Takayasu’s arteritis associated with idiopathic ulcerative colitis

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Abstract
Two patients with ulcerative colitis associated with Takayasu’s arteritis are described. Gangrene of a limb was the presenting feature in one patient and renovascular disease in the other. Angiography showed vascular occlusions affecting several medium or large sized vessels in both patients.

Inflammatory bowel disease is often complicated by intestinal and extra-intestinal manifestations. Common extra-intestinal complications include arthritis, episceralitis, erythema nodosum, and pericholangitis. Vascular complications are uncommon and include arterial and venous thrombosis, polyarteritis nodosa, and stenosing large vessel disease, and have been reported more frequently with Crohn’s disease than with ulcerative colitis. This report describes two unusual cases of idiopathic ulcerative colitis in patients who developed occlusive large vessel disease.

Case 1
A 35 year old Indian woman of Aryan descent had idiopathic ulcerative colitis. She had been treated with sulphasalazine, prednisolone enemas, and oral steroids for the past four years but the disease had not remitted. At the time of admission to hospital she was passing 10 to 12 stools a day mixed with blood and mucus. Two months before admission she had developed a painful swelling of the entire left leg which was diagnosed as iliofemoral venous thrombosis. This was followed a few days later by the abrupt onset of shooting pain and bluish discoloration of the left forearm and fingers. After heparin treatment at another hospital there was an improvement in the blood supply and colour of the skin of the left arm and resolution of venous thrombosis in the left leg. Two weeks before admission to this hospital, she had severe pain in the right leg which was followed by intermittent claudication.

There was no past history of diabetes, hypertension, or thromboembolism. She had not been on oral contraceptives.

At admission to hospital, the patient had moderate pallor. There was no clubbing, pedal oedema, or lymphadenopathy. Examination of the peripheral arteries showed absent pulses in the left radial, brachial, subclavian, and the right popliteal, posterior tibial, and dorsalis pedis arteries. Pulses were reduced in the left popliteal artery. In the right radial, both carotid and femoral pulses were normal. Blood pressure measured in the right arm was 130/75 mmHg. Systemic examination was normal. No bruit was heard over the abdomen.

Laboratory investigations showed a haemoglobin concentration of 6·7 g/dl, a total leucocyte count of 7·8×109/l, and a platelet count of 300×109/l. The erythrocyte sedimentation rate was 55 mm in the first hour, the serum creatinine concentration was 79·6 µmol/l, and the fasting blood sugar was 5·06 mmol/l. The serum cholesterol concentration was 4·29 mmol/l, triglycerides 0·77 mmol/l, and phospholipids 1·4 g/l. The clotting time was four minutes, the prothrombin index was 93%, and the serum fibrinogen value was 2·1 g/l. A test for antinuclear antibodies was negative. Stool examination showed numerous pus cells and red blood cells. Sigmoidoscopy showed a granular haemorrhagic and friable mucosa with loss of vascular pattern. A rectal biopsy specimen showed evidence of acute on chronic colitis. A double contrast barium enema x ray showed that the entire colon was affected, with loss of haustral markings and backwash ileitis (Fig 1). Echocardiography did not show any vegetations. Aortic arch and lumbar angiography showed occlusion of the left subclavian artery near its origin, non-visualisation of the superior mesenteric artery, and a block in the right internal iliac artery near its origin and in the left internal iliac artery near its expected division into further branches. A right femoral angiogram showed complete obstruction at the femoro-popliteal junction without any distal run off (Fig 2).

Treatment for ulcerative colitis with sulpha-

Figure 1: Barium enema x ray of patient 1 showing loss of haustra and involvement of the entire colon.
salazine, steroid enemas, and oral prednisolone was continued, and intravenous heparin 5000 U six hourly was begun. Despite anticoagulation, gangrenous changes developed in the right foot and leg necessitating an above the knee amputation.

Examination of the amputated leg showed a thrombus in the popliteal artery and extensive thrombosis in the superficial and deep leg veins. Histopathological examination of the vessels showed organising thrombi and focal medial calcification in the arteries.

The patient made an uneventful recovery. Pancreatectomy for ulcerative colitis and a left subclavian bypass graft have been advised.

Case 2
A 16-year-old Indian male of Aryans descent presented with a three-year history of loose stools with blood and mucus and a four-month history of headaches. Hypertension had been detected two months before admission to hospital. There was no history of fever, arthralgia, skin rashes, pedal oedema, or haematuria.

On examination, there was moderate pallor. All his peripheral pulses were normal and his blood pressure, measured in right arm was 150/104 mmHg. There was tenderness over the colon. A systolic bruit was heard just above the umbilicus. Examination of the fundus showed moderate arteriolar narrowing.

The patient’s haemoglobin was 7.5 g/dl, total leucocyte count 10 × 10⁹/l, platelet count 400 × 10⁹/l, serum creatinine 123.8 μmol/l, fasting blood sugar 5.2 mmol/l, and serum cholesterol 4 mmol/l. Urine analysis showed no abnormality. Stool examination showed a cellular exude and numerous erythrocytes. Sigmoidoscopy, rectal biopsy specimen, and barium enema x ray confirmed the diagnosis of ulcerative colitis. Ultrasonic scan showed a right kidney of 9 cm and a left kidney of 8-2 cm. Aortography (Figs 3 and 4) showed irregularity of the abdominal aorta and complete occlusion of the superior mesenteric artery with a large collateral (artery of Drummond) arising from the inferior mesenteric artery. There was stenosis of the first cm of the renal arteries, which was more noticeable on the left side. The aortic arch was normal.

The patient’s hypertension was controlled with nifedipine. He was put on oral sulphasalazine and prednisolone enemas and his bowel symptoms improved. A percutaneous transluminal angioplasty of the renal arteries is planned.

Discussion
The two patients reported here had occlusive disease affecting several medium sized arteries that was associated with active idiopathic ulcerative colitis. Patient 1 also developed gangrene of the right leg as a result of arterial and venous thrombosis.

The occurrence of thromboembolism in patients with inflammatory bowel disease has been reported to be 1-2-6-4%. In a recent review of 7199 patients from the Mayo Clinic, 92 (1.3%) patients were found to have arterial or venous thromboembolic complications. Twenty four of these had thrombosis in the arterial tree and the sites affected included mesenteric, cerebral, coronary, and limb vessels.

An association of Takayasu’s disease with inflammatory bowel disease has also been documented, and five patients have so far been described. The simultaneous involvement of several arteries with the site of stenosis close to their origin from the aorta strongly supports the possibility of Takayasu’s disease in both these patients.

The association of Takayasu’s arteritis with inflammatory bowel disease prompts speculation on a common pathophysiology. Antibodies to aorta and colonic mucosa are often present in both Takayasu’s disease and inflammatory bowel disease. The association may be coincidental only, however, since Takayasu’s disease is not uncommon in this country.

Hypercoagulability associated with high values of coagulation factors V, VIII, and fibrinogen, low antithrombin III, thrombocytosis, bacterial endotoxaemia, and dehydration is thought to be responsible for the increased tendency to vascular thrombosis in patients with ulcerative colitis. It is possible that some of these factors predisposed to thrombotic occlusion of the right leg arteries and veins in patient 1. The absence of arteritis at the site of thrombosis is not surprising since the lesions of Takayasu’s disease are likely to have been present at a more proximal site.
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