Aseptic Liver Abscesses as an Exceptional Finding in Cogan’s Syndrome

Aseptic abscesses of deep organs are very rare, and their pathogenesis is unclear. Most reported cases affect spleen or liver and are associated with inflammatory bowel disease, Behçet’s disease, and/or other cutaneous or arthritic manifestations. Here, we report a case of aseptic liver abscesses diagnosed in a patient with Cogan’s syndrome, a rheumatic disease primarily affecting the inner ears and eyes.

Case Presentation

A 41-year-old Caucasian woman presented with fever (39.1°C), abdominal pain, and unintended weight loss. Alongside, the patient reported a progressive hearing loss for 1 year and several episodes of eye redness. She was on no regular medications and there were no known allergies.

DIAGNOSTIC EVALUATION

Laboratory blood investigations showed increased erythrocyte sedimentation rate (53 mm in 1 hour) and C-reactive protein levels (133 mg/L). Differential blood count exhibited microcytic, hypochrome anemia (hemoglobin 10 g/dL), lymphocytopenia (10.1%), neutrophilia (80.4%), and thrombocytosis (430 × 10⁹/L). Gamma-glutamyltransferase (255 U/L) and alkaline phosphatase (336 U/L) were elevated. Abdominal sonography showed multiple hypoechoic liver lesions, but no peripheral hyperenhancement in the arterial phase as a feature of abscesses (Fig. 1A). Repeated ultrasound-guided liver biopsy showed abundance of neutrophils and macrophages, but no indication of bacteria, parasites, fungi, or malignancy (Fig. 1B). Extended microbiological investigations were carried out and included eubacterial and panfungal PCR of liver specimens and screening for viral hepatitis, Echinococcus, Treponema, Brucella, Toxoplasma, and Entamoeba species. All results including repeated blood cultures and urine and stool analyses were negative. Likewise, upper and lower endoscopy were without pathological findings. Magnetic resonance and computed tomography (CT) confirmed multiple intrahepatic lesions (Fig. 2A). Additionally, significant infrarenal wall thickening of the abdominal aorta was detected (Fig. 2B).

CLINICAL COURSE AND FOLLOW-UP

Initially, the patient was treated with piperacillin/tazobactam and metronidazole. As fever continued and...
C-reactive protein levels increased up to 250 mg/L, antibiotic therapy was escalated to meropenem. However, the patient did not respond to extended therapy with meropenem for 10 days. Fluorodeoxyglucose-positron emission tomography (FDG-PET) CT scanning was conducted to detect potential further inflammatory foci and confirmed inflammatory activity in several of the intrahepatic lesions along with infrarenal aortitis and periaortitis (Fig. 2C, left panel). Thus, we diagnosed sterile hepatic abscesses. Moreover, concomitant aortitis, hearing loss, and ocular symptoms were indicative of Cogan’s syndrome, even though aseptic liver abscesses have never been reported in this rare inflammatory disease so far.

After starting intravenous prednisolone treatment (1 mg/kg body weight [BW]), the inflammatory markers dropped promptly and decreased to normal levels within 1 week. Additionally, the patient reported complete recovery of general condition and improvement of hearing ability. Sonographically, the index lesions were continuously monitored and showed a rapid volume reduction.

A maintenance therapy with infliximab (5 mg/kg BW) and azathioprine (100 mg/day) was commenced.

**FIG. 1.** Liver ultrasound and biopsy specimen of liver abscess. (A) Initial liver ultrasound showing multiple hypoechoic liver lesions. Right picture: Contrast-enhanced sonography of liver lesion. (B) Hematoxylin and eosin-stained section from liver biopsy specimen showing infiltration by neutrophilic granulocytes; original magnification ×400. Abbreviation: HE, hematoxylin and eosin.
In further follow-up visits, the patient presented asymptomatic. FDG-PET-CT scanning after 4 months showed persistent response, with reduced metabolic activity of both periaortic and hepatic lesions (Fig. 2C, right panel).

**Discussion**

Cogan's syndrome is mostly diagnosed in early adulthood by characteristic ocular and audiovestibular symptoms. Additionally, systemic manifestations were reported, such as aortitis and arthromyalgia. Here, we describe a case of aseptic liver abscesses in Cogan's syndrome. In Cogan's syndrome, infliximab was identified as a promising therapy, enabling steroid tapering and improving vestibular-auditory outcomes. Because of the high risk of recurrence, maintenance therapy appears reasonable in aseptic abscesses as well, and efficacy of tumor necrosis factor-α inhibitors, azathioprine, and sulfasalazine has been reported. Thus, we started infliximab and azathioprine as a maintenance therapy in the reported case.

From a clinical point of view, noninfectious hepatic abscesses should be considered in patients with systemic inflammatory disorders who do not respond to antibiotics. Due to diagnostic challenges, the actual incidence of aseptic liver abscesses might be underestimated.
Acknowledgment: The authors thank Julia Schwörer (Schwörer Optik & Hörakustik GmbH, Leutkirch, Germany) for the delivery of hearing test results and Frank Leithäuser (Institute of pathology, Ulm University) for the provision of microscopical images of the histological stainings. Open access funding enabled and organized by Projekt DEAL.

Author Contributions: S.H. was responsible for drafting the manuscript and designing the figures with input and under supervision by T.S., M.M., and A.K. All authors were involved in diagnosis and treatment of the reported clinical case and critically revised the manuscript. M.M. acquired the patient’s consent for publication of the case.

REFERENCES

1) Bollegala N, Khan R, Scaffidi MA, Al-Mazzouki A, Tessolini J, Showler A, et al. Aseptic abscesses and inflammatory bowel disease: two cases and review of literature. Can J Gastroenterol Hepatol 2017;2017:5124354.
2) Andre MF, Piette JC, Kemeny JL, Ninet J, Jego P, Delevaux I, et al. Aseptic abscesses: a study of 30 patients with or without inflammatory bowel disease and review of the literature. Medicine (Baltimore) 2007;86:145-161.
3) Grasland A, Pouchot J, Hachulla E, Bletry O, Papo T, Vinceneux P, et al. Typical and atypical Cogan’s syndrome: 32 cases and review of the literature. Rheumatology 2004;43:1007-1015.
4) Durlette C, Hachulla E, Resche-Rigon M, Papo T, Zenone T, Lioger B, et al. Cogan syndrome: characteristics, outcome and treatment in a French nationwide retrospective study and literature review. Autoimmun Rev 2017;16:1219-1223.