CASE REPORT

Middle lobe syndrome: a singularly rare presentation of complicated pulmonary hydatid disease

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SUMMARY

Imaging is crucial to the diagnosis of pulmonary hydatid disease, as it is often the first modality that raises suspicion of the disease. Middle lobe syndrome (MLS) as a presentation of this disease is a distinct rarity. A 45-year-old woman, a never-smoker, presented with cough and streaky haemoptysis. Imaging demonstrated a trapezoidal opacity with its base towards the hilum and contiguous with the right cardiac border. The reformatted sagittal view confirmed the diagnosis of MLS. Fiberoptic bronchoscopy (FOB) revealed an avascular white membranous structure wholly occluding the medial segment of the middle lobe. This was completely removed through gentle suction. Bronchial aspirate revealed hooklets of hydatid and echinococcal serology was positive. Subsequently, three cycles of albendazole were administered with remarkable clinical and radiological improvement. To the best of our knowledge, this is the first detailed description of MLS caused by pulmonary hydatidosis that was completely removed through FOB.

BACKGROUND

Hydatidosis, a zoonotic disorder, is caused by the tapeworm of the *Echinococcus* species and is not uncommon in India.¹ Humans, the accidental host, are infested when they come in direct contact with primary hosts, such as dogs and cats, as well as sheep and goats. The parasite predominantly affects the liver and lung.² Pulmonary hydatid disease often remains asymptomatic for a long time. Radiological imaging and serological tests are crucial to the diagnosis.³ Fiberoptic bronchoscopy (FOB) is usually not needed for the diagnosis of pulmonary hydatid disease. This may be required when the presentation is unusual and diagnosis is not suspected.

Middle lobe syndrome (MLS) as a presentation of pulmonary hydatid disease is a distinct rarity with only six patients being documented in four reports^{4–7} since 1977. In none of the documented patients was pulmonary hydatid disease suspected. The term ‘middle lobe syndrome’ was coined by Graham *et al*⁸ in 1948 to describe chronic or recurrent collapse of the right middle lobe. The first description of recurrent collapse of right middle lobe due to enlarged tuberculous lymph nodes was documented by Brock *et al*⁹ in 1937. MLS due to tuberculous aetiology is still widely known as ‘Brock’s syndrome’.⁹ This distinct clinical entity, though uncommon, continues to tantalise clinicians, as it is caused by a wide spectrum of respiratory disorders, and is yet to be fully understood.

The paucity of literature on the subject prompted this report of a middle-aged woman with hydatid lung disease who presented as a MLS. The diagnosis was suspected during FOB and the membranous structure blocking the bronchus was successfully removed by gentle suction.

CASE PRESENTATION

A 45-year-old HIV-negative woman, a farm worker, was referred to our Institute for evaluation of cough and streaky haemoptysis for the last 1 year. She had never smoked but had a significant history of exposure to biomass fuel smoke for more than 30 years. Based on her symptomatic and radiological profile, though sputum stains and cultures were repeatedly negative for *Mycobacterium tuberculosis*, she had received antituberculous therapy for 9 months without relief. The persistence of haemoptysis along with the right lower zone opacity on the chest radiograph for 9 months prompted a referral to our institute.

Examination revealed a middle-aged woman in no acute respiratory distress. There was no evidence of pallor, no cyanosis, no clubbing and no lymphadenopathy. She was afebrile with a respiratory count of 18 breaths/min. Diaphragmatic excursion was comparable on both sides. Vesicular breath sounds were audible but the intensity was reduced on the right lower hemithorax.

INVESTIGATIONS

Oxygen saturation at room air was 98%. The patient’s haemoglobin was 11.6 g% and total leucocyte count was 8300 cells/mm³ with neutrophilic predominance. ECG, urine analyses, renal and

Figure 1 Chest X-ray showing ill-defined opacity in the right mid and lower zones abutting the right cardiac border with loss of cardiac silhouette suggestive of right middle lobe syndrome.

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Figure 2 (A and B) High-resolution CT of the thorax (mediastinal and lung window) showing a trapezoidal opacity with its base towards the hilum and contiguous with the right cardiac border.

hepatic functions were within normal limits. Sputum stains and culture for *M. tuberculosis* and other aerobic organisms were negative. Fungal culture did not reveal any growth. The chest radiograph (figure 1) showed an ill-defined opacity in the right mid and lower zones abutting the right cardiac border with loss of cardiac silhouette suggestive of right MLS. High-resolution contrast-enhanced CT of the thorax (figure 2A, B) revealed a trapezoidal opacity with its base towards the hilum and contiguous with the right cardiac border, while the reformatted sagittal view too showed a wedge-shaped density extending from the hilum anteriorly and inferiorly towards the chest wall, confirming the presence of MLS (figure 3A, B).

In view of the MLS, the patient's consent was taken for a FOB, which visualised an avascular white membranous structure completely occluding the orifice of the medial segment of right middle lobe (figure 4A, B). The white membranous structure was slowly and completely removed through FOB by repeated but gentle suction. After suction, the medial segment of middle lobe bronchus opened completely (figure 5). The removal of the membranes was uneventful with no evidence of anaphylaxis. Stains and cultures of bronchial aspirate were negative for mycobacterial and other organisms. Bronchial aspirate revealed hooklets of hydatid. Serum sample sent for echinococcal serology was also positive. Ultrasonography of the abdomen did not

show any hepatic involvement. On subsequent interrogation, the patient admitted to having dogs as pets, as well as keeping goats at the farm.

DIAGNOSIS

A diagnosis of complicated endobronchial hydatid cyst presenting as a MLS was made.

TREATMENT

In view of the complete removal of the membranes and full opening of the bronchus after FOB, conservative management was considered and the patient was initiated on oral albendazole. Three cycles of albendazole 200 mg three times a day were given (cycle of 6 weeks: drug for 4 weeks with an interval of 2 weeks) with no untoward reaction.

OUTCOME AND FOLLOW-UP

Remarkable clinical and radiological improvement was observed and after completion of the three treatment cycles, the patient was completely asymptomatic.

DISCUSSION

Hydatid disease occurs worldwide but is endemic in certain regions such as the Mediterranean, the Middle East, Australia

Figure 3 (A and B) High-resolution CT of the thorax reformatted sagittal view (mediastinal and lung window) showing a wedge-shaped density extending from the hilum anteriorly and inferiorly towards the chest wall, confirming the presence of middle lobe syndrome.

Figure 4 (A) Fibreoptic bronchoscopic (FOB) image showing an avascular white membranous structure completely occluding the orifice of the medial segment of right middle lobe. (B) FOB image showing the white membranous structure being slowly and completely removed through FOB by repeated but gentle suction.

and South America.¹⁰ Pulmonary hydatid disease is often encountered by pulmonologists and imaging plays a crucial part in the diagnosis. Radiologically, pulmonary hydatid disease has a protean presentation and can be thought of as a 'picturesque' clinical entity. Although on imaging pulmonary hydatid disease has myriad appearances, MLS as a specific manifestation of this zoonotic ailment has rarely been documented. A search of the literature on the subject using the PubMed, IndMed and other databases revealed, to date, four reports documenting six patients of pulmonary hydatid disease who presented as MLS (table 1).^{4–7} However, there is only one detailed case report available in the English language literature.⁶ Furthermore, a scan of six major review articles on MLS^{11–16} since 1982, including a recent review in the paediatric population,¹⁶ found no mention of hydatidosis as a cause of MLS.

MLS is classified as being either of an obstructive or non-obstructive type. Obstructive MLS is characterised by intraluminal or extraluminal compression of the middle lobe bronchus while in the non-obstructive type, a patent middle lobe bronchus is visualised on bronchoscopy.¹⁵ The obstructive form of MLS is usually caused by endobronchial tumours, lymphadenopathies, a foreign body or granulomatous infection, while benign inflammatory causes are implicated in 62% of the non-obstructive variant.¹² Primary tumours as a cause of MLS are seen in 24% of patients while tuberculosis is responsible for 8–10%.¹² Other less common causes of MLS include allergic bronchopulmonary aspergillosis,^{17–19} bronchial anthracofibrosis²⁰ and even endobronchial tuberculosis.²¹ The middle lobe has a greater

propensity to collapse as the bronchus is narrow at its origin, has a long length and an angular take-off from the bronchus intermedius. To add to this, the collateral ventilation is poor due to its anatomical isolation from the upper and lower lobes.¹⁵

Imaging is the initial diagnostic modality for MLS and the condition presents as a homogeneous opacity abutting the right cardiac border with a loss of cardiac silhouette. The lateral view shows a characteristic wedge-shaped opacity extending anterior and inferior to the hilum. On CT, MLS appears as a trapezoidal opacity with its base towards the hilum and contiguous with the right cardiac border,¹⁵ as was seen in our patient.

The typical radiological appearance of an uncomplicated hydatid cyst is a well-defined, round to oval homogeneous opacity surrounded by normal parenchyma. The various classical signs include 'air-crescent sign or Monod sign' suggestive of impending rupture while 'serpent sign', 'water-lily sign or Camelotte sign', 'onion peel appearance or Cumbo sign' are seen in partially ruptured or ruptured pulmonary hydatid cysts. However, in patients with ruptured or secondary infection of these cysts, atypical radiological features can lead to diagnostic delay.³ CT scans also help in localising and confirming the cystic nature of the lesion as well as in determining the wall thickness.²²

Serology has emerged as an important diagnostic modality of pulmonary hydatidosis with sensitivity of serum antibody detection using indirect haemagglutination, ELISA, or latex agglutination, varying between 85% and 98% for hepatic cysts, 50% and 60% for lung cysts and 90% and 100% in cases of multiple organ cysts.²³ Our patient also had a positive echinococcal serology and the diagnosis was confirmed on detection of hooklets in the bronchial aspirate.

Bronchoscopy in pulmonary hydatid disease is usually performed when the radiological appearance is atypical and the diagnosis is not suspected,²⁴ as in our patient. This generally occurs in complicated pulmonary hydatidosis, which may mimic tuberculosis, malignancy, empyema or lung abscess.²⁵ Opinion regarding the role of FOB for the diagnosis of pulmonary hydatid disease is divided, as the risk of rupturing the uncomplicated cyst and the subsequent possible anaphylaxis remains.²⁴ However, recent literature suggests that FOB may be useful in the diagnosis as well as in the treatment, as was seen in our patient.

We also reviewed the existing literature regarding the use of FOB as a therapeutic modality in pulmonary hydatidosis and found four reports documenting five patients (table 2).^{26–29} Of the five patients, two were females^{28, 29} and one was male,²⁶ while demographic details were not available for the remaining two.²⁷ As in our patient, the diagnosis of pulmonary hydatid disease was not considered in any of the five patients prior to

Figure 5 Fibreoptic bronchoscopic image (postsuction) showing the medial segment of middle lobe bronchus opened completely.

Table 1 Summary of the six documented patients with pulmonary hydatid disease presenting as MLS along with the patient being reported

Serial number	Author, year of publication, country	Number of patients/ age/sex	Symptoms	Radiology	Bronchoscopy findings	Hydatid serology	Histopathology	Management
1	Jelić, <i>et al</i> , ⁴ 1977, Croatia (only abstract in English)	1/NA/NA	NA	NA	NA	NA	NA	NA
2	Modarres, <i>et al</i> , ⁵ 2008, Iran (only abstract in English)	1/71/M	Chronic cough, haemoptysis	CT chest: RML—consolidation	RML and RLL: inflammation, erythema	NA	Hydatid cyst	Surgical removal
3	Sokucu <i>et al</i> , ⁶ 2011, Turkey	1/50/F	Haemoptysis: 3 months	CXR: right paracardiac infiltrate CT chest: RML—consolidation with air bronchogram, acinar nodules, ground glass opacities	Medial segment of RML: whitish membranous material	Indirect Haemagglutination positive	Homogeneous, eosinophilic, lamellar cuticle membrane with chronic inflammatory cells.	Right middle lobectomy
4	Pejhan <i>et al</i> , ⁷ 2015, Iran	3/NA/NA	NA	NA	NA	NA	NA	Surgical removal
5	Present Study	1/45/F	Cough, haemoptysis	CXR: right mid and lower zones opacity abutting the right cardiac border, loss of cardiac silhouette Chest CT: MLS	Medial segment of RML: whitish membrane	Positive: <i>E. granulosus</i>	Bronchial aspirate: Hooklets	Completely extracted by FOB: albendazole—3 cycles

CXR, chest X-ray; *E. granulosus*, *Echinococcus granulosus*; FOB, fiberoptic bronchoscopy; MLS, middle lobe syndrome; NA, not available; RML, right middle lobe.

FOB. In all the five patients, FOB visualised a complicated pulmonary hydatid cyst, which was completely and successfully removed by gentle suction through FOB. Anaphylaxis was not documented in any of the four reports.

Chemotherapy with albendazole is indicated in non-operable cases, and also in preoperative and postoperative patients, to

lower the risk of recurrences caused by spillage of cyst contents. Chemotherapy, in our patient, was also administered to decrease the risk of recurrence.²³

Hydatid as a cause of MLS is a diagnostic rarity and in certain situations can be completely removed through FOB, obviating the need for surgery.

Table 2 Summary of the five documented patients with pulmonary hydatid disease with complete extraction through FOB

Serial number	Author, year of publication, country	Number of patients/ age/sex	Symptoms	Radiology	Bronchoscopy findings	Hydatid serology	Histopathology	Management
1	Bousnina <i>et al</i> , ²⁶ 2005, France (only abstract in English)	1/74/M	Cough, haemoptysis	CXR: upper third of left lung—heterogeneous opacity	Left superior bronchus: hydatid membrane	NA	NA	Completely aspirated by FOB
2	Racil <i>et al</i> , ²⁷ 2009, France (only abstract in English)	2/NA/NA	NA	NA	NA	NA	NA	Complete bronchoscopic extraction: two patients
3	Alavia <i>et al</i> , ²⁸ 2010, Iran	1/26/F	Cough, yellowish brown expectoration, exertional dyspnoea, night sweats, fever: 5 months	CXR: alveolar opacity—lower zone of right lung CT chest: RLL—alveolar involvement, mild pleural effusion and thick wall cystic lesion: posterior surface of liver	Posterior basal segment of RLL: whitish-yellow bright gelatinous membrane	NA	Membranous lesion with laminated layer and germinal covering	Therapeutic FOB: forceps fragmentation, complete suction albendazole
4	Sharif <i>et al</i> , ²⁹ 2011, Iran	1/22/F	Cough, haemoptysis: 2 months	CXR: left lower zone—homogeneous opacity CT chest: superior segment of LLL—consolidation	Superior segment of LLL: whitish endobronchial mass	Positive	Two layers of cyst wall with histopathological confirmation	Completely aspirated by FOB

CXR, chest X-ray; FOB, fiberoptic bronchoscopy; LLL, left lower lobe; NA, not available; RLL, right lower lobe.

Learning points

- Hydatid disease is a major public health issue throughout the world, especially in endemic countries.
- The myriad radiological appearances of pulmonary hydatid disease makes it a 'picturesque' clinical entity.
- Pulmonary hydatid disease presenting as middle lobe syndrome is a distinct diagnostic rarity.
- Middle lobe syndrome continues to intrigue physicians with its characteristic radiological picture caused by a wide spectrum of pulmonary diseases.
- The role of fibreoptic bronchoscopy in diagnosis and management of pulmonary hydatid disease is yet to be defined.

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