CARDIOLOGIE TUNISIENNE

CASE REPORT

Unusual etiology of spontaneous pneumothorax: A case report of primary heterotopic pleural hydatidosis

Étiologie inhabituelle du pneumothorax spontané : A propos d'un cas d'hydatidose pleurale hétérotopique primaire

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Summary

Introduction : Primary heterotopic pleural hydatidosis (PHPH) is a rare condition where an intact hydatid cyst grows in the pleural cavity after the rupture of its pericyst in the lung. This distinguishes PHPH from secondary pleural hydatidosis, which results from cysts rupturing in other locations like the liver or diaphragm. PHPH often presents with nonspecific symptoms, making diagnosis challenging.

Presentation of Case: A 28-year-old woman from a rural area presented with dyspnea and right basithoracic pain. Imaging revealed a right-sided hydropneumothorax and a ruptured hydatid cyst in the right upper lobe, with an intact cyst in the pleural cavity. Surgical intervention via posterolateral thoracotomy was performed, involving extraction of the intact cyst and capitonnage of the residual cavity. Post-operatively, the patient was managed with albendazole (400 mg twice daily) for 6 months, and follow-up imaging showed no recurrence.

Conclusion: This case highlights the importance of considering PHPH in the differential diagnosis of spontaneous pneumothorax in endemic regions. Early diagnosis, surgical intervention, and post-operative anti-helminthic therapy are key to successful management and prevention of complications.

Keywords

Hydatid cyst; Pleura; Lung; Imaging; Surgery

Résumé

Introduction : L'hydatidose pleurale hétérotopique primitive est une affection rare, caractérisée par la croissance d'un kyste hydatique intact dans la cavité pleurale après la rupture du péri-kyste pulmonaire. Ceci la distingue de l'hydatidose pleurale secondaire, qui résulte de la rupture de kystes à d'autres endroits, comme le foie ou le diaphragme. Elle se manifeste souvent par des symptômes non spécifiques, ce qui rend le diagnostic difficile.

Cas clinique : Une femme de 28 ans, d'origine rurale, a consulté pour dyspnée et douleurs basi-thoraciques droites. L'imagerie a révélé un hydro-pneumothorax droit et une rupture de kyste hydatique du lobe supérieur droit, avec un kyste intact dans la cavité pleurale. Elle a été opérée par thoracotomie postéro-latérale. Elle a eu une extraction du kyste et un capitonnage de la cavité résiduelle. En post-opératoire, la patiente a été traitée par albendazole pendant 6 mois, et l'imagerie de suivi n'a montré aucune récidive.

Conclusion : Ce cas clinique souligne l'importance de différencier l'hydatidose pleurale hétérotpique primitive du pneumothorax spontané en région d'endémie. Un diagnostic précoce, une intervention chirurgicale et un traitement anti-helminthique post-opératoire sont essentiels pour une prise en charge efficace, et une prévention des complications.

Mots-clés

Kyste hydatique, plèvre, poumon, imagerie, chirurgie.

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INTRODUCTION

Hydatid disease, caused by Echinococcus granulosus, is endemic in many regions, particularly those where sheep and cattle are raised, such as the Mediterranean countries (1). The lung is the second most common site of hydatid cysts after the liver (2). While secondary spontaneous pneumothorax due to ruptured pulmonary hydatid cysts is not uncommon, primary heterotopic pleural hydatidosis (PHPH) is an exceptionally rare condition. PHPH occurs when an intact hydatid cyst grows in the pleural cavity following the rupture of its pericyst in the lung, releasing the hydatid vesicle into the pleural space (3). This case report describes a rare instance of PHPH and emphasizes the importance of early diagnosis and surgical intervention. This work has been reported in line with the SCARE criteria (4).

PRESENTATION OF CASE

A 28-year-old woman of rural origin, with no significant medical history, presented to the emergency department with a 3-day history of dyspnea and right basithoracic pain. Initial outpatient treatment with analgesics provided no relief. On admission, clinical examination revealed superficial polypnea, with a pulse of 120 beats per minute, blood pressure of 110/70 mmHg, temperature of 37.2°C, respiratory rate of 22 breaths per minute, and oxygen saturation of 93-94% on room air. Respiratory examination revealed reduced chest expansion on the right side, decreased tactile fremitus over the right hemithorax, dullness on percussion over the right lower lung field, and decreased vesicular murmurs with absent breath sounds at the right base, consistent with hydropneumothorax. Abdominal examination was unremarkable. Laboratory tests showed an elevated C-reactive protein (CRP) level of 167.7 mg/L and mild leukocytosis (white blood cell count: 12,000/mm^3). Liver function tests (LFT) and renal function tests (RFT) were normal. Chest X-ray revealed a right-sided hydropneumothorax with a well-defined cavity in the right upper lobe. A chest CT scan (Figure 1) confirmed a hydatid cyst in the ventral segment of the right upper lobe, complicated by rupture into the pleural cavity. The cyst appeared as a well-defined, fluid-filled lesion without enhancement after contrast injection. No other hydatid cysts were identified in the liver, diaphragm, or other regions, confirming the diagnosis of primary heterotopic pleural hydatidosis.

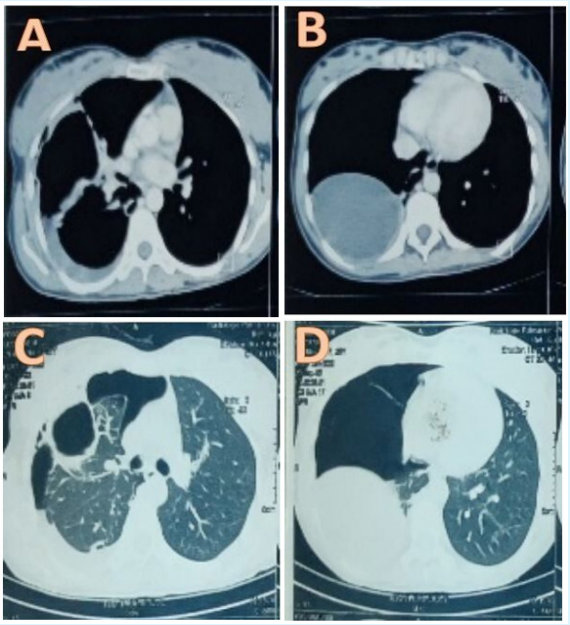

Figure 1. A. Mediastinal window of the chest CT scan showing the residual cavity of the primary heteropic pleural hydatid cyst.

B. Mediastinal window of the chest CT scan showing the primary heteropic pleural hydatid cyst.

C. Parenchymal window of the chest CT scan showing the residual cavity of the primary heteropic pleural hydatid cyst.

D. Parenchymal window of the chest CT scan showing the primary heteropic pleural hydatid cyst.

The patient was initially managed with oxygen therapy and analgesics (paracetamol 1 g every 8 hours) for pain relief. A chest tube was placed on the right side to manage the hydropneumothorax. The chest tube remained in place for 2 days, during which the patient was closely monitored for signs of infection or further complications. The chest tube was removed once the pneumothorax resolved, and the patient was stabilized for surgery.

Surgical exploration via posterolateral thoracotomy revealed an intact hydatid cyst in the pleural space (Figure 2) and a residual cavity in the right upper lobe. The surgical field was protected using compresses soaked in hypertonic saline to prevent spillage and potential anaphylactic shock. The cyst was carefully extracted intact (Figure 3), and capitonnage of the residual cavity was performed. The surgical site was thoroughly irrigated with betadine solution to minimize the risk of infection. The patient was closely monitored for any signs of anaphylaxis during the procedure.

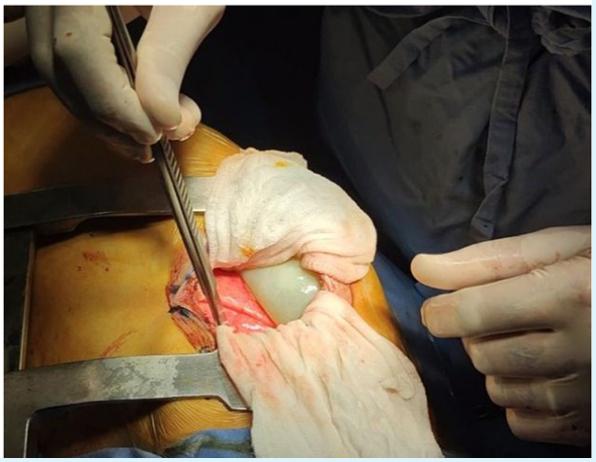

Figure 2. A posterolateral thoracotomy revealing an intact hydatid cyst in the pleural space.

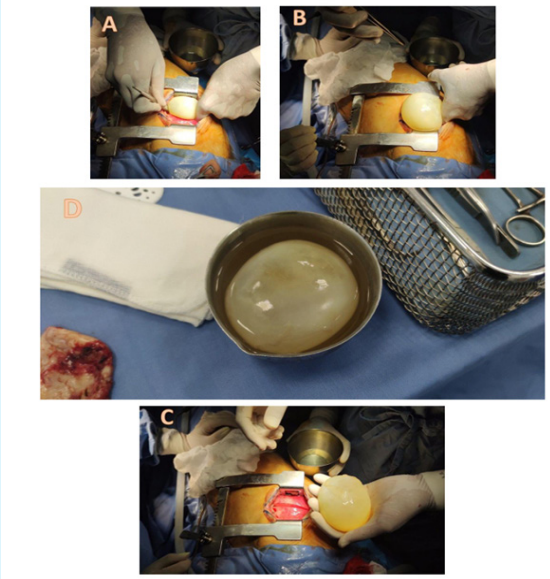

Figure 2. Steps showing the extraction of the hydatid cyst intact from A to D.

Postoperatively, the patient was started on albendazole (400 mg twice daily) to prevent recurrence of the hydatid disease. The anti-helminthic treatment was continued for 6 months. The patient was discharged on the third postoperative day after an uneventful recovery. Follow-up chest X-rays (PA view) were performed at 1,3 and 6 months post-surgery, showing no signs of recurrence or complications. The patient remained asymptomatic during the follow-up period.

DISCUSSION

Primary heterotopic pleural hydatidosis (PHPH) is an exceptionally rare condition, even in regions where hydatid disease is endemic. PHPH occurs when the pericyst of a pulmonary hydatid cyst ruptures, releasing the hydatid vesicle into the pleural cavity, leading to the formation of an intact hydatid cyst in the pleural space (3). This distinguishes PHPH from secondary pleural hydatidosis, which arises from the rupture of cysts in other locations, such as the liver or diaphragm (5). In this case, the absence of hydatid cysts in other regions, such as the liver or diaphragm, confirmed the diagnosis of PHPH (6).

Given the rarity of PHPH, only a few cases have been reported in the literature. Table 1 summarizes the clinical presentation, imaging findings, surgical management, and outcomes of reported cases of primary pleural hydatidosis. These cases highlight the variability in clinical presentation, ranging from pneumothorax to pleural effusion, and underscore the importance of early diagnosis and surgical intervention (7, 8).

Table 1. Summary of Reported Cases of Primary Pleural Hydatidosis

Study	Age/Gender	Clinical Presentation	Imaging Findings	Surgical Management	Outcome
Rakower & Milwidsky (9)	not available	not available	Imaging: Heterotopic and primary origin	Cyst removal, capitonnage	not available
Bouassida et al. (2021) (10)	30/Female	Pneumothorax	CT: 11 cm cyst in left lobe	Cyst removal, capitonnage	No recurrence at 6 months
Belliraj et al. (2019) (11)	28/Female	Dyspnea, chest pain	CT: Hydropneumothorax	Cyst extraction, irrigation	Asymptomatic at 1 year
Scremini (11)	3/male	Tension pneumothorax	Chest X-ray: Spherical opacity in pleural cavity	Cyst removal, lung cavity sutured	No recurrence at 6 months
Current Case	28/Female	Dyspnea, chest pain	CT: Ruptured cyst in right upper lobe	Cyst extraction, capitonnage	No recurrence at 6 months

In this case, the patient presented with dyspnea and right basithoracic pain, which are nonspecific symptoms often associated with pneumothorax or pleural effusion. The diagnosis of PHPH was confirmed by imaging and surgical exploration. The chest CT scan revealed a hydatid cyst in the right upper lobe with rupture into the pleural cavity, consistent with PHPH. Surgical intervention was performed to extract the intact cyst and treat the residual cavity, preventing complications such as secondary pleural hydatidosis and intraoperative anaphylactic shock (9, 10).

The rarity of PHPH makes it a diagnostic challenge, particularly in non-endemic regions. Imaging plays a crucial role in the diagnosis, with chest X-ray and CT scan being the most commonly used modalities. On chest X-ray, PHPH may present as a hydropneumothorax or pleural effusion, while CT scan typically shows a well-defined fluid lesion without enhancement after contrast injection (11). In this case, the CT scan findings were instrumental in confirming the diagnosis.

Surgical intervention remains the mainstay of treatment for PHPH. The goal of surgery is to remove the intact cyst and treat the residual cavity to prevent recurrence and complications. In this case, the cyst was extracted intact, and capitonnage of the residual cavity was performed. Post-operative anti-helminthic therapy with albendazole was administered to prevent recurrence, as recommended in the literature (12). Albendazole, a benzimidazole derivative, is the drug of choice for post-operative management of hydatid disease due to its ability to inhibit parasite microtubule formation, leading to impaired glucose uptake and eventual parasite death (13). The recommended duration of treatment is 6 months, as prolonged therapy is necessary to ensure complete eradication of the parasite and prevent recurrence (13). In this case, the patient was started on Albendazole postoperatively and completed a 6-month course without complications, demonstrating the importance of adherence to anti-helminthic therapy in the management of hydatid disease.

The prognosis for PHPH is generally favorable with timely diagnosis and appropriate treatment. However, delayed diagnosis or inadequate treatment can lead to complications such as secondary pleural hydatidosis, anaphylactic shock, or recurrence. Therefore, it is essential to consider PHPH in the differential diagnosis of spontaneous pneumothorax, particularly in endemic regions.

CONCLUSION

Primary heterotopic pleural hydatidosis is a rare but serious condition that should be considered in the differential diagnosis of spontaneous pneumothorax in endemic regions. Early diagnosis and surgical intervention, combined with post-operative anti-helminthic therapy, are crucial for preventing complications and recurrence. This case highlights the importance of maintaining a high index of suspicion for PHPH in patients presenting with nonspecific respiratory symptoms in endemic areas.

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