Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

Isolated visceral manifestation of Buerger's disease presenting as intestinal obstruction: a case report

Ramesh Rana, MD[a], Satyadeep Bhattacharya, MD[b], Baishali Bhattacharya, MD, MPH[d], Ram K. Ghimire, MD[c], Neeraj Joshi, MD[a,*]

Introduction and importance: Buerger's disease is an uncommon segmental nonatherosclerotic vasculitis essentially affecting small to medium-sized arteries and veins of upper and lower extremities and can lead to limb amputation. Visceral vessel involvement is quite rare accounting for 2% of cases presenting with acute abdomen due to mesenteric ischemia. Moreover, isolated visceral involvement is even rare.

Case presentation: A 42-year-old gentleman, a chronic smoker, presented with abdominal pain associated with nausea and vomiting and loose stool of 2 months duration. Magnetic resonance enterography revealed segmental circumferential wall thickening with stricture in the mid part of the jejunum with lymphadenopathy features of possible inflammatory bowel disease (Crohn's disease). Furthermore, intraoperative surgical findings were also suggestive of Crohn's disease. However, histologic findings were consistent with thromboangiitis obliterans.

Discussion: Thromboangiitis obliterans can present with inflammatory vascular lesions without necrosis in the early stage to varying degrees of recanalisation, gangrene, and amputation in the late stage. It rarely involves the brain, heart, and abdominal viscera. The visceral involvement may be in the form of intestinal obstruction or mesenteric ischemia or can mimic Crohn's in a background of smoking.

Conclusion: This case report will help to learn more about the rarer intestinal presentation of intestinal Buerger's disease. It can present with features of bowel ischemia, obstruction or Crohn's. So, histology would play a pivotal role in differentiating the diagnostic dilemma.

Keywords: Buerger's disease, case report, ischemia, thromboangiitis obliterans, visceral complications

Introduction

Buerger's disease is a relatively rare segmental nonatherosclerotic vasculitis predominantly affecting small to medium-sized arteries and veins, especially of upper and lower extremities, and can lead to limb amputation. It has a lower prevalence than peripheral arterial disease, even in countries where it is prevalent. Visceral vessel involvement is quite rare, accounting for 2% of cases presenting with acute abdomen due to mesenteric ischemia[1–4]. The etiology is unknown and no definite treatment protocol is available. However, smoking is a known risk factor and smoking cessation prevents disease progression[2,3].

We report a very uncommon case of visceral manifestation of Buerger's disease mimicking features of Crohn's disease presenting as intestinal obstruction.

HIGHLIGHTS

- Buerger's disease is a rare segmental nonatherosclerotic vasculitis of small and medium-sized vessels of extremities and can lead to limb amputation.
- Visceral involvement is relatively rare accounting for 2% of cases presenting with acute abdomen due to mesenteric ischemia. Isolated visceral involvement is even rare.
- Our case is a 42-year-old gentleman, a chronic smoker who presented with clinical features suggestive of partial bowel obstruction, radiological imaging, and intraoperative findings were in favor of Crohn's disease. However, histopathological findings revealed Buerger's disease. So histopathological correlation has great importance in case of dilemma.

Case presentation

We report a 42-year-old gentleman who presented with chief complaints of abdominal pain and distension, associated with nausea and vomiting for 2 months. Sometimes pain was aggravated by food intake. Stool consistency was loose with a

[a]Department of Gastroenterology and Hepatology, [b]Department of Colorectal Surgery, [c]Department of Radiodiagnosis and Interventional Radiology, Nepal Mediciti Hospital, Bhaisepati, Lalitpur and [d]Department of GI and Liver Pathology, Samyak Pathology Lab, Baneshor, Kathmandu, Nepal

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

*Corresponding author. Address: Department of Gastroenterology and Hepatology, Nepal Mediciti Hospital, Bhaisepati, Lalitpur, Nepal. Tel.: +977 980 203 5500. E-mail: neerajjoshi@hotmail.com (N. Joshi).

Annals of Medicine & Surgery (2024) 86:3770–3775

Received 24 February 2024; Accepted 17 April 2024

Published online 29 April 2024

http://dx.doi.org/10.1097/MS9.0000000000002111

fluctuating frequency of 4–6 episodes/day, sometimes associated with mucus without blood, and one episode of black stool 2 months before presentation. He also gave a history of pain and tightness over his left lower leg on walking long distances or brisk walks, which relieved by rest. Furthermore, there was a history of left lower leg discolouration and decreased sensation and was advised to get the opinion of a vascular surgeon 7–8 years ago. However, he did not seek a vascular opinion and the symptoms improved later, with no similar issues to date. He has a history of binge drinking and smoking since the age of 20 amounting to 20 pack years. On examination, his vitals were stable (pulse 80 beats per minute, blood pressure 139/90 mmHg, afebrile, respiratory rate of 18/min), with normal bilateral dorsalis pedis and popliteal arterial pulsation. On abdominal examination, multiple small lipomas with mild epigastric tenderness observed in a non-distended abdomen. Respiratory and cardiac examinations were normal. Laboratory investigation showed hemoglobin mildly elevated at 18.4 gm/dl and C-reactive protein of 22.6 mg/dl, otherwise lab values were within the normal limits (Table 1). Initial ultrasonography showed mural thickening and dilatation of jejunal loops in the left hypochondrium and epigastric region with normal peristalsis suggesting enteritis. Furthermore, magnetic resonance enterography in our center revealed segmental circumferential and symmetrical minimal wall thickening with stricture in the mid part of the jejunum with features of small bowel obstruction, prominent vasa recta, and few enlarged lymph nodes in mesentery suggesting probable inflammatory bowel disease (IBD-Crohn's disease). The celiac trunk, proximal superior mesenteric artery (SMA) and inferior mesenteric artery were normal, however, few beaded appearances of the distal branches of SMA were seen without occlusion (Fig. 1). Given the history suggestive of recurrent subacute bowel obstruction, laparoscopic-assisted resection anastomosis of the jejunum was performed. Intraoperative findings showed dense adhesion between the jejunal loop (10 cm), omentum and transverse colon, circumferential stricture in the jejunum in the adhesed loop 80 cm from the duodenojejunal flexure, and dilated proximal loop with collapsed distal loop. Multiple enlarged mesenteric lymph nodes were observed adjacent to the stricture. Therefore, the post-operative first differential was IBD-Crohn's. However, the histology revealed a denuded ulcerative mucosa with acute and chronic inflammation, hemorrhage, and necrosis extending to the submucosa. No granulomas were seen. There was marked fat necrosis in mesenteric fat with histiocytic reaction, giant cells and broad areas of sclerosis. Submucosa also showed sclerosis. Blood vessels, especially arterioles showed intimal thickening with edema and fibrosis and many with near occlusion, rare small vessel inflammation. Fibrinoid necrosis was not seen and the internal elastic lamina was intact. Adjacent mucosa showed regenerative villi. Lymph nodes showed reactive changes. The severe mesenteritis could be related to a healed perforation/mural necrosis. The features were consistent with thromboangiitis obliterans or Buerger's disease given the histological findings and history of heavy smoking (Fig. 2).

The patient was advised to strictly quit smoking. Further investigations were done to rule out Buerger's disease-related other organ involvement. Both venous and arterial Doppler scans of bilateral lower limbs were normal. Mild mitral regurgitation was seen on echocardiography without other abnormalities, which have no clinical significance with Buerger's disease. The patient was asymptomatic at 6 weeks follow-up and the hemoglobin dropped to 16.7 g/dl on subsequent consultation.

This work has been reported in line with the Surgical Case Report (SCARE) 2023 criteria[5].

Table 1

Routine laboratory findings

	Lab parameters	Count	Range	Unit
Hematology	*White blood cells*	6280	4000–11 000	cells/cumm
	Red blood cells	5.63	4.44–5.61	millions/cumm
	Hemoglobin	18.4	13.5–16.9	gms%
	Packed cell volume	51.4	40–50	%
	Platelets	339000	150 000–450 000	cells/cumm
	MCV	91.3	81.8–95.5	fl
	MCH	32.7	27–32.3	pg
	MCHC	35.9	32.4–35	gm/dl
	ESR	11	< 15	mm/hr
	CRP	22.6	< 10	mg/dl
	Prothrombin time	13.10	11–16	sec.
	INR	1.01		
Biochemistry	Fasting blood sugar	93.00	74–106	mg/dl
	Urea	16	15–45	mg/dl
	Creatinine	0.9	0.66–1.25	mg/dl
	Sodium	135	137–145	mmol/l
	Potassium	4.30	3.5–5.1	mmol/l
	Total cholesterol	174	< 200	mg/dl
	Triglycerides	69	< 150	mg/dl
	HDL	34	< 40	mg/dl
	LDL	126.15	< 100	mg/dl
	VLDL	13.80	< 30	mg/dl
	Albumin	4.20	3.5–5.0	g/dl

CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; HDL, high-density lipoprotein; INR, international normalizing ratio; LDL, low-density lipoprotein; MCH, mean corpuscular hemoglobin; MCHC, mean corpuscular hemoglobin concentration; MCV, mean corpuscular volume; VLDL, very low-density lipoprotein.

Discussion

Thromboangiitis obliterans, also known as Buerger's disease, is a nonatherosclerotic, segmental inflammatory and obliterative vascular disease of small to medium-sized arteries, veins, and nerves. It has three phases: i) acute phase: characterized by acute inflammation involving all layers of the vessel wall in association with occlusive cellular thrombosis containing polymorphonuclear leukocytes, multinucleated giant cells, and micro-abscess evolved in the distal circulation. In contrast, the internal elastic lamina appears relatively spared in other systemic vasculitis. ii) Subacute phase: there is a progressive organization of the occlusive thrombus in the arteries and veins with the persistence of inflammatory cells. And iii) Chronic phase: characterized by complete organization of the occlusive thrombus with extensive recanalization, prominent vascularization of the media and adventitial and perivascular fibrosis with no presence of inflammatory cells. In all three stages, the normal architecture of the vessel wall including the internal elastic lamina remains intact, which distinguishes it from atherosclerosis and other systemic vasculitis. Extensive arterial occlusion accompanied by the development of corkscrew collateral vessels is characteristic of angiographic findings but not pathognomonic[6,7]. In the late stage, the thrombus becomes organized leading to varying degrees of recanalization and subsequent gangrene and amputation[8]. The involvement of the brain, heart, and

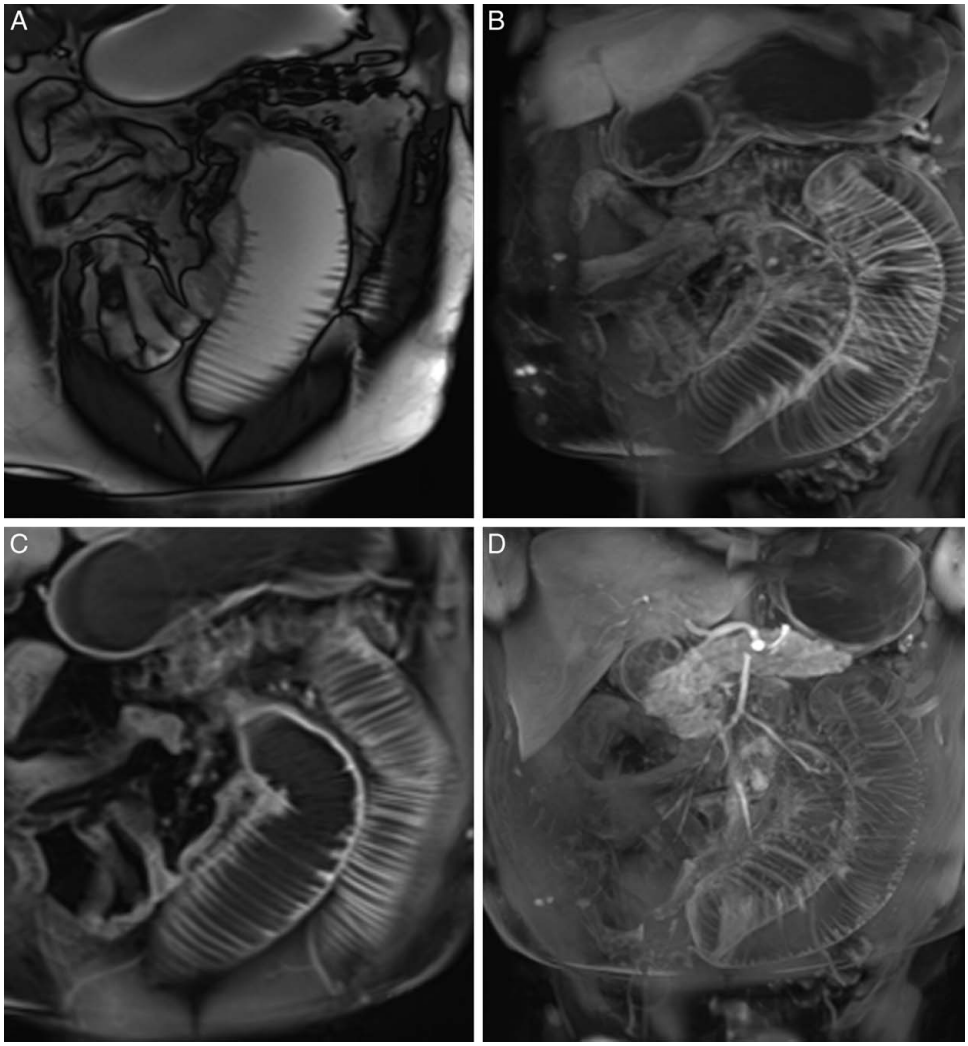

Figure 1. A, B, C: Magnetic resonance enterography showing circumferential wall thickening with a stricture of mid jejunum with proximal dilatation of the lumen. D: magnetic resonance angiography showing few beaded appearances of the distal superior mesenteric arterial branches without occlusion and with the normal celiac trunk, proximal superior mesenteric artery, and inferior mesenteric artery.

abdominal viscera is very rare; however, it can have visceral involvement before involvement of the extremities[3,8]. It is most common in the Orient, Southeast Asia, India, the Middle East and the Far East, but least common in North America and Western Europe. It affects predominantly men than women, especially young male smokers[8–10]. However, its prevalence is rising due to increased smoking in women as well[11,12]. They can present with symptoms of abdominal pain or postprandial pain, weight loss, and bloody stool. Diarrhea can present in 25% of cases[13]. Smoking has a strong correlation with disease involving direct idiosyncratic toxicity caused by a component of tobacco or an immune response to the same agents that have modified host vascular wall proteins[8,10,14]. There is no definite treatment for this disease; however, administration of injectable iloprost, a synthetic analog of prostacyclin PGI2 over 28 days was superior to aspirin and had better results in relieving the pain and complete healing of all trophic changes[10]. Smoking cessation has a high preventive value in disease progression rate with only 5% of ex-smokers developing disease in another site compared to 100% in those who continue smoking[4].

We found 45 case reports regarding intestinal Buerger's disease in the literature and those with full details are shown in Table 2. However, some case reports were in non-English language and the details were inadequate[12,13,15–26].

In our case, the patient presented with abdominal pain and distension associated with nausea and vomiting. The abdominal imaging and intraoperative findings were suggestive of probable IBD-Crohn's. However, the histologic finding was consistent with the Buerger's disease. According to the systematic review, the mean number of cigarettes smoked daily was 32 ± 12 and the duration of smoking before mesenteric ischemia was 22 ± 8 years[3]. In our case, the average number of cigarettes smoked per day was 20, especially in the last 10 years and the duration of smoking was 22 years before abdominal symptoms. In the majority, the SMA involvement was seen (53%), followed by both (22%), inferior mesenteric artery (12.5%), and celiac artery and its branches (12.5%). There was only visceral

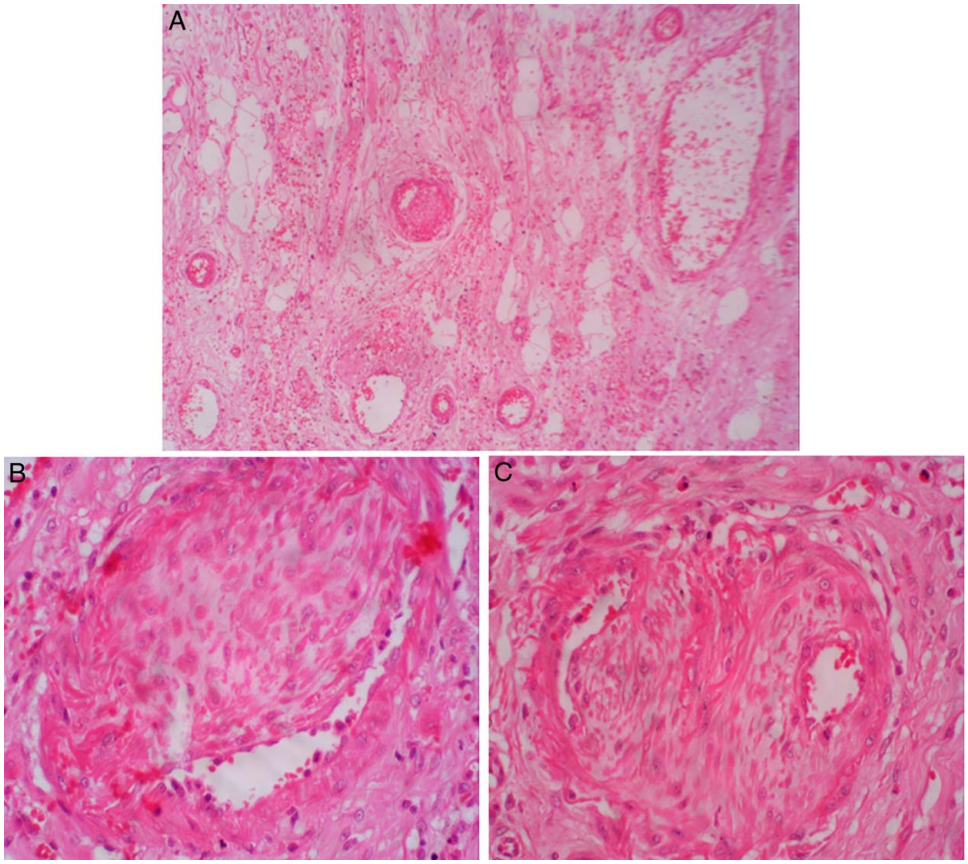

Figure 2. A, B, C: Histopathological picture showing the foci of ulcerative mucosa with acute and chronic inflammation, hemorrhage and necrosis. Arterioles showing intimal thickening with edema and fibrosis and many with near occlusion, rare small vessel inflammation.

Table 2

Buerger's disease case reports with gastrointestinal manifestations

Authors	Age (years)/sex	Peripheral involvement	GI symptoms	Site of GI involvement
Bhushan M *et al.*[7]	45/female	Toe and finger amputation	Pneumo peritoneum	Ascending colon predominant colon
Mishra SV *et al.*[4]	45 years	Right midfoot amputation	Pain abdomen, vomiting, loose stool	Jejunum and appendix
Naqvi H A *et al.*[1]	38 years/male	Lower leg amputation	Intermittent rectal bleeding	Sigmoid colon
Darshan J *et al.*[15]	48 years/male		Abdominal pain and diarrhea	Proximal jejunum
Lee KS *et al.*[8]	65 years	Migratory thrombophlebitis s/p RLL amputation	Periumbilical and RUQ pain	Sigmoid colon
	39 years	Toe amputation	RLQ pain, bloody stool	Sigmoid colon
Edo N *et al.*[16]	54 years/male	Below knee amputation	Severe abdominal pain and vomiting	Superior mesenteric artery
Magalhães ED *et al.*[17]	41 years/male		Periumbilical and right lower quadrant pain, vomiting	Ileum
Cho YP *et al.*[18]	37 years/male	Claudication of left hand	Diffuse abdominal pain	Jejunum and proximal ileum
Medlicott SA *et al.*[11]	43 years/female	Right leg claudication	Abdominal pain	jejunum
Kobayashi M *et al.*[19]	42 years/male	Left leg amputation	Abdominal pain	Terminal ileum, cecum and part of ascending colon
Kurata A *et al.*[20]	35 years/male	Gangrenous left toe with ulcer	Abdominal pain	Ileum
Adem C *et al.*[21]	37 years/male	Distal limb	Unexplained abdominal pain	Mesenteric infarction
Arkkila PE *et al.*[22]	50 years/male			Perforation of colon
Ziad H *et al.*[12]	50 years/male	History of Iterative amputations	Postprandial epigastric pain, vomiting I& diarrhea	Preterminal Small bowel
MZ Siddiqui *et al.*[23]	51 years/male	Peripheral artery disease		Recurrent intestinal ischemia
Ito M *et al.*[24]	42 years/male	Below knee amputation		Cecum and proximal ascending colon
Michail PO *et al.*[25]	42 years/male	Upper limb venous thrombosis	Abdominal pain and vomiting	Terminal ileum
Herrington JL *et al.*[14]	33 years/male		Left lower quadrant abdominal pain	Sigmoid colon
	42 years/male		Abdominal pain, anorexia, weight loss	Distal ileum

involvement, beaded appearance in the distal branches of SMA without occlusion and obvious peripheral vascular involvement in our case as reported in the literature, which is extremely rare[3].

Our patient is unique with features of Buerger's disease. While only abdominal symptoms suggesting subacute intestinal obstruction were there. The initial thought was probably IBD-Crohn's until the histology report confirmed the diagnosis and retro-prospectively we evaluated the patient to see if other sites of ischemia were involved or not. However, he did not have significant involvement of other sites except visceral involvement leading to intestinal obstruction besides his smoking habit.

There were some limitations of our case report as well. We did not rule out other vasculitis which can mimic Buerger's disease such as Behcet's syndrome, rheumatoid arthritis, and systemic lupus erythematosus[27].

Conclusions

Intestinal Buerger's disease without peripheral involvement is very uncommon. It can mimic Crohn's or present with features of bowel ischemia or obstruction. Therefore, in that case, Buerger's disease should be kept in the differential. Moreover, histopathology would play a pivotal role in differentiating the diagnostic dilemma. As in our case, the histopathology revealed Buerger's disease where the primary differential was Crohn's in a background of clinical findings, radiological imaging, and intraoperative findings.

Ethical approval

Ethical approval was taken, Institutional Review Committee (IRC) Nepal Mediciti- A Unit of Ashwins Medical College and Hospital Pvt. Ltd. Ref No: IRC-CR-2080/81-04.

Consent

Patient informed consent was obtained.

Sources of funding

Not applicable.

Author contribution

All the authors contributed equally in drafting, editing, revising, and finalizing the case report.

Conflicts of interest disclosure

Not applicable.

Research registration unique identifying number (UIN)

Not applicable.

Guarantor

Neeraj Joshi and Ramesh Rana.

Data availability statement

Not applicable.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Acknowledgements

Not applicable.

References

[1] Naqvi HA, Bilal M, Yousuf S. Ischemic colitis in buerger's disease: case presentation and review. Cureus 2020;12:e8303.
[2] Bourgeau ME, Key PG, Chou J, *et al*. Left carotid artery thrombosis due to thromboangiitis obliterans. Am J Forensic Med Patholo 2021;42: 297–300.
[3] Fakour F, Fazeli B. Visceral bed involvement in thromboangiitis obliterans: a systematic review. Vasc Health Risk Manag 2019:317–53.
[4] Mishra SS, Mishra TS. Intestinal thromboangiitis obliterans: a case report. J Med Case Rep 2021;15:215.
[5] Sohrabi C, Mathew G, Maria N, *et al*. The SCARE 2023 guideline: updating consensus Surgical Case Report (SCARE) guidelines. Int J Surg 2023;109:1136–40.
[6] Aqel MB, Olin JW. Thromboangiitis obliterans (Buerger's disease). Vasc Med 1997;2:61–6.
[7] Olin JW. Thromboangiitis obliterans (Buerger's disease). N Engl J Med 2000;343:864–9.
[8] Bhushan M, Betzold RD, Al Diffalha S. Intestinal manifestation of Buerger's disease in a middle-age female with subsequent transverse colon perforation: a case report and review of literature. SAGE Open Med Case Rep 2023;11:2050313x231175295.
[9] Lee KS, Paik CN, Chung WC, *et al*. Colon ischemia associated with Buerger's disease: case report and review of the literature. Gut liver 2010; 4:287–91.
[10] Bogacz R, Gaik M, Uram E, *et al*. Thromboangiitis obliterans (Buerger's disease)-a review. J Educ Health Sport 2023;36:124–34.
[11] Puéchal X, Fiessinger J-N. Thromboangiitis obliterans or Buerger's disease: challenges for the rheumatologist. Rheumatology 2007;46:192–9.
[12] Medlicott SA, Beaudry P, Morris G, *et al*. Intestinal thromboangiitis obliterans in a woman: a case report and discussion of chronic ischemic changes. Can J Gastroenterol 2003;17:559–61.
[13] Hassoun Z, Lacrosse M, De Ronde T. Intestinal involvement in Buerger's disease. J Clin Gastroenterol 2001;32:85–9.
[14] Roberts JA, Meyer JP. Buerger's disease presenting as a testicular mass: a rare presentation of an uncommon disease. Urol Ann 2016;8:249–51.
[15] Edo N, Miyai K, Ogata S, *et al*. Thromboangiitis obliterans with multiple large vessel involvement: case report and analysis of immunophenotypes. Cardiovasc Pathol 2010;19:59–62.
[16] Magalhães Ede P, Trevisan M, Mochizuki M, *et al*. Intestinal ischemia as a single manifestation of thromboangiitis obliterans–a case report. Angiology 2005;56:789–92.
[17] Cho YP, Kwon YM, Kwon TW, *et al*. Mesenteric Buerger's disease. Ann Vasc Surg 2003;17:221–3.
[18] Kobayashi M, Kurose K, Kobata T, *et al*. Ischemic intestinal involvement in a patient with Buerger disease: case report and literature review. J Vasc Surg Jul 2003;38:170–4.
[19] Kurata A, Nonaka T, Arimura Y, *et al*. Multiple ulcers with perforation of the small intestine in buerger's disease: a case report. Gastroenterology Sep 2003;125:911–6.
[20] Adem C, Benamouzig R, Royer I, *et al*. [Buerger's disease or thromboangiitis obliterans revealed by an enteric ischemia. Case report and literature review]. Gastroenterol Clin Biol 2002;26:409–11.
[21] Arkkila PE, Kahri A, Färkkilä M. Intestinal type of thromboangiitis obliterans (Buerger disease) preceding symptoms of severe peripheral arterial disease. Scand J Gastroenterol 2001;36:669–72.
[22] Siddiqui MZ, Reis ED, Soundararajan K, *et al*. Buerger's disease affecting mesenteric arteries: a rare cause of intestinal ischemia–a case report. Vasc Surg 2001;35:235–8.

[23] Ito M, Nihei Z, Ichikawa W, *et al*. Intestinal ischemia resulting from Buerger's disease: report of a case. Surg Today 1993;23: 988–92.
[24] Michail PO, Filis KA, Delladetsima JK, *et al*. Thromboangiitis obliterans (Buerger's disease) in visceral vessels confirmed by angiographic and histological findings. Eur J Vasc Endovasc Surg 1998;16:445–8.
[25] Herrington JL Jr, Grossman LA. Surgical lesions of the small and large intestine resulting from Buerger's disease. Ann Surg 1968;168:1079–87.
[26] Darshan J, Shaw E, Green B, *et al*. Intestinal ischaemia and mesenteric necrosis in a heavy smoker. QJM 2012;106:183–6.
[27] Khat'kov IE, Rotin DL, Parfenov AI, *et al*. [Mesenteric inflammatory veno-occlusive disease is a rare nosological entity: literature review and the authors' clinical observation. Ter Arkh 2014;86:26–30.