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CASE REPORT

A 12-year-old farmer's boy presented with 4 weeks of left flank pain. On examination, a palpable mass below the right costal margin was noted. Investigations, including full blood count, C-reactive protein, and kidney and liver tests, were normal, except for mild eosinophilia (0.53×10^9/L). Immunological work-up was normal including immunoglobulins, lymphocyte subsets and serology for HIV. Ultrasonography and abdominal CT revealed an extensive lesion originating from segment V/VI to segment VIII with encasement and stenosis of the portal vein (figure 1). The boy reported contact with animals on the family's farm, such as dogs and cattle and foxes living in the surrounding area. ELISA and indirect haemagglutination for echinococcosis were 2.01 optical density (OD; normal <0.50 OD) and 1:512 (normal <1:128), respectively. Infection with *Echinococcus multilocularis* was confirmed by specific bands for 7, 16, 18 and 26–28 kDa in Western blot and specific antibodies against EmG11 and Em18.[1] On positron emission tomography (PET)-CT, metabolic activity was noted in the liver suggesting parasite viability[2] (figure 2), and staging of the disease was P3N0M0 according to WHO criteria.[3] Curative right extended hepatectomy with portal vein reconstruction and biliary-enteric anastomosis was offered, but the family preferred oral albendazole treatment.[3] Albendazole (8 mg/kg/day) was well tolerated with intermittent mild elevation of liver function tests. Regular therapeutic drug monitoring showed albendazole sulfoxide concentrations of 2.5–3.0 µmol/L (therapeutic range 2.0–5.0 µmol/L). The lesions decreased in size at 1.5 years' follow-up PET-CT, though persistent metabolic activity was present.

Alveolar echinococcosis is mainly found in temperate climates of the northern hemisphere and has an incubation period of 5–15 years.[4] It has high fatality rates and poor prognosis if managed incorrectly. The prognosis has improved considerably with albendazole, especially in cases where surgery is high risk.

Figure 1 Abdominal CT revealing a cystic, partly calcified lesion of about 12 cm size originating from segment V/VI with extension into the liver hilum, encasing the portal vein and leading to a stenosis (diameter prestenosis 13 mm and 7 mm intrastenosis, not shown). The lesion filiformly extends into segment VIII.

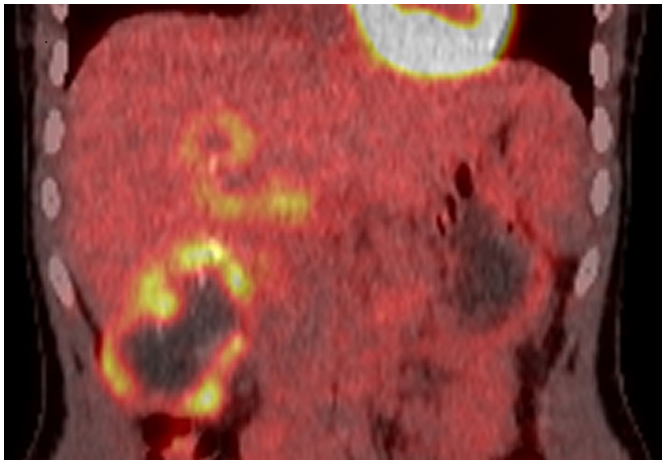

Figure 2 Cystic lesion with central calcifications showing enhanced fluorodeoxyglucose (FDG) uptake at the edge in segment V/VI and plane tracer uptake in the liver hilum and segment VIII.

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REFERENCES

1 Siles-Lucas M, Casulli A, Conraths FJ, *et al*. Laboratory diagnosis of echinococcus spp. in human patients and infected animals. *Adv Parasitol* 2017;96:159–257.
2 Ammann RW, Stumpe KD, Grimm F, *et al*. Outcome after Discontinuing long-term benzimidazole treatment in 11 patients with non-resectable alveolar echinococcosis with negative FDG-PET/CT and anti-EmII/3-10 serology. *PLoS Negl Trop Dis* 2015;9:e0003964.
3 Brunetti E, Kern P, Vuitton DA, *et al*. Expert consensus for the diagnosis and treatment of cystic and alveolar echinococcosis in humans. *Acta Trop* 2010;114:1–16.
4 Schmidberger J, Kratzer W, Stark K, *et al*. Alveolar echinococcosis in Germany, 1992-2016. An update based on the newly established national AE database. *Infection* 2017.