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Case Report

An unusual cause of pulmonary hypertension: Mediastinal hydatid cyst ☆,☆☆

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ABSTRACT

Hydatid disease is a parasitic zoonosis prevalent in sheep-rearing areas. The lung is the second most commonly involved organ following the liver. Intra-thoracic and extra-pulmonary hydatid disease is rare and can affect the mediastinum, heart, diaphragm, pleura, and chest wall. Uncommon locations represent a diagnostic challenge. We report clinical and imaging manifestations of a compressive mediastinal hydatid cyst revealed by pulmonary hypertension. Radiologists must be familiar with the imaging features of hydatid disease and contemplate the diagnosis when facing atypical chest cystic lesions, especially in patients from endemic regions or with evidence of hydatidosis in a different anatomical location.

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Introduction

Human hydatid disease is endemic in Morocco. It is occasioned by the *Echinococcus granulosus* larval form. Primary hosts are dogs, wolves and coyotes, while the intermediate hosts are sheep, cattle, and deer. Humans are infected from contaminated water, food, and direct contact with carnivores. The lung is the second most commonly involved organ following the liver [1]. Intrathoracic extrapulmonary disease can involve the pleura, pericardium, diaphragm, and chest wall. Mediastinal cysts are exceptional [2]. We report clinical and imaging manifestations of a compressive mediastinal hydatid cyst revealed by pulmonary hypertension.

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Case presentation

A 56-year-old female living in a rural area of northern Morocco with unremarkable medical and surgical history, presented to the emergency department with dyspnea that has been evolving for the past 4 months. The patient was afebrile. General status was conserved. There was right upper quadrant tenderness upon abdominal palpation. Auscultation detected a loud P2 sign. No other abnormalities were objectified. Chest radiography revealed mediastinal enlargement with an abnormal convex bulge representing the enlarged main pulmonary artery (Fig. 1). ECG showed right bundle branch block. Transthoracic echocardiography identified a cystic mass externally compressing the right pulmonary artery and left atrium. Continuous Doppler of the main pulmonary artery recorded a 27-mmHg mean pressure. CT-angiography of the chest showed a middle mediastinal cystic mass (Figs. 2,3,4, A-B). The included upper abdominal CT slices revealed two



Fig. 1 – Frontal chest x-ray demonstrating an enlarged mediastinum with an abnormal convex bulge representing an enlarged main pulmonary artery (Black arrow).



Fig. 2 – Coronal reconstruction of a chest CT angiogram showing a well-defined thin walled cystic lesion with no calcifications nor enhancement following contrast administration, occupying the middle mediastinal compartment, measuring 39 × 40 mm and exerting a mass effect on the left atrium, the right superior pulmonary vein(*) and the right pulmonary artery ().**

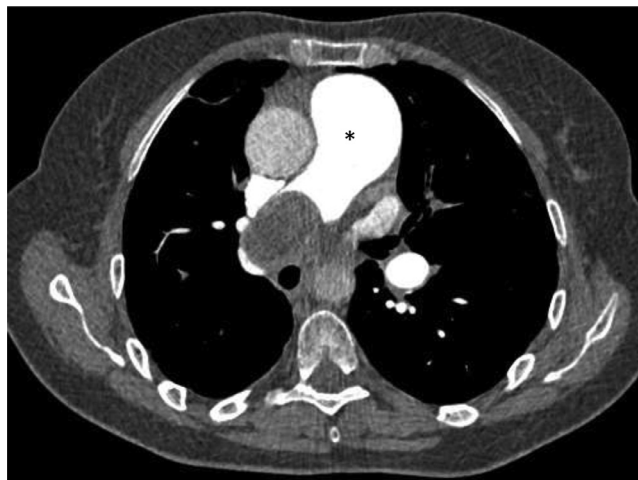


Fig. 3 – Axial chest CT angiogram image showing the middle mediastinum cyst compressing the right pulmonary artery, responsible for the main pulmonary artery enlargement (MPA diameter * = 42 mm).

multi-loculated hepatic cystic masses with peripheral calcifications, adjacent to the inferior vena cava (Figs. 5,6). Blood work detected no abnormalities notably no eosinophilia. Indirect hemagglutination test for echinococcosis was positive. The retained diagnosis was a mediastinal hydatid cyst. The patient was lost to follow-up.

Rationale for final diagnosis

Morocco is an endemic country for hydatidosis. The most important findings to support hydatidosis diagnosis in our patient's case are residency in a rural area, with frequent contact with sheep and dogs, and concomitant hepatic multiloculated cysts with wall calcifications.

Our patient's mediastinal cyst presented all CT features of benignity including: an oval shape with a smooth, thin, well-defined wall enhancing following contrast administration, homogeneous water density (0 - 20 HU), no enhancing components and no contiguous structures infiltration [3]. This allowed distinction from cystic degenerated tumors that demonstrate mixed solid and cystic component. Mediastinal abscess and pancreatic pseudocyst were ruled out for the patient had no infectious nor pancreatitis clinical history [4]. Duplication cysts are usually detected in infancy and are adjacent to or within the esophageal wall [5]. This localization allowed diagnosis exclusion. Lymphangiomas are uni or multi-loculated septated masses that usually mold adjacent structures whereas our patient's mediastinal cyst was compressive and responsible for pulmonary hypertension [6]. Pericardial cyst's attachment to the pericardium is constant; however, its predominant location is the right anterior cardiophrenic angle and it's not compressive [7]. Bronchogenic cysts are localized near the carina in the middle or posterior mediastinum [8]. In

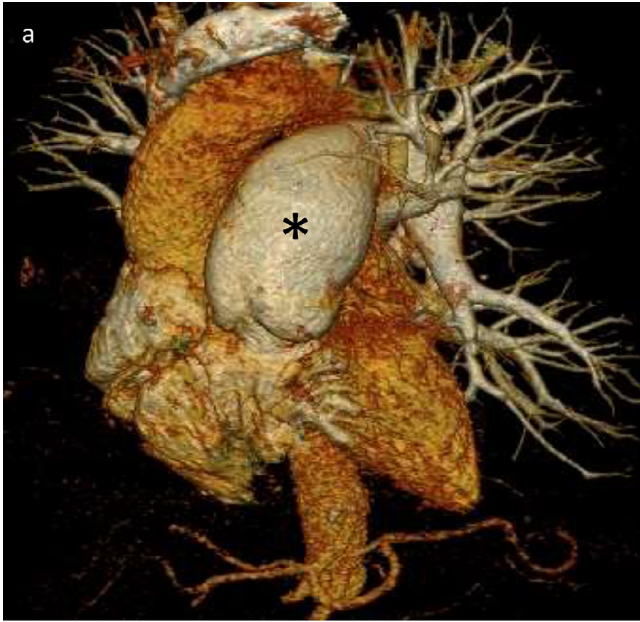


Fig. 4 – A,B – 3D volume rendered reconstructions showing (A) the main pulmonary artery dilatation and (B) the mass effect of the mediastinal cystic mass on the left atrium and superior right pulmonary vein.

our case, no intimate contact with the tracheo-bronchial tree was observed.

Discussion

Clinical presentation & general epidemiology

Hydatid disease is a cosmopolitan cyclozoonotic infection mainly due to *Echinococcus granulosus* and *E. multilocularis*; less common but more invasive causing alveolar echinococcosis (AE). *E. granulosus* for which dogs and sheep serve as

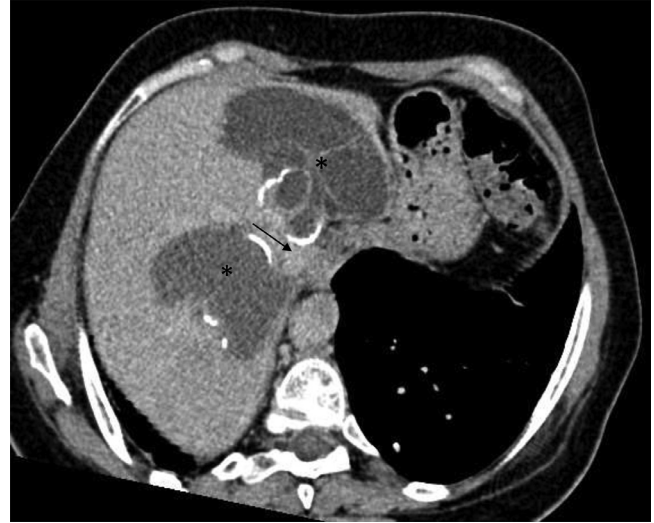


Fig. 5 – Axial CT image of the abdomen in the venous phase showing the two multiloculated cystic hepatic masses with peripheral calcifications (*) adjacent to the inferior vena cava with no evidence of its invasion (Black arrow).



Figs. 6 – Sagittal chest CT angiogram reconstruction showing the cystic mediastinal lesion (), alongside with 2 hepatic multiloculated cysts (*) presenting peripheral wall calcifications, adjacent to the inferior vena cava (Black arrow).**

definitive and intermediate hosts respectively, is endemic in the Mediterranean region, Africa, South America, the Middle East, Australia and New-Zealand, and responsible for the most commonly encountered type of hydatidosis in humans: cystic echinococcosis (CE). The liver is humans' first defense line. It is involved in 75 % of cases, lungs in 15 % and other localizations in 10 % [1]. Mediastinal echinococcosis is rare. Primary or concomitant to other organs' involvement; it repre-

sents 0.1% of all anatomic localizations and 1 % of thoracic locations [2]. Advanced pathogenesis hypothesis include: Fissuring of a hepatic or pulmonary hydatid cyst into the systemic circulation, Transdiaphragmatic dissemination [9], Hydatid solices hematogeneous spread bypassing liver and lungs filters or lymphatic diffusion via intercostal and parasternal lymph nodes [10]. Solitary or multiple; mediastinal hydatid cysts can involve all mediastinal compartments; the posterior mediastinum being the most frequently affected [9].

Mediastinal hydatid cysts' presenting signs are non-specific and depend on the size, location, involvement and mass effect on adjacent structures. Pulmonary hypertension caused by E. granulosus is exceptional. Cough, dyspnea, dysphagia and chest pain are commonly reported [11]. Rupture can cause anaphylactic shock and death.

Imaging features

Hydatid cysts are to be considered in the differential diagnosis of cystic mediastinal lesions particularly in patients from endemic regions or concomitant hydatid cysts in other anatomic locations [9]. Echocardiography, CT, and MRI depict the cystic nature of the mass and its relation to the cardiac chambers. CT and MRI dispense invaluable diagnostic informations concerning the cyst's morphology, attenuation, signal, localization and connections to contiguous structures. Imaging aspects range from uni to multilocular with or without detached membranes or calcifications. CT best shows calcifications whereas MRI's high resolution for cystic lesions assesses the presence of daughter cysts.

Based on imaging features; hydatid cysts are classified into 4 types:

Type I (Simple cysts): Well-defined anechoic masses including hydatids' sand or septa on ultrasound with a homogeneous fluid attenuation on CT, low signal on T1 and high signal on T2 weighted MRI images with a dark rim on both sequences.

Type II (Cysts with daughter cysts and matrix): Daughter cysts appear as smaller cysts, commonly at the periphery of the large mother cyst, lower in attenuation on CT and iso- or hypointense to the matrix. Occasionally; larger irregularly shaped daughter cysts occupy most of the mother cyst.

Type III (Calcified/Dead cysts): Calcifications appear echogenic on ultrasound with posterior shadowing, hyperattenuating on CT and hypointense on MRI.

Type IV (Complicated cysts): Hydatid cysts' complications include rupture -in response to degeneration, treatment, trauma ...- and/or superimposed infection. When rupture is contained, floating membranes appear as serpentine structures of low attenuation on CT and low signal on MRI within the matrix 'Water-lily sign'. Intra-cystic air suggests a communicating rupture and/or superinfection [11].

Prognosis, treatment or therapeutic options

Prognosis depends on the infestation type. Morbidity is due to cysts' rupture or infection and involved organs' dysfunctions (Cirrhosis, biliary obstruction, bronchial obstruction ...). Mortality is secondary to anaphylaxis, systemic (sepsis, respiratory failure ...) or operative complications.

CE's prognosis is normally good, and cure possible with complete surgical excision and no spillage. Chemotherapy's (Albendazole/Mebendazole) indications include: Inoperable primary liver or lung cysts, Cysts in 2 or more sites and peritoneal cysts. In AE, prognosis is poor. Cure is only possible with early detection and radical surgical excision. When the latter is unfeasible, a long-term chemotherapy substantially decreases mortality [12].

Treatment of mediastinal hydatid cysts' is classically surgical and consists of cystectomy with total or partial pericystectomy. The surgical approach depends on the cyst's location and surrounding structures' involvement [9].

Conclusion

Although uncommon, mediastinal hydatid cysts should be considered in the differential diagnosis of mediastinal cystic lesions, particularly in endemic regions. CT is the chief modality for diagnosis and possible extension study. Early surgical removal is the treatment of choice, and prevents complications. Adjuvant medical therapy averts recurrence.

REFERENCES

- [1] Pedrosa I, Saiz A, Arrazola J, Ferreirós J, Pedrosa CS. Hydatid disease: radiologic and pathologic features and complications. *Radiographics* 2000;20(3):795–817. doi:10.1148/radiographics.20.3.g00ma06795.
- [2] Oğuzkaya F, Akçali Y, Kahraman C, Emiroğullari N, Bilgin M, Sahin A. Unusually located hydatid cysts: intrathoracic but extrapulmonary. *Ann Thorac Surg* 1997;64(2):334–7. doi:10.1016/S0003-4975(97)00521-3.
- [3] Jeung MY, Gasser B, Gangi A, Bogorin A, Charneau D, Wihlm JM, et al. Imaging of cystic masses of the mediastinum. *Radiographics* 2002;22 Spec No:S79-S93. doi:10.1148/radiographics.22.suppl_1.g02oc09s79.
- [4] Kawashima A, Fishman EK, Kuhlman JE, Nixon MS. CT of posterior mediastinal masses. *Radiographics* 1991;11(6):1045–67. doi:10.1148/radiographics.11.6.1749849.
- [5] Salo JA, Ala-Kulju KV. Congenital esophageal cysts in adults. *Ann Thorac Surg* 1987;44(2):135–8. doi:10.1016/s0003-4975(10)62023-1.
- [6] Miyake H, Shiga M, Takaki H, Hata H, Onishi R, Mori H. Mediastinal lymphangiomas in adults: CT findings. *J Thorac Imaging* 1996;11(1):83–5. doi:10.1097/00005382-199601110-00009.
- [7] Rogers CI, Seymour EQ, Brock JG. Atypical pericardial cyst location: the value of computed tomography. *J Comput Assist Tomogr* 1980;4(5):683–4. doi:10.1097/00004728-198010000-00020.
- [8] Mendelson DS, Rose JS, Efremidis SC, Kirschner PA, Cohen BA. Bronchogenic cysts with high CT numbers. *AJR Am J Roentgenol* 1983;140(3):463–5. doi:10.2214/ajr.140.3.463.
- [9] Eroğlu A, Kürkçüoğlu C, Karaoğlanoğlu N, Tekinbaş C, Kaynar H, Onbaş O. Primary hydatid cysts of the mediastinum. *Eur J Cardiothorac Surg* 2002;22(4):599–601. doi:10.1016/s1010-7940(02)00398-6.

- [10] Isitmangil T, Toker A, Sebit S, Erdik O, Tunc H, Gorur R. A novel terminology and dissemination theory for a subgroup of intrathoracic extrapulmonary hydatid cysts. *Med Hypotheses* 2003;61(1):68–71. doi:[10.1016/s0306-9877\(03\)00108-7](https://doi.org/10.1016/s0306-9877(03)00108-7).
- [11] Saeedan MB, Aljohani IM, Alghofaily KA, Loutfi S, Ghosh S. Thoracic hydatid disease: A radiologic review of unusual cases. *World J Clin Cases* 2020;8(7):1203–12. doi:[10.12998/wjcc.v8.i7.1203](https://doi.org/10.12998/wjcc.v8.i7.1203).
- [12] Manterola C, Otzen T, Muñoz G, Alanis M, Kruuse E, Figueroa G. Surgery for hepatic hidatidosis. Risk factors and variables associated with postoperative morbidity. Overview of the existing evidence. *Cirugía de la hidatidosis hepática. Factores de riesgo y variables asociadas al desarrollo de morbilidad postoperatoria. Revisión global de la evidencia existente.* *Cir Esp* 2017;95(10):566–76. doi:[10.1016/j.ciresp.2017.08.009](https://doi.org/10.1016/j.ciresp.2017.08.009).