Mediastinal hydatidosis with an unusual presentation

A Rare Case Report

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Abstract

Hydatidosis is a common zoonotic disease with high prevalence in developing countries. While solitary cyst with unilateral lung involvement is common, bilateral involvement and multiple cysts are rare, seen in 20% and 30% of the cases. Likewise, extensive involvement of extrapulmonary tissues and mediastinum is rare. We report an unusual case of mediastinal hydatidosis mimicking an intrathoracic malignancy in a 24-year-old female. She presented with a history of left-sided chest pain and heaviness in the left hemithorax for a period of two months. Diffuse, multiple fluid-filled cystic lesion with internal echoes throughout the mediastinum, lung, pericardium, diaphragm and chestwall were observed in contrast-enhanced computed tomography of the thorax. An incidental cystic lesion in liver was noted. Since serology for echinococcosis was negative, a differential diagnosis of intrathoracic malignancy was considered. However, intraoperative and histopathologic findings were suggestive of hydatidosis.

Keywords: Hydatid cyst; intrathoracic malignancy; computed tomography; magnetic resonance imaging; mediastinum
Introduction

Hydatid disease caused by larval stage of cestode echinococcus is one of the major zoonotic diseases of public health significance with a reported mortality rate of 2-4%.\(^1\) Although lungs are the second most commonly involved organ, intrathoracic extrapulmonary involvement is uncommon, seen in 5-7% of the cases.\(^1\)\(^,\)\(^2\) Hereby we report a rare presentation of mediastinal hydatidosis with extensive involvement resembling malignancy.

Case report

Hemodynamically stable, healthy 24-year-old female with no significant medical history presented to our Department with heaviness and left-sided chest pain for two months. Respiratory symptoms including fever, breathlessness, cough, hemoptysis, dysphagia, or dysphonia were absent. Besides, weight or appetite were unaffected. Decreased left sided chest movement, deviation of trachea to right side, dullness over the left hemithorax and decreased vocal fremitus were observed during chest examination. Also, diminished breath sounds with decreased vocal resonance was noted on left side. Routine blood investigations, liver and renal function tests were within standard limits. While, the pulmonary function test showed a restrictive pattern and sinus tachycardia was noted on the electrocardiogram, the findings on echocardiogram were normal. The chest x-ray posterior-anterior view showed opacity on the left hemithorax (Figure 1). Contrast-enhanced computed tomography (CECT) of the thorax showed multiple fluid-filled cystic lesion throughout the mediastinum as well as within the lung parenchyma with atelectasis, largest measuring 10x7cms (Figure 2a, 2b, 2c). An incidental hepatic cyst measuring 4x3cms was also noted (Figure 2). Fiber optic bronchoscopy showed a deviated trachea with compressed left upper lobe bronchus and no endobronchial growth. Although the findings were suspicious of hydatid cyst, IgG titers for hydatid serology was negative and needle aspiration was inconclusive. Hence, differential diagnosis of various intrathoracic malignancy were considered.

She underwent left posterolateral thoracotomy under general anesthesia, double-lumen intubation. Intraoperatively glistening white fluid filled cystic lesions resembling hydatid cyst were present throughout the lung parenchyma and mediastinum, with dense vascular adhesions between cyst wall and thoracic cage. Parenchymal preserving cyst excision of the lung
parenchyma with captionage and cystectomy for the cysts in the mediastinum, diaphragm, pericardium, and the chest wall were performed (Figure 4a). Histopathology was suggestive of multiple daughter cysts with germinal layer and scolices suggestive of hydatid disease (Figure 4b). Postoperative course was uneventful and patient was prescribed albendazole 400 mg twice daily for a period of three months. Postoperative chest X-ray as well as computed tomography (CT) chest after three months of follow up did not show any recurrence (Figure 3a, 3b, 3c, 3d). She underwent laparotomy and cyst excision after 6 months for a hepatic cyst. Inform consent was obtained from the patient for the publication purpose.

**Discussion**

Hydatidosis is a zoonotic disease transmitted via feco-oral route to humans from the animal hosts including dogs and sheep. Echinococcus granulosus is the most common source of infection accounting for 95% of 2-3 million cases reported globally. The prevalence of infection varies widely by region, with higher prevalence in Mediterranean regions. Following liver, lungs are the second common organ involved with higher predilection for right lower lobe. However, in our case extensive involvement of left side was present. Often, the cysts are solitary and unilateral in distribution, while bilateral involvement and multiple cysts are not unusual, seen in 20% and 30% of cases respectively. Moreover, extensive mediastinal cystic involvement is rare, seen in less than 4% of the cases. The presenting symptom depends on the size, location of the cyst and degree of compression of the mediastinal structure. Vertebral destruction, superior venacava syndrome and Bernard Horner’s syndrome have been reported previously with mediastinal cysts. Apart from the chest pain and heaviness of chest, no other associated symptoms were present in our patient.

Considering the age, presence of diffuse involvement of lung parenchyma and opacified left hemithorax on CECT thorax and chest x-ray and negative serology for hydatid cyst, a wide variety of differential diagnosis were considered including mediastinal hydatidosis, germ cell tumor, pulmonary sarcoma and diffuse pulmonary metastases. High attenuation wall and a low-density content are characteristic of Hydatidosis. Germ cell tumors constitute 1-3% of intrathoracic malignancies affecting males with mean age of 25-35 years. Lobulated heterogeneous mass containing soft tissue elements with fluid and fat has been seen with
immature teratoma with calcification in 20-40% of the cases. On the other hand, pulmonary synovial sarcoma characterized by well-circumscribed heterogeneous mass occurs between 16 to 77 years of life with equal sex predilection. Although well circumscribed, rounded soft-tissue attenuation noted with diffuse pulmonary metastases is reported in young patients was considered one among the differential diagnosis, however is unlikely in our case as the patient did not have any symptoms of metastases or malignant disease.

Although, magnetic resonance imaging (MRI) is superior in differentiating between solid and cystic lesions, its role in hydatidosis is very minimal. They appear as low signal intensity on T1 and high signal intensity on T2 weighted images. Similarly, cystic components of teratoma results in high signal intensity on T2 weighted images and low signal intensity on T1 weighted images with magnetic susceptibility artifacts in the presence of calcification. Seminomatous germ cell tumors have a hypointense mass on T2 weighted images with relatively homogeneous enhancement. In pulmonary synovial sarcoma, there is a heterogeneous signal intensity on T1 weighted images while in pulmonary metastases, diffusion-weighted images show high signal intensity. Considering the feasibility MRI was not suggestive. In our case, considering the above mentioned differential diagnosis, posterolateral thoracotomy of left side was planned. Intra-operatively, glistening white fluid filled cystic lesions were noted suggestive of hydatid disease which was later confirmed by histopathology.

Surgery is the most accepted treatment of choice for hydatid disease, however in patients with recurrent cysts, multiorgan disease, poor general conditions and those who refuse to undergo surgery, medical management is the next best option. Albendazole (10-15 mg/kg/day) is the drug of choice in hydatid disease. It inhibits microtubular assembly within the parasite resulting in glycogen depletion and finally autolysis of cell. Preoperatively albendazole intake helps to soften the cyst and reduce the intracystic pressure, while postoperatively it prevents recurrence of the disease. In our case albendazole was prescribed for 3 months and in follow up no recurrence was observed.
Conclusion
Due to its variable presentation, diagnosis based only on clinical and radiographic findings of mediastinal hydatidosis can be challenging. Although surgical excision remains the treatment of choice without extensive resection, however, a thorough understanding of all the radiographic differential diagnosis while encountering such an extensive disease of the mediastinum is vital to devise a treatment plan.

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References


**Figure 1:** Chest X-ray PA view showing left opacified hemithorax with contralateral tracheal deviation
**Figure 2a:** CECT thorax coronal view shows multiple cystic lesions with thickened septa

**Figure 2b:** CECT thorax axial view shows interconnected cystic lesions with mediastinal shift to the opposite side
Figure 2c: Axial view of upper lobes with lung window shows consolidation of the left lung with minimally visible parenchymal markings

Figure 3a: Immediate postoperative chest X-ray
**Figure 3b:** Postoperative chest X-ray PA view after 3 months shows completely resolving cystic lesion

**Figure 3c:** Postoperative CT thorax at the level of carina shows no recurrent cystic lesions
Figure 3d: Postoperative CT thorax below the level of carina shows no recurrent cystic lesions

Figure 4a: Innumerable hydatid cysts with daughter cysts with typical glistening white appearance
**Figure 4b:** Histopathology (hematoxylin and eosin staining) shows laminated and nucleated germinal layer giving rise to brood capsule. Protoscolices are seen within brood capsule.