Primary necrosis of the round ligament in adults: A new case and literature review

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Abstract

Introduction: The necrosis of the round ligament is an exceptional entity. The clinical presentation is nonspecific and can lead us to call to mind other diseases. The diagnosis is suspected by an abdominal ultrasound and CT scan showing lesions between the liver and the abdominal wall with fat density that extends to the umbilicus. It is confirmed by the surgical exploration of the abdomen performed by classical or laparoscopic approach. Case Report: We are reporting the 16th case of this entity which presents a 56-year-old women who presented with right upper quadrant pain. The diagnosis of round ligament necrosis was suspected on abdominal ultrasound and the CT scan findings. It was then confirmed by laparoscopy which showed a gangrene of the round ligament. Conclusion: Primary necrosis of the round ligament of the liver is an extremely rare cause of acute abdominal pain. The diagnosis is tricky in spite of the contribution of the abdominal CT scan. The treatment is surgical.

Keywords: Gangrene, Laparoscopy, Necrosis, Round ligament

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Introduction

The round ligament is a fibrous remnant of the fetal umbilical vein. It stretches from the left part of the liver to the anterior abdominal wall. In adults, it can exceptionally form a gangrene and present with an acute abdomen. The underlying pathology is a primary ischemic necrosis of the connective tissue causing an inflammatory response in the peritoneal cavity. The subsequent translocation of enteric bacteria through the intestinal wall gives rise to diffuse infectious peritonitis. No triggering factor has been identified. Clinically, patients present with nonspecific peritoneal irritation signs. The diagnostic challenge has been overcome thanks to the contribution of the CT scan which demonstrates a necrotic mass between the liver and the abdominal wall.

In this article, we have compiled 15 cases from bibliographic search. We were able to access the medical observation details in 14 of them. We also report a new case diagnosed in Tunisia; the 16th in the world-review of literature.

This work has been reported in line with the SCARE criteria [1].
CASE REPORT

A 56-year-old woman with a history of high blood pressure presented to the emergency department complaining of right upper quadrant pain over the past week. Physical examination showed a temperature of 38°C and guarding of the right upper quadrant. Laboratory results showed WBC of 13100/mm³, HB of 9.8 g/dl, the rest was normal.

Abdominal ultrasound showed an oblong hyperechoic mass between the 3rd and 4th hepatic segments and a gallstone. A CT scan was performed. It showed infiltration and thickening of the round ligament and a cholecystitis (Figure 1). The patient was operated on by laparoscopy. Intra-operatively we discovered an inflammatory cholecystitis and a gangrene of the round ligament (Figure 2). A cholecystectomy and a total removal of the round ligament were performed with sub-hepatic drainage. Follow-ups were simple and complication-free. The patient was discharged from the hospital six days after the surgery. The anatomic pathology examination was in favor of an abscess of the round ligament.

DISCUSSION

The primary necrosis of the round ligament is an exceptional entity. The preoperative diagnosis is difficult due to the lack of specific signs. The symptoms consist of abdominal pain, vomiting, nausea and fever. It can mislead us to think of other diseases such as cholecystitis [2], pancreatitis [3] and a perforated ulcer [4]. The pathophysiology is still uncertain. Two theories have been developed; The first considers the origin to be infectious with a starting point in the umbilicus [5]; The second considers the origin to be ischemic similar to appendicitis [6]. The first theory is more likely in the case of our patient because of the association with a cholecystitis.

The ultrasound can show a heterogeneous hyperechoic mass between the liver and the anterior abdominal wall which is variable in size. The CT scan is more accurate and specific but the interpretation of morphological data can be distorted due to the lack of knowledge of the disease. The treatment of primary necrosis of the round ligament is surgical involving a resection of the ligament at variable levels and a potential drainage. Some authors suggest a medical treatment if the diagnosis is uncertain [7]. Through a systematic review of literature, we have compiled only 15 cases (Table 1) [8–20].

After analyzing the data from literature, we have concluded that, like in our case, this disease affects mostly women (10 women) with ages varying between 18 and 86 years old. In all the cases reported, the clinical presentation was an acute abdomen that evoked at first other diagnoses. The Ultrasound and CT scan showed pathognomonic signs which are of a heterogeneous and hyperechoic mass with fat density situated between the

| Author and Year | Age and Gender | Symptoms          | Exploration | Treatment Technique | Gesture | Anatomic Pathology | Germs |
|-----------------|---------------|-------------------|-------------|---------------------|---------|--------------------|-------|
| Charuzi 1976 [8] | F             | Acute abdomen     | N/A         | Laparotomy          | resection | N/A                | N/A   |
| Waston 1988 [9] | F             | Acute abdomen     | N/A         | Laparotomy          | resection | N/A                | N/A   |

Figure 1: Gangrene of the round ligament.

Figure 2: Abscessed and necrotic round ligament: intra-operative view (A), Extracted specimen (B).
| Author and Year | Age and Gender | Symptoms | Exploration | Treatment Technique | Gesture | Anatomic Pathology | Germs |
|----------------|---------------|----------|-------------|---------------------|---------|------------------|-------|
| Pans 1999 [10] | 69 y.o F      | Epigastric pain, vomiting | CT scan: Hypodense infiltration of the junction between the 3rd and 4th liver segment. | Laparotomy resection | Necrotic-inflamatory and purulent lesions | E.coli, K.pneumoniae enterococcus |
| Goti 2000 [4]  | 32 y.o F      | Right upper quadrant pain | CT scan: heterogeneous mass of the round ligament | Laparoscopy Resection | Hemorrhagic infarction | N/A |
| Losanoff 2002 [11] | 18 y.o M | Epigastric pain | N/A | Laparotomy Resection | N/A | N/A |
| Tison 2005 [12] | 86 y.o M | Pain, jaundice | Ultrasound: perihepatic effusion CT scan: perihepatic and sub-diaphragmatic collections with fat density | Laparotomy Resection drainage | Necrotic and inflammatory transformation | N/A |
| Aoun 2006 [13] | 62 y.o M | Epigastric pain, vomiting | CT scan: a heterogeneous image of the round ligament with air bubbles | Laparotomy Resection Drainage | Gangrene with purulent and inflammatory necrosis | E.coli |
| Tsukuda 2008 [14] | 70 y.o F | Right upper quadrant pain, fever | CT scan: abscess of the round ligament | N/A resection | Necrosis | S.epidermidis |
| Ghariani 2009 [15] | 62 y.o M | Acute abdomen | CT scan: infiltration and thickening of the round ligament | Laparotomy Resection Drainage | Gangrene with hemorrhagic necrosis and calcification of the umbilical vein | N/A |
| Czysmek 2010 [16] | 44 y.o F | Epigastric pain | Ultrasound: sub-parietal lesion, gallstone CT scan: damage of round and falciform ligaments | Laparoscopy Resection | Hemorrhagic necrosis | S.epidermidis |
| Ghadouani 2011 [17] | 50 y.o F | Epigastric pain | Ultrasound: interhepatoportal heterogeneous and hyperechoic mass CT scan: a mass of fat that stretches from the cracking of the round ligament to the umbilicus | Laparoscopy Incomplete resection | Necrosis | N/A |
| Bourguiba 2014 [18] | 76 y.o F | Sub-umbilical pain | Ultrasound: gallstone CT scan: a Hypodense infiltration of all the round ligament | Laparotomy Resection drainage | N/A | N/A |
liver and the umbilicus. The abdominal CT scan seems to be the standard imaging technique to diagnose this entity. The treatment was surgical in all reported cases. The laparoscopy was used five times including our case.

CONCLUSION

The primary necrosis of the round ligament is an exceptional cause of acute abdomen. Preoperative diagnosis is difficult to establish but it can be evoked based on many imaging data. Surgery allows the confirmation of diagnosis and the treatment especially laparoscopy which has many advantages.

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Author Contributions
Wissem Triki – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Baraket Oussema – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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Guarantor of Submission
The corresponding author is the guarantor of submission.

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Written informed consent was obtained from the patient for publication of this case report.

Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

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