Ectopic liver tissue in two distinct anatomical regions : a case report

T. Zwaenepoel[1], D. Dierickx[2], W. Laleman[3], R. Sciot[4]

Department of (1) General Internal Medicine, (2) Haematology, (3) Liver and Biliopancreatic disorders and (4) Pathology, University Hospitals Leuven, Leuven, Belgium.

Abstract

Extrahepatic liver tissue (ELT) is a rare clinical finding. Few cases are described. The reported location is almost exclusively confined to the subdiaphragmatic region with the gallbladder being the most frequent localisation. This paper describes a unique case with not only two localisations of ectopic liver tissue, but also in anatomical regions where ELT has never been described before. (Acta gastroenterol. belg., **2014**, 77, **68-70**).

Key words : extrahepatic liver tissue, ectopic liver tissue.

Ectopic Liver Tissue In Two Distinct Anatomical Regions : A Case Report Extrahepatic liver tissue is a rare developmental anomaly. A series of 5500 autopsies in 1940 reported an incidence of 0.23% (1). Ever since, several cases have been reported. The diagnosis is mostly made incidentally during medical imaging of the hepatobiliary tract, regional surgical procedures or autopsy (2-9), as often these lesions are clinically asymptomatic. Seldom, problems arise due to compression, pain or bleeding (6,10-13).

This paper describes a unique case with not only two localisations of ectopic liver tissue in the same patient, but even more but also in anatomical regions where ELT has never been described before.

Case report

A 63-year old woman was referred to the emergency department of our hospital, because of an infectious syndrome. Three days before presentation, she suddenly experienced a twinge on the right hemithorax. Two days later, a dark red plaque appeared under the right armpit, which extended to the ventral thoracic region. At presentation at the emergency department, cellulitis was diagnosed. Clindamycin (600 mg tid) and levofloxacin (500 mg qd) were initiated. On the fourth day of hospital admission, another red, painful plaque had developed in the right gluteal region. Despite ultrasonography of the regions of interest, it remained unclear whether these lesions were to be considered as cellulitis or an atypical haematoma.

At presentation, biochemical analysis revealed deep leukopenia (300/mm^3, ref. range 4500-11,000/mm^3), neutropenia (26%, ref. range 40-70%) and thrombopenia (136,000/mm^3, ref. range 150,000-400,000/mm^3). Because of persistence of these latter, a bone marrow aspirate was obtained. Bone marrow analysis showed no signs of underlying haematological disorder. Additional history did not reveal any explanatory factor. In the following days, a spontaneous recovery both biochemically as well as clinically was observed so that the cellulitis was considered the most likely diagnosis. The patient was discharged from the hospital. Two weeks later, at follow-up consultation, the patient mentioned fatigue, though denied any more episodes of fever since discharge. During clinical examination, none of the above mentioned inflammatory signs could no longer be detected or had reappeared, although a painless bulge remained palpable in the subaxillary region. A computer tomography was scheduled. However, before this exam could be performed, the patient was readmitted through the emergency department because of a sudden painful and increased swelling of the subaxillary bulge. During clinical examination, a warm, red and painful mass was withheld at the known subaxillary site. Additionally, another palpable lump was found at the right hip. CT scanning of the thorax and abdomen revealed two hypodense collections, both with a thickened contrast enhanced wall, which led to the conclusion of abscess formation. The first collection was located in the lateral posterior thoracic wall, between the lateral margin of the serratus anterior muscle and the anteromedial margin of the latissimus dorsi muscle. The second collection was found against the right gluteus medius muscle, and reached caudally as far as the greater trochanter. Antibiotics were restarted (flucloxacillin 1 g q4h intravenously), pending surgical drainage. Endocarditis was excluded. Cultures grown from an evacuation of 150 millilitres of wound fluid yielded presence of multisensitive Staphylococcus aureus (MSSA). During surgical drainage, aberrant tissue of unknown origin was detected in the margin of both abscesses, of which biopsies were obtained. A complete surgical resection has not been performed. Histologically, the biopsies were composed of strands of haemorrhagic inflammatory fibrous tissue. Within the inflamed areas nodules of liver tissue were present, consisting of muralia of hepatocytes and occasional ductular structures in portal tract-like areas (Fig. 1). On immunohistochemistry, the hepatocytes expressed low molecular weight keratins

Correspondence to : Tom Zwaenepoel, Department of Internal Medicine, University Hospitals of Leuven, Herestraat 49, 3000 Leuven, Belgium.
E-mail : tom.zwaenepoel@uzleuven.be

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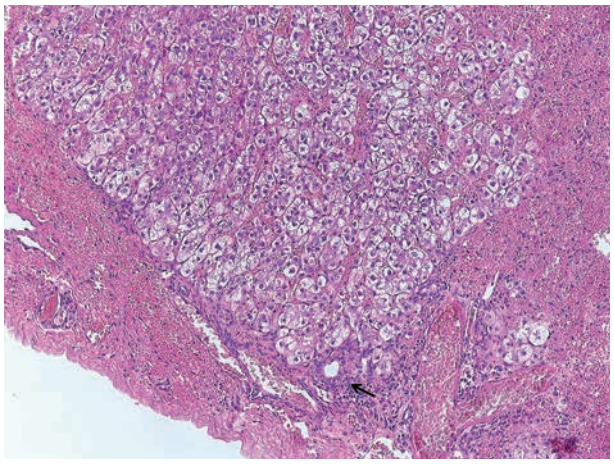

Fig. 1. — Ectopic liver tissue, consisting of muralia of polygonal hepatocytes as well as ductular structures (arrow). Haematoxylin and eosin stain.

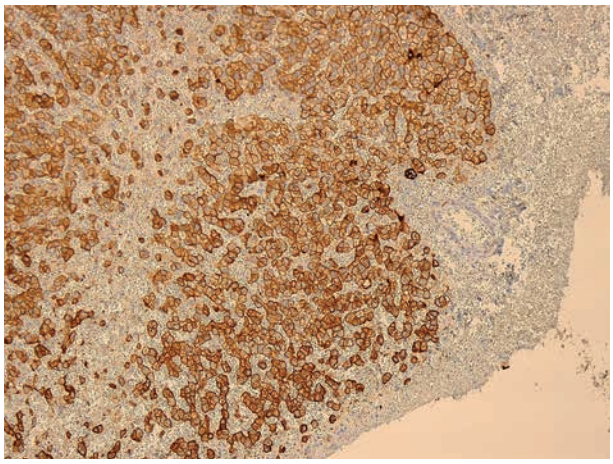

Fig. 2. — The hepatocytes express low molecular weight cytokeratins.

(CK8/18), as well as glypican 3 (Fig. 2). Alfa fetoprotein was weakly expressed in the hepatocytes, polyclonal CEA antibodies stained the liver cells in a canalicular pattern. Cytokeratin 7 labelled the bile ductules. Cytokeratin 20, S-100, melan-A, inhibin, calretinin, synaptofysin, CD31 and CD34 were negative in the hepatocytes. Two days after surgical drainage, the patient was discharged and antibiotic therapy (flucloxacillin 500 mg qid, oral intake) was prescribed for another two weeks. Since then, the patient remained without complaints. No relapse has been reported to our knowledge.

Discussion

Collan *et al*. (14) classified anatomical anomalies with regard to hepatic tissue into four categories : (1) an accessory liver lobe that can reach a considerable size and is attached to the liver by a stalk, (2) a small accessory liver lobe which is attached to the liver but is usually small, about 10-30 g in weight, (3) a macroscopic ectopic piece of the liver without any connection to the original liver and (4) microscopic ectopic liver tissue. The majority of reported ectopic liver tissue has been described to be attached to the gallbladder, with over 20 cases reported in the literature (15-19). Other locations include the adrenal glands, the pancreas, the spleen, the falciform ligament, the pylorus, the umbilicus, the retroperitoneal and pleural space, the oesophagus and the pericardium (5,6,10,11,13,20). Interestingly enough, hepatocellular carcinoma was involved in about one third of the reported cases of ELT (21), suggesting that ELT has an increased propensity of undergoing hepatocarcinogenesis (22,23).

The final diagnosis in our patient was made following the occurrence of abscesses out of microscopic ectopic liver tissue, confirmed by histological and immunohistochemical staining on tissue biopsies, taken during surgical drainage of the described abscesses.

No clear pedicle to the liver was noticed. Notably, this tissue was found in two anatomically distinctive regions, one of which was the right gluteal region. No other loci were identified in this patient, though no additional investigations, other than medical imaging via CT, were performed. We did a thorough study of the limited literature available on this subject. However, in the more than one hundred articles we reviewed, no previous case of two anatomically distinctive regions has been reported or occurrences in the gluteal region have been published. It is still unclear to us as to why abscess formation has occurred nearly simultaneously in these regions with ELT. However, as mentioned before, hepatocellular carcinoma was involved in a significant portion of the reported cases. In the majority of these cases, no clear known risk factor (e.g. chronic viral hepatitis B or C) has been identified, though data on serologic testing is mostly lacking. Nevertheless, this seemingly increased propensity of ELT undergoing hepatocarcinogenesis, before or in absence of liver cirrhosis, has been ascribed to the lack of normal vascular and ductal systems, possibly leading to prolonged exposure to carcinogenic factors and persistent cellular stress, coupled with cell death and cell proliferation (21,22). Theoretically, these anomalies can also explain a crippled drainage of haematogenic spreading microbial agents, leading ELT to become a sensible region, which these agents can then colonize and form abscesses in. Perhaps one can consider this case as a peculiar form of community acquired MSSA, disseminated to two loci minoris resistentiae. We think these findings might be of relevance to the omnipracticus upon confrontation of anomalous tissue.

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