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CHRONIC COUGH IN A 10- YEAR OLD BOY AS A FIRST PRESENTATION OF INFLAMMATORY MYOFIBROBLASTIC TUMOR (IMT): A CASE REPORT

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a rare benign tumor with an unknown origin. Its clinical and radiological manifestations are variable and non specific, also half of them are asymptomatic while cough, hemoptysis, dyspnea are possible to see. Therefore diagnosis is too hard to establish unless an exactly evaluation by an expert pathologist on a biopsy from surgical resection .The standard treatment for diagnostic and therapeutic reasons is a complete resection whereas incomplete resection increases the risk of recurrence. Here we report a 10-year old boy with prolonged cough and collapse-consolidation in his chest x-ray who referred to our pediatric center in north east of Iran.

Key words: Inflammatory myofibroblastic tumor, cough, collapse, pediatric.

CASE PRESENTATION

A 10- year old boy referred from Nishabur in north east of Iran to our center in Dr.Sheikh pediatric hospital in Mashhad, Iran in January of 2015, who was suffering from chronic cough from one month ago without any response to medical treatment and he has had a history of choking 20 days ago. He had received variable antibiotic, mucolitics and a broad spectrum of anti cough drugs and he was admitted to a local hospital due to pneumonia that a segmental collapse in left lung was seen in his chest-x-ray. In past medical history he had a history of tonsillectomy his. His growth and developmental process was normal up to that time. In his family history he was the first child of a normal family and the second child was healthy up to now. His parents were relative. His grandmother has had asthma. He had received common protocol of national vaccination in Iran.

Therefore he was referred to our hospital for evaluating prolonged cough and collapse in left lung. In order to roll out of cystic fibrosis sweet test was done and normal results were obtained. According to diagnostic evaluations also any immunodeficiency was not seen as his Cell Blood Count (CBC), Nitroblue tetrazolium (NBT) test were normal. In addition assessment of immunoglobulins such as IgE, IgA, IgM, IgG revealed normal values. Anti HIV antibody was negative. Complement rout evaluated by measurement of CH50 which was in normal range. Erythrocyte Sedimentation Rate (ESR) was a little high and non-quantitive C - reactive protein was negative. No organism was detected in blood cultures. Routine laboratory data like serum level of sodium, potassium, calcium, magnesium, phosphor and blood gas analysis, liver and renal function test also showed normal results. There was not any coagulopathies in our evaluations.

In chest imaging collapse and consolidations existed in left lung (figure3). Mediastinal and lung CT scan with intravenous contrast injection revealed reduced volume of left lung and mediastinal and cardiac shift to left in addition to collapse-consolidation in whole left lung expect to apicoposterior segment in superior lobe. Mass like consolidation in left hill associated with enhancement had cut the left main bronchus. Adenopathy, plural and pericardial effusion were not seen .Soft tissue, chest wall, some cuts of upper abdomen were normal (figure3, 4).

Finally diagnostic rigid bronchoscopy was performed particularly for roll out of foreign bodies or any other complications in his air ways. It was notable that there was a mass in left lung with a compression effect on left main bronchus. According to later thoracotomy, superior left lobectomy

(Sleeve surgery), left lung tumor were resected and some biopsies were sent to a professional pathologist who was specialist in pediatric tumors. A chest tube was inserted to left hemi thorax. The pathologist reported Inflammatory myofibroblastic tumor (inflammatory pseudotumor) as the definite diagnosis (figure1, 2). Results of immunohistochemical evaluation also approved the diagnosis of Inflammatory myofibroblastic tumor (inflammatory pseudotumor) as below: Actin : focally positive weakly ,desmin: negative , synaptophysin : negative, chromogranin: negative.

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare benign tumor, accounting for 0.7% of all lung tumors (1). The frequency of this lesion in the lung is 0.04–1.0% among the general population (2).
Its origin is unknown, but recent studies have shown that it is a true tumor rather than a reaction process (1). It should be considered when dealing with primary lung tumors in children, adolescents, and nonsmoking adults. It is, from a pathologic point of view, a benign tumor composed of a spindle cell proliferation and inflammatory cells, but, its clinical behavior, however, is variable with a benign evolution at one, and a malignant evolution with recurrent and metastatic disease at the other end of the spectrum (3). Therefore it is usually benign, although rare cases of malignant behavior have been reported (4).

The first description of this tumor was given by Brunn in 1994((1, 5). In 1955, this tumor was given the name plasma cell granuloma by Lane *et al*. since plasma cells were predominant in the tumor (5). Thereafter many different terms were used to describe this tumor, the most common being inflammatory pseudotumor (5). Childhood inflammatory pseudotumor is the most common isolated lung lesion in children, usually is asymptomatic (4).
Inflammatory myofibroblastic tumor is defined by WHO as a myofibroblastic spindle cell soft tissue tumor with infiltrative plasma cells, lymphocytes, and eosinophils. It has been classified as an intermediate neoplasm in the current WHO histological typing of tumors of soft tissue and bone (5).
Most of the reported cases are between 27 and 50 years of age, and under 40, no geographic or ethnic predisposition has been reported (2). It is predominantly seen in older children, without any sex predilection (1, 6).

Its clinical and radiological manifestations are diverse and non specific. That's why diagnosis is difficult to establish unless a surgical resection is performed (1).

Inflammatory myofibroblastic tumor has a number of pseudonyms. Some of them are inflammatory pseudotumor, plasma cell granuloma, fibroxanthoma, histiocytoma, sclerosing hemangioma, hyalinizing granuloma, etc (6).

The other synonyms used were post inflammatory tumor, xanthofibroma, xanthogranuloma, fibrohistiocytoma, plasma cell granuloma, pseudosarcomatous myofibroblastic tumor, and invasive fibrous tumor of the tracheobronchial tree (5, 7). Inflammatory myofibroblastic tumor is now considered a more appropriate term as it takes into consideration the ultrastructural and immunohistochemical properties of this tumor (5).

IMTs are benign solid tumors that arise in many anatomic sites. The lung is the most common site, but other sites are also reported, including the small and large bowel mesentery, mediastinum, retroperitoneum, omentum, spleen, spermatic cord, prostate, peripheral nerves, soft tissue, orbit and diaphragm (8). IMT is considered a benign tumor with local aggressive course but is not a malignancy in itself (8).

Because of similar morphology of theses lesions, only immunohistochemical investigations allowed the correct final diagnoses. Histological examination was necessary for diagnosis (1).

PATHOGENESIS

There are many uncertainties about the pathogenesis of IMT. Several hypotheses have been proposed, most of which postulate an unchecked or exaggerated immunologic response of myofibroblasts to a viral or foreign antigen-antibody reaction (6, 7).

Indeed, 30% of cases are closely related to recurrent respiratory infections which are caused by several microorganisms such as Mycoplasma, Nocardia, Actinomycetes (1). Also HHV-8 and EBV DNA sequences have been documented in some tumors indicating that viral infection may have some role in pathogenesis of this tumor (5).

Other studies, however, suggest that it might be a true neoplasm due to the presence, at the myofibroblastic component, of a fusion gene involving the ALK gene, a tyrosine kinase oncogen located on chromosome 2p23, initially found to be arranged in anaplasic large cell lymphomas (1). This fusion leads to constitutive over expression of the ALK, causing cell proliferation (1). Coexistence of gene susceptibility and recurrent infection have also been reported (9).

CLINICAL MANIFESTATIONS

Approximately half of the patients are asymptomatic, whereas 26–56% of the patients have symptoms including cough, hemoptysis, dyspnea and chest pain (2). Nearly, 70% patients will be symptomatic with complains of fever, cough, dyspnea, and rarely hemoptysis (5). However, most patients are asymptomatic and the tumor is discovered incidentally on a chest X-ray performed for another reason. Weight loss and anorexia are rare (1). It was notable that chronic cough was the only symptom in our patient

The presentation of IMT varies depending on the site and includes fever, weakness, abdominal pain, upper GI bleeding, weight loss, vomiting, poor appetite, gastroesophageal reflux, pallor, growth retardation and ascites. These symptoms can appear either alone or in a combination (8). Clubbing, and arthralgias may be noted. Generally no specific findings on physical or laboratory examinations exist (7).

DIAGNOSIS

Radiographic images and invasive diagnostic procedures, including bronchoscopy and percutaneous fine needle aspiration biopsy, are considered insufficient for diagnosis (7) as they may contain only inflammatory cells (9). Therefore, open lung biopsy or videothoracoscopic resection is often necessary (2).

Consequently, surgery is crucial for both diagnostic and therapeutic reasons and frozen section histological examination is also subject to errors (7). Early diagnosis is crucial for the possibility of tissue-sparing treatment; most patients are diagnosed at an advanced stage (9). Only about 6% of patients are diagnosed on the basis of a biopsy specimen (9).

Radiological Assessment

CXR

Radiological aspects are variable and nonspecific. Most patients (87%) have a mass or a pulmonary nodule, solitary, rarely multiple (5%), measuring 1 to 10 cm in diameter, sharply limited, smooth or bumpy. The lesion is located on the periphery with a predilection for the lower lobes (1, 6, 7). The mass lesion is discovered incidentally in chest x rays (2).

Multiple lung masses, pneumonic consolidation, atelectasis, hilar or mediastinal masses, pleural effusion, and cavitation are unusual (7).

CHEST CT SCAN

Thoracic computed tomography reveals a single nodule and mass in 90% of these patients, and multiple nodules in 5%. Secondary infiltration of the hilus, mediastinum and airways were reported in 16% of the patients (2). It specifies the tissue or cystic nature of the tumor, its vascular behavior and it assesses the locoregional extension. Calcifications and cavitations are rare. Pleural effusion is seen in less than 10% and atelectasis in 8% of cases (1). Most inflammatory myofibroblastic tumors are well-circumscribed, nonencapsulated parenchymal masses, with less than 5% invading the mediastinum and chest wall (5).

In a study a case of pulmonary IMT in a 15-year-old male with malignant features on radiographic and F-Fluoro-deoxyglucose positron emission tomography imaging was reported (3)**.**

Laboratory investigations

Laboratory investigations are not specific. The most common finding is microcytic hypochromic anemia refractory to iron therapy (90.9%), but other laboratory findings, such as a high erythrocyte sedimentation rate, thrombocytosis, eosinophilia and hypergammaglobulinemia, may be seen (8).

PATHOLOGY

The pediatric primary pulmonary neoplasms represent a wide range of pathology from benign to malignant end of the spectrum. They are quite different in their histopathologic distribution from that of adults (6).

However, in children, the major bulk of benign tumors consists inflammatory myofibroblastic tumors, chondromatous hamartomas, granular cell myoblastomas, leiomyomas, bronchial chondromas and teratomas (6), whereas the list of malignant lung tumors constitutes of carcinoids, mucoepidermoid carcinomas, pleuropulmonary blastomas (PPBs), pulmonary blastomas (PBs), adenoid cystic carcinomas, sarcomas like fibrosarcomas, leiomyosarcomas, rhabdomyosarcomas (6).

Gross description of inflammatory myofibroblastic tumors includes Solitary, small peripheral nodules, yellow, firm, covered by intact pleura or polypoid bronchial mass (4).The tumor is well circumscribed but not encapsulated, firm and homogeneous, with sometimes some foci of necrosis (1).

Macroscopically it is containing variable inflammation, haemorrhage, calcification, and rarely cavitation (7).

Micro description includes Plasma cells (often abundant), lymphocytes, histiocytes and myofibroblasts. May have vascular proliferation, collagenous or hyalinized stroma, myxoid change, xanthoma cells, hemosiderin May resemble nodular fasciitis, fibrous histiocytoma or fibromatosis (4). Histologically there is fibrosis and intra-alveolar fibrosis, especially at the margin of the lesion (6). Inflammatory pseudotumours typically consists of variable amounts of stromal and cellular elements, with the myofibroblast, a cell involved in tissue repair, recognized as the principal cell-type (7). In fact a lesion formed of varying proportions of spindle cells of myofibroblastic type, arranged in a fibrous, myxoid or calcified stroma, associated with an inflammatory component predominantly lymphocytic and plasmacytic, with a variable component of eosinophils (1).

Because of the diversity of clinical and radiological manifestations of IMT, diagnosis is difficult to establish without chirurgical management (1).

Pathological subtypes

IMT includes three histological subtypes (1).

- A richly vascularized and myxoïde resembling fasciitis or granulation tissue.
- Onother is a more compact fascicular spindle cell proliferation with variable collagenized regions and lymphoid nodules, resembling fibromatosis
- A very sclero-hyaline, slightly cellular pattern, looking more like a desmoid tumor (1).

Another different histologic patterns have been described (8).

- Myofibroblasts have a spindle appearance and are distributed loosely in a myxoid stroma.
- The second is more densely packed stromal cells intermingled with an inflammatory component.
- The third is characterized by hyalinized, hypocellular stroma.

These three patterns can be found in a single tumor intermixed closely . All of these features resolve after resection of the tumor (8).

Matsubara et al (2) categorized inflammatory pseudotumor into three groups based on cellular types and main histologic properties:

A) Organized pneumonia formed by gradual healing of the intraalveolar exudation (44%),

B) Fibrous histiocytoma formed by storiform proliferation of plasmocyte and lymphocyte aggregates (44%), and

C) lymphoplasmocytic type formed by the aggregation of both plasmocyte and lymphocytes (12%)(2).

Depending on the major histopathologic features, inflammatory pseudotumours are divided into the following types:fibrous histiocytoma, lymphoplasmacytic, and organising pneumonian (7).

IMMUNOHISTOCHEMISTRY

Immunohistochemistry showed reactivity for vimentin and smooth muscle actin. Immunohistochemical positivity for anaplastic lymphoma kinase(ALK) is detectable in just over half of the cases with cytoplasmic staining, more rarely at the nuclear membrane (1).

Immunohistochemistry has demonstrated the polyclonal nature of plasma cells with immunoglobulin G predominance (7).

Some studies have suggested that IMT is a true neoplasm on the basis of characteristic gene alterations such as fusion of the gene encoding anaplastic lymphoma kinase (ALK) on chromosome 2p23 and tropomyosin 3 (TPM3), which results in a functional chimeric protein (9). This phenomenon (TPM3-ALK) is detected in 30% of paediatric IMT cases (9).

In a study (10) Anaplastic lymphoma kinase (ALK) expression was negative in all pulmonary samples by immunohistochemistry (IHC), however, rearrangement for ALK locus by fluorescence in situ hybridization was found in one lung and in two CNS samples. These findings may reflect higher sensitivity of the molecular biologic procedure compare to traditional IHC practice. In our pediatric experience, 25% of patients with lung IMT developed CNS lesions; therefore they consider that CNS screening in these patients should be considered, at diagnosis and later during follow up (10).

The genetic landscape of this tumor is incompletely understood and therapeutic options are limited. Although 50% of IMTs harbor anaplastic lymphoma kinase (ALK) rearrangements, no therapeutic targets have been identified in ALK-negative tumors (11) .A study in 2014 reported for the first time that IMTs harbor other actionable targets, including ROS1 and PDGFRβ fusions. Molecular tumor profiling revealed a ROS1 fusion, and he had a dramatic response to the ROS1 inhibitor crizotinib (11).

PATHOLOGIC DIFFERENTIAL DIAGNOSIS

Pathologic differential diagnosis includes organized pneumonia, lymphoma and solitary fibrous tumor. The mode of tumor growth, the low mitotic index, the polyclonality of lymphoid markers and the negativity of CD34 usually remove most of these diagnoses. Other differential diagnoses are desmoid fibromatosis, angiomyofibroblastoma, fibrosarcoma, leiomyoma, and malignant fibrous histiocytoma (1).

Differentiating IMT from sarcoma may be challenging because of the local invasiveness of both pathologies. The lack of mitosis and nuclear atypia in IMT can differentiate it from sarcomas (12). The tumor mainly needs to be differentiated from lymphoma, plasmacytoma, organizing pneumonia and the rare malignant fibrous histiocytoma (6).

TREATMENT

The choice treatment for diagnostic and therapeutic reasons is a complete resection. An incomplete resection increases the risk of recurrence (1). While complete resection, wherever possible, is the standard therapy and results in excellent survival (4, 6).

Wedge resection is adequate treatment if removal is complete. Lobectomy should be performed if it is required for complete resection and if the patient's pulmonary reserve is adequate. Non-surgical treatment modalities including radiotherapy, chemotherapy, and corticosteroids may have a place in the setting of incomplete surgical resection, multifocal disease, postoperative tumour recurrence, or contra-indications to lung resection (7). Although spontaneous regression may occur, local expansion may cause significant morbidity and occasional death (7).

Controversy still exists regarding the role of chemotherapy in IMT patients. According to Kovach *et al.*, chemotherapy has been reserved for patients for whom resection is neither complete nor possible. The dosage and regimen of chemotherapy should be adjusted according to the biologic aggressiveness of the tumor, and there is no good evidence to support chemotherapy after complete resection regardless of tumor biology (13). Chemotherapy is useful in cases of multifocal, invasive lesions or in cases of local recurrence (1).

Radiation, steroids and non steroidal anti inflammatory drugs have been used to treat inflammatory myofibroblastic tumors with variable and inconsistent results. Hence, surgical excision still remains the treatment of choice (5).

Radiation treatment showed some benefit in pulmonary IMT .On the other hand, failure of radiation therapy in treating IMT has been reported, confirming surgical excision to be the treatment of choice(8). Radiation therapy is used in palliative treatment of this tumor, decreasing the mass effect of IMT and in patients whose tumors are not resectable (13).

Corticosteroids are generally not useful in adults, although good results have been reported in children in cases of unresectable tumors or hilar and mediastinal invasion (1). The use of steroids is also recommended to reduce the inflammatory process that is surrounding the tumor, especially if the tumor is in the central nervous system (13).

Using nonsteroidal antiinflammatory drugs as a conservative measure for treatment of patients with IMT whose tumors are not amenable to surgical resection (8).Of late, nonsteroidal anti-inflammatory drugs are being put to good use in large lesions (6).

TUMOR BEHAVIOR AND RECURRENCE

Most are parenchymal but some are endobronchial and may cause airway obstruction. Less than 5% invade the mediastinum and/or chest wall. Local recurrence is attributed to incomplete resection of the primary lesion (7). Metastasis of the tumor to mediastinum or the brain even many years after complete resection has been described. Rarely, simultaneous intra- and extrathoracic locations may occur. Association with other malignancies in sporadic cases has been reported (7) .Some of patients with lung IMT developed CNS lesions; therefore CNS screening in these patients should be considered, at diagnosis and later during follow up (10).

About 18% to 40% of IMTs recur, and most recurrences appear in extrapulmonary lesions that are larger than 8 cm and are locally invasive .Retroperitoneal and mesenteric IMT seem to be associated with more frequent recurrences (8). Recurrence rates are 1.5% in tumors confined to the lung and 46% in locally invasive tumors with extension beyond the lung (5). The frequency of recurrent disease is increased in cases with incomplete resection, larger tumours, deep location, presence of the susceptibility gene (ALK), and in younger patients (9).

Recurrence is rare in cases with complete resection of the tumor (5). Long-term follow-up is recommended, especially in cases of incomplete resection since recurrence following incomplete resection has been reported (6) .Patients with recurrent disease should undergo re-resection (7). Based on an article in 2014 the recurrence rate after surgery ranges from 1.5 to 4%. Long-term

follow-up of patients after IMT resection is always required as recurrences have been reported 11 years after primary tumour resection (9).

Karnak *et al* reported a case with very high leukocyte count and suggest that this could be a good marker for recurrence (14).

PROGNOSIS

Despite IMT is a benign tumor, it is considered by some authors as a low grade tumor because of malignant features such as local invasiveness, recurrence (4% in cases of incomplete resection) or malignant transformation (exceptional). The evolution depends on the tumor size (less than or equal to 3 cm) and the quality of surgical resection. The 5-year survival is 91.3% (1).
The prognosis of these rare tumours is excellent after complete surgical excision (7) .Five-year survival after resection exceeds 91% (9).

CONCLUSION

Inflammatory myofibroblastic tumor is a rare benign tumor (1) .The most common clinical picture is one of an asymptomatic, well-circumscribed lung mass that mimics cancer (7). Clinical and radiological presentation is variable and nonspecific (1). Clinicians need to bear in mind the diverse clinical presentations. Surgical excision is usually indicated to reach a firm diagnosis and cure (7). Only histological and immunohistochemical study can confirm the diagnosis. Despite being a benign lesion, its potential for recurrence and local invasion requires complete surgical resection (1).

Because there is no "standard-of-care" therapy for IMT, the identification of actionable genomic alterations, in addition to ALK, is expected to redefine management strategies for patients with this disease (11).

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REFERENCES

1. Nawal Hammas, Laila Chbani1, Mohammed Rami, Meryem Boubbou, Sara Benmiloud, Youssef Bouabdellah, et al. A rare tumor of the lung: inflammatory myofibroblastic tumor. Diagnostic Pathology 2012, 7:83.

2. Ozkan Demirhan, Selvinaz Ozkara, Mustafa Yaman, and Kamil Kaynak . A rare benign tumor of the lung: Inflammatory myofibroblastic tumor – Case report. espir Med Case Rep. 2013; 8: 32–35.

3. Van den Heuvel DA, Keijsers RG, van Es HW, Bootsma GP, de Bruin PC, Schramel FMet al. Invasive inflammatory myofibroblastic tumor of the lung. J Thorac Oncol. 2009 Jul; 4(7):923-6.

4. Lung tumor,Benign tumors,Inflammatory pseudotumor Reviewer: Deepali Jain, M.D. (see Reviewers page) Revised: 7 January 2013, last major update September 2012 Copyright: (c) 2003-2013, PathologyOutlines.com, Inc.

5. Prabudh Goel, Veereshwar Bhatnagar, Vishesh Jain, Ajay Verma, Monica Breta, Manoj K. Singh. Locally invasive pulmonary inflammatory myofibroblastic tumors in children. J Indian Assoc Pediatr Surg. 2012 Jul-Sep; 17(3): 135–137.

6. Subhalakshmi Sengupta, Uttara Chatterjee, Ranjana Bandyopadhyay, Kuntal Bhowmick, andSugata Banerjee. Primary pulmonary neoplasms in children: A report of five cases. Indian J Med Paediatr Oncol. 2011 Oct-Dec; 32(4): 223–226.

7. R. Morar, A. Bhayat, G. Hammond, H. Bruinette, and C. Feldman. Inflammatory Pseudotumour of the Lung: A Case Report and Literature Review. Case Reports in Radiology Volume 2012 (2012), Article ID 214528, 4 pages http://dx.doi.org/10.1155/2012/214528

8. Ohammad Salameh, Iyad Sultan, Maha Barbar, Maysa Al Hussaini, Abdullah Jameel, Khalil Ghandour, et al. Inflammatory myofibroblastic tumor causing unexplained anemia in a toddler: a case report. Journal of Medical Case Reports 2011 (feb), 69(5) : doi:10.1186/1752-1947-5-69. http://www.jmedicalcasereports.com/content/5/1/69

9. Barbara Cuch, Urszula Opasała, Paweł Nachulewicz, and Justyna Szumiło. Inflammatory myofibroblastic tumour in a 5-year-old child – a case report and review of the literature. Wideochir Inne Tech Maloinwazyjne. 2014 Dec;

10. Siminovich M, Galluzzo L, López J, Lubieniecki F, de Dávila MT. Inflammatory myofibroblastic tumor of the lung in children: anaplastic lymphoma kinase (ALK) expression and clinico-pathological correlation. Pediatr Dev Pathol. 2012 May-Jun;15(3):179-86.

11. Lovly CM, Gupta A, Lipson D, Otto G, Brennan T, Chung CT, et al. Inflammatory myofibroblastic tumors harbor multiple potentially actionable kinase fusions. Cancer Discov. 2014 Aug;4(8):889-95.

12. Sanders BM, West KW, Gingalewski C, Engum S, Davis M, Grosfeld JL: Inflammatory pseudotumor of the alimentary tract: clinical and surgical experience. *J Pediatr Surg* 2001**,** 36(1)**:**169-173.

13. Kovach SJ, Fischer AC, Katzman PJ, Salloum RM, Ettinghausen SE, Madeb R, et al: Inflammatory myofibroblastic tumors. *J Surg Oncol* 2006, 94(5)**:**385-391.

14. Karnak I, Senocak ME, Ciftci AO, Cağlar M, Bingöl-Koloğlu M, Tanyel FC, et al: Inflammatory myofibroblastic tumor in children: diagnosis and treatment. *J Pediatr Surg* 2001, 36(6)**:**908-912.

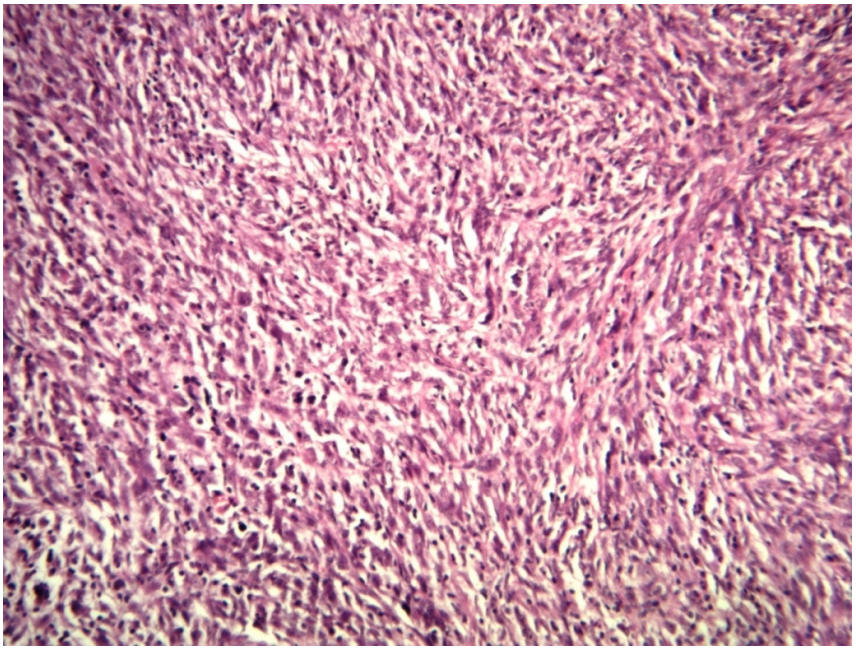

Figure1: pathological picture of lung biopsy

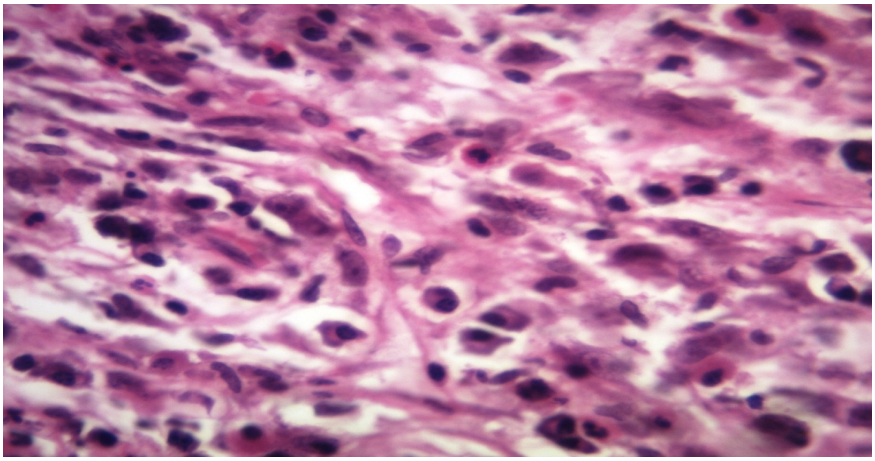

Figure2: pathological picture of lung biopsy

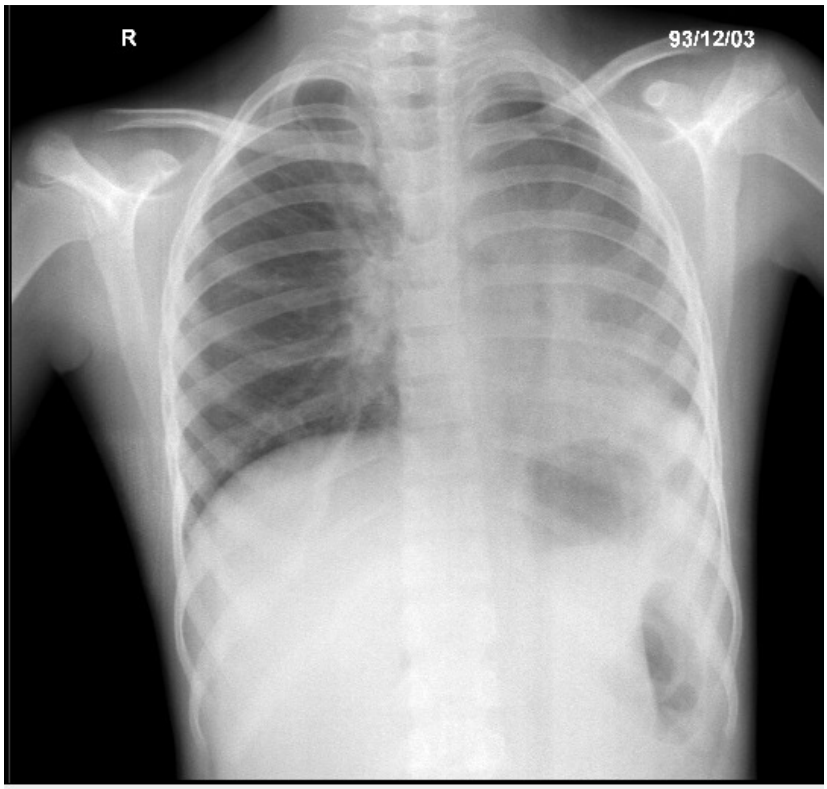

Figure3 :collapse consolidation in chest x- Ray

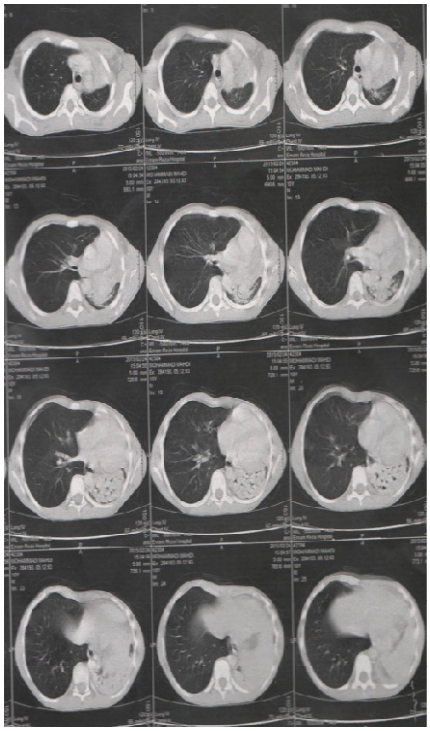

Figure4 :Mass in chest CT scan

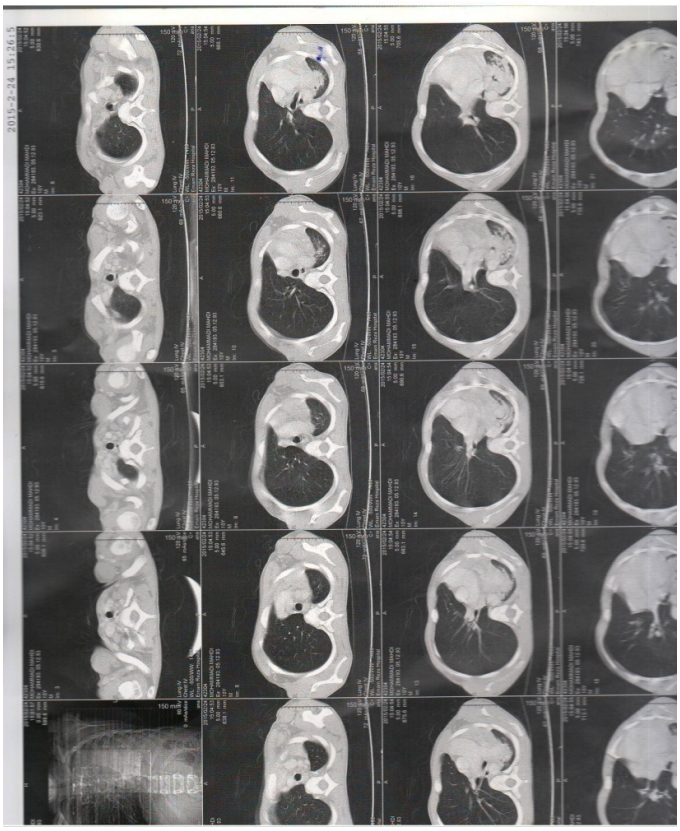

Figure5 :Mass in chest CT scan