

Received: 2021.07.25

Accepted: 2021.10.12

Available online: 2021.10.25

Published: 2021.11.30

Hydatid Pulmonary Embolism: A Case Report and Literature Review

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

AEF 1 **Adila Aili**
A 2 **Liqing Peng**
B 1 **Jiarui Zhang**
B 1 **Yu Liu**
B 1 **Lige Peng**
A 1 **Qun Yi**
A 1 **Haixia Zhou**

1 Department of Respiratory and Critical Care Medicine, West China Hospital, Sichuan University, Chengdu, Sichuan, PR China
2 Department of Radiology, West China Hospital, Sichuan University, Chengdu, Sichuan, PR China

Corresponding Author: Haixia Zhou, e-mail: zhouhaixia@wchscu.cn

Financial support: None declared

Conflict of interest: None declared

Patient: Female, 58-year-old
Final Diagnosis: Pulmonary embolism caused by hydatid cyst
Symptoms: Chest and back pain • hemoptysis • shortness of breath
Medication: —
Clinical Procedure: —
Specialty: Infectious Diseases • Pulmonology • Radiology

Objective: Rare disease

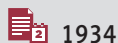
Background: Infection with *Echinococcus granulosus* is endemic in sheep and dogs in Central Asia, including Tibet. In humans, ingested parasites from the gastrointestinal system enter the liver via the portal vein. Rarely, hepatic hydatid cysts can rupture into the portal vein and embolize to the lungs. This report is of a 58-year-old woman with liver cysts and a pulmonary embolism due to hydatid disease.

Case Report: We present a rare case of a pulmonary embolism caused by a hydatid cyst. A 58-year-old woman from the Tibet Autonomous Region of China was admitted to the hospital with symptoms of chest and back pain and shortness of breath within the previous 6 months. She had a 5-year history of hepatic echinococcosis. During hospitalization, the patient reported having aggravated chest and back pain and she developed a new symptom of hemoptysis. A pulmonary embolism was confirmed by computed tomography pulmonary angiography. After a multidisciplinary consultation, and based on the patient's medical history, clinical manifestations, laboratory test results, and imaging findings, a diagnosis of a pulmonary embolism caused by a hydatid cyst was established.

Conclusions: This report shows the importance of imaging findings in diagnosing a non-thrombotic pulmonary embolism due to hepatic hydatid disease. In this case, early and accurate diagnosis resulted in appropriate treatment with multidisciplinary patient management.

Keywords: Echinococcosis • Membranous Obstruction of Inferior Vena Cava • Pulmonary Artery • Pulmonary Embolism

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/934157>



1934



1



3



16



Background

Echinococcosis is a parasitic disease caused by a tapeworm and is responsible for affecting human health worldwide. Although it occurs globally, it is found mainly in countries dominated by agriculture and animal husbandry, and in China it occurs mostly in the northwest [1]. Echinococcosis is mainly caused by the larva of *Echinococcus granulosus* (also called hydatid cyst), and humans can be infected after ingesting water or animal livers contaminated with the parasite's eggs or after contact with animals infected with hydatid eggs. After entry into the digestive tract of humans, the eggs can further enter the portal vein system or lymphatic system through the intestinal mucosa. Most eggs then arrive in the liver capillaries, parasitize the liver, and develop into larvae or die. The eggs can also parasitize other organs of the body through the blood circulation. However, the liver is the most frequently infested organ, followed by the lungs, whereas the cardiovascular system is less affected [2,3]. Pulmonary emboli caused by hydatid cysts are extremely rare; however, here, we report a rare case and present a thorough review of the literature to investigate the clinical characteristics of this disease (Table 1). This report is of a 58-year-old woman with liver cysts and a pulmonary embolism due to hydatid disease.

Case Report

A 58-year-old woman from the Tibet Autonomous Region of China was admitted to the hospital with symptoms of chest and back pain and shortness of breath within the last 6 months. The patient reported that she had been diagnosed with hepatic hydatid disease 5 years prior. On physical examination, venous distension along the neck veins and diminished breath sounds of the right lower lung were noted.

On laboratory examination, the eosinophil counts were in the normal range. D-dimer was positive, with an increased value of 1.58mg/L fibrinogen equivalent units. Serologic testing was positive for antibodies against cystic hydatidosis (IgG 3.68 S/cut-off). No other abnormal laboratory findings were observed.

The patient received an unenhanced computed tomography (CT) scan of the chest at a local hospital, showing pleural effusion in the right lung and ground-glass opacities on both sides of the lung. Abdominal ultrasonography showed an irregular echo zone of about 11.8×10 cm within the parenchyma of the right liver lobe, which was the hepatic echinococcosis. During hospitalization, the patient had aggravated chest and back pain and developed a new symptom of hemoptysis.

Because a pulmonary embolism was suspected, CT pulmonary angiography (CTPA) was performed, showing a filling defect in

the trunk of the right pulmonary artery and in the basilar artery of the left lower lobe (Figure 1A, 1B). Cystic solid density shadows with separation inside could also be seen in the right hilum, right lower posterior mediastinum, and left lower lobe, with no obvious enhancement. The boundary between the lower edge of the lesion and the liver was not clear, and similar density shadows were seen in the liver (Figure 2A, 2B). The cystic solid lesion in the liver seemed to extend into the inferior vena cava.

The echocardiography revealed right heart enlargement and severe pulmonary arterial hypertension (78 mmHg). Chest radiography demonstrated an enlarged pulmonary tract and dilation of the right ventricle (Figure 3). Color Doppler ultrasound of the lower extremity vein found no abnormality. After multidisciplinary consultation, which was based on the patient's medical history, clinical manifestations, laboratory test results, and imaging findings, the patient was diagnosed as having hydatid disease with liver, lung, and mediastinum involvement and a pulmonary embolism caused by hydatid cyst. Surgery was advised, but the patient refused surgery and was therefore discharged with albendazole tablets. The patient died of pulmonary hypertension and right heart failure 6 months later.

Discussion

A pulmonary embolism caused by a hydatid cyst is a type of non-thrombotic pulmonary embolism with low morbidity but high mortality. It has been reported that echinococcosis of the pulmonary artery is mainly derived from hepatic hydatid cysts, and when these rupture in the liver or other organs in the abdomen, they invade the inferior vena cava or the hepatic vein. This enables the cyst contents to enter into the blood circulation and cause a pulmonary embolism after entering into the pulmonary artery and its branches through the right atrium and right ventricle [4-6]. By the time this situation occurs, the hydatid cyst is not usually seen in the right atrium or right ventricle, and most patients will have a history of hepatic hydatid disease. In our patient, as determined by the CTPA, cystic solid lesion in the liver extended into the inferior vena cava. Furthermore, echocardiography failed to detect a hydatid cyst in the right atrium or right ventricle. Taken together, these findings suggest it is possible that such a mechanism occurred in our patient, which could also have been the situation for 7 of the 12 cases we reviewed. Another less common mechanism of a pulmonary embolism caused by a hydatid cyst is the complication of cardiac hydatid disease, which occurs when hydatid cysts parasitizing the right atrium and the right ventricle fall off or rupture, and the contents within the cysts enter the pulmonary artery and its branches [7]. Three of the 12 cases reviewed here were confirmed to have cardiac hydatid disease. The rarest condition is when this parasite within

Table 1. Reported cases of pulmonary embolism due to hepatic hydatid disease.

Study	Patient age (years)/sex	Clinical presentation	Imaging manifestation	Location of hydatid cyst	Treatment	Outcome
Yague et al (1998) [3]	44/Male	Hemoptysis	CT: 2 oval masses of 40 and 20 mm located in the pulmonary arteries; MR: hypointense in T1-weighted images and hyperintense in T2-weighted images	Right, left pulmonary artery	Surgery: thoracotomy	Not mentioned
Yuan et al (2014) [4]	70/Male	Dyspnea, intermittent cough	Enhanced CT: low-density filling defect in the pulmonary artery, complete occlusion of the lumen	left lung lower lobe segmental pulmonary artery	Surgery	Not mentioned
Lioulias et al (2001) [5]	57/Male	Left chest pain, dyspnea, cyanotic	CTA: cystic lesions, complete and partial occlusion of the relevant pulmonary arteries; MR-angiography: multiple cysts in the pulmonary artery	Left pulmonary artery, distal branches of the right pulmonary artery	Surgery: thoracotomy	After 42 months the patient is asymptomatic
Alper et al (1995) [7]	55/Male	Dyspnea	CT: low-density lesions in the pulmonary artery, well-defined oval shape and with water density. MRI: a cystic lesion, hypointensity on T1-weighted images and hyperintensity on T2-weighted images; a thrombosis lesion, moderately hyperintense on all sequences. Conventional pulmonary angiography: total occlusion of the relevant arteries	Right main pulmonary artery and left lower lobe artery	Surgery: Dissected right pulmonary artery	Not mentioned
Karantanas et al (2000) [8]	67/Male	Dyspnea, cough, hemoptysis	Enhanced CT: multiple echinococcal cysts in both lungs and a hypodense mass located in the left pulmonary artery compatible with intra-arterial hydatid cyst. MRI: multiple cysts sub-pleurally with mild wall enhancement and a similar cystic lesion in the lumen of the left pulmonary artery	Left pulmonary artery	Mebendazole	After 19 months there has not been deterioration
Kokasal et al (2006) [9]	24/Male	Cough, hemoptysis	CT: a para-hilar cavitated mass-like lesion, a cavitary lesion in the upper lobe and disseminated parenchymal infiltration in the right lung; right pulmonary artery was occluded with a hypodense lesion. MRI: complete occlusion of the right pulmonary artery	Right pulmonary artery	Surgery: Dissected right pulmonary artery	After 14 months no pathology has been detected
Akgun et al (2011) [10]	43/Male	Hemoptysis, abdomen pain, chest pain, dyspnea	CT: multiple cystic emboli in the pulmonary arteries, vessel enlargement, multiple cystic parenchymal nodules in the lung lobes, and cystic embolus in the right atrium; the border between IVC and cystic component of the mass was undetermined Sonographic images: the border between the wall of the hydatid cyst and IVC was missing	IVC, pulmonary artery, right atrium	Albendazole	After 2 months the patient is asymptomatic

Table 1 continued. Reported cases of pulmonary embolism due to hepatic hydatid disease.

Study	Patient age (years)/sex	Clinical presentation	Imaging manifestation	Location of hydatid cyst	Treatment	Outcome
Herek et al (2012) [11]	31/Female	Chest pain, dyspnea, cough	Ultrasonography: a giant hydatid cyst lesion in the liver measuring 15×10 cm and compressing the portal and hepatic veins, germinal membrane of the cyst extending into the IVC and right atrium. Enhanced CT: a giant hydatid cyst extending into the right atrium via the IVC, embolization of the cyst contents into the pulmonary arteries, near-complete occlusion of the lumen	Right atrium, IVC, left main and left lower lobe pulmonary arteries	Surgery and albendazole	Not mentioned
Abid et al (2011) [13]	16/Male	Hemoptysis, chest pain, dyspnea	CT: distended distal branches of the right and left pulmonary artery, partial occlusion by cystic lesions and multiple segmental defects; an intra-right atrial mass; echocardiography: a large cystic mass measuring 19×22 mm, with a large implantation basis adhesive to the right side of the inter atrial septum with an extension to the right ventricle	Distal branches of the right and left pulmonary artery, right atrium and ventricle	Surgery: thoracotomy, albendazole	After 18 months the patient is asymptomatic
Mahouachi et al (2007)[14]	51/Male	Hemoptysis, chest pain	Enhanced CT: multiple echinococcal cysts in both lungs and partial occlusion of a distal branches of the left pulmonary artery by cystic lesions, residual cavity in the liver	Distal branches of the right and left pulmonary artery	Albendazole	After 12 months the patient is asymptomatic
Savaş et al (2017) [15]	48/Female	Dry cough, chest pain, dyspnea	Echocardiography: right chamber dilatation with moderate tricuspid regurgitation and elevated pulmonary artery systolic pressure (75 mmHg). Unenhanced CT: multiloculated cystic lesions in the pulmonary artery; a large filling defect by contrast-enhanced CT. MRI: multiloculated cystic lesions, hyperintense in T2-weighted images	Right pulmonary artery	Surgery: embolectomy	Dead
Asri H et al (2019) [16]	73/Male	Dyspnea	Echocardiography: enlarged pulmonary arteries, enlarged chambers of the right heart and elevated pulmonary systolic pressure (80mmHg). CT: a cystic filling defect of the pulmonary artery	Main and right pulmonary artery	Albendazole	Not mentioned

CT – computer tomography; MRI – magnetic resonance imaging; CTA – computed tomography angiography; IVC – inferior vena cava.

the pulmonary artery wall or in the nearby lung parenchyma gradually grows and ruptures the arterial wall and then enters and blocks the pulmonary artery [8,9]. The pulmonary emboli caused by a hydatid cyst in 2 of the 12 cases we reviewed probably developed through this mechanism.

The clinical manifestations of a pulmonary embolism caused by a hydatid cyst are not specific, and relevant symptoms occur only when hydatid worms develop to block or puncture blood vessel walls. Hemoptysis may be the earliest and most common symptom for patients with the disease. In addition

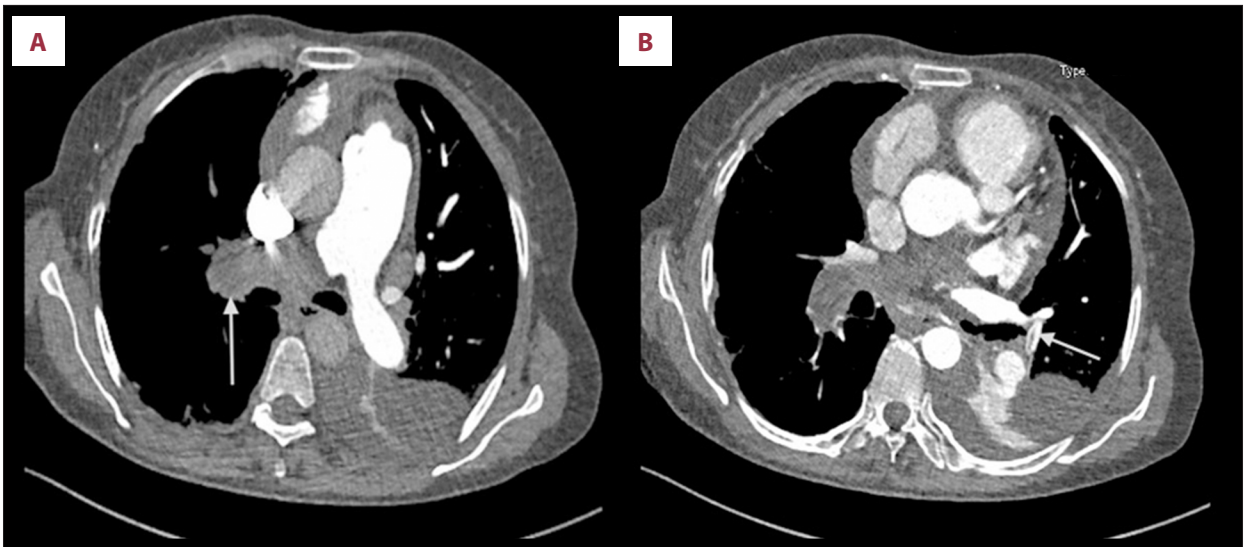


Figure 1. Computed tomography pulmonary angiography showed (A) filling defects in the trunk of the right pulmonary artery and (B) in the basilar artery of the left lower lobe.

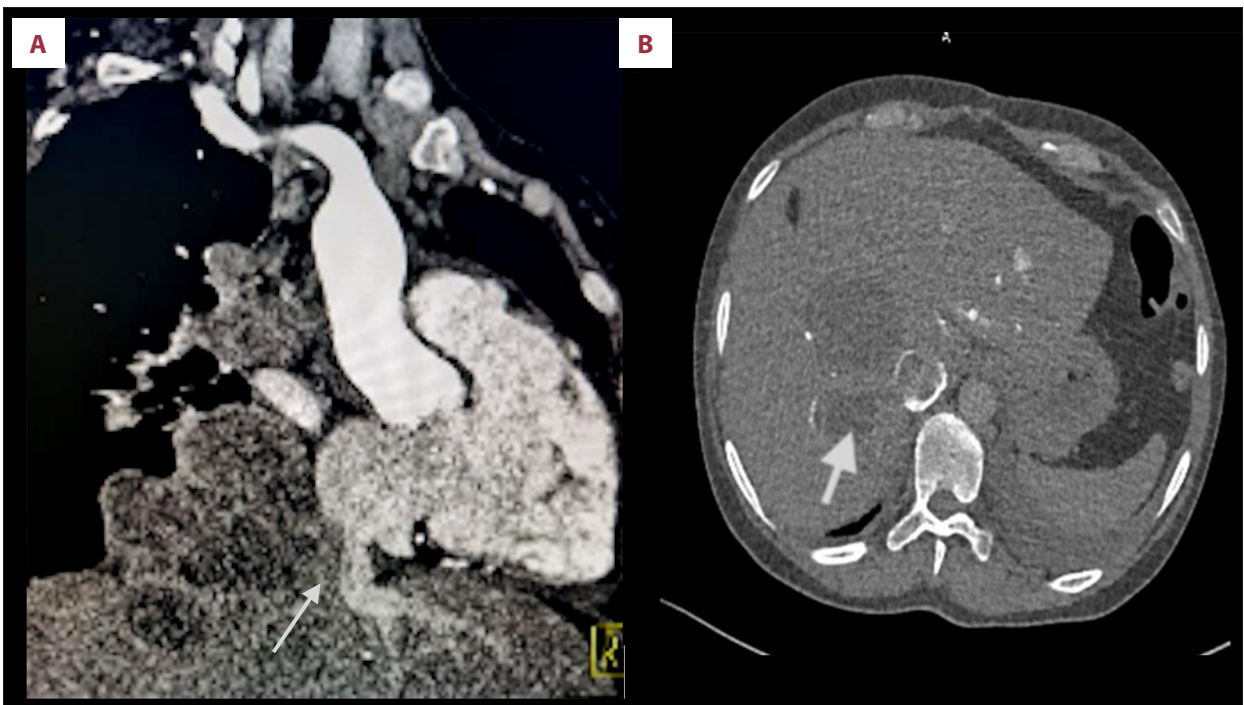


Figure 2. Enhanced computed tomography of the chest revealing (A) the lesion in the right hilum, right lower posterior mediastinum and left lower lobe, and (B) similar density shadows were seen in the liver. (A) Boundary between the lower edge of the lesion and the liver was not clear. Cystic solid lesion in the liver extended into the inferior vena cava (A and B, indicated by the arrow).

to hemoptysis, dyspnea, chest pain, and coughing, expectoration and allergic reactions can occur [10]. In the 12 cases we reviewed, hemoptysis was present in 6 cases, chest pain was presented in another 6 cases, and dyspnea was presented in all cases. There are mainly 3 outcomes of a pulmonary embolism caused by a hydatid cyst: (1) sudden death due to

massive embolization; (2) death within 1 year due to pulmonary hypertension; and (3) long-term pulmonary hypertension [11,12]. Most patients appear to have chronic pulmonary hypertension accompanied by an acute pulmonary embolism. In some acute cases, hemorrhagic shock due to massive hemoptysis, hemorrhage due to rupture of a vessel wall, or



Figure 3. Chest radiography showing enlarged pulmonary tract and dilation of the right ventricle.

anaphylactic shock because of the material in the cysts entering the bloodstream were the common causes of sudden death. In our case, the patient reported having chest and back pain and dyspnea 6 months previously, and the patient's echocardiography showed severe pulmonary hypertension. While hospitalized, her chest pain was aggravated, and hemoptysis occurred. All these symptoms suggested that the patient may have had chronic pulmonary hypertension accompanied by an acute pulmonary embolism, both caused by hydatid cyst, and the patient died of pulmonary hypertension and right heart failure 6 months later.

Early diagnosis of a pulmonary embolism caused by a hydatid cyst is critical for early treatment and reduction of mortality. However, it is often difficult to diagnose due to the absence of specific symptoms or symptoms that do not support a pulmonary embolism. The disease tends to be confirmed only when the patient develops the relevant symptoms or experiences acute episodes, and, in some patients, it is only confirmed on autopsy. Laboratory tests for a pulmonary embolism caused by echinococcosis are not specific. However, the blood test for anti-hydatid antibodies is usually positive and the number of eosinophils tends to increase significantly [7,13]. In terms of imaging examinations, a chest X-ray is not helpful. Ultrasonography can show hydatid emboli in the blood vessels, atria, and ventricle, as well as hydatid cysts in major organs. Upon CT, non-complicated cysts usually show low levels

of attenuation and well-defined limits, with the possible presentation of calcium on the periphery, depending on their developmental state. When using enhanced CT with contrast, only the wall is highlighted, whereas the appearance of the hydatid cyst on magnetic resonance imaging (MRI) is usually characteristic and can be observed as an oval lesion, which is hypointense on T1-weighted images and hyperintense on T2-weighted images. A typical image is represented by a peripheral ring and hypointense T2-weighted images, which represent the pericyst (a dense fibrous capsule from the reactive host tissue) [3,8]. Unfortunately, in our case, the patient refused MRI for economic reasons. At present, CTPA is the most commonly used method for the diagnosis of a pulmonary embolism. It can accurately show the blocked pulmonary artery and filling defect at corresponding vascular segments but cannot discern the part below the blocked vascular segment, and when the vessel is completely blocked, it is sometimes difficult to distinguish the type of pulmonary embolism. Therefore, a diagnosis of pulmonary embolism caused by a hydatid cyst should be based on a comprehensive consideration of medical history, clinical manifestations, and imaging results. The patient in our case had a 5-year history of hepatic echinococcosis and presented 3 typical signs of pulmonary embolism with chest pain, dyspnea, and hemoptysis. Also, a CTPA revealed filling defects in some pulmonary arteries, and a hydatid cyst lesion extended into the inferior vena cava. Thus, a pulmonary embolism caused by a hydatid cyst was considered, based on this comprehensive analysis.

Pulmonary embolism caused by a hydatid cyst generally has a poor prognosis, and surgery remains by far the best treatment option. The specific surgical method is selected according to the location of the hydatid embolus in the vascular lumen and its anatomical position. Thrombectomy is the most common surgical method, but the risk of perioperative death is relatively high. During surgery, hydatid cysts or arteries can rupture and lead to severe complications, such as anaphylactic shock, aneurysm, new embolus formation, or hydatid dispersion to other tissues and organs [9,14]. In the cases reviewed here, 8 of 12 received surgery. Among the 4 cases whose outcome was mentioned, 3 patients survived and 1 patient died after surgery. Medication is not very effective for pulmonary emboli due to the presence of hydatid cysts. However, it can be used in the following circumstances: (1) recurrent echinococcosis, (2) disseminated disease, including secondary lung or pleural hydatidosis, and (3) contraindication of surgery or high risk of hydatid fluid leakage during surgery. For medication, mebendazole and albendazole are mainly used and these medications can prolong survival time and improve quality of life of patients [13,14]. In the cases reported here, 4 out of 12 patients received only medication and after several months of follow-up, 3 patients were asymptomatic, and the outcome from 1 case was not recorded. In our case, the patient refused

surgical and anticoagulation treatment and was discharged with albendazole tablets. Unfortunately, the patient died of pulmonary hypertension and right heart failure 6 months later.

Conclusions

For the diagnosis of pulmonary embolism caused by a hydatid cyst, careful evaluation of clinical manifestations and regular laboratory test results are necessary because of the lack of specific symptoms. However, the history of the parasite exposure in endemic areas and findings from imaging analyses

can provide strong supporting evidence for a diagnosis. This report has shown the importance of imaging findings in diagnosing a non-thrombotic pulmonary embolism due to hepatic hydatid disease. In this case, early and accurate diagnosis resulted in appropriate treatment with multidisciplinary patient management.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

References:

1. World Health Organization (WHO). Echinococcosis. 17 May 2021. Available from: <https://www.who.int/news-room/fact-sheets/detail/echinococcosis>
2. Polat P, Kantarci M, Alper F, et al. Hydatid disease from head to toe. *Radiographics*. 2003;23(2):475-94
3. Yagüe D, Lozano MP, Lample C, et al. Bilateral hydatid cyst of pulmonary arteries: MR and CT findings. *Eur Radiol*. 1998;8(7):1170-72
4. Yuan Z, Maihesuti M, Wenya L, et al. Rare reason for pulmonary embolism: One case of pulmonary hydatid cyst and review of the literature. *J Thromb Thrombolysis*. 2015;40(1):126-29
5. Lioulias A, Kotoulas C, Kokotsakis J, et al. Acute pulmonary embolism due to multiple hydatid cysts. *Eur J Cardiothorac Surg*. 2001;20(1):197-99
6. El Hadj Sidi C, Mgarrech I, Alimi F, et al. Four pathways for hydatid pulmonary embolism. *J Card Surg*. Aug 2020;35(8):1877-84
7. Alper H, Yünten N, Scener RN. Intramural hydatid cysts of pulmonary arteries: CT and MR findings. *Eur Radiol*. 1995;5(6):666-68
8. Karantanas AH, Bitsios G, Karaïskou E. Echinococcus of the pulmonary artery: CT, MRI and MRA findings. *Comput Med*. 2000;24(4):265-67
9. Koksall C, Baysungur V, Okur E, et al. A two-stage approach to a patient with hydatid cysts inside the right pulmonary artery and multiple right lung involvement. *Ann Thorac Cardiovasc Surg*. 2006;12(5):349
10. Akgun V, Battal B, Karaman B, et al. Pulmonary artery embolism due to a ruptured hepatic hydatid cyst: Clinical and radiologic imaging findings. *Emerg Radiol*. 2011;18(5):437-39
11. Herek D, Karabulut N. CT demonstration of pulmonary embolism due to the rupture of a giant hepatic hydatid disease. *Clin Imaging*. 2012;36(5):612-14
12. Kardaras F, Kardara D, Tselikos D, et al. Fifteen year surveillance of echinococcal heart disease from a referral hospital in Greece. *Eur Heart J*. 1996;17(8):1265-70
13. Abid L, Laroussi L, Abdennadher M, et al. A cardiac hydatid cyst underlying pulmonary embolism: A case report. *Pan Afr*. 2011;8(1):12
14. Mahouachi R, Berraies A, Taktak S, et al. Pulmonary hydatid cyst embolization successfully treated with albendazole. *Respir Med Extra*. 2007;3(4):192-94
15. Savaş G, Coşgun MS, Karabiyik U, et al. A rare cause of pulmonary embolism: hydatid cyst. *Am J Respir Crit Care Med*. 2017;196(3):386
16. Asri H, Zegmout A, Abid A. A rare cause of chronic pulmonary embolism: Hydatid cyst. *Arch Bronconeumol*. 2018;55(2):101