

Case Report

Childhood Hydatid Disease at the Pietersburg Hospital in Polokwane, Limpopo Province, South Africa: A Case Presentation

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Abstract

Hydatid disease caused *Echinococcus* species is a problem in many countries, especially in the developing world. We report 3 cases of 3, 8 and 10 years old respectively. The first case is a 3 year old girl who presented with cough, chest pains, abdominal pains and difficulty in breathing. On radiological imaging she had extensive bilateral pulmonary cystic lesions, with some liver involvement as well. Serology (indirect haemagglutination) was positive for *Echinococcus* with a titre of 1:1024. The second case involves an 8 year old boy who presented with cough and fever. Radiological imaging showed a single giant pulmonary cystic lesion on the right. *Echinococcus* serology (indirect haemagglutination) was negative, however fluid aspirated from the cyst was found to have *Echinococcus scolices* on histology. The last case is a 10 year old girl who presented with chronic cough associated with vomiting, chest pains, fever and difficulty in breathing. Radiological imaging showed a well-defined pulmonary soft tissue dense lesion. Serology (indirect haemagglutination) was positive for *Echinococcus* with a titre of 1; 1024. All the 3 cases are from rural areas with livestock subsistence farming with dogs kept as pets. Two cases were treated medically with albendazole and responded well. The other case was treated with a combination of albendazole and surgery with good outcome.

Conclusion: A high index of suspicion is required when dealing with cystic lesions in organs, especially for those working in rural areas like ourselves.

INTRODUCTION

Hydatid disease is a disease caused by infestation with the larval stage of *Echinococcus granulosus* and the clinical disease is then characterized by cystic lesions [1,2] Cystic *Echinococcus* is a zoonosis with the highest incidence reported from sheep-rearing countries [2].

There are four species in the genus *Echinococcus*; *E. granulosus*, *E. multilocularis*, *E. vogeli* and *E. oligarthus*; with *E. granulosus* being the most widespread and cosmopolitan of the species [2]. Infestation by this parasite occurs on every continent where domestic animals, the definitive host, occur [3]. Man is an accidental or intermediate host, and we play no role in the

biological cycle. In adults the liver is the commonest site, lung involvement occurs less but can be more common in children [1,3]. Man is infested through ingestion of food contaminated with dog faeces or by direct contact with dogs [2]. The embryos of the organism that are liberated in the duodenum, are transported to the liver by the portal circulation, with the liver and the lungs acting as the primary and secondary filters respectively [2].

Echinococcus is recognized as a clinical problem in Southern Africa, although information on the epidemiology of the disease is not well known [4]. The number of cases in South Africa (SA) is estimated from National Health Laboratory Service (NHLS) data, to be 137 cases per year, in eight of the nine provinces of South Africa [4].

Pietersburg hospital is a tertiary referral hospital for the Limpopo province, a largely rural province of South Africa. The hospital treats many children and adults with hydatid disease and this article describes three such children.

CASE PRESENTATION

Case 1

Patient TM, a 3 year old female, presented to a regional hospital with difficulty in breathing, cough, chest pain, fever, loss of weight, loss of appetite, night sweats on occasions and abdominal pain over a 2 month period. There was no other relevant medical history. Importantly, the family has dogs in the home. She was born and bred in a village where there are cattle, goats and sheep. Her birth history was uneventful and her immunization record was up to date. Her developmental milestones were normal. She was treated for pneumonia at the regional hospital without improvement and was subsequently referred to our tertiary institution for further management.

On examination the child was noted to be underweight for age with a Z-score of <-3. Her respiratory rate was 44 breaths/minute, her pulse rate was 120 beats/min and her peripheral oxygen saturation was 99% on nasal prong oxygen. Her temperature was 37.7°C. She was acutely ill looking and in moderate respiratory distress. There was no evidence of digital clubbing and no overt generalized lymphadenopathy. Examination of her respiratory system revealed alar nasal flaring, intercostals and sub costal recessions and bilateral coarse crackles. On auscultation of her cardiovascular system no abnormalities were revealed. Abdominal palpation revealed a 2-3 cm non-tender hepatomegaly with no splenomegaly. Examination of her central nervous system found no pathology.

On blood investigations human immunodeficiency virus (HIV) infection and tuberculosis were excluded. Hematological testing demonstrated a leucocytosis of $19.2 \times 10^9/L$ (with marked eosinophilia of $3.57 \times 10^9/L$, a microcytic hypo chromic anemia and a thrombocytosis (hemoglobin of 9.9 g/dl, MCV of 65, 2 fland MCH of 18, 4 pg, platelet count of $754 \times 10^9/L$, C-reactive protein (CRP) was noted to be 35 mg/L.

Serological test for *Echinococcus* (indirect haemagglutination) was positive with a titre of 1:1024.

Radiological imaging's of the three cases were as follows:

Radiological imaging of the first patient

Figure 1 of the chest radiograph demonstrates multiple, bilateral, rounded, homogeneous and well defined soft tissue lesions of varying sizes, suggestive of cysts. These lesions are bilateral and diffuse involving mostly the left lung, silhouetting the cardiac shadow and both hemi-diaphragms, with no parenchymal reactions or lymphadenopathy. These radiological features are highly suggestive of pulmonary echinococcosis.

Figure 2 is the subsequent X-ray taken at 3 months post initiation of therapy, demonstrating progressive resolution of these pulmonary lesions.

Figure 3 is a radiograph taken 18 months from the initial radiograph and demonstrates almost complete resolution of the



Figure 1 Chest radiograph of the first patient demonstrates multiple, bilateral, rounded, homogeneous and well defined soft tissue lesions of varying sizes, suggestive of cysts. These lesions are bilateral and diffuse involving mostly the left lung, silhouetting the cardiac shadow and both hemi-diaphragms, with no parenchymal reactions or lymphadenopathy. These radiological features are highly suggestive of pulmonary Echinococcus.

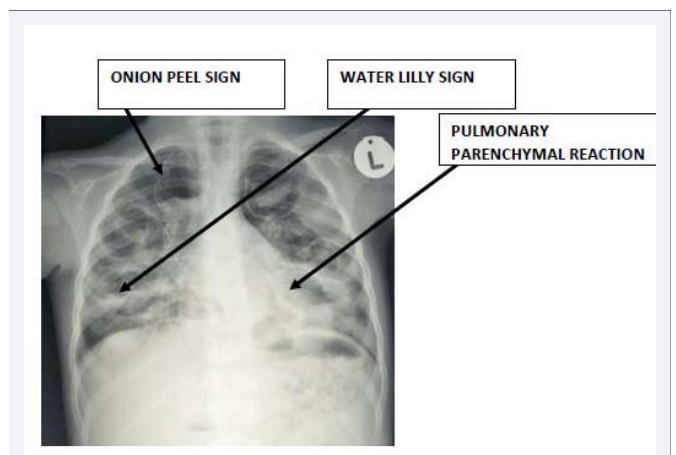


Figure 2 The subsequent X-rays taken at 3 months post initiation of therapy demonstrate progressive resolution of these pulmonary lesions.



Figure 3 Radiograph taken 18 months from the initial radiograph demonstrates almost complete resolution of the disease process with no evidence of cysts. There are, however, residual reticular parenchymal changes.

disease process with no evidence of cysts. There are, however residual reticular parenchymal changes.

The CT scan in Figure 4 confirms the radiographic findings as described in figure 1 above. The largest pulmonary cyst is visualized in the left hemi thorax and measures approximately 8x4 centimeters in size. The features above are in keeping with multi-organ echinococcosis.

Figure 5 is a contrasted computerized axial tomography CT scan demonstrating cystic infiltrations in most segments of the liver.

This child received six cycles of albendazole with 2 weeks drug free intervals in between the cycles. She was successfully weaned off oxygen a week after commencing albendazole and was discharged to continue therapy at home. The cystic lesions completely resolved radiologically.

Case 2

Patient MN is an 8 year old boy referred to us from a regional hospital with 'non-resolving pneumonia'. He presented to the regional hospital with cough and fever of two weeks duration. There was no significant previous medical history of these symptoms. Importantly, however, the family owns dogs. His birth history was uneventful and immunization record was up to date. Growth and development were normal in this boy. He has been staying in a village, where they practice subsistence livestock farming, since birth.

Initial management at regional hospital

An assessment of community acquired pneumonia was made which was treated with a 7 day course of ampicillin and gentamycin. Pulmonary tuberculosis and HIV infection were excluded on further investigations. There was clinical improvement with no radiological improvement. The child was transferred to our tertiary hospital for further management.

Examination findings at our Tertiary hospital

The vital data were normal with a weight of 18.7 kg, height of 121 cm, temperature of 36.8°C respiratory rate of 20 breaths/min, Pulse of 82 beats/min, and Saturation of 98% in room air. There was generalized lymphadenopathy, with no digital clubbing.

Examination of the respiratory system revealed percussion dullness and decreased air entry on the right lung anteriorly, with bilateral coarse crackles. All other systems were normal.

Human immunodeficiency virus was excluded on investigation. Full blood count only showed leucocytosis of $41.69 \times 10^9/L$, with marked neutrophilia of $37.45 \times 10^9/L$. Serology for *Echinococcus granulosus* (indirect haemagglutination) was negative.

Watery fluid with whitish materials was aspirated from the cyst. The chemistry of the fluid was normal with negative bacterial and tuberculosis culture. *Echinococcus granulosus* scolices were however present on histology. Liver function tests were normal.

Chest radiograph in Figure 6 demonstrates a unilateral rounded middle lobe homogeneous and well defined pulmonary soft tissue dense lesion suggestive of a cyst in the lung fields.

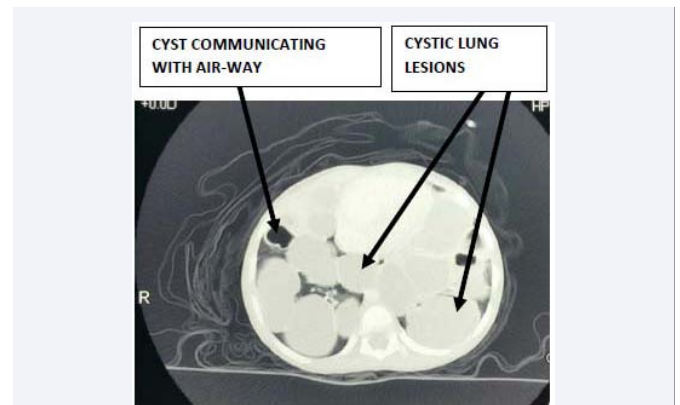


Figure 4 Chest Computed Tomography (CT scan).

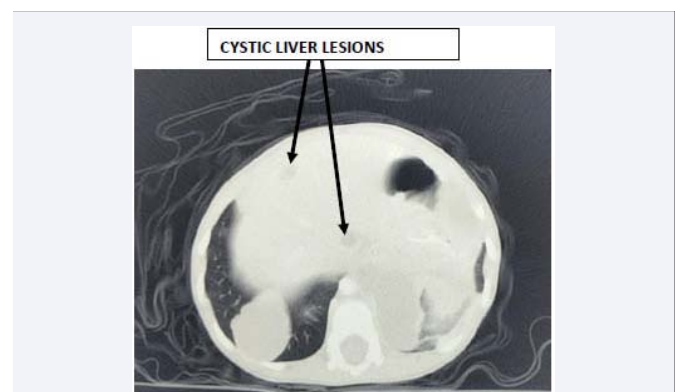


Figure 5 The largest pulmonary cyst is visualised in the left hemi-thorax and measures approximately 8x4 centimeters (cm) in size. The kidneys, pancreas and the spleen are not obviously affected. No ascites or lymphadenopathy is visualised. These features are in keeping with multi-organ Echinococcal infection.

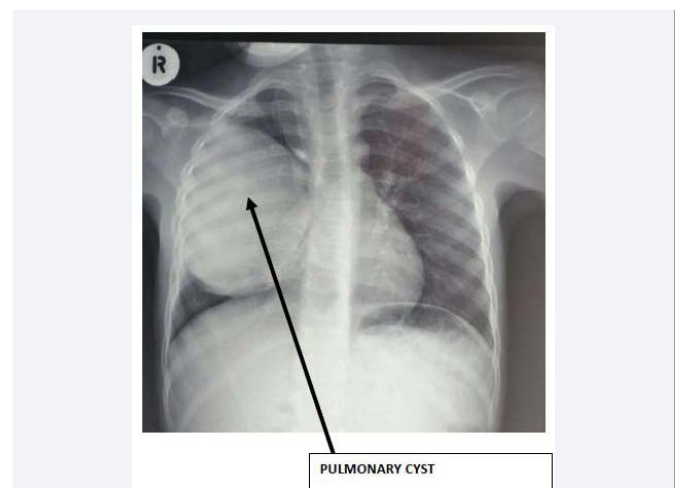


Figure 6 Radiograph on presentation: The lung fields demonstrate a unilateral rounded, middle lobe homogeneous and well-defined pulmonary soft tissue dense lesion suggestive of a cyst. There is associated mass effect on the mediastinum with slight shift to the left and compression of the right main bronchus and bronchus intermedius with associated tapering and/or narrowing and resulting in right lower lobe collapse. These features are in keeping with pulmonary Echinococcal infection.

There is a mass effect on the mediastinum with slight shift to the left and compression of the right main bronchus and bronchus intermedius, with associated tapering and resulting in right lower lobe collapse. These features are in keeping with pulmonary echinococcal infection.

Figure 7 is a subsequent chest radiograph taken seven months post the initial radiograph and post aspiration, demonstrating a sharp fluid level in this cyst with smooth thick walls and no obvious change in size.

Figure 8 is a post-surgical radiograph which demonstrates successful excision of the cyst and resultant lung contusions. Intercostals drain is in situ with no obvious rib fractures

Figure 9 is a contrasted CT of the chest which confirms the radiographic findings described in the first chest X-ray, locating the cyst in the lateral segment of the middle lobe and collapse of the segment. The cyst is measuring approximately 15x10 centimeters largest diameters with an onion peel sign on its lateral wall (detaching membrane) in keeping with pulmonary echinococcal infection.

An assessment of pulmonary hydatidosis was made and albendazole was commenced. During the course of treatment the child developed nosocomial sepsis with non-responding temperature spikes, which warranted thoracotomy and cystectomy. Post-surgery the temperature settled and the child was discharged to home with albendazole.

Case 3

MH is a 10 year old girl referred to us from Lebowakgomo, one of our district hospitals. The child presented with a chronic cough for a month which was associated with vomiting, chest pains, fever and difficulty in breathing. The birth history was uneventful and the developmental milestones were normal. The child was brought up in rural settings where there is livestock farming. The family also keeps dogs which the child used to play with. The child is from a low income family.

The child was admitted with a diagnosis of pneumonia requiring nasal prong oxygen and intravenous antibiotics.

The chest radiograph showed a well circumscribed round lesion not responding to antibiotic therapy and warranted referral for further management.

The following were findings on arrival at our hospital: Clinically respiratory symptoms had resolved and no supplemental oxygen was required.

Investigations done at our hospital revealed a positive HIV ELISA and negative TB tests. Serology for *Echinococcus granulosus* was positive by indirect haemagglutination with a titre of 1:1024.

In both Figures 10,11 a unilateral oval, homogeneous, well-defined pulmonary soft tissue dense lesion in the lateral segment of the middle lobe is visualized suggestive of pulmonary echinococcosis. No obvious parenchyma reaction is surrounding the lesion. The rest of the lung-fields are grossly intact.

The child was treated with four 28 days cycles of albendazole with a two weeks drug free interval between the cycles. She was also started on antiretroviral therapy. She responded well



Figure 7 Subsequent Follow up Chest Radiograph: Subsequent chest radiograph taken seven months post the initial radiograph and post aspiration demonstrates a sharp fluid level in this cyst with smooth thick walls and no obvious change in size.



Figure 8 Post-surgical radiograph demonstrates successful excision of the cyst and resultant lung contusions. Intercostals drain is in situ with no obvious rib fractures.

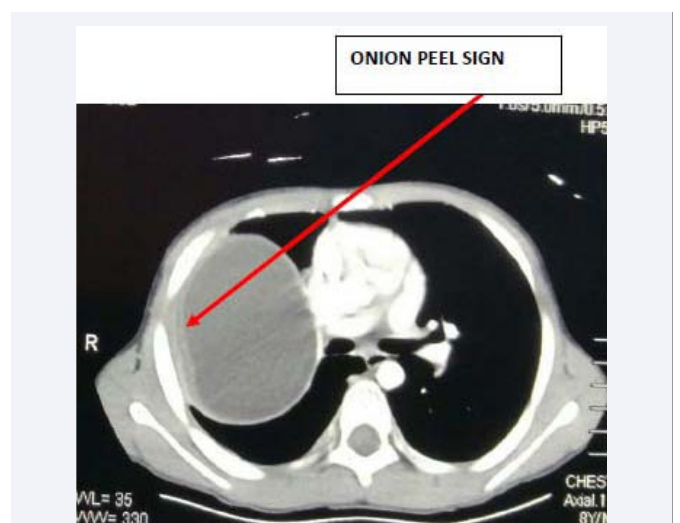


Figure 9 Contrast CT scan of the Chest.



Figure 10 The radiological features confirm the radiographic findings described for the chest X-Rays, locating the cyst in the lateral segment of the middle lobe and collapse of the segment. The cyst is measuring approximately 15x10 largest diameters with an onion peel sign on its lateral wall (Detaching membrane) in keeping with pulmonary Echinococcal infection.



Figure 11 Chest Radiograph.

to the treatment and is still being followed up in our pediatric pulmonology clinic.

DISCUSSION

Our three pediatric cases highlight that the hydatid disease should also be considered as a differential diagnosis in cystic and solid lesions of the lungs in our rural settings. Our first case presented with multiple lung and liver cystic lesions on imaging. The diagnosis was confirmed by serology (indirect haemagglutination) test.

Our second case demonstrated a unilateral giant cyst, defined as lung hydatid cyst measuring more than 10cm in the largest diameter [1]. The diagnosis was confirmed by the presence of scoleces in the cyst aspirate. There is no collective agreement on the precise definition of a giant pulmonary hydatid cyst size in literature; however the size of 10cm in the largest diameter

is used by consensus [1]. Our third case was diagnosed by chest radiograph and serology. The cyst showed a marked decrease in size with medical treatment. However our first chest radiographs was misfiled and hence they were not included in the pictures in this case report.

The areas from which our 3 patients come are rural, where they mostly practice subsistence farming in maize, cattle, goats and sheep. They also keep domestic dogs as pets. According to literature *E. granulosus* cysts are acquired in childhood, although they present in the 3rd and 4th decade [3]. Clinical symptoms normally occur some years after infection because the hydatid cyst grows slowly [1]. Our first case was failing to thrive as her weight fell below-3 standard deviation on the Z-score. This could explain early presentation due to rapid growth of cysts in malnourished and therefore immunosuppressed patients [5]. In case two the patient had a giant cyst at the age of eight years despite normal nutritional and immune status. This could be explained by the fact that the lung tissue in children is highly elastic and the rate of growth of the cyst in children is much higher ranging from a few millimeters to 5 centimeters per year [1].

Hydatid disease is endemic in cattle-grazing areas where there is dominance of sheep-dog cycle [5]. This disease has also been known since the time of Hippocrates [6]. Apart from lung and liver involvement, there are case reports of other organ involvement such as the heart, spine, orbit of the eyes, peritoneum and thigh muscles [6].

If the hydatid cysts are not treated, they may progressively grow and result in life-threatening complications, especially if they rupture as this may result in anaphylaxis [5,7]. There have been instances where spontaneous collapse, regression and a dead calcified cyst have been noted [5].

The clinical presentation of this disease is protean depending on which organ is involved. The clinical symptoms and signs differ according to the size, age and location of the cyst [7]. Pulmonary hydatid disease has predilection for the left lung although it may involve any part of the lung [8]. It may present with cough, purulent sputum, and fever and chest pain. In some cases haemoptysis and allergic reaction can occur [5]. The cyst may rarely rupture onto the bronchial tree and cause hydatidoptysis [5]. The diagnosis of hydatid disease is made complex by multiple presentations and the fact that the cyst might be an accidental discovery. Consideration of the condition is one of the keys to the diagnosis [9]. Clinical presentation depends on the organ involved, knowledge of the patient's background and the possibility of exposure to *Echinococcus species*. Clinical findings on examination may be nonspecific and related to the effect of the cyst on the anatomy and function of affected organs [9]. Unlike the clinical presentation, radiological signs are clear [10], as demonstrated by reports in our 3 cases. Chest radiographs, sonar and contrast CT of the chest are very useful in the diagnosis [8].

Laboratory results may be non-specific. Elevated liver enzymes may suggest involvement of the liver, with leucocytosis suggesting infection of the cyst. Eosinophilia may be present in 25% of all infected patients [9].

Percutaneous drainage of the cyst may be helpful in the diagnosis as happened in our second case where scoleces were

identified. It is however important to avoid spillage of the aspirated fluid onto the patient's tissues as this might lead to anaphylaxis and may also cause other organ involvement.

ELISA, indirect haemagglutination (IHA) and fluorescent antibody tests are helpful, although they may be negative in 10-50% of patients with active cystic hydatid disease [9]. Serological tests using indirect haemagglutination assay (IHA) is mainly used for *Echinococcus* serology in our public sector hospitals. Titers of 1:32-1:128 are regarded as low positive and non-specific. Titers above these are regarded as strong positive, like in two of our patients [4]. In the Western Cape province of South Africa, an enzyme immunoassay (ELISA) which is a qualitative test reported either as positive or negative is used [4]. The serological tests may have some limitations in monitoring patients after surgery, percutaneous drainage or drug therapy, with the antibody titers starting to fall at 3 months after therapy and becoming negative in a period of 12-24 months [9].

Treatment of hydatid disease includes both medical and surgical interventions. Surgery however remains the mainstay of treatment which offers hope for complete cure [9]. The other treatment modalities include chemotherapy or a combination of surgery and chemotherapy [9]. Hydatid cyst drainage is performed using a procedure popularly known as PAIR (puncture, aspiration, instillation of scolicalid agent, reaspiration) which is a technique that has recently gained acceptance [11]. Chemotherapy is effective in small thin-walled cysts <4cm in diameter and in younger patients [11]. Primary prevention of hydatid disease can be done by educating people, treatment and control of all dogs and proper hand washing after contact with dogs or their faeces [5]. If the cyst is not amenable to complete excision, fluid aspiration in conjunction with injection of scolicalid agent into the cyst is done [5]. Anthelmintic treatment with benzimidazoles is reserved for inoperable cases, poor surgical candidates, disseminated and recurrent disease [5]. Recommendations are that albendazole should be given in cycles of 4 weeks with a 2 week drug-free interval [12]. This is because albendazole partially inhibits microsomal function while inducing its own [12]. Duration of treatment with chemotherapy should be individualized for each patient [12]. There is some evidence to support a role of praziquantel in combination with Albendazole in pre and post-intervention chemotherapy for hydatid disease [6].

CONCLUSION

We have presented three pediatric cases with cystic *Echinococcus* disease. The first patient presented with multiple lung cysts as well as liver involvement. The second patient presented with one giant cyst and the third patient presented

with a small round pneumonia-like lesion. One of our cases was diagnosed in 2014 and the other two in 2015. We have a few adult patients who have been treated by our cardiothoracic surgeons that have not been reported. The cases that reach us from the periphery are a tip of the iceberg, as we expect more cases because Limpopo is mainly a rural province. We need a high index of suspicion and the use of diagnostic tools, especially imaging in our province to make an early diagnosis. This will lead to early management of the disease and prevention of complications [9]. The combination of medical and surgical intervention needs to be used in endemic areas [9], although in some cases it might not be possible to resect multiple lesions.

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