Which Is The More Advisable Treatment For Recurrent Inflammatory Pseudotumour Of The Liver? A Case Report.
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Citation

Abstract
In this report the authors describe the case of a recurrency of a liver pseudotumor previously described from the same authors, characterised by an hepatic mass in a woman aged 62. The infiltrative pattern of the mass at the US scan suggested a malignant hepatic neoplasm. The histologic examination of the needle biopsy of the liver suggested the diagnosis of an inflammatory lesion of the liver having the features of an inflammatory pseudotumour. The diagnostic and therapeutic implications of this rare pathology are here considered.

INTRODUCTION
As previously described in a prior paper by the same authors, the inflammatory pseudotumor of the liver is a heterogeneous group of disease, a complex of lesions of unknown aetiology and pathogenesis which occupy space and mimic a neoplasm. This quite rare lesion (lesser few than 140 cases reported in the world literature) can frequently arise in subjects affected by systemic diseases such as rheumatoid arthritis, or in subjects who use drugs.

Asia is the geographical center for this pathology, with prevalence for male gender. Usually this condition onset with dyspeptic symptoms and laboratory findings of increased ESR, increased CRP, leucocitosys, slightly elevated activities of the aminotransferase, increased bilirubin and variable alterations of the electrophoretic pattern.

The inflammatory pseudotumor shows nodular lesions characterised by wide sclero-hyaline bands, sometimes in vorticoid structures, and an extremely variable inflammatory infiltrate.

Sometimes the histologic differential diagnosis may be quite difficult especially in discriminating pathologies such as chronic granulomatous hepatitis, sarcoidosis and tuberculosis affecting the liver.

Although rare, recurrences of this pathology are reported. These recurrences may either occur in operated patients or in patients treated by drugs administration.

CASE REPORT
A 62 years old woman was admitted to the General Surgery Department of Policlinico of Bari because of well defined abdominal pains in the right hypocondrium, digestive disorders and moderate elevation of the body temperature (37.4 - 37.6 C°) during the night.

The past medical history showed a serious form of long standing scoliosis, and allergic asthma lasting 15 years. She was also treated with specific drugs for rheumatoid arthritis for three years. Ten years before being hospitalised, she underwent to hysteroadnexectomy, because of an acute metrorrhagia due to uterine leiomyoma, six years later she underwent cholecystectomy for cholelithiasis. Two years ago she underwent to atypical bisegmentectomy of the 5th and 6th hepatic segments because an inflammatory pseudotumor of the liver.

The patient underwent laboratory tests, which revealed increased ESR (1st h =23) and increased hepatic function activities (ALT=63 U/l; AST=100 U/l; -GT=87 U/l; tot. Bil.=1.1 mg/dl). An ultrasound scan of the upper abdomen showed an increase of hepatic volume and the presence of an echographically heterogeneous area measuring about 2.5 cm in diameter in the right lobe of the liver. Colonoscopy and skin tests for tuberculosis were negative.

By using contrast, an abdominal CT scan confirmed the presence, in the residual right liver, of a heterogeneous hypodense expansive mass with a diameter of 4 cm, the dilatation of the extra-hepatic biliary tract, the enlargement
of the pancreas head, and a “minus” image (reduction of
lumen diameter) of the second segment of the duodenum. In
addition, several intra-hepatic lesion, very little in diameter,
were observed. The kidneys, the spleen and the other
intrapertoneal organs appeared normal. The enlargement of
the pancreas had an inflammatory origin, probably.

After hospital admission she underwent other clinical tests
to evaluate some viral and bacterial markers (HBV, HCV,
CMV, HSV1-2, S. tiphy, etc.) and the tumoral markers of
the digestive tract (-FP, CEA, TPA, CA19.9), which proved
to be negative, excepted S. tiphy.

Under US guidance was performed a needle biopsy of the
main lesion. Hepatic tissue and liquefactive material was
obtained.

The samples, fixed in 10% buffered neutral formalin, were
paraffin embedded and the sections were stained for H/E and
Alcian blue/PAS at pH 2.5.

Histologically focal areas of colliquative necrosis were
detected, which accounted for the main lesion observed at
US and CT scans. Little foci of chronic inflammation with
fibrous bands, giant and granulomatous epithelioid cells
were found in the surrounding normal tissue. The
inflammatory cells were mainly lymphocytes, plasma cells
and foamy histiocytes.

However, a lymphoid inflammatory infiltrate of the portal
tract and around the bile ducts was observed.

This pattern and the past medical history of the patient are
consistent with the hypothesis of a recurrent inflammatory
pseudotumor of the liver.

High doses of co-trimoxazole (Bactrim™, 20 mg/kg, per os
every 6 hours) were administered for 7 days, until dosage
was reduced to 600 mg/qd, per os. The body temperature
became normal after 3 days.

After 20 days the patient was discharged from hospital and
the treatment was again continued for 25 days.

During the treatment were performed four US scans at the
inpatient, and three at the outpatient. Two US scans were
performed two months and five months after the end of the
treatment.

The imaging study by US, during and after the treatment,
demonstrated a slow and continuous regression of the lesions
of the liver. These lesions have completely disappeared after
three months from the onset of the therapy

DISCUSSION

The inflammatory pseudotumor of the liver is a condition of
great interest, because it maybe easily confused with other
pathologies such as granulomatous hepatitis, sarcoidosis,
hepatic tuberculosis or hepatocarcinoma.

Though the inflammatory pseudotumor onset as clinically
silent hepatitis, it could insidiously mimic a malignant
neoplasm (sudden and progressive weight loss, elevated
activities of the aminotransferase, increase of CRP, of ESR
and leucocitosis) that, if confirmed by the radiodiagnostic
exams, may lead to an excessively aggressive therapeutic
treatment. Otherwise, the negativity of tumoral markers can lead to the
diagnosis of a non-neoplastic condition, which may have
relief - in any case - from the surgical removal of the mass
that acts as a further irritative stimulus for the surrounding
parenchyma, and can cause an extended fibrosis of the
organ, which determine a progressive hepatic failure and the
death of the patient. It represents therefore a pathology
which is quite difficult to describe clinically, whose nature
can be exclusively revealed by histologic study after mass
removal. Liver biopsy could be useful for diagnosis in
compliant patients, avoiding major surgery.

According to some studies this pathology is due to
bacteria from foods or chronic appendicitis or cholecystitis
whose would seed in the hepatic parenchyma through the
bloodstream of the portal vein, subsequently eliciting
inflammatory lesions with obliterating phlebitis and
granulomatous inflammation. Some Authors support the
hypothesis of an EBV infection.

Optimum management of this disease has not yet been
standardized. The majority of patients are treated by hepatic
resection, although spontaneous regression has also been
described.

According to the world literature, the best therapeutic option
was been liver resection in 93 cases, medical therapy in 16
cases and a “watchful waiting” in the other cases. In fact, in
patients whose underwent to surgical liver resection were
reported 6 deaths after 72 hours and 17 after 60 months from
the surgical intervention. The patients of the other two
groups had high rate of mortality (totally 8 deaths due to
hepatic failure) and morbidity (persistence and/or progression of the disease).

The reported case, considering that at follow up four years after surgery the patient is healthy, confirms the utility and necessity of surgery in individuals presenting large-sized lesions (more than 5 cm in diameter) and suggests the usefulness of antibiotic therapy in patients with smaller lesions (less than 5 cm in diameter) and histology confirmation of the diagnosis.

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