

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Medical Imaging Quiz – Case 38

A 5-year-old girl presented to the Emergency Department due to fever for two days, dyspnea and thoracic pain. Her parents revealed that the girl had dyspnea for at least three weeks. From her medical history, they referred only one trip 6 months ago to a rural area. Medical examination revealed mild dehydration and severe respiratory distress (SpO₂ 90%), with reduced auscultatory sounds over the left lung. Laboratory investigation showed elevated total leukocytes count of 27.3 cells/ μ L (neutrophils 70%, eosinophils 9%) and increased C reactive protein of 90 mg/L (normal: <5). Chest x-ray revealed a well-defined large cystic lesion involving the left upper lung zone. She immediately started antimicrobial therapy and underwent a computed tomography (CT) scan that revealed a round well circumscribed fluid filled cystic lesion suggestive of the diagnosis (fig. 1). Albendazole was added to her treatment protocol. Serological examination for *Echinococcus* was negative.

Comment

Invasion of the human lungs by the larvae of the dog tapeworm Echinococcus granulosus (pulmonary cystic echinococcosis, PCE) is an incapacitating disease, frequently found across a wide geographic range involving Europe, Africa, America, Asia, Australia, and Europe.

In adult humans, cysts occur more frequently in the liver, followed by the lungs. On the other hand, in children, lung is the predominating site for cyst detection apparently by allowing faster growth of the cyst due to its compressible nature, vascularization, and negative pressure. The majority of lung cysts are located to the right lung and are primary cyst formed by a filled cavity and comprise three

layers: The pericyst, of host origin and consisting of compressed lung tissue with an associated inflammatory host reaction evoked by the parasite and fibrosis; ectocyst (laminated layer, or hyaline membrane); and endocyst (germinal layer). Protoescolices located on the inner surface of the germinal layer deposit as a sediment (hydatid sand) or form daughter cysts. When cysts rupture, either by spontaneous trauma or during medical intervention, protoscolices contained in the cyst fluid may disseminate, developing secondary cysts in the surrounding tissues. A high proportion of lung cases may be discovered incidentally on a routine x-ray evaluation. Apparently most individuals harboring small lung cysts often remain asymptomatic five to twenty years after infection until the cyst enlarges sufficiently to cause symptoms.

Most symptoms of pulmonary cystic echinococcosis are caused by mass effect from the cyst volume, which exerts pressure on the surrounding tissues. The most common symptoms described by the literature include: Cough (53–62%), chest pain (49–91%), dyspnea (10–70%) and hemoptysis (12–21%). Other symptoms described less frequently include dyspnea, malaise, nausea, as well

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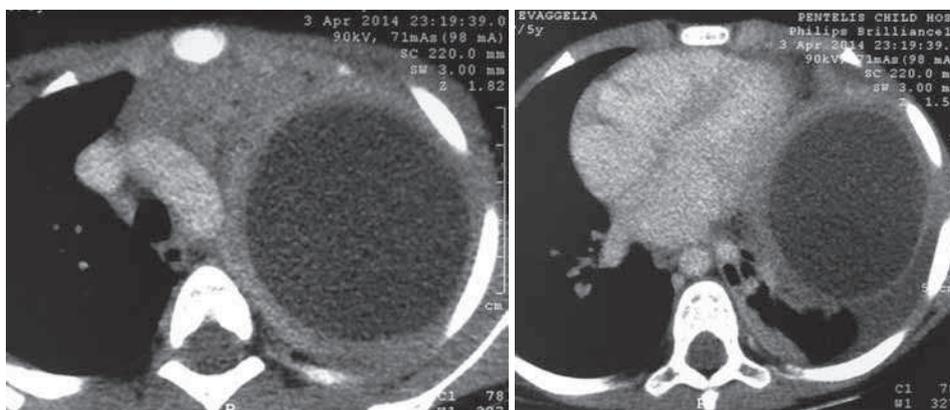


Figure 1. Round well circumscribed fluid filled cystic lesion of the upper left lobe.

as vomiting and thoracic deformations. The majority of children and adolescents with lung lesions are asymptomatic despite having lesions of impressive size, assumedly because of a weaker immune response and the relatively higher elasticity of the lung parenchyma in children and teenagers.

Cysts can break or become infected with an aggregated bacterial infection. The presence of any of these complications changes the clinical presentation, either causing new symptoms or increasing the severity of those already existent. The principal complication is cyst rupture. Lung cysts may break and cyst material containing fragments of larval tissue and protoscolexes spilled from the ruptured cyst may flow either into bronchial tree producing cough, chest pain, hemoptysis, or vomica; or into the pleural cavity, causing simple or tension pneumothorax, pleural effusion, or empyema. Another important complication is the infection of the cyst, manifesting as a pulmonary abscess with poorly defined margins.

The primary diagnosis is obtained by imaging, with the role of serology mostly limited to case confirmation. Radiological studies are the primary step in the detection and evaluation of pulmonary CE cysts. By reasons of cost and availability, chest x-rays are still the most used examination. In chest x-rays, cyst are well defined as a rounded mass of uniform density that occupies a part of one or of both hemithorax. When a cyst is broken, endocyst detachment is seen as floating membranes within the cyst. While CT is not required to establish a presumptive diagnosis, it better recognizes certain details of the lesions and their surrounding structures, helping to exclude alternative differential diagnoses and can also uncover additional smaller cysts that were not detected by conventional chest x-ray. The better imaging definition of CT is particularly useful in the case of complicated cysts, for example to identify a cyst wall defect in a ruptured cyst. Infected cysts show in CT as poorly defined masses with an increased internal density and contrast enhancement around the cyst wall (ring enhancement sign) after the injection of contrast substance.

The use of ultrasound in lung lesions is quite limited. It may provide good imaging in lesions close to the thoracic wall. Most importantly, however, ultrasound examination of the liver may reveal concomitant liver involvement in up to 15% of individuals with lung CE.

Immunodiagnostic tests are used to support the clinical diagnosis of CE. The principal factor related to a positive serology is the presence or absence of complications (rupture and infection/abscess) because of the release of parasite antigens. Most of the literature describes lower sensitivity of serology in lung CE compared to liver CE, but more recent studies with more sensitive assays seem to find similar proportions of seropositive individuals.

Most patients with lung CE come to be treated after many years of infection and close to 50% of them present with complications, mainly infection. Surgery is the main therapeutic approach. Surgical treatment of CE has two goals: (a) To safely remove the parasite, preventing intraoperative dissemination and (b) to treat the bronchopercyst pathology and other associated lesions.

Surgery may involve excision of the cyst or resection of the cyst and the immediate surrounding parenchyma. Despite the lack of consensus, the currently most accepted surgical treatment for lung CE is complete excision using parenchyma-preserving methods, such as cystostomy, intact cyst enucleation or removal after needle aspiration preserving as much lung parenchyma as possible. World Health Organization (WHO) guidelines for hydatid disease state that chemotherapy using benzimidazoles is the preferred treatment when surgery is not available, or complete removal is not feasible. Medical treatment can result in reduction of the cyst size. Long courses (several months) of albendazole is somewhat efficacious for pulmonary cysts.

About the prognosis using surgical treatment, it has changed during the last few years, and results are currently commonly satisfactory. The most frequent complications are pleural infection and prolonged air leakage, whereas operative mortality rate does not exceed 1% to 2%.

Recurrence of lung CE is observed in those cases where cyst content was spillage during the surgery. To reduce the number of recurrent cases, it is recommendable the adequate use of scolicalid agents during cyst evacuation in all CE cases, and the concomitant use (pretreatment or post treatment) of benzimidazole compounds in complicated/giant cases.

References

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Diagnosis: Infected pulmonary cystic echinococcosis