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Syphilitic aortic aneurysm — a source of fatal intestinal bleeding (clinical case)

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ABSTRACT
An observation of late visceral syphilis in a senile patient complicated by syphilitic abdominal aortic aneurysm with aortoduodenal fistula and fatal intestinal bleeding is presented. The patient was taken to the hospital by an ambulance with complaints of severe weakness, dizziness, and black loose stool in the last few days. The patient was hospitalized in the intensive care unit. A history of gastrointestinal bleeding diagnosed several months ago. Repeated esophagogastroduodenoscopy and colonoscopy revealed twice-resuming gastrointestinal bleeding during 5 days of hospital stay. The source of bleeding has not been found. CT made it possible to diagnose aneurysm (mycotic?) of the infrarenal section of the abdominal aorta and suggest the presence of an aortoduodenal fistula. On the 5th day of hospitalization, the patient died. Syphilis is diagnosed pathomorphologically posthumously. Syphilitic gummas were found in the walls of the aorta, duodenum, and adjacent retroperitoneal tissue, and productive and granulomatous inflammation was found in the liver, lungs, heart, and pia mater. The diagnosis of tertiary visceral syphilis was confirmed histologically, including the finding of pale treponema during staining by Levaditi. The aortoduodenal fistula was formed by the fusion and decay of the gummas of the adjacent to each other the walls of the aorta and duodenum. During the life of the patient, serological tests for syphilis were not carried out.

Keywords: late visceral syphilis, gummas of the internal organs, abdominal aortic aneurysm, aortoduodenal fistula (fistula), recurrent intestinal bleeding.

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Introduction

The most formidable complication of syphilitic mesaortitis is aortic aneurysm, while the process localization in its abdominal part is atypical. Syphilitic aneurysms are most often located in the aortic root region and ascending aorta, then (as the frequency of occurrence decreases) in the arch of aorta, descending aorta, and abdominal region [1–5]. In the described case, the aneurysm of the abdominal aorta was located infrarenally, i.e., as low as possible.

Aortic aneurysms cause compression and impaired function of nearby organs of the respiratory and digestive systems. One of the serious complications ending, as a rule, fatally, is the aneurysm rupture with bleeding into the lung parenchyma or bronchi and trachea, as well as into the pleural cavity, mediastinum, esophagus and casuistically rarely into the duodenal lumen [6–9].

The presented case is unique not only by the fact of the aneurysm rupture and bleeding into the duodenum, but also by the formation of multiple, partially merging gummas in the aorta, small intestine, and retroperitoneal tissue with the formation of the aortoduodenal fistula through which blood periodically entered the duodenum. The wide dispersion of multiple specific changes in the internal organs and soft tissues is noteworthy.

Case history

A 85-year-old patient, female, was urgently hospitalized in the intensive care unit of a surgical hospital in the Moscow region with suspected coronary artery disease, unstable angina and complaints of weakness, dizziness, and black loose stools. From the anamnesis it is known that an episode of gastrointestinal bleeding, the cause of which has not been established, was registered a year ago. When examining data indicating acute cardiac pathology was not detected. Obviously, the clinical picture is due to recurrent gastrointestinal bleeding.

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The results of laboratory and instrumental methods of research (are abbreviated)

Hyperglycemia, glucose level 6.5-15.7 mmol/l with a tendency to decrease; a decrease in the level of total protein and albumin, respectively, from 50.51 to 33 g/l and from 23 to 16 g/l; periodic increase in the concentration of both total (up to 24.1 mmol/l) and direct (up to 9.3 g/l) bilirubin; ALT up to 70.6 U/l; increase in troponin level to 2.76 ng/ml.

Decrease in the Hb level from 81 to 62 g/l; leukocytosis (up to $17.3 \cdot 10^9$/l); increasing thrombocytopenia ($89\text{-}40 \cdot 10^9$/ l); ESR 60–67 mm/h.

Urine protein 0.2 g/l, leuocytes 30-40 in the field of view, erythrocytes 9-10 in the field of view, urine ketones 2 mmol/L.

A gradual decrease in prothrombin level from 73.4 to 45% (Quick's value) and an increase in prothrombin time from 13.7 to 19.5 s were observed.

Echocardiography: aortic compaction and valvular calcification with the formation of moderate stenosis. Diastolic dysfunction; myocardial contractility saved. Mitral regurgitation (grade 1), tricuspid regurgitation (grade 1).

Esophagogastroduodenoscopy:

1st study – the stomach mucous membrane is pale with confluent erosion ... a small amount of transparent liquid is in the lumen. The pylorus is free to pass ... The mucous membrane of the duodenal bulb is swollen, hyperemic with erosion, and freely expands with air. Postbulbar part is without pathology. There are no signs of bleeding.

2nd study (after a few hours) – below the Z-line ... an injected vessel is determined – a likely source of bleeding. Hemostasis was performe... A large amount of blood is in the stomach lumen. Incomplete inspection.

3rd study – an ulcer defect covered with fibrin is determined below the Z-line, with a diameter of up to 0.3 cm with an injected vessel and signs of chipping and electrocoagulation ... There is no evidence of a recurrence of bleeding.

The 4th study (in a day) – the esophagus ... contains blood with clots in the form of reflux ... The stomach contains a large amount of wash water, fresh blood and ... blood clots. Lesser curvature, body and partially pyloric section were examined. The mucous membrane of these areas is pale. In the prepiloric zone, erosions up to 3-4 mm in size are determined, from which blood leaks. Blood reflux from the duodenum. Hemostasis is without effect ... The pylorus mucous membrane is not visualized due to profuse reflux of blood from the duodenum. Its bulb is passable, the lumen is filled with blood clots; the mucous membrane is pale, erosion up to 2 mm in size, covered with fibrin, is determined. Postbulbar sections are not examined, since the lumen is filled with blood clots. Conclusion: signs of active gastrointestinal bleeding from an unknown source. Endoscopically bleeding is not stopped.

Colonoscopy: the apparatus is carried out to the upper third of the sigmoid colon, then a large number of blood clots is in the lumen. Inspection is impossible. In the examined area, the intestinal lumen is free, not deformed, freely spreads by air ... Conclusion: signs of intestinal bleeding from an unknown source ... Incomplete examination.

Bronchoscopy (2 studies): diffuse bilateral tracheobronchitis of the 1–2 degree of intensity of inflammation was found.

CT scan of the abdomen and pelvis: the abdominal aorta with a diameter of 20 mm at the level of the diaphragm and 16 mm at the level of bifurcation. Multiple atherosclerosis calcific plaques are in the wall. S-shaped tortuosity of the abdominal aorta. In the infrarenal segment, 50 mm below the right renal artery, diffuse expansion of the aortic lumen up to a maximum of 21 mm long before bifurcation. A contrasting protrusion of the finger-shaped lumen with a wide base is determined in the initial section of this extension along the right side wall of the aorta (50 mm below the right SA). Size 9×11 mm. The anterior and lateral walls of the aorta over a length of 35 mm are clutch-like thickened to 13–16 mm and cover a finger-like protrusion. Density 30–50 HU. In the root of the mesentery of the small intestine, a drop-shaped formation with a density of 50 HU, not amplified by contrast, directed by the narrow isthmus towards the aortic wall. The muft-like strata on the aortic wall are closely adjacent to the horizontal and ascending parts of the duodenum. Extravasation of radiopaque substances was not detected. The walls of this duodenum parts are thickened, the adjacent fiber is moderately striated ... Conclusion: condition after gastrointestinal bleeding. CT signs of aortic atherosclerosis with *dilatation of the infrarenal and abdominal aortic pseudoaneurysm (mycotic?)* adjacent to the horizontal and ascending parts of the duodenum; consequences of inflammatory changes? or old hematoma? in the aortic wall and paraaortic tissues. Thickening of the duodenum walls. The presence of an *aortoduodenal fistula* cannot be ruled out.

Surgery: laparotomy, gastroduodenotomy, stitching of erosion of the duodenum, Finney pyloroplasty.

Treatment in the intensive care unit included hemostatic, antiulcer and blood transfusion therapy. The course of the disease was complicated by hemorrhagic shock, which was the direct cause of death on the 5th day of hospital stay.

Final clinical diagnosis (abbreviated)

Underlying disease. Atherosclerotic aneurysm of the infrarenal segment of the abdominal aorta. Aortoduodenal fistula?

Complications of the underlying disease. Recurrent gastric and duodenal bleeding. Operations of endoscopic and abdominal hemostasis. Hemorrhagic shock. Pulmonary edema. Cerebral edema.

Comorbidities. Atherosclerosis of the aorta.

Postmortem examination results (only significant abnormal changes in abbreviated form are given)

In the cortex and white matter of the right temporal lobe, multiple point hemorrhages were detected on an area of 1.2×1.2 cm. In both pleural cavities and abdominal cavity, 300 ml of transudate and 300 ml of sacred contents were detected, respectively. Liquid dark blood with a volume of 50 ml is in the lumen of the duodenum, and 300 ml in the lumen of the jejunum and colon. The mucous membrane of the stomach body posterior wall is ulcerated in two places on the area of 0.6×0.6 and 0.7×0.7 cm. On the back wall of the horizontal part of the duodenum, there is a round defect in the wall with a diameter of 1.3 cm with smooth soft edges.

Intima of the aorta is yellow in color with the flat plaques of pale yellow color, in some places with calcification and surface destruction. The lumen of the abdominal part is somewhat widened (perimeter 7 cm with a norm of up to 6 cm) with aneurysmal protrusion in the infrarenal zone over an area of 4×2.5 cm, which is partially covered by thrombotic masses. When removal of parietal thrombi, sections of wall necrosis and a perforation hole with a diameter of 0.7 cm were determined (**Fig. 1**), communicating with the lumen of the duodenum (aortoduodenal fistula; **Fig. 2**). The lumen of the fistula is thrombosed, the walls are loose with necrosis patches of a gray-brown color, and in some places they are imbibed with blood. The aortic wall in the fistula area is thickened to 0.9 cm. Adjacent retroperitoneal tissue is with necrosis foci and areas imbibed with blood.

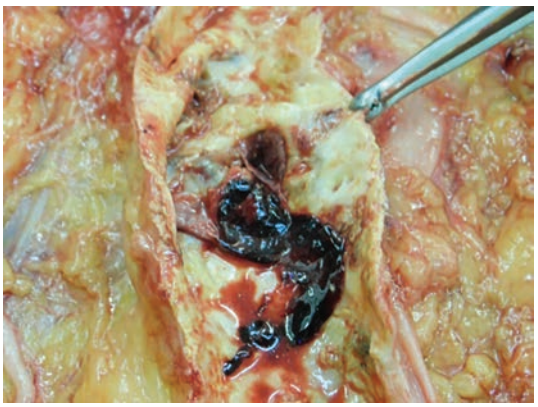


Fig. 1. **Organ specimen of the abdominal aorta.**
Distension of the aorta is minimal. Visualized aortoduodenal fistula, partially covered by thrombotic masses.

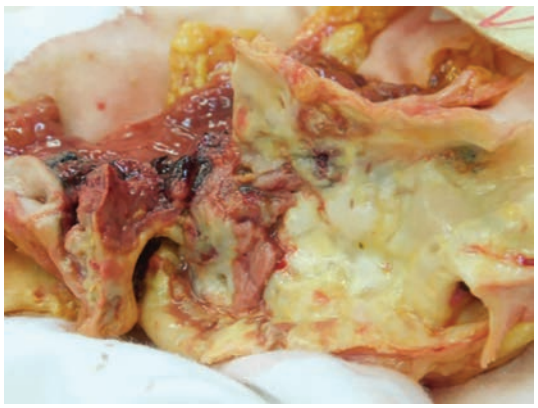


Fig. 2. **Organ specimen of the autopsy aortoduodenal fistula.**
In the foreground, an autopsy aorta with multifocal necrotic changes of intima, in the background on the left is the mucous membrane of the duodenum.

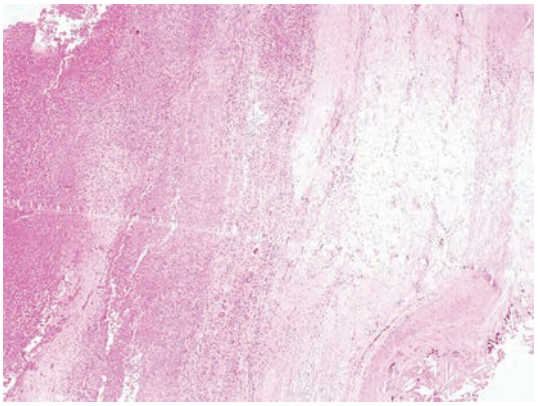


Fig. 3. **A fragment of a histological preparation of the aortic wall with signs of productive mesaortitis.**
In the center of the photo is an aortic wall with inflammatory infiltration; on the left is necrotic adventitia and paraaortic fiber; on the right is the aortic intima with minimally pronounced atherosclerotic changes. Hematoxylin-eosin stain. Zoom 40.

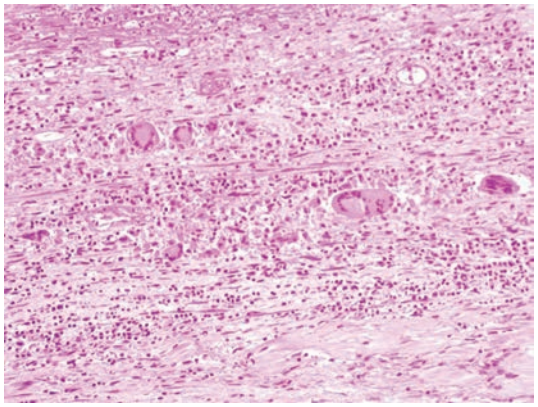


Fig. 4. **A fragment of a histological preparation of the aortic wall with signs of productive mesaortitis with a large zoom.**
Severe productive inflammation — intramural lymphoplasmacytic infiltration with giant multinucleated cells. Segmented neutrophils are not detected. Hematoxylin-eosin stain. Zoom 400.

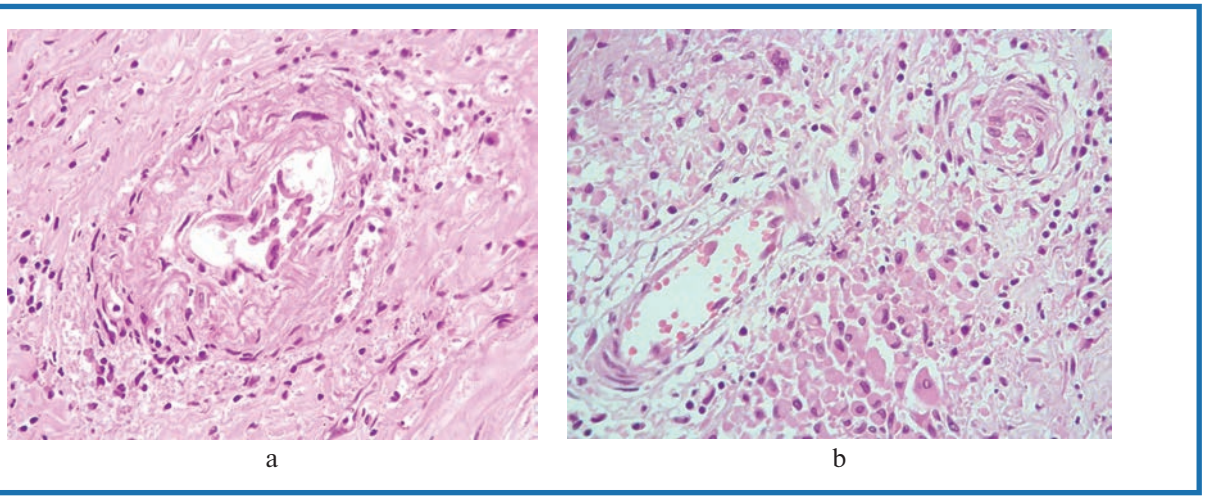


Fig. 5. **Fragments (a, b) of histological preparations of the aortic wall (represented by Vasa-vasorum).**
Productive vasculitis is determined. Hematoxylin-eosin stain. Zoom 400.

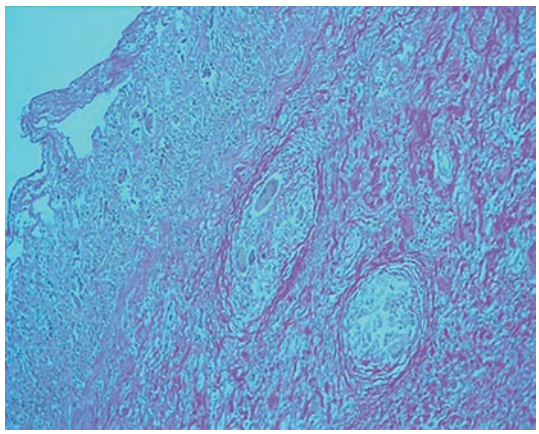


Fig. 6. **A fragment of a histological preparation of the wall of the duodenum.**
In the center, gumma and epithelioid cell granuloma with giant multinucleated cells. On the left mucous membrane. Staining for connective tissue according to Van Gieson. Zoom 100.

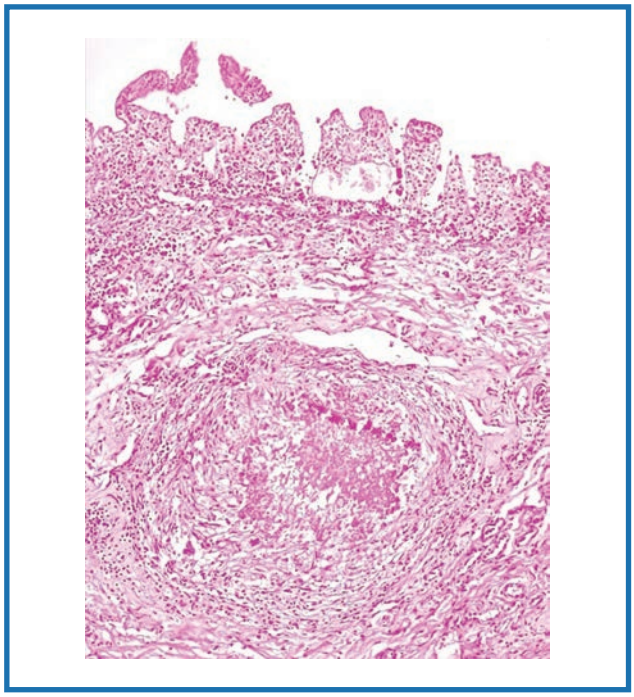


Fig. 7. **A fragment of a histological preparation of the wall of the duodenum.**
In the center is gumma. Hematoxylin-eosin stain. Zoom 100.

The endocardium is unevenly fibrosed, heart valves are sclerotic and calcified. The lungs are motley, heavy, densified, tissue pieces sank in formalin solution. The remaining organs and tissues are without macroscopically significant pathological changes.

It should be emphasized that suspicions regarding the syphilitic origin of the pathology arose on autopsy, since changes in the ascending and abdominal aorta did not fully correspond to the atherosclerotic genesis of the lesion, which is clinically suspected. In particular, the intima of the ascending aorta had a rough appearance ("shagreen skin") with lipid spots and plaques, and the expansion of the abdominal aorta and damage to its intima were insufficient for this pathology. In addition, the causes of a defect in the wall of the duodenum and necrotic changes in the retroperitoneal tissue remained unclear, and the mechanism of aortoduodenal fistula formation was not understood.

Answers were received at the microscopic stage of the pathological study. Late visceral syphilis with a primary

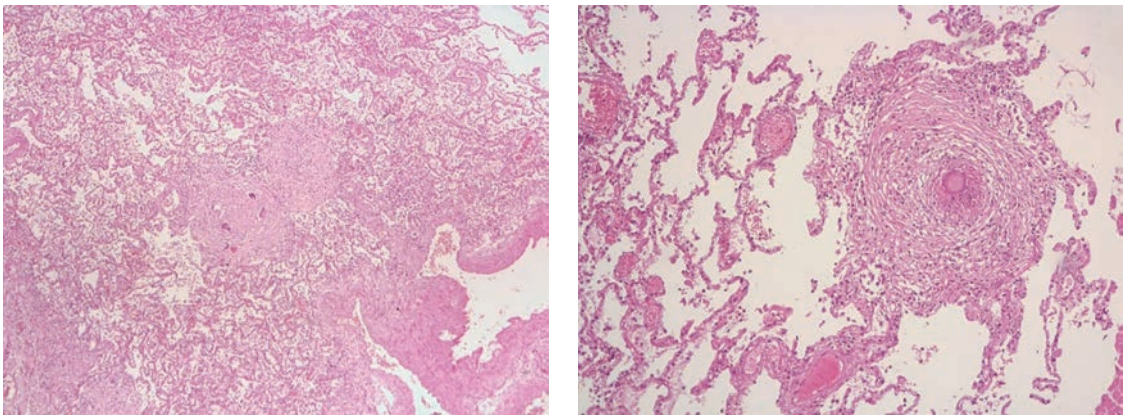


Fig. 8. **Fragments of histological preparations of lung tissue.**
Productive inflammation. Epithelioid cell granulomas with giant multinucleated cells are visualized. Hematoxylin-eosin stain. a — zoom 100, b — zoom 200.

lesion of the aorta, duodenum, lungs and liver was diagnosed histologically and bacterioscopically.

Destructive syphilitic changes were noted only in the aortic wall (**Fig. 3–5**) and the duodenum (**Fig. 6, 7**), as well as in the adjacent retroperitoneal adipose tissue. Merging gummas with extensive destructive changes and extensive zones of tissue necrosis without purulent inflammation were found. Moreover, the signs of productive mesaortitis in the areas adjacent to the fistula were maximal. Thus, a pathomorphological substrate for the aortoduodenal fistula formation was revealed.

Productive and granulomatous inflammation is noted in other organs and tissues. Multiple confluent epithelioid cell or sclerotic granulomas are found in the lungs and liver (**Fig. 8, 9**). At the same time, minimal productive inflammation was detected in the brain and heart (productive meningitis and vasculitis) (**Fig. 10, 11**).

To confirm the syphilitic origin of the disease, an additional histochemical study of tissues using specific Levaditi stain was performed. Isolated positively stained microorganisms with the structure of Treponema pallidum were identified in the aortic adventitia (**Fig. 12**).

Thus, according to the results of macro- and microscopic examination, a pathological diagnosis is formulated (given in abbreviated form).

Underlying disease. Late visceral syphilis: productive granulomatous inflammation, solitary and merging gummas in the wall of the aorta and small intestine, retroperitoneal adipose tissue, liver, lungs, heart and pia mater.

Aneurysmal expansion of the infrarenal section of the abdominal aorta with parietal thrombosis.

Background disease. Essential hypertension. Hyperglycemia.

Complications of the underlying and comorbid diseases. Destruction of gummas in the area of the infrarenal segment of the abdominal aorta, in the adjacent retroperitoneal adipose tissue and the wall of the horizontal part of the duodenum with the aortoduodenal fistula formation and bleeding. Hemorrhagic shock. Diffuse alveolar damage. Cerebral edema.

Comorbidities. Atherosclerosis of the aorta.

Discussion

Comparing the final clinical and pathological diagnoses, we are convinced of their divergence. During the patient's life, the diagnosis of syphilis was not verified, since a serological study was not performed. Also, the true source of fatal intestinal bleeding has not been established.

As you know, a specific treponemal antigen test is included in the list of compulsory studies for all hospitalized. The probable reasons for its failure could be, firstly, the venerable age of the patient, secondly, the need for urgent medical measures, and thirdly, the possible connection of intestinal bleeding with syphilis seemed extremely unlikely.

It is well known that syphilis is a "great mimic" that can "conduct" any clinical symptoms. In addition, Treponema pallidum, having infiltrated the human body, is able to maintain viability in at least some patients, even in cases of specific treatment. It is stored in the tissues and fluids that are the most inaccessible for medical preparations (in the brain and spinal cord, cerebrospinal fluid) in survival forms (cysts, L-forms), which are capable of reversing into pathogenic ones.

Would reconstructive surgery on the aorta be indicated in case of treponemal antigen test? Reconstructive surgery on the aorta is known to be a serious surgical procedure that would be performed in old age. Obviously, the likelihood of a fatal outcome in this situation would also

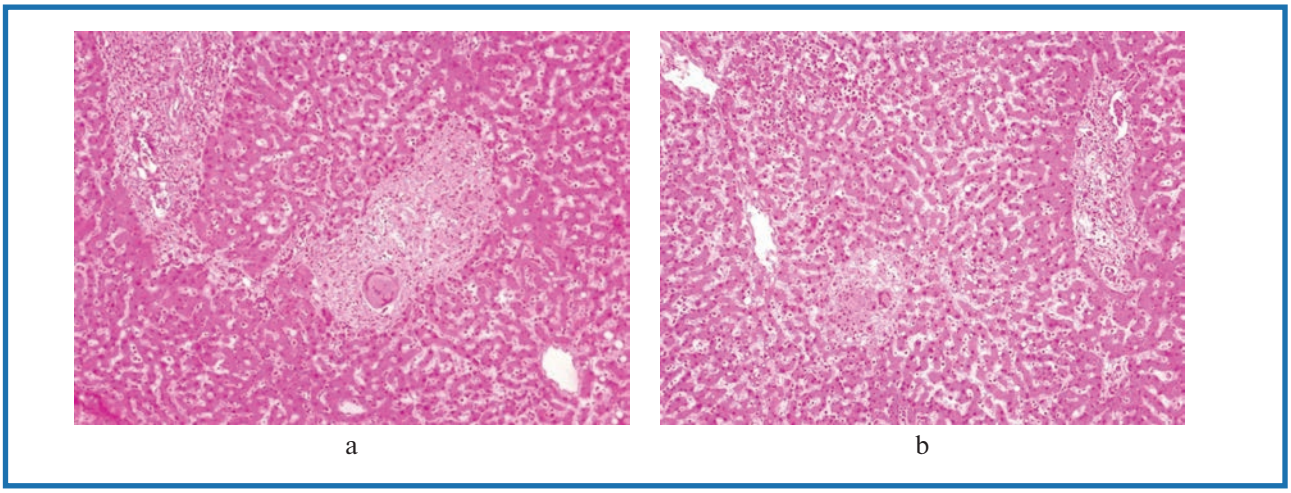


Рис. 9. **Fragments (a, b) of histological preparations of liver tissue.**
Productive inflammation. Epithelioid cell granulomas with giant multinucleated cells are visualized. Hematoxylin-eosin stain. Zoom 100.

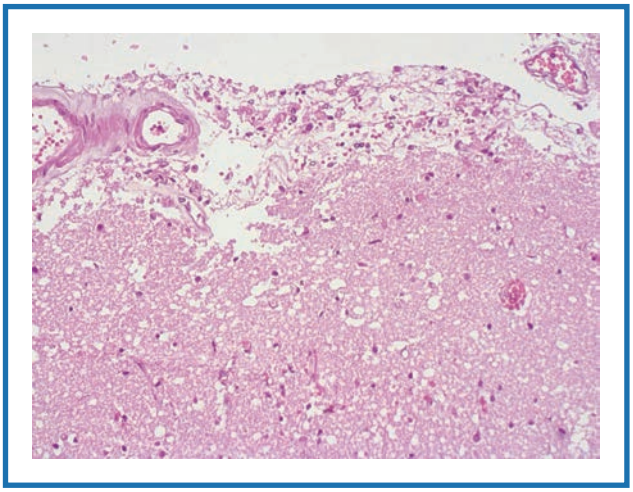


Fig. 10. **Fragments of a histological preparation of brain tissue with pia mater.**
Productive meningitis. Lymphoplasmohistiocytic infiltration of the pia mater and swelling of the substance of the brain are determined. Hematoxylin-eosin stain. Zoom 100.

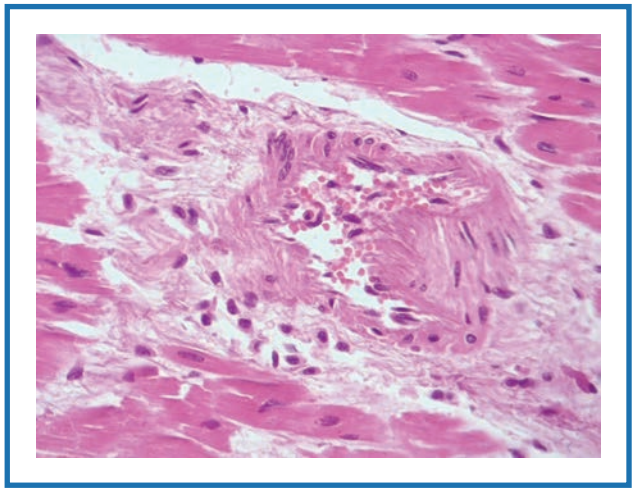


Fig. 11. **A fragment of the histological preparation of myocardial tissue.**
Productive vasculitis in the myocardium. Plasma-histiocytic infiltration is visualized. Hematoxylin-eosin stain. Zoom 400.

be extremely high, but the direction of clinical thinking would be correct and there would still be a certain chance for the patient to survive.

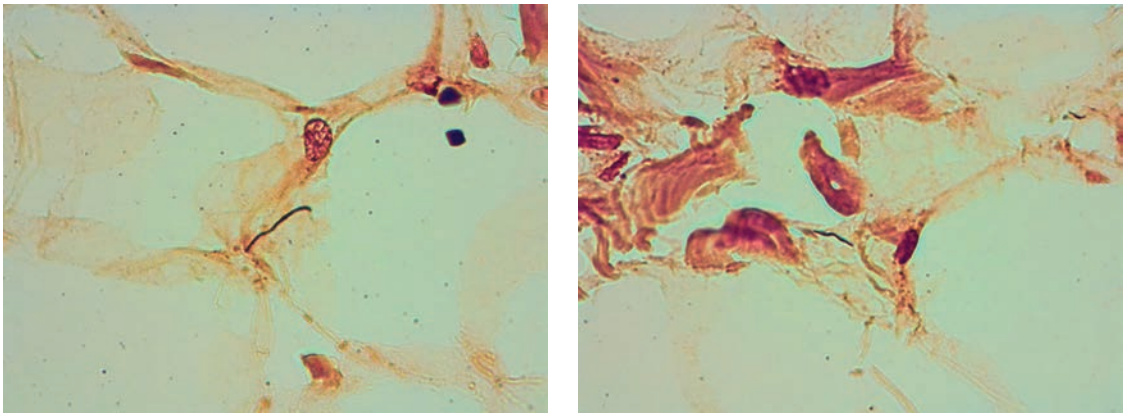
If we evaluate the contribution of specialists to the struggle for the patient's life, we should first of all single out the surgeons who performed repeated endoscopic and abdominal operations aimed at finding the source of bleeding – and they were farthest from the true cause of the disease. A CT scan, interpreted as a "mycotic" aortic aneurysm, the assumption of an aortoduodenal fistula, is a serious advance to the truth of the doctor of functional diagnostics after a single study.

The presented observation is instructive not only for doctors of functional diagnostics and surgical specialties, but also for dermatovenerologists, who, as a rule, consider visceral syphilis to be characteristic of middle-aged men [10]. At the same time, publications have recently appeared on observations of late visceral syphilis in women, including those with a fatal outcome [11].

Thus, the authors of the article remind doctors of all specialties about the need for a mandatory serological examination for syphilis. There will be less tragic deaths!

Conclusions

1. Syphilis is an interdisciplinary problem, and doctors of all specialties should be aware of its possible participation in a variety of clinical pathologies. This is espe-



а/a б/b

Рис. 12. **Fragments (a, b) of histological preparations of adventitia tissue of the abdominal aorta.**
A pale treponema is visualized in the center. Levaditi's stain. Zoom 1000.

cially true of late forms of the disease, which are currently much more common than early ones, and which are much more difficult to diagnose.

2. Observing the mandatory serological examination of all inpatients, it is possible to accidentally identify syphilis not as the cause of the underlying pathology, but as a concomitant disease. This will allow the patient to complete treatment and avoid serious complications in the future.

The authors declare no conflict of interest.

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