CASE REPORT

Echinococcus multilocularis infection of the liver presenting as abdominal wall fistulaZygimantas Juodeikis,¹ Tomas Poskus,¹ Dmitrij Seinins,² Kestutis Strupas¹

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SUMMARY

Echinococcus multilocularis causes infection where the most commonly affected organ is the liver, followed by the lung, kidney, bone and the brain. Other sites such as the heart, spleen, pancreas and soft tissues are very rarely affected. Surgical treatment combined with chemotherapy using various technical approaches remains the main therapeutic modality for echinococcal liver disease. To the best of our knowledge there are less than five clinical cases of cutaneous presentation of liver alveolar echinococcosis described. We present a unique case of liver echinococcosis presenting as recurrent abdominal wall fistula and abscess in a 29-year-old man. Diagnosis was based on CT imaging, serological analysis and histological findings from the fistula. Medical treatment with albendazole was initiated and liver resection was performed. The patient has no symptoms and signs of recurrence 1 year after operation, while still on albendazole therapy. This case description highlights the importance of early suspicion and treatment of unusual echinococcosis clinical presentations.

BACKGROUND

Metacestode of *Echinococcus multilocularis* initially develops almost exclusively in the liver, more often in the right lobe, from foci of a few millimetres to areas of 15–20 cm in diameter.^{1,2} About one-third of the patients have simultaneous lesions in one or more extrahepatic organs.¹ Concomitant lesions may be formed in adjacent organs through local spread or through a lymphatic or haematogenous route.¹ The management of the disease consists of surgical and medical treatment. We present a unique case of *E. multilocularis* infection presenting as recurrent abdominal wall fistula.

CASE PRESENTATION

A 29-year-old man was admitted to our hospital with fever, abdominal pain and yellowish drainage from the right lower lateral abdominal wall for the past 2 days. He had previously undergone three consecutive abscess drainage procedures in another institution on the same site with recurrent abscess formation over the past year. Cultures were taken from the abscesses, but there was no bacterial growth. The patient also mentioned that he had suffered blunt abdominal trauma 3 years ago and was treated conservatively in his local hospital.

Physical examination revealed two external fistula openings located 4–5 cm above the anterior superior iliac spine and 3–4 cm above the posterior superior iliac spine, approximately 1 cm in size

Figure 1 Soft tissue around fistula (H&E staining, ×200). Necrotising granuloma with irregular cysts (arrows) and numerous macrophages.

each. Revision of the affected area showed a communication between the anterior and posterior abdominal wall fistula openings. Both had no relation with abdominal cavity, with fistula tracks in the subcutaneous tissue.

INVESTIGATIONS

Fistula exploration was performed under anaesthesia, biopsies were taken. Histological findings revealed necrotising granuloma with irregular cysts and numerous macrophages. Cysts had germinal membrane and PAS-positive laminated membrane (figures 1 and 2).

Abdominal ultrasound revealed a hyperechoic structure of 2×3 cm, located between the sixth and seventh segments of the liver. Abdominal CT confirmed the presence of the aforementioned structure in the liver.

ELISA test was performed to detect IgG against *E. multilocularis* and *Echinococcus granulosus* (Bordier Affinity Products SA, Switzerland). Result

Figure 2 Cysts have germinal membrane and Periodic acid-Schiff (PAS)-positive laminated membrane (arrows; PAS, ×200).

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Figure 3 Liver resection specimen (ruler in cm), *Echinococcus* tissue appears whitish in the parenchyma of the liver (arrows). Margin of 1 cm of healthy liver around echinococcal tissue is visible.

for *E granulosus* was positive at 1.412 (cut-off value >0.806) and result for *E multilocularis* was negative at 0.507 (cut-off value >0.822). No further serological confirmation was pursued, as histological diagnosis was available.

ELISA test for IgG against *E granulosus* is 90 % cross reactive to *E multilocularis* infection and *E multilocularis* test is negative in up to 10% of infections.

TREATMENT

With these findings, medical treatment with albendazole (400 mg twice daily) was initiated and a radical treatment option was chosen.

Atypical liver resection of sixth and seventh liver segments was performed (figure 3) with 1 cm clear resection margin around the echinococcal tissue to be achieved. No communication of the liver with abdominal wall or no evidence of penetration of echinococcal tissue from the liver to the abdominal wall was visible. Complete excision of the fistula tracks was performed at the same procedure. There were no complications in the postoperative period. The patient was discharged on the seventh postoperative day. Postoperative histopathological findings confirmed the presence of *E multilocularis* and clear resection margins in the liver (figures 4 and 5).

Figure 4 Liver (H&E staining, $\times 20$). Numerous necrotising granulomas with identical irregular cysts (arrows) and extensive peripheral rim of granulomatous inflammation.

Figure 5 Cysts have germinal membrane and Periodic acid-Schiff (PAS)-positive laminated membrane (arrow; PAS staining, $\times 200$).

OUTCOME AND FOLLOW-UP

After 1 year of treatment with albendazole the patient is asymptomatic, with no evidence of fistula or *Echinococcus* recurrence.

DISCUSSION

There are only a few reports in the literature of the *Echinococcus* infection in the skin.^{3–5} In our case the skin infestation could have resulted from blunt abdominal and liver trauma and proctoscolices migration from primary lesion in the liver to contiguous abdominal wall; as previous abdominal wall abscess operations did not cure the disease. Liver mass was evident on the ultrasound scan, *Echinococcus* infection should have been suspected earlier.

The diagnosis of *E multilocularis* infection requires at least two of the four parameters:

1. Typical organ lesion detected by imaging techniques.
2. Detection of specific serum antibodies for *Echinococcus* species.
3. Histopathology compatible with metacestode of *E multilocularis*.
4. Detection of *E multilocularis* nucleic acid in clinical specimen.^{1 2}

Unusual sites of this disease can frequently cause diagnostic problems and so this can give rise to an increased risk of diagnostic delay and many potentially serious complications.

Chemotherapy with benzimidazoles is mandatory in all patients: medication can be used temporarily in those, who underwent curative radical resection; however, they should be continued indefinitely in cases where radical resection is not possible. Surgical treatment should be attempted, when radical resection is possible. In those patients, on whom radical surgery is not possible, minimally invasive palliative surgery procedures are preferable.^{1 2 6}

As there were no other foci of infestation, radical surgery under the benzimidazole coverage for liver lesion was chosen. Preoperative benzimidazoles were administered because infestation had spread to the abdominal wall.

Learning points

- ▶ *Echinococcus* infection is increasing.
- ▶ The infection can spread into most tissues of the body—suspicion is important.
- ▶ Serological diagnosis is very helpful.
- ▶ Surgical treatment together with benzimidazoles can provide a long-term remission or cure.

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