Pulmonary Tuberculosis in a Child with Mediastinal Inflammatory Myofibroblastic Tumor : A Rare Case

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ABSTRACT

Background : Tuberculosis is one of the most important global health problems in children. Mediastinal masses in pediatric age patients have a wide range of differential diagnoses, including benign and malign tumors and chronic infectious process. Mediastinal mass is a rare presentation of tuberculosis in children.

Objective : To report a rare case of pulmonary tuberculosis in a child with mediastinal inflammatory myofibroblastic tumor.

Case Report : A 4 year 2 month-old boy with chief complaint of cough since less than one month before admission. The cough accompanied with greenish yellow sputum, no coughing of blood, the cough during morning, noon, or night, and did not affected by cold air and there were no causative factors causing of coughing. The coughing did not improve by taking medication. The patient also complained of shortness of breathing since three days before admission accompanied by chest pain in the right side. The initial examination of the patient alert with a weakness of general condition and dypneu. Anthropometric examination obtained patient weight 13 kg, height 97 cm (WAZ =-2, LAZ >-2, WLZ > -2, WHO Child Growth Standards). Laboratory result on admission were leukocytes 10.120/uL, hemoglobin 11,3g/dL, hematocrit 36,9%, trombocyte 517.000, BUN 5mg/dL, SCr 0,36mg/dL, ALT 46U/L, AST 41U/L, Na 136nmol/L, K 4,3mmol/L, Cl 104mmol/L, Ca 8,7mmol/L, CRP 14,8mg/L. The thoracic MSCT result showed the tuberculosis

process. The Gene X-pert examination revealed MTB detected positive rifampicin susceptable The guiding Thoracic CT Scan FNAB result showed mediastinal mass with size 4.6 x 5.5 cm (benign spindle mesenchymal tumor, according with inflammatory myofibroblastic tumor). The patient given first line tuberculosis treatment, for a while the mediastinal inflammatory myofibroblastic tumor not be treated, will be reevaluated after the tuberculosis treatment was completed.

Conclusions : This case report describes a pulmonary tuberculosis with mediatinal inflammatory myofibroblastic tumor

Keywords : Pulmonary Tuberculosis, Chidren, Mediastinal Inflammatory Myofibroblastic Tumor

Introduction

Tuberculosis is one of the most important global health problems. There were 8.6 million new TB cases in 2012 and 1.3 million TB deaths worldwide. Pulmonary tuberculosis most frequently presents as patchy or lobar consolidation, it may present as community acquired pneumonia.[1] The mediastinum is the most common location of chest masses in the pediatric population. Mediastinal masses may be nonvascular or vascular masses and represent congenital anomalies, infections, benign and malignant neoplasms, and pseudomasses.[2] Tuberculosis is a rare cause of mediastinal masses in children. Tuberculosis was not found as a mediastinal mass in some previous case series. However, there are at least five cases of tuberculosis mediastinal masses in the literature. Clinical and radiological findings may be very similar among these entities.[3] Inflammatory myofibroblastic tumor (IMT) is a rare tumors of intermediate biological potential, it occurs rarely in the mediastinum.[4,5] There are no specific signs, radiologic manifestations, or symptoms related to IMT. It is difficult to make an accurate diagnosis just on the basis of the clinical presentation and radiologic manifestations. An accurate diagnosis is made based on the histopathologic and immunohistochemical study from a resected tumor.[5]

The most frequently reported site of IMT is in the lung, while it occurs less commonly in the liver, spleen, stomach, orbit, urinary bladder, and rarely in the mediastinum.[6] Of the 38 reported patients, 29 (76%) were children between 2 months and 16 years of age. Eighteen children (47%) were younger than 5 years. The children had mesenteric(17), retroperitoneal (4), gastric (2), omental (2), hepatic (l) , mediastinal (l), and diaphragmatic (1) tumors.[7] Five-year survival in case of inflammatory myofibroblastic tumors is 91.3%, but the recurrence rate after resection is 4% and

appears in locations of incomplete resection.[8]

The cause of inflammatory myofibroblastic tumors is uncertain. Some of the cases of inflammatory myofibroblastic tumors are thought to result from excessive inflammatory response following infectious disease, trauma or surgery.[9,10] Mediastinal IMT has been described to give rise to cough, dyspnea, and thoracic discomfort and pain, or may be discovered as an incidental finding on chest radiography.[4] In most cases, an accurate diagnosis is made based on the histopathologic and immunohistochemical study from a resected tumor. [5]

Case Report

A 4 year 2 month-old boy with chief complaint of cough since less than one month before admission. The cough accompanied with greenish yellow sputum, no coughing of blood, the cough during morning, noon, or night, and did not affected by cold air and there were no causative factors causing of coughing. The coughing did not improve by taking medication. The patient also complained of shortness of breathing since three days before admission accompanied by chest pain in the right side. The shortness of breathing is not associated with cold air. There were no history easily becoming short of breath during exercise or activity, no swelling in the hands, ankles or feet. The patient also complaining of fever since less than one weeks before admission, fever is subfebris sometimes accompanied with sweating cold at night. The patient had no history of seizure during fever. The patient also no history of allergy, there was no decreased of appetite and loosing weight.

From chest x-ray examination : suspecious pneumonia, right pleural effussion, suspecious lung consolidation dd mediastinum tumor. Family history of disease, his parents there were no history of allergy or asthma. There was no TB contact in the family, but his neighbour was infected with tuberculosis and had received anti tuberculosis treatment for six months.

The grandmother of the patient has been diagnosed as breast cancer and his aunt has been diagnosed as lymph node cancer. Patients had BCG immunization, there was BCG scar.

At the time of the initial examination of the patient alert with a weakness of general condition, he looks dypneu. Vital signs were blood presure 90/60, heart rate was 112 beat per minute, respiratory rate was 40 times per minute, temperature was 38^{o}C, oxygen saturation 98%. Examination of her skin reveals normal, no anemic on both conjunctiva, no cyanosis. From the head examination reveals normal, from the neck examinations there was no enlargement of neck

lymph nodes. Chest was symmetric and there was intercostal retraction. The heart sound was normal, without murmur neither gallop sound. The breath sound in the right and left chest was vesicular, there were rhonki wed rough in the right and left chest, and no wheezing were heard. The abdomen was flat with normal bowel sound, mass did not found, the liver and spleen were not enlarged, and no pain on palpation. The extremities were warm, with a capillary refill time of less than two seconds, and no edema in the extremities.

Anthropometric examination obtained patient weight 13 kg, height 97 cm (WAZ = -2, LAZ > -2, WLZ > -2, WHO Child Growth Standards). Laboratory result on admission were leukocytes 10.120/uL, hemoglobin 11,3g/dL, hematocrit 36,9%, trombocyte 517.000, BUN 5mg/dL, SCr 0,36mg/dL, ALT 46U/L, AST 41U/L, Na 136nmol/L, K 4,3mmol/L, Cl 104mmol/L, Ca 8,7mmol/L, CRP 14,8mg/L. Thoracic radiological picture showed consolidation in the right paracardial, infiltrate in the right parahiler, reticulo granuler pattern in left lung, The thickening of the left hilus. There appears a homogeneous in the lower right to upper lateral hemithorax (right pleural effusion). Opacity firmly on the right side hemithorax superior with differential diagnose of suspecious lung consolidation and tumor mediastinum. The thoracic MSCT result showed multiple lymphnode which partially conglomerated accompanied by calcification therein which causes narrowing of the right lobar medius bronchus and obstructive atelectasis of the right lung medius lobe, giant cavity with a solid portion therein in the superior lobe of the right lung may be a giant bullae with the pulmonary collapse portion of the fungus ball, multiple nodules In the superior segment of the right lung inferior lobe may be tuberculoma, the right organural pleural effusion that has been organization, thickening accompanied by upper right pleural irregularity. All of the above findings are illustrative of the tuberculosis process.

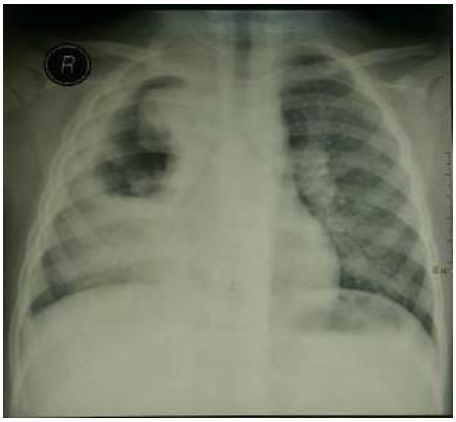

Figure 1. Chest Xray’s examination

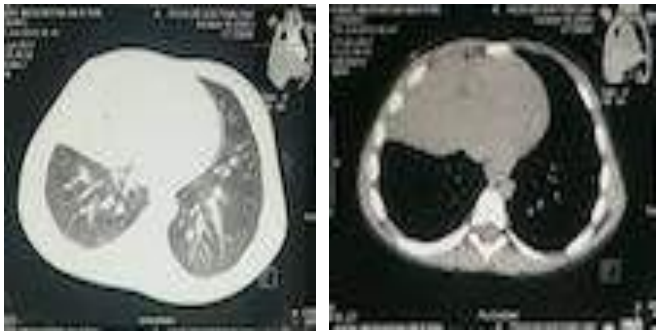

Figure 2. Thoracic MSCT of the patien

The result of thoracic radiological evaluation after completed of intensive phase treatment. It shows consolidated in the right suprahiler, looking right hilar thickening, according to the description of pulmonary tuberculosis.

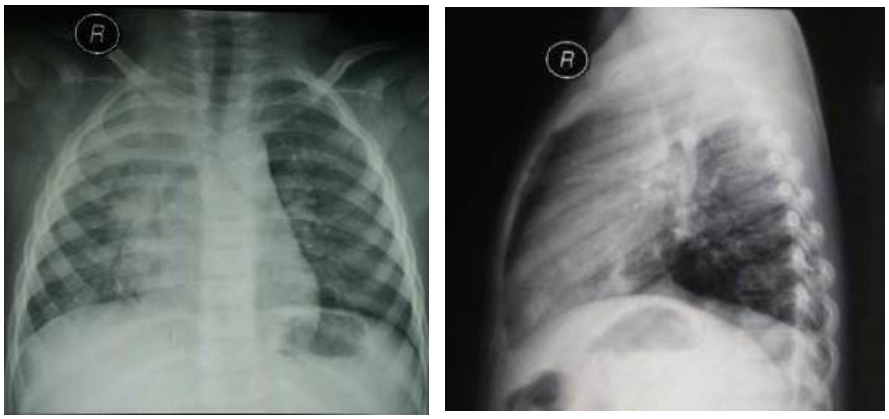

Figure 3. Chest Xray's evaluation after completed of intensive phase treatment

Discussion

This case present a 4 year 2 month-old boy came with chief complains of cough since less than one month before admission accompanied by chest pain in the right side and dyspnea. The patient also complained of fever since one weeks before admission. From thoracic radiological picture showed right pleural effusion, suspecious lung consolidation differential diagnosis tumor mediastinum.

There are various clinical, laboratory and radiological methods currently used to diagnose respiratory diseases. Tuberculosis and pneumonia can overlap, it can sometimes be difficult to differentiate between the two entities both clinically and radiologically.[12] Pneumonia and tuberculosis are both diseases of the lungs. Pneumonia is an inflammatory condition within the

lungs produced as a result of infection that primarily affects the alveoli. It is usually caused by viral or bacterial infections. The common signs of pneumonia include fever, chills, productive cough, and chest pain. The most common bacteria involved are *Streptococcus pneumoniae, Staphylococcus aureus, Escherichia coli,* and *Haemophilia influenzae*. Tuberculosis is an infection of the lungs caused by Mycobacterium species, the most common pathogen being *Mycobacterium tuberculosis.* The symptom of tuberculosis include rapid and frequent breathing, chronic cough, haemoptys, weakness, and fatique.[13]

Table 1. Comparison of Pneumonia and Tuberculosis

Clinical Feature	Pneumonia	Tuberculosis
Type of Microorganisms Involved	Bacteria, Virus, Fungi	Bacterial
Species of Microorganisms Involved	Streptococcus, Staphylococcus, Escherichia, Chlamydia, Legionella	Mycobacterium Tuberculosis
Organ System Affected	Lungs	Lungs, Skeletal System, & Genito-Urinary System
Physical Signs	Fever, productive cough, dyspnea	Chronic Cough, Fever,Weakness, Haemoptysis
Quantity and Nature of Sputum	Chest Radiographs	Mantoux Test,sputum examination, Chest Radiographs
Radiological presentation	Consolidation and infiltration, with or without pleural effusion	Granulomas, consolidation and cavitation.

In this patient according from clinical symptoms and chest X-ray result with differential diagnose of pneumonia and pulmonary tuberculosis. This patient given ampicillin sulbactam intervenous and gentamicine intervenous.

Antibiotic therapy improves the outcome of pediatric bacterial pneumonia; because it is difficult to define the etiological agent, however, treatment is almost always empirical. Ampicillin/sulbactam has a broad spectrum of antimicrobial activity, being effective against Gram-positive (e.g. Staphylococcus aureus, coagulase-negative staphylococci, enterococci, S. pneumoniae), Gram-negative (including H. influenzae, Moraxella catarrhalis and E. coli), and anaerobic (Bacteroides fragilis, Bacteroides spp).[14]

Pleural effusions (liquid in the pleural space), which occur less frequently in children than in adults, can be caused by a variety of infectious and noninfectious diseases. Pleural effusions in children most commonly are infectious (50% to 70% parapneumonic effusion); congestive heart failure is a less frequent cause (5% to 15%), and malignancy is a rare cause. [15]

The clinical picture and presenting symptoms of pleural effusion depend on the underlying disease and the size and location of the effusion. In this regard, the recent history of upper respiratory tract infection, bronchitis, or pneumonia is expected in effusion due to infectious pneumonia that can be manifested by persistent fever, cough, anorexia, malaise, tachypnea, dyspnea, and chest pain. The most common manifestations of Pleural effusion with tuberculosis basis include cough, pleuritic chest pain, dyspnea, night sweats, fever, hemoptysis, and even weight loss. In malignancies, some patients maybe asymptomatic that manifested only by cough and low grade fever, however in higher stages, respiratory distress or mediastinal mass can be observed. In pleural effusion due to congestive heart failure or nephrotic syndrome, the symptoms range from asymptomatic status to diseases specific manifestations. [16]

Table 2. Common Causes of Pleural Effusions in the Pediatric Population[15]

Cause	**Incidence**
Pneumonia (parapneumonia effusion)	50% - 70 %
Renal disease	9%
Trauma	7%
Viral disease	7%
Malignancy	5% - 10%

Congenital heart disease	5% - 11%
Other (liver failure, sickle cell anemia, meningitis)	3%

The Gene X-pert examination in this patient revealed MTB detected positive rifampicin susceptable. Currently available diagnostic tools for tuberculosis rely on AFB microscopy, culture growth, and molecular DNA detection (eg, Xpert MTB/RIF test) of M. tuberculosis in specimens, largely in sputum. Clinical diagnostic approaches using symptoms and signs, including scoring systems, have been used to facilitate the diagnosis of tuberculosis in children. However, they are not standardized, validated, nor adapted to malnourished children or children living with HIV.[18]

The patient got firstline tuberculosis treatment with Rifampicin 200mg daily, Pirazinamide 500mg daily, Isoniazid 150mg daily, Ethambutol 200mg daily and Prednisone 10mg-10mg-5mg (~ 2mg/kgBW/day) during 1 month with tappering off. After intensive phase for 2 months and will continue to the continuation phase of 4 months with 2 drugs.

According to *Petunjuk teknis manajemen dan tatalaksana TB anak 2016* as children with positive smear tuberculosis disease were recommended to given 4 kinds of OAT drugs in intensive phase followed by Rifampicin and INH for 2 months continuation phase. Corticosteroid can be given for meningitis TB, endobronchial TB, pericarditis TB, milliary TB, pleural effusion TB and abdominal TB. Prednison can be given 2-4 mg/kgBW/day. Tappering off after 2 weeks of drug administration.[20]

The guiding Thoracic CT Scan FNAB result showed benign spindle mesenchymal tumor, according with inflammatory myofibroblastic tumor. Inflammatory myofibroblastic tumor (IMT) is a rare disease. The most frequent site of IMT is in the lung, while it occurs rarely in the mediastinum. We report an IMT in the mediastinum, which showed a heterogeneously enhanced irregular mass in the anterior mediastinum and a small pericardial effusion on computed tomography (CT). The mass was resected under a thoracoscopic surgery. [5]

Surgical resection is the treatment of choice. Complete excision seems to be curative and prevents recurrences. Radiotherapy, chemotherapy or corticosteroid therapy can also be given where surgical intervention is contraindicated, and where there is multifocal disease or incomplete removal of the mass. Treatment with non-steroidal anti-inflammatory drugs (NSAIDs) has also been reported. These non-surgical treatments can achieve tumour shrinkage or even resolution. Operative technique depends on tumour location, the existence of local invasion and the patient's

age and condition. Surgical management takes the form of mass excision, lobectomy or pneumonectomy, depending on tumour size and location. The most controversial aspect of surgery, especially in small children, is the resection of a mass arising from the main stem of the bronchus. Tumors in this location mostly require a total pneumonectomy; this usually has serious consequences for children. They may be afflicted with life-threatening right post-pneumonectomy syndrome. The reduction of pulmonary tissue is usually significant and after pneumonectomy patients may develop chest or spinal deformities and fail to thrive. Recent standards recognise the utility of a less destructive sleeve lobectomy in cases with nodal status limited to N2. The operative technique is more difficult than pneumonectomy, and should only be carried out by experienced surgeons, especially in small children and infants.[27]

Conclusions

This report describes the case of a child with pulmonary tuberculosis with mediastinal inflammatory myoblastic tumor. The patient came with chief complaint of cough since one month, fever since one weeks before admission, and chest pain in the right side accompanied by dyspnea. There were no history of blood cough, decreased of appetite and loosing weight. From laboratory result was normal limit. A chest radiograph revealed right pleural effusion, opacity firmly on the right side hemithorax superior suspecious mediastinum tumor. The result of the first sputum smear showed negative and the second sputum smear also showed negative, tuberculin test was negative without induration. The Gene X-pert examination revealed MTB detected positive rifampicin susceptable. The guiding Thoracic CT Scan FNAB result showed benign spindle mesenchymal tumor, according with inflammatory myofibroblastic tumor. The patient given firstline tuberculosis treatment, and for the inflammatory myofibroblastic tumor while not be treated will be reevaluated after the tuberculosis treatment was completed.

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