enables views of the colon and rectum).
- CT (computed tomography) scanning with contrast. This enables views of whole bowel and may help to identify severely involved areas.
- Barium 'follow through' (barium is swallowed and X-rays are taken as it travels through the small intestine, an area that is not examined by colonoscopy or gastroduodenoscopy).
- Ultrasound (this enables examination of the major structures in the abdomen).

Treatment

Treatment with bowel-specific anti-inflammatory drugs, such as mesalazine, 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. Intestinal complications of Behçet's disease may recur after surgery. As the chance of recurrence (something that happens again) is not reduced by removing a larger area of intestine, the smallest amount of intestine necessary for control of the problem should be removed.