

Case No. 10

Female - 62 years old

Long-term medical history - Treated and controlled diabetes

Recent medical history

For nearly four months complaints of a dull pain in the right hypochondrium, fever of up to 38° C. and weight loss (approximately 10 kg in recent weeks). Medical therapy induced no effects.

Physical Examination

Febrile state without any other particular features. Abdomen distended, no pain on palpation, hypochondriac organs normal, no palpable masses.

Diagnostic Workup

WBC 13,700, of which 67% were neutrophil granulocytes, 23% lymphocytes, 2.6% eosinophils and 7% monocytes. Glycemia within normal limits. Blood tests for HBV, HCV, HIV and hydatidosis were negative. A slight increase was present in alkaline phosphatase (435 U/l) and γ GT (83 U/l), as well as in globulin compared to normal albumin levels; CEA, CA19-9 and α -fetoprotein levels were all normal.

Abdominal ultrasonography (US) revealed a hypoechoogenic, irregular mass approximately 6 cm. in diameter, without a peripheral capsule, in the right lobe of the liver (Fig. 1).



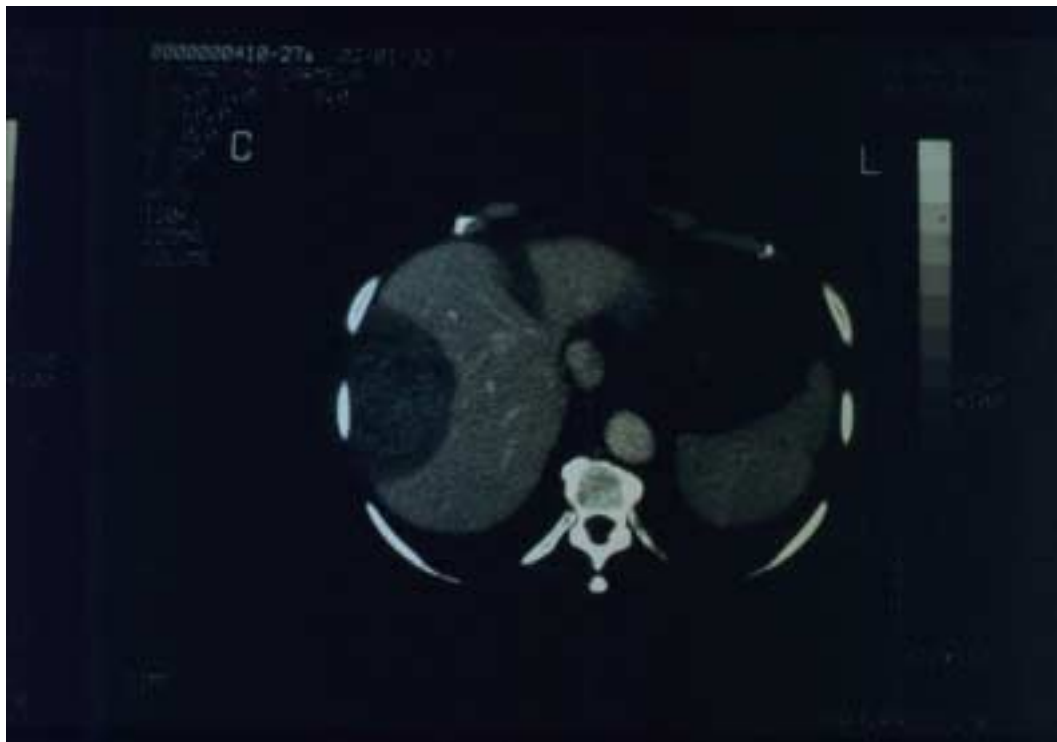
Fig. 1 - see text

Chest X-ray and esophago-gastroscopy were negative.

Computerized tomography (CT) confirmed the presence of a rounded, high-density area with poorly defined margins in the subcapsular area of the VIII hepatic segment. After IV contrast medium administration the lesion showed weak and heterogeneous enhancement, with some central more rarified areas and without peripheral enhancement (Fig. 2).

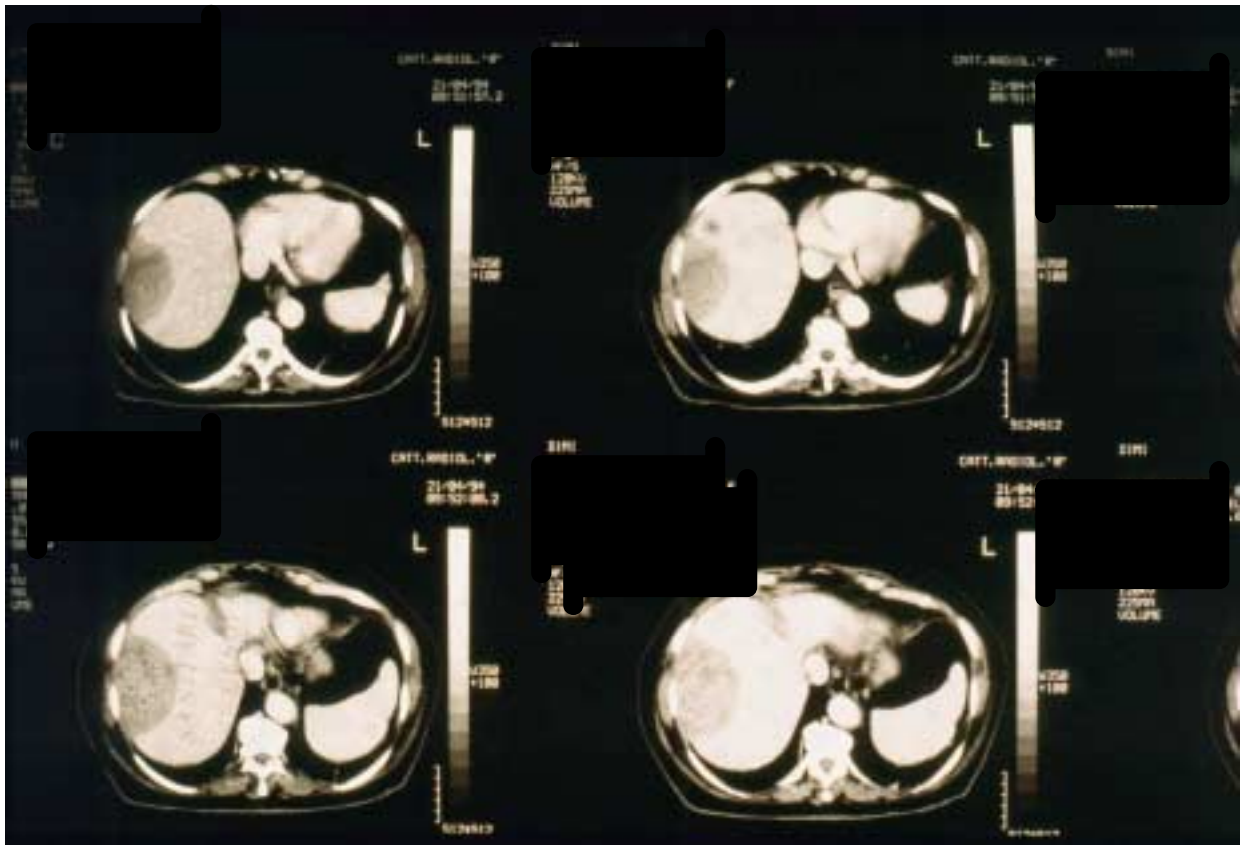


a



b

Fig. 2 - see text



c

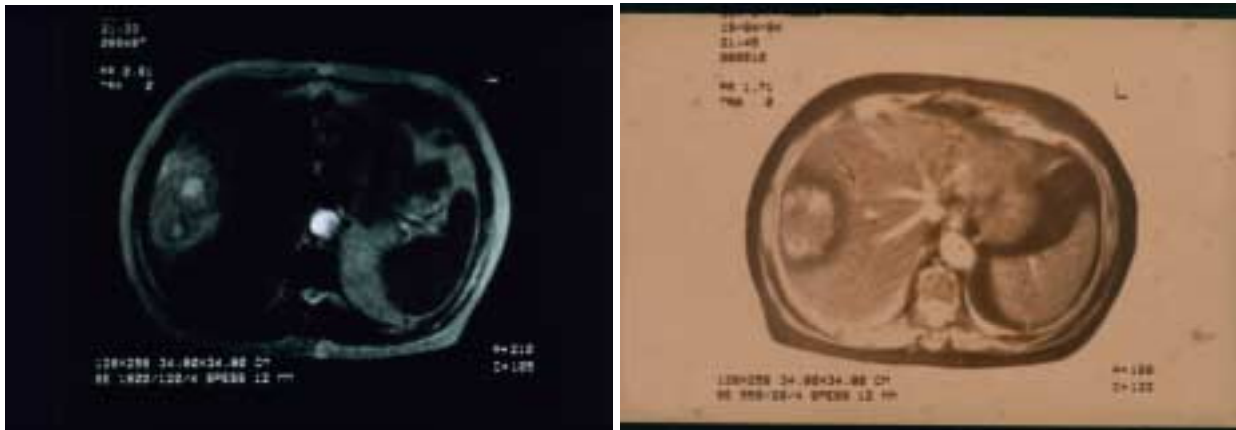
Fig. 2 - see text

Magnetic resonance imaging (MRI), with T₁-weighted (TR/TE 550/20) and T₂-weighted (TR/TE 1800/120) spin-echo sequence, followed by a T₁-weighted sequence after IV Gd-DTPA administration, was performed.

Imaging revealed weak T₁ intensity, intense T₂-weighted enhancement and central areas of hyperintensity corresponding to those seen with CT with contrast medium. A thick peripheral area around the hypodense central mass was observed after Gd-DTPA administration (Fig. 3).



Fig. 3, a - see text



b

c

Fig. 3 - see text

Cytological examination performed by US-guided fine needle aspiration (FNA) yielded no useful diagnostic elements.

Because the battery of the above clinical and instrumental examinations could not afford a reliable diagnosis, and given the expansive and seemingly neoplastic features of the lesion, liver resection was decided.

Surgical Procedure

Right, S-shaped, subcostal laparotomy prolonged to the left of the median line. The right liver presents a tumor-like mass approximately 6 cm in diameter, pinkish-yellow in color, umbilicated at the center, rather embedded in the surrounding hepatic parenchyma, from which it is separated by a circumferential furrow (Fig.4). The remaining hepatic areas appear normal, and perioperative US confirms the already known features of the lesion and the absence of other signs of disease. In view of the lesion's size and tumor-like appearance, as well as of the fact that a definitive diagnosis could not be established, a regulated right hepatectomy was decided.

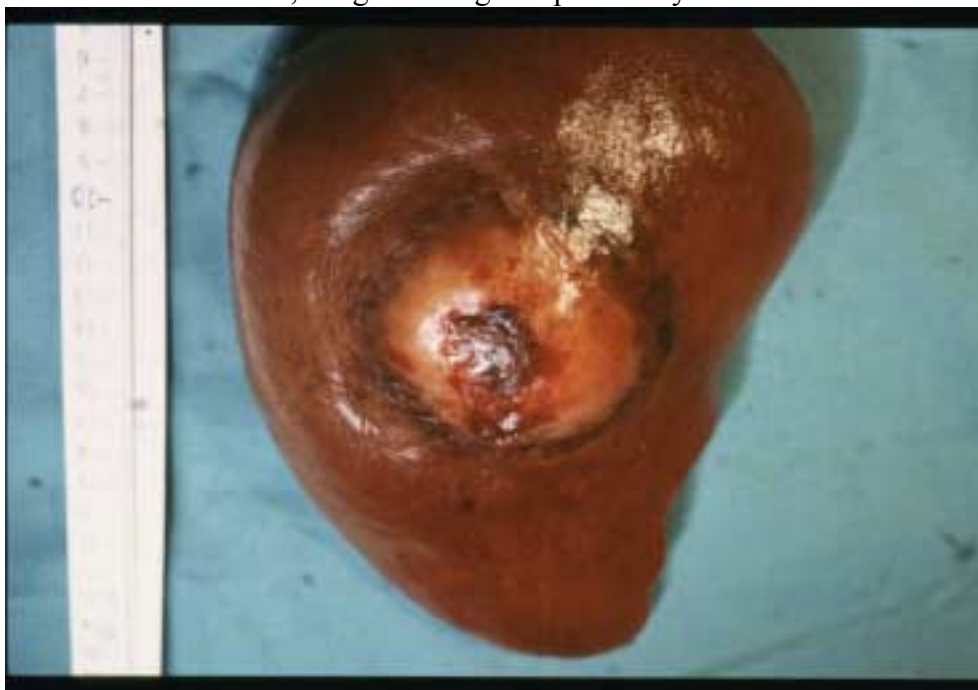


Fig. 4 - see text

Pathology

Macroscopic - the features described above are confirmed: the mass is circumscribed, dense and a maximum of 6 cm in diameter. On sectioning it is compact, grayish-yellow in color, with hemorrhagic and necrotic areas. A peritumoral capsule is not visible. The adjacent liver appears normal (Fig. 5).



Fig. 5 - see text

Microscopic - specimens prepared with hematoxylin-eosin, periodic acid-Schiff, Ziel-Nielsen and Grocott staining. The lesion contains a large area of dense, fibrous tissue, with spiraling laminated features, and inflammatory infiltrates made up of plasma cells, lymphocytes, macrophages, polymorphonucleated leukocytes and foamy cytoplasmic histiocytes. Numerous spindle-shaped or oval fibroblasts are visible. Extensive necrotic areas are also present. The formation's margins are indistinct, and an incomplete fibrous capsule traps inflamed biliary ducts. Foci of obliterating phlebitis are evident. The bile ducts in the surrounding hepatic parenchyma also display signs of inflamed walls. Anomalous mitoses are not visible. The whole mass is finely vascularized. Staining for microorganisms is negative.

Diagnosis: inflammatory pseudotumor of the liver.

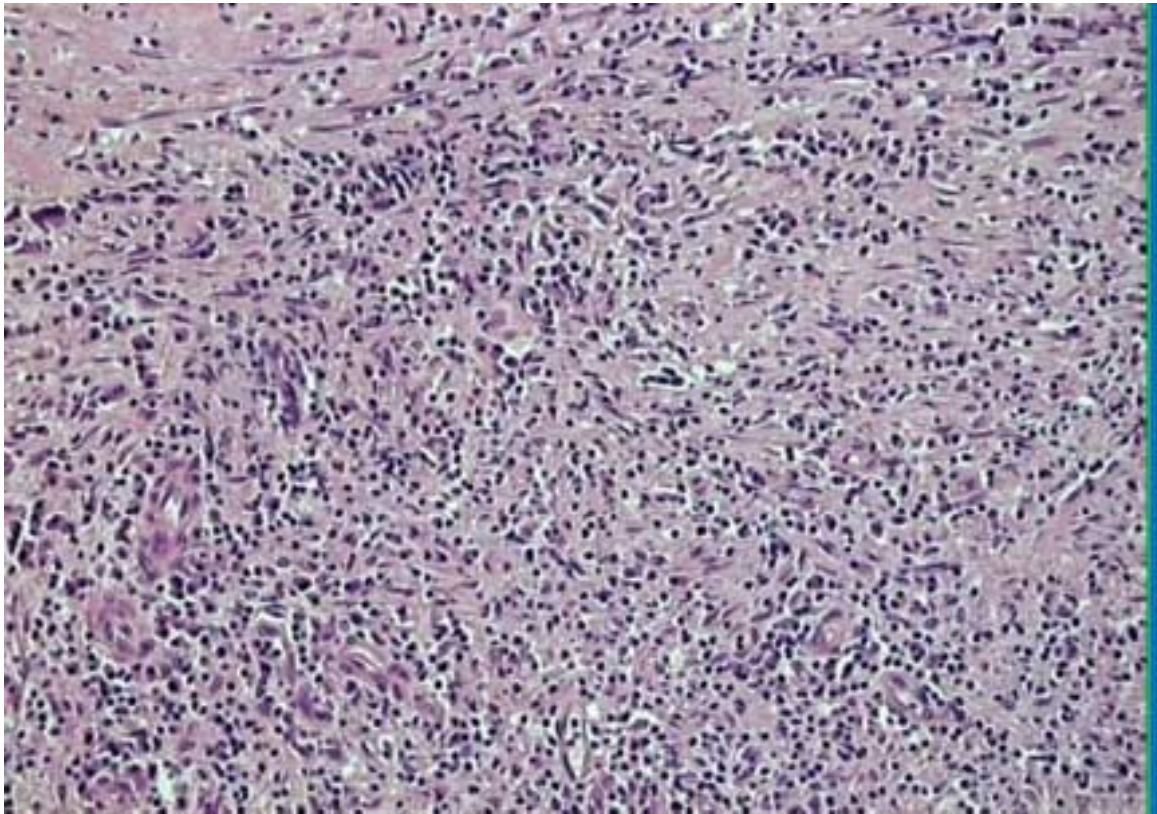


Fig. 6 - Dense fibrous tissue with an inflammatory infiltrate made up of various types of cells.

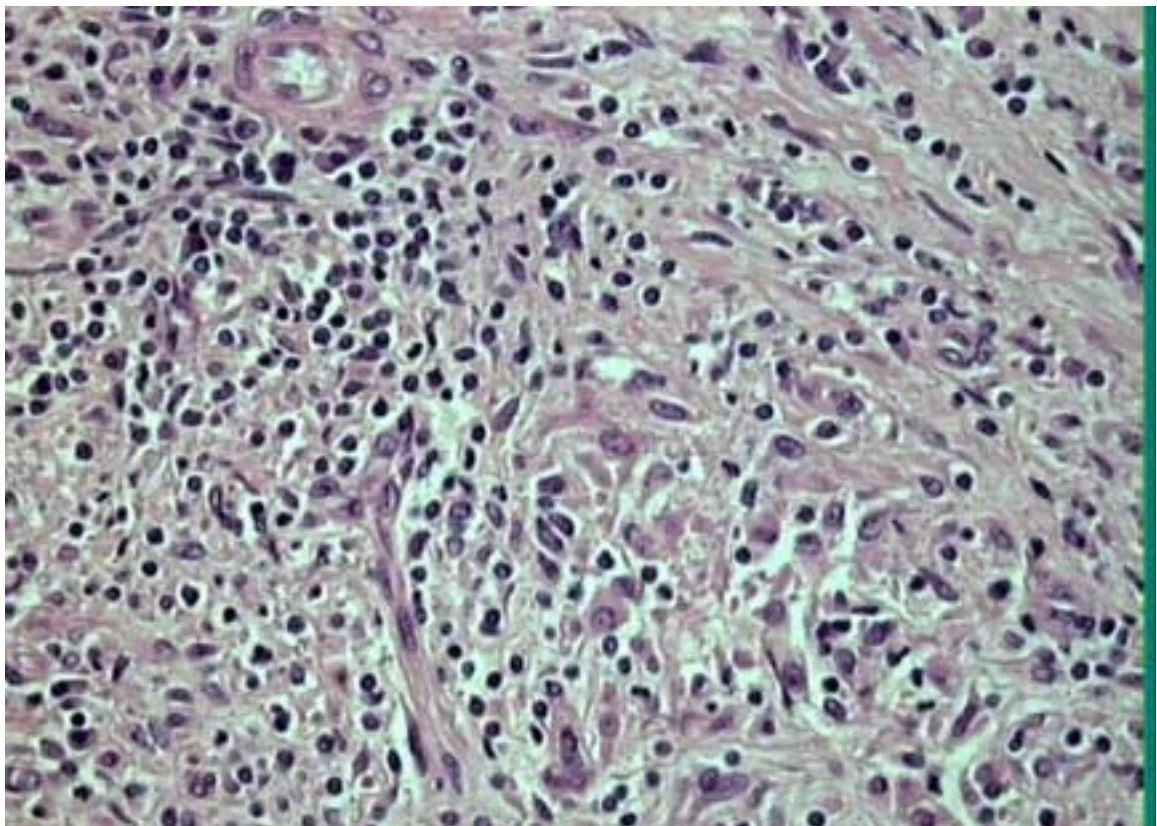


Fig. 7 - Inflammatory infiltrate mostly composed of lymphocytes and plasma cells, with activated fibroblasts and myofibroblasts.

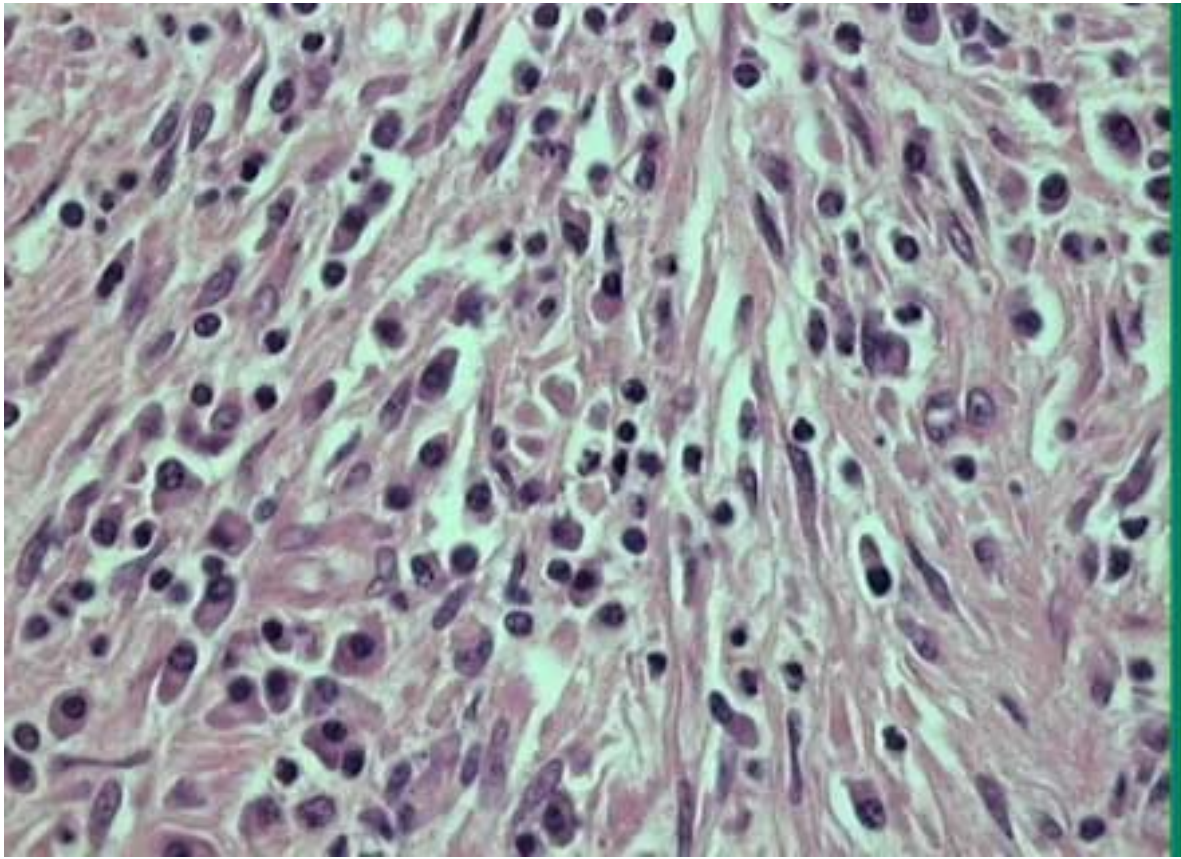


Fig. 8 - Lymphocytes, plasma cells, activated fibroblasts and myofibroblasts

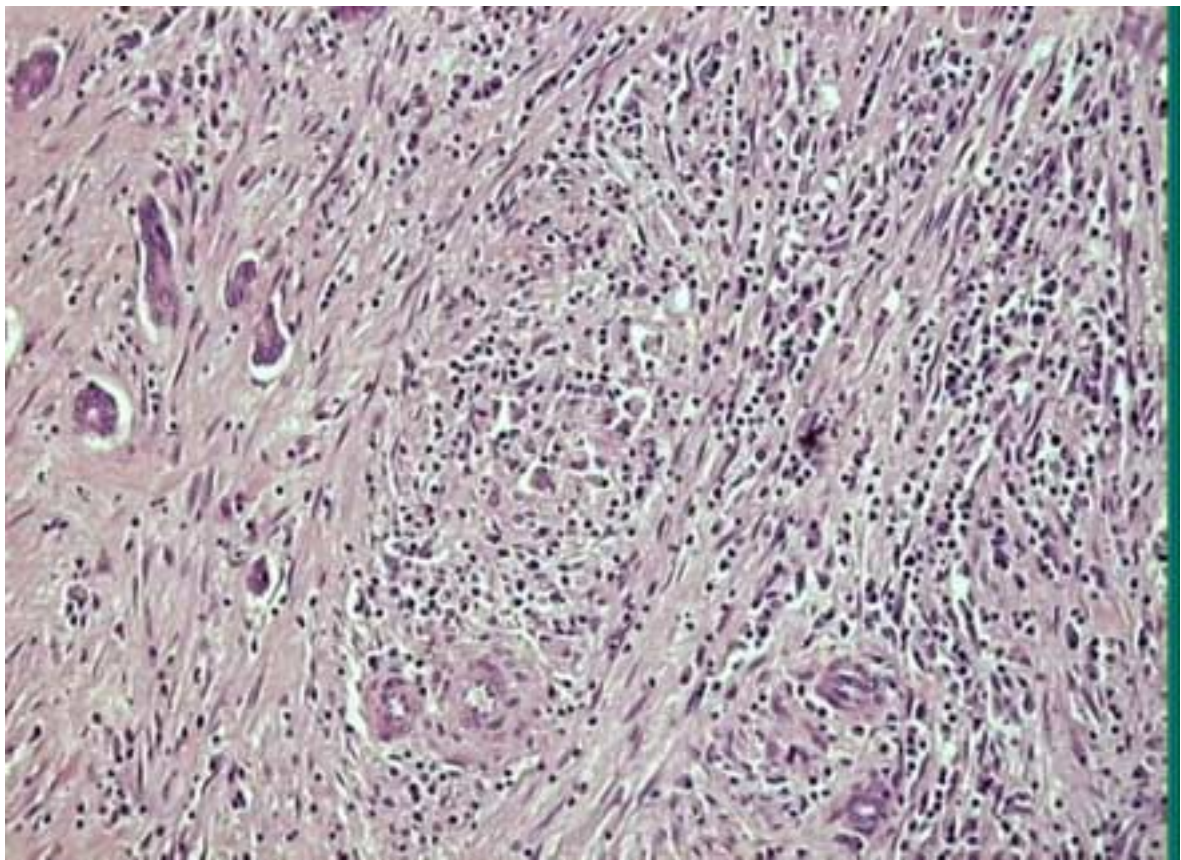
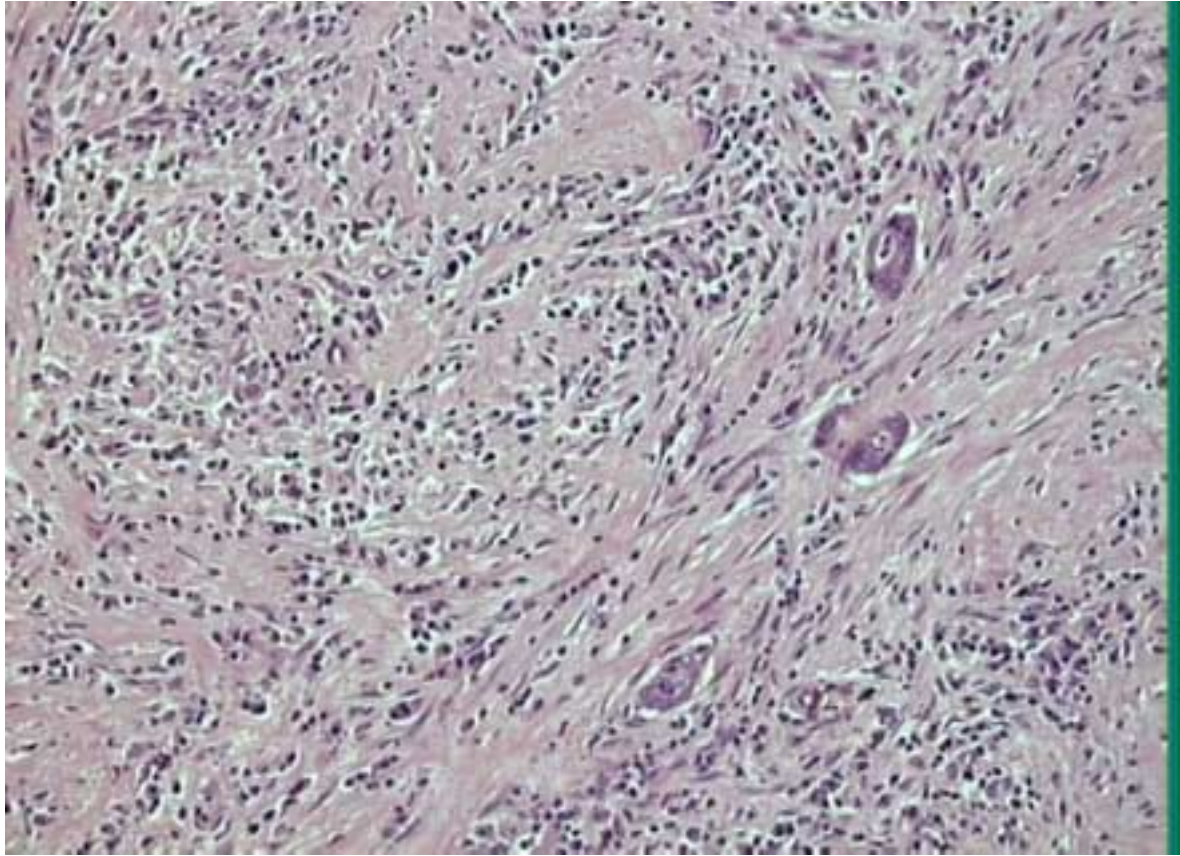
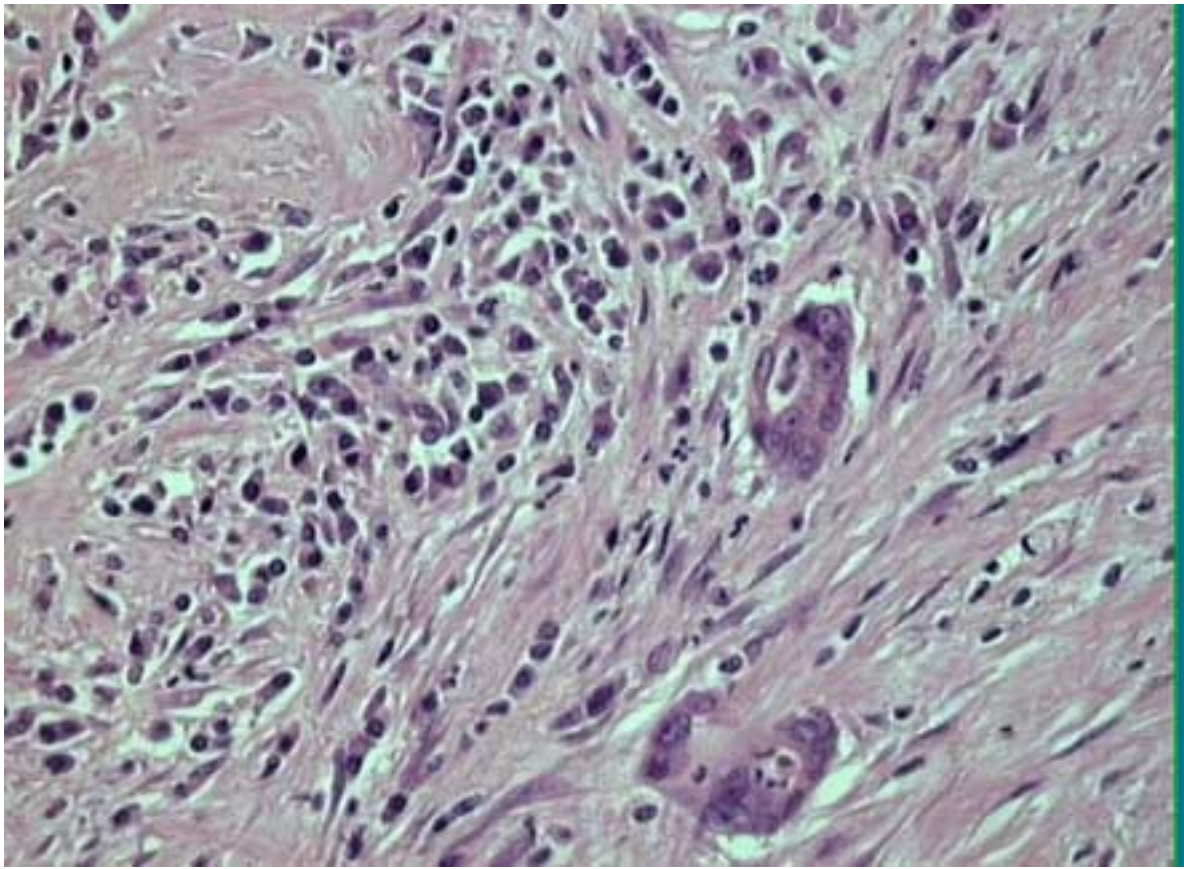


Fig. 9 - Residual bile ducts and vessels with pattern of obliterating vasculitis.



a



b

Fig. 10 a, b - Phlogistic and fibrous elements trap distorted biliary ducts.

The postoperative period is regular; long-term follow up confirms definitive healing and optimal hepatic regeneration.

Remarks

Here we have the case of diabetic woman with well-controlled glycemia, who for some months had complained of moderate pain in the right hypochondrium, weight loss and fever. Blood tests revealed a slight neutrophil leukocytosis, and physical examination, particularly of the abdomen, was negative.

At this stage, postulating any diagnosis is difficult to say the least, so vast is the array of lesions that could account for such a clinical picture. Not even the laboratory findings are helpful.

Only diagnostic imaging studies are able to reveal the putative cause of symptoms; even these, however, do not reliably identify their nature. If made only on statistical grounds, the first diagnostic hypothesis is of an expansive neoplastic lesion, namely hepatocarcinoma. Not all of the features, however, not even the results of FNA cytology, reflect or confirm this conclusion. Only histological examination affords the true diagnosis as inflammatory pseudotumor of the liver.

At this point we can raise a few questions:

- Was a precise preoperative diagnosis possible? Inflammatory pseudotumor is a rare condition, and the literature reports only few cases. For this reason, imaging and cytology studies (these latter hampered by inadequate sampling due to extensive necrotic areas in a neoplastic context) may likely not be able to demonstrate the typical pattern of such liver masses, granted that their features can be classified.
- Was a right hepatectomy in the case in question the correct procedure? Together with our pathologists, we believed that, given the lesion's features, perioperative findings would not have yielded a reliable diagnosis. On the other hand, because of the lesion's size, if malignancy had been detected, biopsy or even partial treatment would have induced a risk of seeding. In a patient without any particular risk (elderly age, concomitant diseases, etc.) right hepatectomy constitutes quick and sure intervention. As such, this seemed to be the preferable alternative.
- In cases in which diagnosis of inflammatory pseudotumor of the liver, i.e., a benign lesion, is certain, is medical therapy possible? The answer is to be sought in the rare observations reported in the literature. Here, there is consensus with regard to the case presented here: because the preoperative differential diagnosis is not only difficult, but in most instances impossible, resection surgery is - by current standards - the ideal therapeutic means both to ascertain diagnosis and to resolve the condition. Moreover, the same literature reports a number of cases not treated surgically with a poor outcome, thus lending credibility to a possible unfavorable evolution.

Note: for further reading on the issue see: "Pseudotumor of the Liver: A Challenging Diagnosis". Borgonovo G., Razzetta F., Varaldo E., Cittadini G., Ceppa P., Torre G. C., Mattioli FP. Hepato-Gastroenterology, 45, Sept. - Oct. 1998.