Pulmonary Inflammatory Myofibroblastic Tumor in a Male Child; a Case Report

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Abstract

Pulmonary inflammatory myo-fibroblastic tumor (IMT) is a rare condition in normal population and specifically in pediatric population. We reported a 9-year-old male child presented by cough and intermittent fever and weight-loss that was mostly suggestive for infectious process. We reviewed the consideration of diagnosis and treatment and other previous cases.

Pulmonary Inflammatory Myofibroblastic Tumor in a Male Child; a Case Report

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Abstract

Pulmonary inflammatory myo-fibroblastic tumor (IMT) is a rare condition in normal population and specifically in pediatric population. We reported a 9-year-old male child presented by cough and intermittent fever and weight-loss that was mostly suggestive for infectious process. We reviewed the consideration of diagnosis and treatment and other previous cases.

Keywords: inflammatory myo-fibroblastic tumor, pulmonary tumor, pediatric population

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Background

Occurrence of primary pulmonary neoplasms in pediatric patients are uncommon. Inflammatory myofibroblastic tumor (IMT) is a rare neoplasm, which has a moderate risk of malignancy appearing mainly in children and young adults (1, 2). Its nature is in majority of cases benign and rarely can metastases however could commonly relapse (3). This tumor is consist of different inflammatory cells, which its diversity is from primarily myo-fibroblastic cells to plasma cells in pathologic exam (4). IMT is defined as "a lesion composed of a proliferation of myo-fibroblastic spindle and stellate cells with abundant eosinophilic cytoplasm mixed with infiltrative plasma, inflammatory cells, lymphocytes and eosinophils" in world health organization (WHO) (5). IMT is known by numerous terms, including plasma cell granulomas, inflammatory pseudo-tumor, fibrous histiocytoma and pseudo-lymphoma (6), and may occur in a wide range of anatomical locations such as the lungs, omentum, bladder, spleen, breast, pancreas, liver, colon, spermatic cord, prostate, peripheral nerves, soft tissue, and orbit. About one-third of these tumors are found in the respiratory tract (6). We here report a case of IMT of the lung.

Case presentation

A 9-year-old male child with chronic productive cough and occasional fever since six months ago referred to our clinic. He had severe weight loss in this period. Two months before admission, he had massive hemoptysis and clubbing. There was no shortness of breath or Respiratory distress. He had a history of tonsillectomy 2 years ago and no exposure to tuberculosis case. Theophylline, Guaifenesin and Cefixime have been administered for him. There no history of familial pulmonary disorder. At admission, his vital signs were stable. In the examination, there was decreased pulmonary sound in right lung. There was clubbing in upper and lower limbs fingers. His chest x-ray is shown in figure1. In his pre-admission computer tomography (CT) scan, a mass-like foci in the right lung was reported.

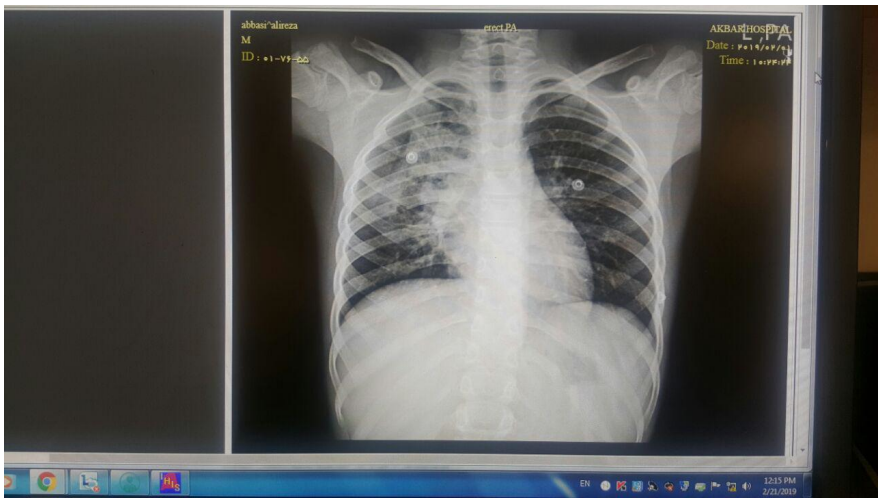

Figure1- Chest X Ray

He was admitted in our center for more evaluation. After admission antibiotic therapy and supportive care was performed. He underwent bronchoscopy. In fibro-optic bronchoscopy, upper division of right bronchus was completely obstructed. After washing, various cystic membranes was seen in the airway. The mucosa was erythematous and inflamed with dense secretions that was suctioned (Figure2).

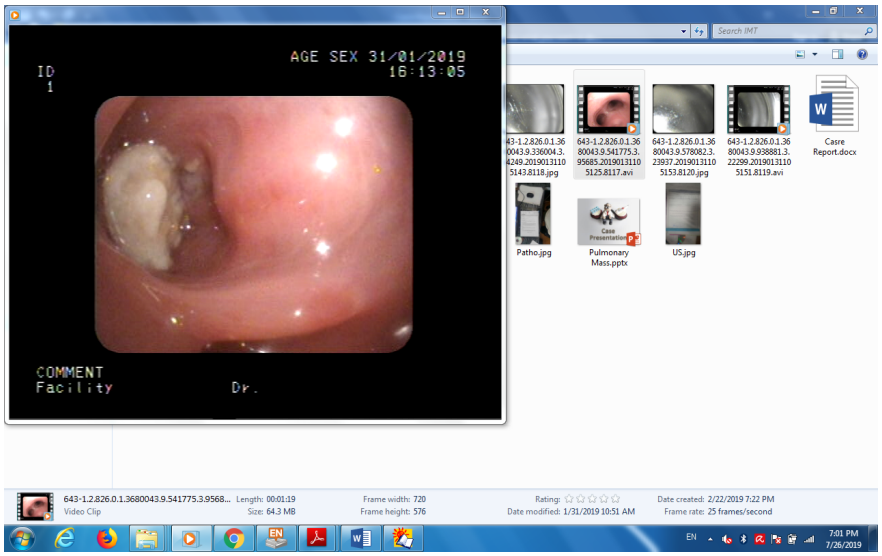

Figure2- Bronchoscopy; the upper division of right bronchus that is obstructed by a mass.

There was no pathologic issue in his chest and abdominal sonography. His first laboratory data showed a leukocytosis (WBC 22,300 with PMN dominant pattern (85%)). ESR and CRP was in nearly normal range. Anti Echinococus antibody was negative. Other laboratory data was in normal range. After surgical consult, he was candidate for surgery and underwent right upper lobectomy and wedge resection. After thoracotomy, mass-like lesion with granulomatous tissue was resected. There was a fistula that was closed by resection. Chest tube size 28 was inserted and the sample was sent for pathologic examination.

Pathologist assessed wedge biopsy of lung, with brown outer surface, measuring 2.5×2 ×1 cm, and a central creamy-yellow nodular lesion, measuring 1 cm in diameter. Sections of the lung tissue with pleural fibrous thickening, revealing prominent interstitial nodular lymphoid proliferations surrounding large epithelioid cells (resembling granulomas), few of them with large nucleolated cells without necrosis. A diffused intra-alveolar infiltration of foamy histiocytes were noted. In addition, a central focus of pulmonary necrosis with mixed acute inflammatory cell infiltration (abscess formation) was present. Foci of hemorrhage was also repoted. In Zhiel-Neelson staining acid-fast bacilli were not seen. PPD test was negative. Because of nodular lymphoid proliferation of pulmonary interstitium with one necrotic area and abscess formation, it was suggestive for interstitial lymphocytic pneumonia or granulomatous disease and we ruled out it by checking CD20, CD3, CD15 and CD30 that shown no definite evidence of Hodgkin lymphoma. Inflammatory myo-fibroblastic tumor (IMT) was diagnosed by its pattern (Figure3). Patient discharged after the surgery and is following up to now for probable relapse.

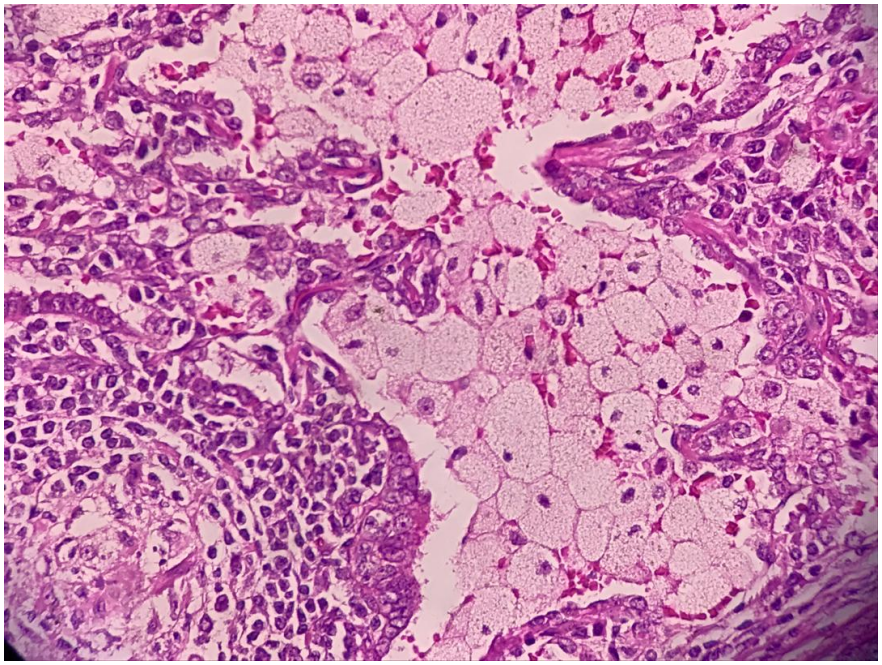

Figure3- Histopathological H&E staining showing inflammatory cells

Discussion

IMT is one of the rare low-to-intermediate grade sarcomas. Initially it was thought to be an inflammatory response to various stimuli, but recent studies have proved IMT to be neoplastic and can recur locally and metastasize (7). It has also been suggested that trauma, surgery, autoimmune etiologies, inflammation, and infections such as Epstein-Barr virus or human herpes virus could result in the development of IMT (8).

IMT was first described in the lungs but later was also found in other sites such as the orbit, spleen, genitourinary tract, mesentery, cardioesophageal junction, breast, central nervous system, and larynx. The larynx has been a very rare site for involvement in IMT (9). Children with IMT may exhibit symptoms of chronic inflammation as a low-grade fever, weight loss, anemia, thrombocytosis, polyclonal hyper-gammaglobulinemia, and elevated sedimentation rate. Several cases are asymptomatic and are detected only incidentally on imaging studies. Among patients with end-obronchial lesions, symptoms of bronchial irritation such as cough and hemoptysis may be accompanied by chest pain (10).

In our case, chest x-ray, history of weight loss and occurrence of intermittent fever and hemoptysis, beside endemic status, made us suspicious to infectious process like tuberculosis or echinococcosis. Because of the negative staining of BAL sample and laboratory data, we thought about the non-infectious process like malignancies because of weight loss. Despite of doing bronchoscopy and HRCT, the diagnosis was last after histopathological assessment. According to previous studies, there are only 26 published cases of pediatric pulmonary IMT (the age between 3 to 13 years), even though the real incidence is presumed to be higher (3, 11). Peripheral lung lesions appear to be more frequent than central and endobronchial tumors that may be present about in 10% of the cases resulting in bronchial obstruction and atelectasis (12).

IMT can be sometimes diagnosed as incidental finding on a routine CXR (13). In all previous cases, at the time of presentation, patients had fever, respiratory distress, arthralgia, clubbing, night sweat, vomiting, and hemoptysis and at the onset, fever and cough were the commonest symptoms (3). There has been an ongoing controversy whether an IMT is a reactive lesion or a true neoplasm (14). Although its incidence is fairly scarce, the existing literature clearly defines its relative similarities in terms of clinico-pathological and radiological findings and almost uniformly favors surgical resection as a mainstay for the most efficient management strategy. The recurrence rate remains low and a 10-year survival rate is around 80% (15).

Treatment is primarily a complete but conservative surgical excision. This approach is necessary to prevent recurrence (16, 17). An appropriate histologic assessment should be obtained before the surgery (needle biopsy by bronchoscopy), in order to avoid an unnecessarily procedure (17).

Conclusion

Pulmonary IMT is a rare disorder with significant complications among the children population. It may be non-specific in presentation in this population and patients undergo different antibiotic treatment before definite diagnosis. While radiological techniques can help in diagnosis, confirmation of the diagnosis should be performed by histopathological assessment. The choice of treatment is based on complete surgical resection.

Conflict of interest

There is no conflict of interest.

We declare that none of the authors listed on the manuscript are employed by a government agency that has a primary function other than research and/or education. None of the authors is submitting this manuscript as an official representative or on behalf of the government.

Acknowledgement

Nothing.

Key clinical message

Cough is a serious sign in children that should be regarded by physicians. Pulmonary IMT may be non-specific in presentation in this population. Despite of its rare prevalence, pulmonary mass like IMT should be considered in the list of differential diagnosis of cough.

Author contribution

Dr. Seyed-Javad Seyedi, Dr. Amin Saeidinia and Dr. Parisa Dehghanian were responsible for the case treatment, development of methodology, writing of the manuscript and confirming final version.

Data availability statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Patient consent

The authors have confirmed that patients' consent have been signed and collected in accordance with the journal's patient consent policy.

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