Pleural Fluid with Hooklets in a 7-year-old Boy with Severe Multisystem Cystic Echinococcosis

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Author's contributions

We were all involved in the patient's care and diagnosis. P.M.M.S. wrote the first draft of the manuscript. We all edited the manuscript and approved the final version. Written consent for publication was obtained from the patient's father.

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A 7-year-old boy from Macedonia who immigrated to Switzerland two years earlier presented to our clinic with a cough since a week and loss of appetite for months. His medical history was unremarkable and he was not exposed to animals. On physical examination, he was tachypneic and had absent breath sounds on the right hemithorax. Laboratory investigations revealed a total leukocyte count of 11,610/ μ L without eosinophilia and a C-reactive protein level of 103mg/L. Chest x-ray showed opacification of the right hemithorax (Figure 1A) with >10mm rim of fluid on sonography pointing at a large pleural effusion (1). A diagnostic pleural puncture for suspected parapneumonic effusion drained 40mL of clear fluid. Gram stain and antigen testing for *Streptococcus pneumoniae* on pleural fluid specimens were negative. Subsequently, the patient developed a pneumothorax (Figure 1B), requiring tracheal intubation and pleural needle decompression. Computed tomography revealed cystic masses in the right hemithorax (Figure 1C) and the liver (Figure 1D).

Pleural fluid microscopy showed protoscoleces from *Echinococcus granulosus* (Figure 1E) with shiny colorful hooklets (Figure 1F and Video 1). Cystic echinococcosis was confirmed by polymerase chain reaction and serology (enzyme-linked immunosorbent assay and immunoelectrotransfer blot) (2). Pleural fluid and blood cultures and a tuberculin skin test were negative. The accidental cyst puncture with spillage of protoscoleces led to anaphylaxis and acute respiratory distress syndrome requiring extracorporeal membrane oxygenation for 4 days. He was started on albendazole, followed by urgent lobectomy of the right upper lobe, total pericystectomy of the right lower lobe at 3 weeks (Figure E1), and puncture, aspiration, injection, re-aspiration (PAIR) technique of the liver cyst at 3 months. Albendazole was administered for total 5 months. A 6-month follow-up revealed

normal level of activity but exertional dyspnea, re-expansion of the right lower lobe, and reduction in the size of the liver cyst.

Cystic echinococcosis is a very rare disease in children but may be considered as differential diagnosis of pneumonia with pleural effusion in migrant children from endemic areas (3-5). Even though the incubation period lasts for years also young children can be affected (3). Our case further highlights that symptoms in children may be sparse or lacking despite enormous mass effect of cysts (3). Hooklets on microscopy can be clues to diagnosis (6, 7). Importantly, spillage of protoscoleces should be avoided to prevent anaphylaxis and severe immunological reactions (8).

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Figure 1. Severe childhood multisystem cystic echinococcosis. (A) Chest x-ray at presentation showing opacification of the right hemithorax, and (B) after pleural puncture showing a thin-walled radiolucent lesion with curvilinear septa (“water lily sign”, red arrow) in the right upper lobe, a radiopaque mass in the right lower lobe, and air trapping in the right pleural space with contralateral mediastinal displacement indicative of a tension pneumothorax. (C) Computed tomographic scan with contrast medium showing detached free-floating membranes within the low-density cystic lesion (“water lily sign”, red arrow), a high-density cystic mass in the right lung (blue arrow), and (D) a high-density cystic mass in liver segment VIII (green arrow). (E) Light microscopy of pleural fluid showing everted protoscolex from *Echinococcus granulosus* (100× magnification, unstained wet preparation) with calcareous corpuscles (black arrow) and row of hooklets (insert shows corresponding area with magnification). (F) Phase-contrast microscopy with detailed view of hooklets (400× magnification, unstained wet preparation).

Video 1. Active movements of hooklets of *Echinococcus granulosus* in pleural fluid (400× magnification, unstained wet preparation).

Figure 1.

242x153mm (300 x 300 DPI)

Video Still

Online Data Supplement

Figure E1. Total pericystectomy of the right lower lobe.

Figure E1.