CASE REPORT

Thromboangiitis obliterans (Buerger’s Disease) in Visceral Vessels Confirmed by Angiographic and Histological Findings

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Introduction

Thromboangiitis obliterans (TAO) differs from arteriosclerotic and other inflammatory vascular diseases in its course, as well as in angiographic and histological patterns. The disease occurs predominantly in young male tobacco smokers, the onset of the disease usually occurring before the age of 40 years. It principally involves the medium-sized and small arteries and veins of both the lower and upper extremities; however, involvement of the coronary arteries, cerebral vasculature, renal arteries, and mesenteric arteries have been documented as single case reports. This report refers to a single case of TAO with angiographic and histologic evidence of superior mesenteric vessel involvement.

Case Report

The patient was a 42-year-old man who had been a heavy smoker since the age of 18. His symptoms included painful restriction of his walking distance to less than 300 metres and chronic abdominal pain. At the age of 27 he sustained an upper limb venous thrombosis which was diagnosed as effort thrombosis and 4 years later he experienced a second episode affecting the same limb. At age 42, he was admitted to our hospital complaining of acute abdominal pain and vomiting. The clinical evaluation and X-rays of the abdomen revealed small bowel obstruction. Colonoscopy did not reveal any pathological findings. Several hours later, the patient experienced acute periumbilical pain. Abdominal X-rays revealed free air under the diaphragm and a laparotomy was performed. Rupture of the terminal ileus was found and a partial enterectomy was performed.

Histologically, the small bowel segment showed a 10 cm long area with congestion, thinning and deep ulceration of the wall. Chronic ulceration penetrating the bowel wall was found within the setting of chronic ischaemic bowel disease. The medium sized mesenteric arteries and veins showed mild inflammatory infiltration and a great number were occluded and recanalised. Inflammatory cells infiltrated throughout the media and adventitia, but no necrotising lesions were found in the media. The vessel wall was generally well preserved and no arteriosclerosis obliterans was detected. Histological findings were consistent with Buerger’s disease (Fig. 1).

A diagnostic evaluation for possible Buerger’s disease was therefore undertaken. An intra-arterial angiogram of the abdominal aorta, visceral vessels, upper and lower limbs and the carotids was performed. The pathological findings were as follows:

Superior mesenteric artery. Collateral vessels with a corkscrew appearance from the arteries of the small bowel to the right colon. Terminal branches of the small bowel were occluded in an abrupt manner. The right colic artery was not visualised (Fig. 2a).

Inferior mesenteric artery. This could not be visualized and catherisation was not feasible. Collaterals from the pancreatic to the left colic artery were present.

Lower limbs. There was focal stenosis of the right common femoral artery and a corrugated appearance of the femoropopliteal segment on the left limb. The
tibial vessels were visualised by collaterals which had a corkscrew appearance. The tibial vessels showed tapering occlusions at the level of the ankles bilaterally (Fig. 2b).

During hospitalisation, the patient’s general laboratory parameters were within normal range except for an ESR of 42 mm/h, c-reactive protein: 19.6 mg/dl and leukocytes: 12.1 × 10⁹ µl. Urine sediment and 24 h urine proteins, electrolytes and creatinine clearance values were normal. Oral glucose tolerance tests initially showed normal values. Immunologic findings, including autoantibodies against anti-ds-DNA, anti-ss-DNA, anti-Sin, anti-Ro, anti-La, anticollagen, antielastin and anticardiolipin were negative. Findings were also negative for antineutrophil cytoplasmic antibody (ANCA), circulating immunocomplexes, rheumatoid factors and the complement factors. C₁, C₂ and C₉. Repeated Quick test and partial thromboplastin time (PTT) were within normal values. Plasminogen, fibrinogen and antithrombin III, proteins C and S were within normal range. Lupus anticoagulant, cold agglutinins, cryoglobulins and cryofibrinogen were not found.

Discussion

The clinical criteria for the diagnosis of TAO as proposed by Shionoya¹ are: (1) history of smoking, (2) onset before the age of 50 years, (3) infrapopliteal arterial occlusive lesions, (4) either upper limb involvement or phlebitis migrans and (5) absence of atherosclerotic risk factors other than smoking. However, many of the clinical signs and symptoms are common to other vascular ischaemic diseases. This has led to an exasperating state of affairs with each investigator choosing a different set of criteria.²³ There is evidence that thromboangiitis obliterans is a true clinicopathologic entity distinct from arteriosclerosis.⁴ Thromboangiitis obliterans is almost exclusively a disease of the blood vessels of the extremities. There
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have been only associated reports, usually without adequate histological proof of involvement of the large arteries. Although it was first mentioned by Buerger in 1908 that vascular obliteration could include blood vessels other than those of the limbs, to date, organ involvement in TAO has been described only in single case reports namely: coronary, pulmonary, cerebral and intestinal involvement.

In our case report, the patient fulfilled all the classic criteria of TAO, the only difference being that the course of the disease was complicated by mesenteric vessel involvement. The patient had a typical arteriogram for TAO with multiple segmental occlusions of distal extremity arteries and an extensive reticular network of collaterals around each occlusion with a characteristic corkscrew or root-like appearance. Unlike arteriosclerosis obliterans, neither calcification of the vessel wall nor plaques were seen. A similar arteriographic appearance was seen in the superior mesenteric artery circulation as described in the previous case report. We also had the opportunity to obtain histopathological confirmation of TAO in the intestinal vessels. Our findings were of organised thrombi in the mesenteric arteries and veins with residual inflammatory reaction and early intimal proliferation i.e. an intermediate-to-chronic stage pattern of Buerger’s disease. The early stage patterns of the disease i.e. granulomatous tissue, polynucleated leukocytes and giant cells were not seen. Although intermediate stage lesions have far fewer characteristic features and are seldom diagnostic, marked cellular proliferation and inflammatory infiltrate are rarely seen in the ordinary arterial and venous thrombi.

Intestinal perforation appeared many years after claudication. This supports the existing evidence that patients can develop intestinal Buerger’s disease at any time during the course of their disease, either early as an initial event or much later. This suggests that gastrointestinal complaints by patients suffering from TAO should be taken seriously and thoroughly investigated. Chronic symptoms such as unexplained malabsorption or diarrhoea suggest the possibility of

Fig. 2. (A) Right superficial femoral artery severe stenosis and extensive reticular network of collaterals, (B) superior mesenteric artery occlusion of the right colic artery and root like appearance of collaterals.
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coeliac disease. Although the relationship between Buerger's disease and coeliac disease remains unknown, an immunological connection is suspected.\textsuperscript{11}

Our case can be added to the small list of six reported cases in the existing literature where there was histologic evidence of TAO in the mesenteric circulation. Although this paucity of reports suggests that mesenteric TAO rarely presents clinically, autopsy studies indicate that 20\% of patients have histological evidence of TAO.\textsuperscript{12} The rarity of organ involvement in TAO makes the diagnosis difficult despite the clearly established diagnostic criteria for TAO of the limbs. However, the following proposals can be made:

1. Existence of all classic clinical diagnostic criteria for TAO of the peripheral vessels.
2. Symptoms and/or signs of arterial ischaemia of the involved organ.
3. Angiographic and histologic findings of TAO in the arterial supply to the involved organ.

References


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