Hepatocardiopulmonary hydatid cysts: A rare paediatric case

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Echinococcus granulosus (CE) is a worldwide public health problem causing considerable human morbidity and mortality. We report on a case of complicated paediatric echinococcosis as evidenced by pulmonary, hepatic and cardiac cysts. Combined surgery and chemotherapy represent an appropriate strategy for managing cardiac and cardiopulmonary echinococcosis. However, there is a pressing need for developing a prevention strategy, collaborative research efforts and establishing a registry for information sharing, particularly in SA and other endemic regions.

S Afr J Child Health 2024;18(2):1851. https://doi.org/10.7196/SAJCH.2024.v18i2.1851

Echinococcus granulosus (CE) is a worldwide public health problem causing considerable human morbidity and mortality. The World Health Organization (WHO) estimates an annual prevalence of 50 per 100 000 people, with 19 300 deaths and 871 000 disability-adjusted life-years, globally.[1]

Though the lifecycle is well described, no epidemiological data is available for paediatric echinococcosis. In South Africa (SA), the epidemiology of CE has not been investigated in detail or in a systematic manner, despite clinical experience in various centres suggesting that SA might be an endemic country.[2]

Previous studies have highlighted the difference in the clinical presentation of adult v. paediatric echinococcosis. Adult echinococcosis commonly presents with liver hydatid cysts, whereas children commonly present with lung involvement or a combination of lung and liver hydatid cysts.[2]

We report on a case of complicated paediatric echinococcosis as evidenced by pulmonary, hepatic and cardiac cysts.

Case

A three-year-old boy presented to his local hospital for the second time with a history of persistent cough for more than 2 months and low-grade fever. He had been previously treated for community-acquired pneumonia based on clinical grounds.

At the current presentation, his clinical symptoms had worsened. He exhibited significant respiratory distress, including tachypnoea, nasal flaring, subcostal and intercostal recessions. He was also hypoxic in room air, with an oxygen saturation of 90%. He had significantly decreased air entry in the right hemithorax with scattered crepitations in both lung fields. A cardiac examination revealed good perfusion and normal blood pressure but a displaced, heaving apex. His liver was enlarged, measuring 4 cm below the costal margin in the midclavicular line, and was firm and non-tender.

A full blood count showed moderate leukocytosis (19.45 x 10^9/L). C-reactive protein (CRP) concentration was elevated at 52 mg/L, and immunoglobulin G (IgG) tested by enzyme-linked immunosorbent assay (ELISA) for echinococcus was positive. Tuberculosis screening was negative (tuberculin skin test, sputum for GeneXpert, microscopy and culture).

Chest radiographs demonstrated multiple, bilateral, oval pulmonary parenchymal densities of varying sizes (Fig. 1). The heart border was not clearly demarcated. Electrocardiogram (ECG) was normal and did not show any chamber enlargement.

Contrasted chest computer tomography (CT) scan showed multiple, bilateral, oval lesions of varying size in the lung parenchyma. The cyst was filled and non-enhancing, consistent with pulmonary hydatid cysts (Fig. 2 and 3). One smaller cyst in the left upper lobe also demonstrated internal gas, suggesting erosion into the bronchus and separation of the endocyst and ectocyst walls. The largest lung cyst, originating from the right lower lobe, measured 49 mm x 72 mm in the axial plane with a craniocaudal extension of 83 mm.

Similar, multiple, large, varying-sized fluid-filled non-enhancing cysts were seen in the liver on the CT scan, compatible with liver hydatid cysts. No cysts were noted in the spleen.

One cyst, measuring 30 mm x 27 mm and displaying similar CT characteristics, was found in the antero-superior portion of the left ventricle, consistent with a cardiac hydatid cyst. Cardiac echocardiography performed after the CT scan confirmed normal cardiac function and a cystic lesion confined to the myocardium.

The patient was treated with a 7-day course of amoxicillin/clavulanic acid and started on albendazole (10 mg/kg twice daily). He completed a 1-week course of albendazole before undergoing cardiac cystectomy via median sternotomy on cardiac bypass. He was discharged with a 3-month supply of albendazole however the patient was lost to follow up. Efforts to trace him were successful after 10 months, during which his pulmonary and hepatic cysts remained intact. He is now scheduled for a staged thoracotomy and PAIR procedure. Written consent was obtained from the patient's parents for the publication of this case.

Discussion

Cystic echinococcal (CE) disease or hydatid disease (HD) is caused by the larvae of *Echinococcus species,* most commonly *E. granulosus*. The primary infection occurs following the ingestion of *Echinococcus* eggs. The larvae enter the circulation through the gut and are hematogenously disseminated leading to distal cyst formation.[1,2]

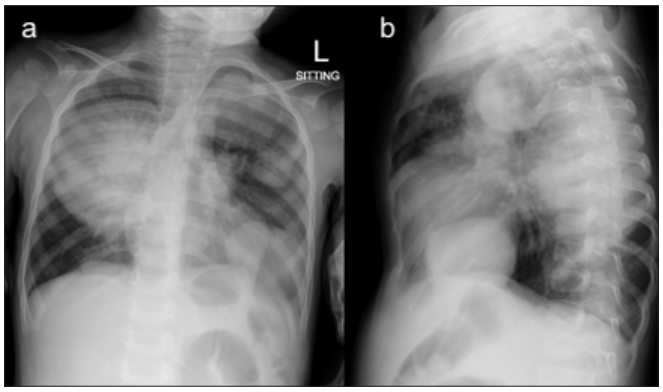

Fig. 1 A and B. Frontal (a) and lateral (b) chest radiographs in a 3-year-old boy demonstrating multiple, bilateral, large, varying-sized oval masses involving the lung parenchyma of the right lower lobe (apical and basal segments) and the left lower and upper lobes. In addition, there is mild cardiomegaly with a displaced cardiac apex.

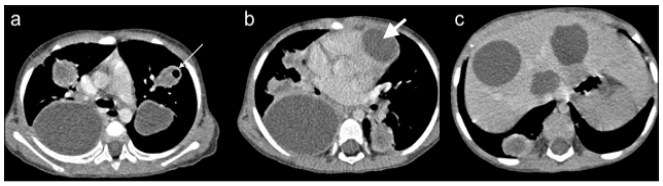

Fig. 2 A-C. Selected slices from axial post-contrast CT-scan of the chest at the level of the (a) carina, (b) the aortic root (c) upper abdomen. Bilateral pulmonary large cysts are confirmed with more smaller lesions, in both upper lobes and the left lower lobe apical segment. The lesions are cystic, and there is no enhancement related to the internal or peripheral aspects of the lesions, in keeping with multifocal lung hydatid disease. (a)There is some gas noted within the non-dependent portion of the left upper lobe lesion suggesting erosion into a bronchus (long arrow). (b) A cardiac hydatid positioned within the left ventricular chamber is demonstrated (short arrow). (c) Multiple, varying-sized, similar appearing, non-enhancing, cystic lesions are noted in the imaged portions of the liver in segments VII, IV and I of the liver, in keeping with hepatic hydatid disease.

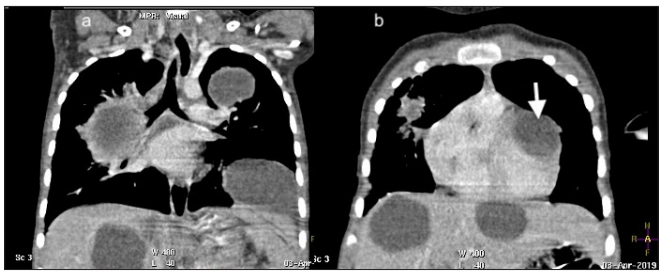

Fig. 3 A and B: Coronal reformat of the CT-chest at the level of the carina (a) and the level of the cardiac chambers (b). The images confirm both the lung parenchymal and hepatic hydatid cysts seen in Figure 2 and also localise the cardiac hydatid in the superior aspect of the left ventricular chamber (arrow) adjacent to the interventricular septum.

The lung is the most common site of paediatric echinococcosis infection, acquired via the lymphatic and haematogenous spread.[2] Pulmonary hydatid cysts (PHC) have a high propensity for the right lung, with the lower lobe accounting for 60% of cases.[3]

PHC vary in diameter from 1 - 20 cm. Children often present with large, rapidly growing cysts owing to the elasticity of their lung parenchyma and their respiratory capacity, which is greater compared with adults.[3] Symptoms may range from asymptomatic to manifestations, such as fever, cough, dyspnoea, haemoptysis and/or acute hypersensitivity reactions, suggesting cyst rupture.[3]

Literature suggests that adult cases of cardiac HC account for 0.5 - 2% of echinococcosis.[3] The most common site for cardiac HC is the left ventricle (LV) accounting for 50 - 60% of reported cases.[3] The propensity for HC cysts to occur in the LV is assumed to be because of its abundant blood supply. The slow growth of cardiac cysts is postulated to be the cause of the variable and uncommon presentation of cardiac HC in adults and paediatrics.[4]

Cardiac HCs are generally asymptomatic but can be fatal owing to their complications. Cysts in the LV may rupture into the pericardium, causing pericardial tamponade, constrictive pericarditis or secondary pericardial cysts.[4]

Cardiac and cardiopulmonary echinococcosis are exceedingly rare, with only case reports of paediatric cardiac and cardiopulmonary HC documented in the literature.

Several complementary investigations are required for the tentative diagnosis of concomitant cardiac and cardiopulmonary echinococcosis. Chest X-ray, though widely available, is non-specific as most cases have a normal cardiothoracic ratio and various lung cysts either oval or circular, with or without a sign of meniscus.[3,4] Transthoracic echocardiography (TTE), CT and magnetic resonance imaging (MRI) remain the mainstays in the diagnosis of cardiac and cardiopulmonary HC, and in the assessment of their relationship to cardiac chambers.[3,4] TTE is accepted as the most effective modality in screening for CE.[4]

Although performed after a chest CT in our case, due to the high complication rate of CE, we advocate that paediatric patients with multiple pulmonary hydatid cysts should be screened for cardiac cysts using TTE.

Limited evidence exists to guide the treatment of CE in children, with only a few randomised trials comparing strategies. Treatment guidelines are largely adult-based, with many originating as far back as 1980. Surgery is considered the mainstay treatment of cardiopulmonary echinococcosis, while medical chemotherapy is touted to have a role perioperatively as adjuvant therapy that provides sterilisation.[4,5]

There are several surgical options available for adults with multiple PHC affecting both lungs, including bilateral simultaneous thoracotomies, bilateral staged thoracotomies separated by a time period, median sternotomy and clamshell incision.[5] However, there is a lack of controlled studies to establish the superior procedure, as each presents some advantages and disadvantages.

When considering surgical management, it is unclear whether pulmonary or cardiac HCs should be removed first.

Future research aimed at understanding the host(s) immunological response to echinococcosis infection, as well as the pharmacodynamics of anthelmintics on intact cysts, could shed insights on the variability of the disease and prove fruitful in discovering novel therapies.

Conclusion

Cardiac and cardiopulmonary echinococcosis are rare conditions in children. Paediatric patients with multiple cysts encompassing one or more organs should be screened for cardiac cyst(s), especially in endemic regions.

Combined surgery and chemotherapy represent an appropriate strategy for managing cardiac and cardiopulmonary echinococcosis. However, the sequence of cardiac or pulmonary cyst removal needs careful consideration.

Given the enormous impact on child health, there is a pressing need for developing a prevention strategy, collaborative research efforts

and establishing a registry for information sharing, particularly in SA and other endemic regions.

Declaration. None.

Acknowledgements. The authors would like to thank the paediatric pulmonology and PICU teams, radiology and cardiothoracic departments at Tygerberg Hospital for their supporting roles in managing these patients.

Author contributions. LM wrote the case presentation and prepared images to be included in the manuscript. PG & LVW reviewed the paper and modified the literature review and discussion section several times. SA reviewed the images and assisted with the interpretation and labelling.

Funding. None.

Data availability statement. The link to the repository cannot be shared as this is a medical file.

Conflicts of interest. None.

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Accepted 31 May 2024.