A rare presentation of multi-organ hydatid disease: Case report and review of the literature

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Hydatid disease involving multiple organs is rare in clinical practice. The most commonly affected organs in adults include the liver and lungs. We report a case of a 54-year-old woman who presented to our hospital with multiple cystic lesions in the liver, lung and spleen. The case highlights the need to have a high index of suspicion for hydatid disease in endemic areas in patients with this clinical presentation.
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Hydatid disease is a zoonotic parasitic infestation prevalent worldwide, which is caused by the larval stages of cestodes (tapeworms) belonging to the genus Echinococcus. Four species of Echinococcus have been recognised to be of public health concern, of which *Echinococcus granulosus* causes the most infestations in humans, manifesting as cystic echinococcosis. Risk factors for hydatid disease include homemaking and an animal farming occupation, residing in or travel to an endemic area, a contaminated drinking water source, ownership/access to dogs or sheep, low socioeconomic status, poor education, age and sex.[1]

Case report

A 54-year-old woman was referred to our hospital for assessment of 'multiple round cystic opacities varying in size' seen on a chest radiograph (Fig. 1A and B) and a concern for possible lung malignancy. She had been a homemaker for many years in a rural area of Eastern Cape Province, with extensive exposure to indoor biomass fuel, and had close contact with dogs, cattle and sheep. She presented with a 6-month history of daily cough, productive of half a cup of yellow/green sputum that had become white over the past month. She had noted haemoptysis on only two occasions during the first month of cough. She described seeing white 'eggshell membrane'-like substances in her sputum over several months. During the same period, she experienced shortness of breath on exertion, wheeze that was worse at night, central sharp chest pain that radiated to both arms, and left upper-quadrant (LUQ) that was worse when lying down on her left side. Constitutional symptoms included weight loss (15 kg), with no associated loss of appetite, and night sweats that were profuse at the start of the symptoms, with associated fever and chills. These symptoms subsided somewhat after two courses of antibiotics.

She was a non-smoker and had no history of tuberculosis (TB); previous investigations for TB were negative (GeneXpert (Cepheid, USA) and auramine stain). She recalled being diagnosed by a local doctor, with two abdominal masses on ultrasound examination, but defaulted on her appointment at a tertiary centre to which she had been referred.

On physical examination, she had a blood pressure of 118/80 mmHg, heart rate of 80 bpm and respiratory rate of 22 breaths per minute, with saturation of 94% in room air. She was comfortable and weighed 50.0 kg (body mass index (BMI) 20.8). A general examination was normal, and her respiratory examination revealed a trachea deviated to the right and dullness in the right upper and lower zones, with decreased breath sounds in the right lower zone. She had bronchial breathing globally with no wheeze or crackles. Her abdomen was soft and non-tender, with no ascites. She did have mild hepatomegaly and an 8 - 10 cm firm mass in the LUQ.

On further investigation, her enzyme-linked immunosorbent assay (ELISA) HIV test was negative and a full blood count revealed a mild microcytic anaemia (Hb 10.7 g/dL, mean corpuscular volume 78.3 fL, mean corpuscular Hb concentration 25.2 pg, white cell count 7 230/μL, platelet count 5 810/μL) and elevated lactate dehydrogenase (248 U/L). She had normal liver enzymes, international normalised ratio (INR) and renal function (total protein 85 g/L, albumin 35 g/L, total bilirubin 4 μmol/L, conjugated bilirubin 2 μmol/L, aspartate transaminase 20 U/L, alanine transaminase 15 U/L, alkaline phosphatase 95 U/L, gamma-glutamyl transferase 19 U/L, INR 1.30, urea 3.7 mmol/L, creatinine 76 μmol/L).

Sputum-microscopy sensitivity and culture revealed +3 neutrophils with normal respiratory flora. GeneXpert was negative for *Mycobacterium tuberculosis*. Pulmonary function tests demonstrated mild obstruction with significant and complete reversibility after administration of a bronchodilator (260 mL; 19%) and normal diffusion capacity (forced expiratory volume (FEV_1) 1.37 L (63% predicted), forced vital capacity (FVC) 2.02 L (79%), FEV_1/FVC 67.69%, and carbon monoxide transfer factor 16.42 mL CO/min/mmHg (75% predicted)).

The chest radiograph (Fig. 1A) showed right upper-lobe volume loss with the trachea deviated to the right, crowding of ribs, right upper-zone patchy opacity with possible intracavity mass with crescent of air and marked pleural thickening, large posterior right lower-lobe round opacity (Fig. 1B) and smaller right middle-lobe round opacity with marked bronchial wall thickening but no pleural effusions. The most likely diagnosis was hydatid disease in an endemic area. Other

diagnostic considerations included lung cancer, sarcoma or aspergilloma, for which a computed tomography (CT) scan of the chest was done as a further investigation. A CT scan of the chest (Fig. 2A) revealed a large right lower-lobe cyst (8 × 8 cm), with significant right middle- and upper-lobe scarring, traction bronchiectasis/bronchiolectasis and lung cavities containing low-density material with gas locules, in keeping with aspergillomas. The left lung had a small well-circumscribed superior lingual cyst (19 mm), while the upper abdomen (Fig. 2B) revealed two large cysts in the liver (9.5 × 7.0 cm) and spleen (15 × 10 cm). The final diagnosis was hydatid disease of the lungs, with a rare giant splenic hydatid, while the right upper- and middle-lobe disease was possibly due to post-TB lung disease with aspergillomas and/or bronchiectasis. The thoracic surgeons recommended surgical removal of the cysts, which the patient declined, citing a need to consult further with her family. A trial of medical treatment (albendazole 400 mg twice daily) was commenced while awaiting surgery. The patient later became loss to follow-up.

Discussion

Distribution

Echinococcosis is one of the 17 neglected tropical diseases recognised by the World Health Organization (WHO), with >1 million individuals infected worldwide each year. There are large regional differences in the prevalence of this disease in Africa; it is most common in north and east Africa (~2 - 3.5%), with few epidemiological data on its prevalence in sub-Saharan Africa.[2] The disease is rare in west Africa, with no data from central Africa. In South Africa, the epidemiology is poorly understood. Retrospective data analysis results of echinococcosis serology, microscopy and histopathology in eight provinces (excluding KwaZulu-Natal) showed an overall positivity rate of 17.0% (*n*=1 056/6 211) in diagnostic samples. Provinces with the highest rates were Eastern Cape (30.4%), North West (19.0%) and Northern Cape (18.0%).[3]

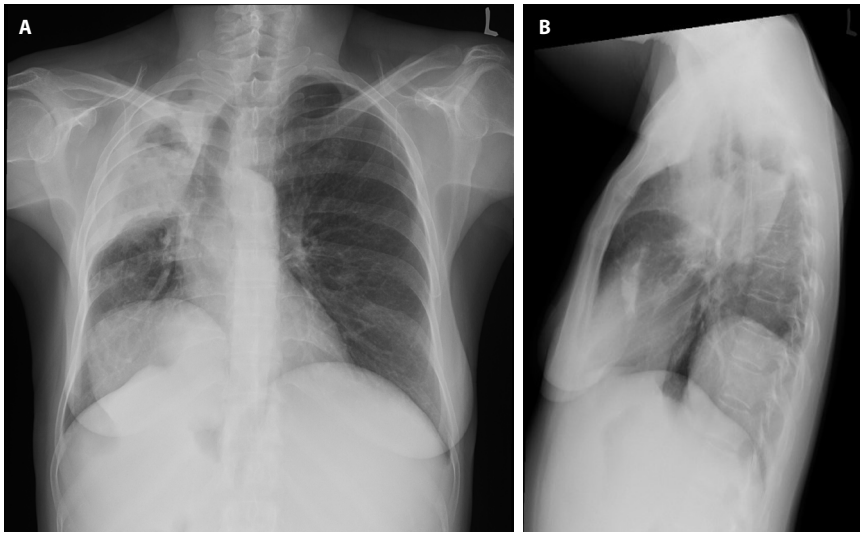

Fig. 1. (A) Chest radiograph, showing right upper-lobe volume loss and intracavity mass, as well as large posterior right lower-lobe cyst. (B) Chest radiograph, showing large posterior right lower-lobe cyst.

Epidemiology and transmission

Hydatid disease is acquired by ingesting food or liquid contaminated by the faeces of a definitive host that contains eggs. After ingestion, the liberated eggs pass through the intestinal wall into the portal system and settle in the liver. The majority of cysts occur in the liver (65%), followed by the lungs (25%) or both. More rarely (2 - 10%), cysts can occasionally be found in the spleen, kidney, peritoneal cavity, heart, brain, vertebral column and ovaries.[1]

The case presented here is rare, with multi-organ involvement affecting the lung, liver and spleen.

Clinical features and diagnosis

The lung is the most frequently reported site of hydatidosis in children and the second most common site in adults.[4] In the initial

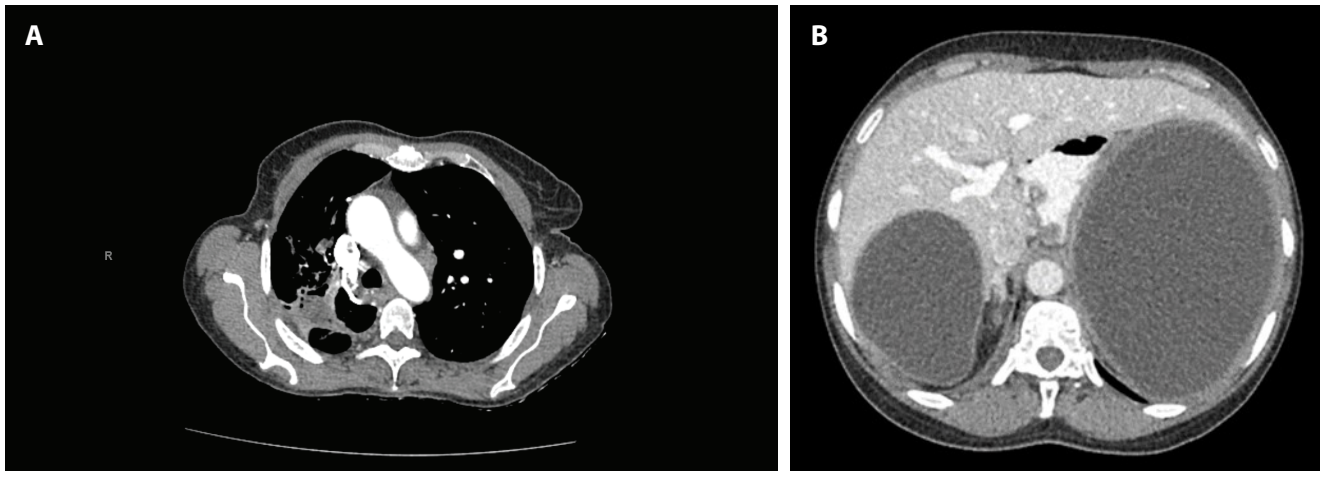

Fig. 2. (A) Computed tomography scan of the chest, showing multiple low-density material with gas locules in the right upper-lobe cavities, in keeping with aspergillomas. (B) Computed tomography scan of the abdomen, showing large cysts in the liver and spleen.

stages of the disease, most patients with pulmonary hydatidosis are asymptomatic, with subsequent complaints of cough (with occasional expectoration of the cyst contents), chest pain, dyspnoea, haemoptysis and fever. Pulmonary cysts may rupture, releasing antigenic material, which may lead to life-threatening complications such as anaphylaxis, tension pneumothorax, secondary hydatid dissemination and secondary infection.[4] Antibodies directed against specific echinococcal antigens are detected in only ~50% of such patients, while <15% have eosinophilia.[4,5] Radiologically, the cysts appear as well-defined solitary or multiple round or oval masses with a propensity for the right lower lobe of the lung.

The liver is the most common site of infestation in adults and the diagnosis is often incidental.[5] Patients usually remain asymptomatic for many years. Presenting symptoms are often nonspecific, including nausea, vomiting, fatigue and epigastric and/or right upper-quadrant abdominal pain. Complications include cholangitis from biliary tree obstruction, with super-added infection, hepatitis, abscesses, peritonitis from intraperitoneal leakage and anaphylactic shock. Ultrasonography of the abdomen is used for staging of the disease and treatment selection, based on the WHO classification of hepatic hydatid cysts. A CT scan of the abdomen and magnetic resonance imaging (MRI) can be used to confirm the diagnosis. Immunological investigations such as *E. granulosus* antigen B can support the diagnosis.

Splenic hydatidosis is rare and patients remain asymptomatic for years.[6] It may present as a left upper-abdominal mass (splenomegaly), with associated tenderness and fever. Rupture of a splenic cyst may extend into surrounding structures and lead to anaphylactic shock and secondary infection. A CT scan has higher sensitivity and specificity than ultrasound in aiding with the diagnosis.[6]

Treatment and prevention

The mainstay of treatment is surgery, particularly for large and/or infected cysts, those likely to rupture or those causing a significant mass effect. Surgery might be impractical where multiple cysts in several organs occur or where there is inadequate expertise; therefore, preoperative chemotherapy is an option.[1] Cysts may also be punctured under ultrasound guidance using the PAIR (puncture, aspiration, injection, re-aspiration) technique, and is recommended for liver cysts. Chemotherapy with the use of antifungal treatment (benzimidazole compounds: albendazole and mebendazole) has been shown to result in the resolution of up to 48% of cysts and a significant reduction in the size of an additional 24%.[1] These measures predominately target active disease, while upstream preventive measures should also be considered in the control of echinococcosis. These include implementing sustained cystic echinococcosis control programmes/policies, anthelminthic treatment of dogs, improved slaughter hygiene, surveillance of high-risk groups such as farm workers, improved hand hygiene and sanitation, safe disposal and incineration of infected organs and health education.[1,2]

Conclusion

This case illustrates the need to have a high index of suspicion for hydatid disease when managing a patient who presents with cystic lesions, particularly when they live in or travel to endemic areas. Surgical treatment alone or in combination with antifungal treatment is an appropriate method to prevent complications. A limitation of the clinical management is the lack of hydatid indirect haemagglutinin assays, which despite the reduced sensitivity in multi-organ hydatid disease, could have been performed and should be considered in endemic areas.

Declaration. None.

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