Egyptian Journal of Chest Diseases and Tuberculosis (2013) 62, 817–824

The Egyptian Society of Chest Diseases and Tuberculosis

Egyptian Journal of Chest Diseases and Tuberculosis

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CASE REPORT

# Unusual pulmonary lesions – A series of rare cases

**Rama Saha** [a,1], **Aruna Bhattacharya** [b,*], **Joydip Deb** [c], **Pamela Nayak** [a,2], **Sumit Ranjan Pramanik** [d,3]

[a] *Department of Pathology, Institute of Postgraduate Medical Education & Research, Kolkata, West Bengal, India*
[b] *Department of Biochemistry, Institute of Postgraduate Medical Education & Research, Kolkata, West Bengal, India*
[c] *Department of Chest Medicine, Bankura Sammilani Medical College & Hospital, Bankura, West Bengal, India*
[d] *Department of Obstetrics & Gynaecology, Institute of Postgraduate Medical Education & Research, Kolkata, West Bengal, India*

Received 26 July 2013; accepted 4 September 2013
Available online 5 October 2013

**KEYWORDS**

Lung tumour;
Hamartoma;
Pulmonary;
Pseudotumour castlemans disease;
Neuroendocrine tumour

**Abstract** In thoracic imaging, mass means any pulmonary, pleural, or mediastinal lesion more than 3 cm in diameter (without regard to contour, border, or density characteristics). Nodule means rounded opacity, well or poorly defined, measuring up to 3 cm in diameter. We report 4 rare cases – presented as mass or nodular lesion in chest imaging.



## CASE 1: (Pulmonary hamartoma)

### *Introduction*

Pulmonary hamartoma is a commonest benign lung lesion and accounts for 3% of all lung tumours. It is a benign lesion composed of cartilage, adipose tissue, fibrous and myxoid connective tissue [1].

### *Case report*

A 50 year old man presented with dry cough over 2 years and one episode of breathlessness and chest pain. Routine haematological examination, USG whole abdomen and Endoscopy were normal. BAL fluid cytological examination was negative for malignancy. Chest X-ray and CT scan (Figs. 1 and 2) thorax revealed a coin like lesion in the right lower lung. Biopsy of the lung nodule could not be performed as patient's relatives refused for the procedure but right lower lobectomy was

* Corresponding author. Address: Institute of Postgraduate Medical Education & Research, 35 A K.G. School Road, East Anandapuri, P.O. Nonachandanpukur, Barrackpur, Kolkata 700122, West Bengal, India. Tel.: +91 9433465339.
E-mail addresses: drrama.saha@gmail.com (R. Saha), draruna.bhattacharya@gmail.com (A. Bhattacharya), jaydip.deb@gmail.com (J. Deb), dr.pml.nayak@gmail.com (P. Nayak), drsrp_73@yahoo.com (S.R. Pramanik).
[1] Tel.: +91 9433127668.
[2] Tel.: +91 9231676063.
[3] Tel.: +91 9830191215.

Peer review under responsibility of The Egyptian Society of Chest Diseases and Tuberculosis.

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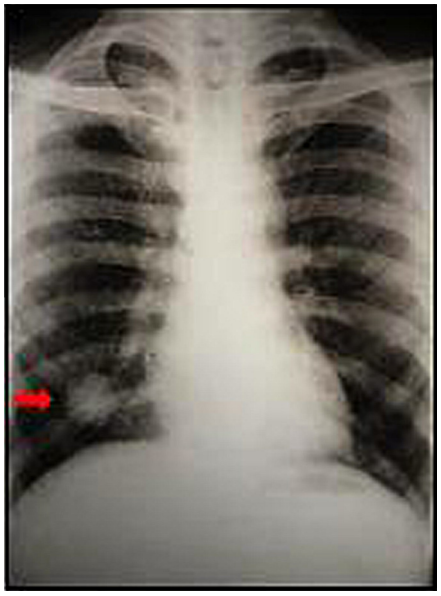

**Figure 1** Chest X-ray PA view, coin lesion right lower lobe.

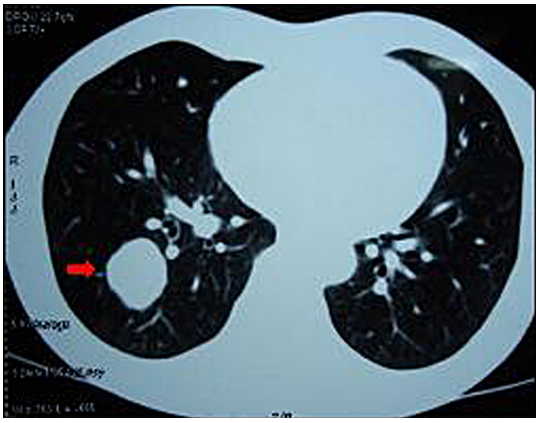

**Figure 2** CT Thorax, well defined pulmonary nodule.

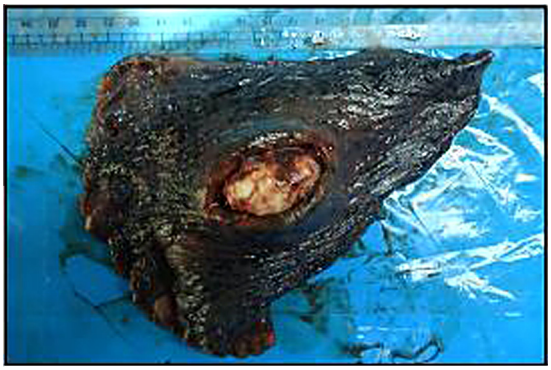

**Figure 3a** Gross, right lower lobectomy specimen, whitish globular mass subpleurally.

performed and histopathological examination of the specimen diagnosed it as pulmonary hamartoma (Fig. 4).

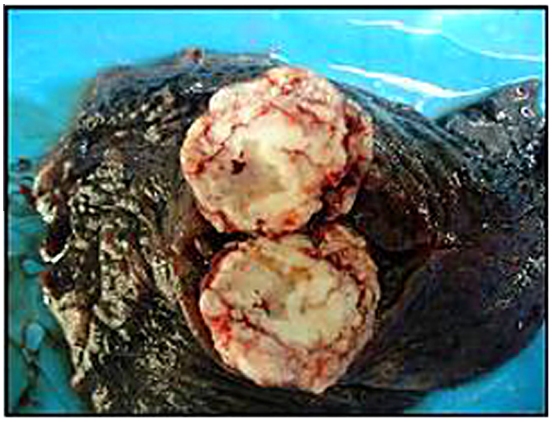

**Figure 3b** Cut section showing cartilaginous tissue.

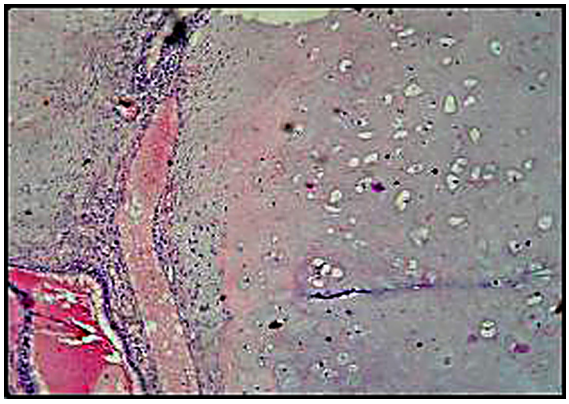

**Figure 4** Lobules of cartilages with cleft like spaces, (H&E stain 400×).

*Discussion*

The clinical importance of diagnosis of pulmonary hamartoma is related to 2 things.

(1) It should be differentiated from malignant lesion.
(2) It may cause secondary symptoms due to compression.

The incidence of pulmonary hamartoma accounts to 6% of solitary pulmonary lesion [7]. It usually is to be differentiated from coin lesion. There is a strong correlation with age. They are rarely seen in childhood and never at the time of birth [8]. They are most common in sixth and seven decade of life. Males are mostly affected [1,3]. 53.8% of the patients show history of heavy smoking [10]. Now it is considered as true neoplasm of primitive bronchial mesenchymal tissue having capacity to differentiate towards multiple mesenchymal components [1,2]. It can be seen in all parts of lung but mostly in periphery and rarely in hilar part [4,5]. Most patients are asymptomatic and when incidentally found on chest X-ray examination [4] only 1/3rd show symptoms [6]. Recurrence after resection is uncommon [9]. Van den Bosch reported 2 cases of recurrence in a series of 154 patients [2].

Our case was of a male nonsmoker patient aged 55 years with a history of dry cough and breathlessness. Chest X-ray and CT (Figs. 1 and 2) showed a coin like lesion in the right lower lobe of lung situated subpleurally. Excision was done. On gross examination there was a greyish white tumour having cartilagenous area with ill defined cleft like spaces (Figs. 3a and 3b). On microscopic examination, there was a lobule of cartilage with cleft like spaces lined by cuboidal epithelium (Fig. 4). Diagnosis was done as pulmonary hamartoma with no recurrence observed till date.

Pulmonary hamartoma is a benign lesion and represents SPN (solitary pulmonary nodule) in imaging. Efficacy of transbronchial biopsy is of limited value in preoperative diagnosis. Definitive treatment is achieved by surgical resection with minimum morbidity.

**CASE 2: (Castleman disease)**

*Introduction*

It is a rare benign and vascular lymphoproliferative disorder which may present as nodal or extra nodal mass. It was first reported by Castleman in 1956 in a report of solitary mass in the mediastinum [11].

*Case-report*

A 26 year old female presented with chest pain and breathlessness for 6 months. Routine haematological examination, USG whole abdomen and Endoscopy were normal. Chest X-ray and CT scan thorax revealed a nodular lesion in the right upper lung. FNAC from the lung nodule was performed followed by excision of the lesion. Gross and histopathological examination of the specimen was carried and it was diagnosed as Castleman disease.

*Discussion*

Aetiology of Castleman disease is unknown. According to some, viral infection may play a role. Commonest site is within the thorax [12]. Pathogenesis is unknown but most people believe defective immunoregulation results in excessive B cell proliferation and plasma cell infiltration in lymphoid organ [13]. There are 2 pathological types – hyaline vascular type which is most common (90%) and plasma cell type which is less common but clinically more aggressive. However, mixed type may occur. Clinically they may be localised or disseminated. Localised ones are benign and disseminated lesions are associated with symptoms like fever, splenomegaly and leucocytosis [14].

Grossly, it is a single rounded mass (size varies from 1.5–16 cm). Microscopically, they show a regressively transformed

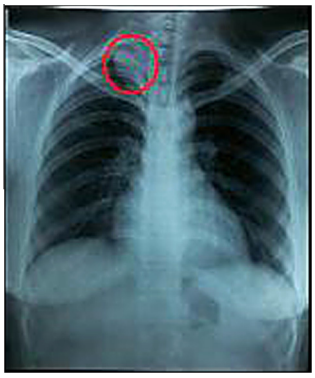

**Figure 5a** Chest X-ray, Nodular opacity, apical region of right lung.

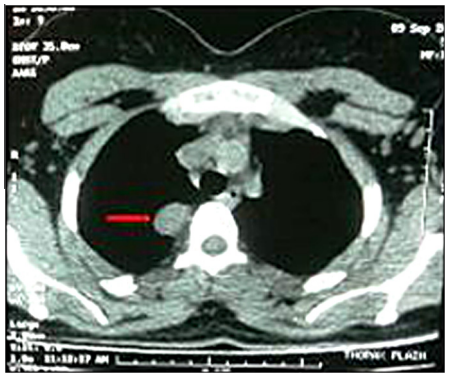

**Figure 5b** CT Thorax, nodular lesion in right thorax.

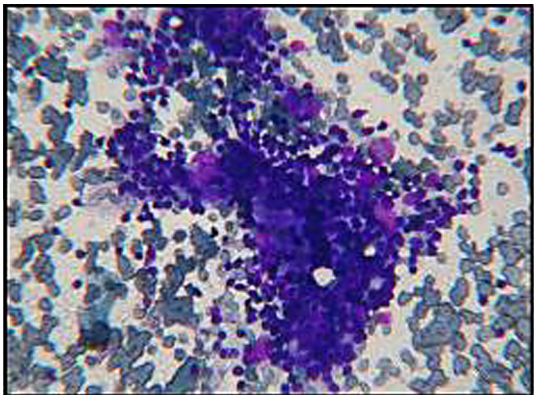

**Figure 6** FNAC, mostly mature lymphocytes.

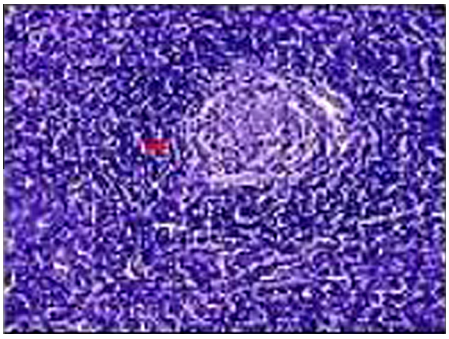

**Figure 7** Lymphoid follicle with hyalinised germinal centre, onion skin layering of mantle zone, and vascular inter follicular area. Radially penetrating sclerotic vessels (lollipop appearance) (H&E stain).

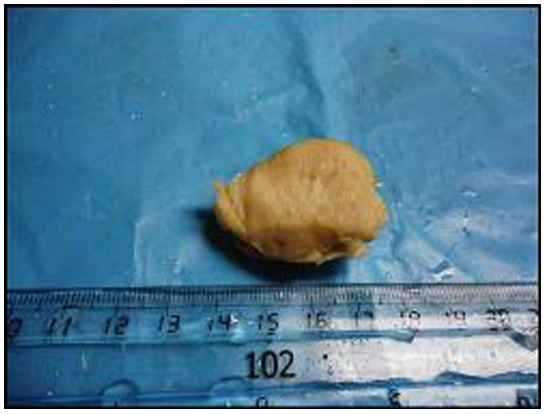

**Figure 8** Whitish globular mass, cut surface whitish homogeneou.

germinal centre surrounded by variably sized mantle zone (onion skin layers) with prominent interfollicular vascularity and absence of sinus. Germinal centre is devoid of lymphocyte and consists of dendritic reticulum cell, endothelial cells with hyalinisation and marked vascular proliferation [16]. The capillaries penetrating the follicles at right angles give rise to "lollipop appearance [15]."

Recurrence is not expected [16,17].

In our case, the patient was a 26 year old female who presented with chest pain and breathlessness for 6 months. On chest X-ray and CT thorax, a nodular opacity is seen in the apical region of the right lung (Fig. 5a and 5b). When FNAC was done, it showed the presence of lymphoid cells only (Fig. 6). Excision of the mass was done. On gross examination, it was a white globular mass with white homogenous cut surface (Fig. 8). Microscopy showed the presence of lymphoid follicle with hyalinised germinal centre, onion skin layering of mantle zone, vascular interfollicular area, radially penetrating sclerotic vessels (lollipop appearance) (Fig. 7). All the findings support diagnosis of hyaline vascular type of Castleman disease. Recurrence is not seen till date.

**CASE 3: (Inflammatory pseudotumour)**

*Introduction*

It is a rare benign lung tumour of unknown aetiology occurring in young patients. They are also known as plasma cell granuloma, histiocytoma and fibroxanthoma depending on predominant cell type. Though benign, they have the capacity to spread locally, grow rapidly and transform into sarcoma [18]. Spontaneous regression even may occur after steroid therapy. This case is reported because of rarity.

*Case report*

A 10 year old female presented with fever with occasional cough and hemoptysis. General examination revealed clubbing, SerumALP – 700 mg/dl, A: G – 1:3.5. Immunoprofile: IgG – 3950 mg/dl (N 694 – 1618)IgA – 359 mg/dl (N 68–378)IgM – 295 mg/dl (N 60–263) Urine for B.J. Protein: Negative. Bonemarrow aspiration revealed hypercellular marrow with M:E = 6:1. Erythropoisis: adequate and normoblastic. Granulocytes are relatively increased with a slight increase in lymphoid cells at places. Plasma cells – 8%. No parasites were seen. All the findings were in favour of chronic disorder.

Debulking of the mass was done through the right posterior intercostal space. Mass could not be removed totally due to adherence to the adjacent structure.

*Discussion*

The various terminologies used have resulted in a great deal of confusion regarding understanding the nature of the disease.

Microscopically, it consists of inflammatory cells with complete maturity of the fibroblastic element with no mitosis. It is a nonneoplastic process characterised by unregulated growth of inflammatory cells which may be due to derangement in the metabolism with viral or immunological origin. Infection may play a role [18].

Gross appearance shows a round well circumscribed nonencapsulated tumour mass in pulmonary parenchyma. Histopathologically, it is a circumscribed mass composed of variable mixture of collagen, benign mesenchymal cell, spindle myofibroblast and fibroblast with inflammatory cells like plasma cell, macrophage, foam cell, giant cell, lymphocytes etc.

It is a benign lesion of childhood. Reported incidence is. 04% to 7% of lung masses [18]. There is no sex predilection [25].

The symptomatology is cough, chest pain, dyspnoea, haemoptysis, clubbing [18].

Most common radiological presentation is solitary well circumscribed round/oval pulmonary mass seen in 50% of patients [19]. Grossly, the tumour size varies from 5–30 cm [21,22]. Age incidence according to Bahaduri and Leivo is 40 cases out of which 16 were child and 25 were adult [23]. Occasionally it may cause obstructive pneumonia [24]. Central cavity formation and hilar lymphadenopathy are very rare. No pleural effusion has been reported till now [20].

It is a relatively uncommon lesion which is typically solitary, peripheral sharply circumscribed lobulated mass. Most patients are symptomatic at diagnosis and have prior history of respiratory tract infection.

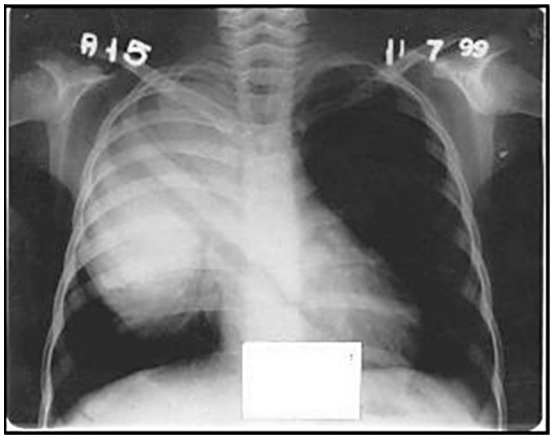

**Figure 9** Chest X-ray PA view-mass, right middle and upper lobe.

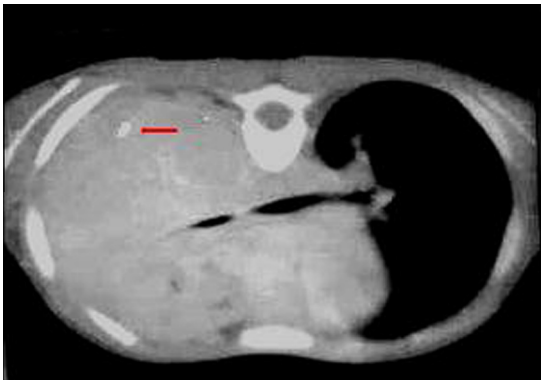

**Figure 10** CT mass right hemi thorax, focal calcification.

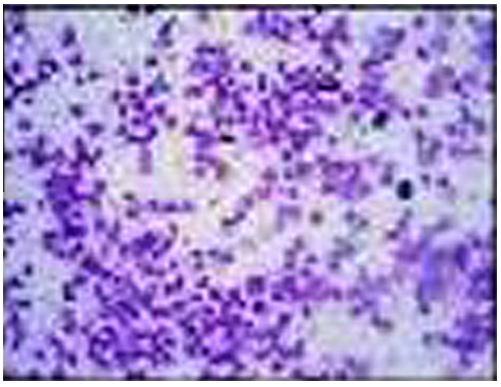

**Figure 11** FNAC, mostly lymphocytes, plasmacytoid cells (MGG STAIN 400×).

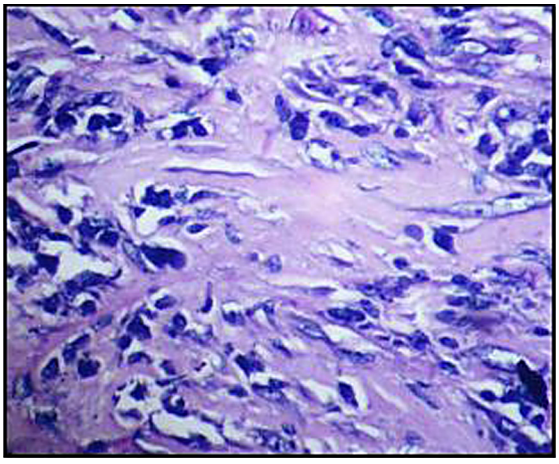

**Figure 12** Spindle shaped fibroblastic cells along with inflammatory infiltrate (H&E STAIN 400×).

It should be differentiated from primary neoplasm, metastatic tumour, hamartoma, chondroma, sclerosing haemangioma and pulmonary granuloma.

Treatment of choice is surgical excision. Nonsurgical treatment includes radiotherapy. Steroid therapy is indicated where there is incomplete surgical removal, recurrence and when patient is unfit for surgery [18].

In our case, the patient was a 10 year old female with fever and cough with haemoptysis occasionally and with clubbing of fingers. X-ray and CT chest showed mass in the right, middle and upper lobe with areas of calcification (Figs. 9 and 10). Debulking of the mass was done through right posterior intercostal space but due to adherence total removal could not be done. FNAC was done which showed lymphocytes and plasmacytoid cells (Fig. 11). Microscopy showed fibroblastic cells with hyalinised collagen with inflammatory and plasma cells (Fig. 12).

**CASE 4: Large cell neuroendocrine tumour**

*Introduction*

Large cell neuroendocrine tumour (LCNET) is categorised under large cell tumour of the lung (WHO).

*Case report*

A 47 year old male habitual smoker presented with fever and shortness of breath. Routine haematological examination showed absolute eosinophil count-856/cumm, other parameters-within normal limit. X-ray chest and CT thorax showed mass in the lower lobe of the right lung. FNAC was done followed by right lower lobectomy. Histopathological examination and IHC were in favour of LCNET.

*Discussion*

Travis et al. in 1991 were the first to categorise LCNET as a separate entity of pulmonary neuroendocrine tumour which is different from typical carcinoid, atypical carcinoid and small cell carcinoma [26]. These tumours have a cell size three times than that of SCC, exhibit an organoid pattern, cellular palisading, high mitotic rate and a variably granular chromatin pattern with absent or inconspicuous nucleolus and patches of geographic necrosis [27].

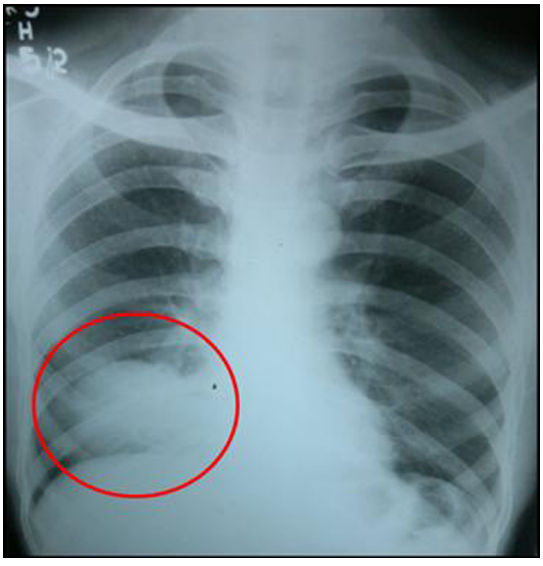

**Figure 13** Chest ray PA view, mass lower lobe right lung.

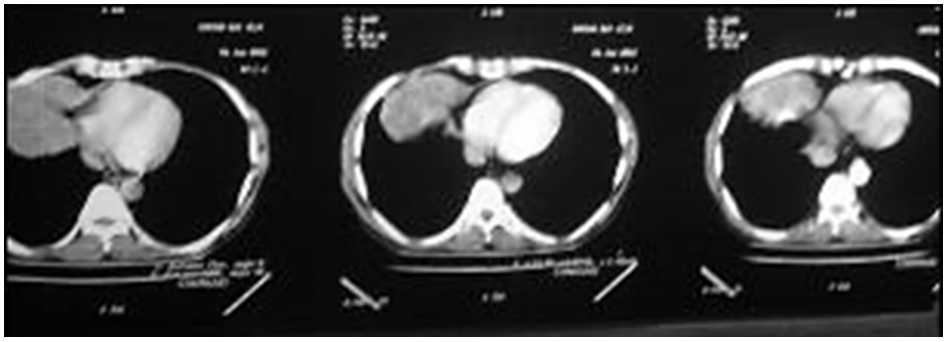

**Figure 14** CT, mass right hemi thorax.

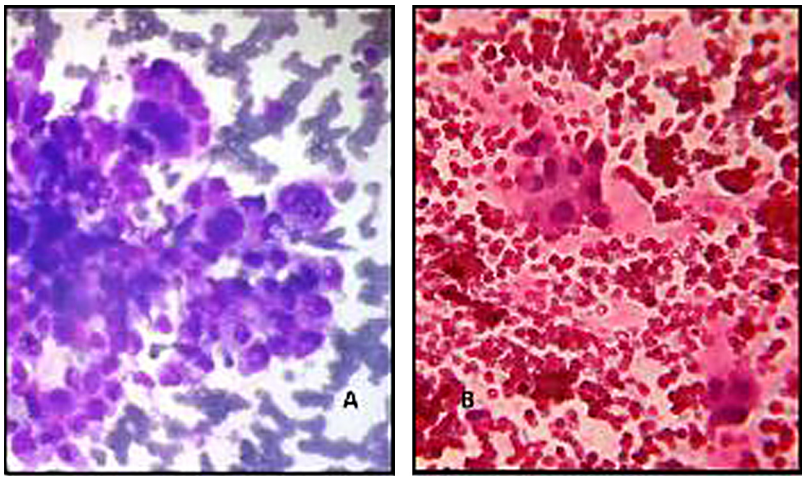


**Figure 15** Pleomorphic large cells, abundant cytoplasm, non small cell carcinoma of lung.

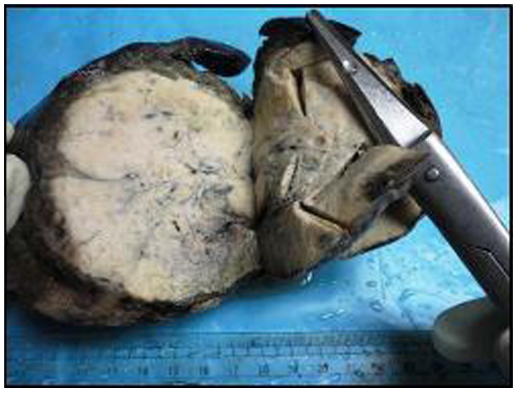

**Figure 16** cut section showing tumour involving lung tissue.

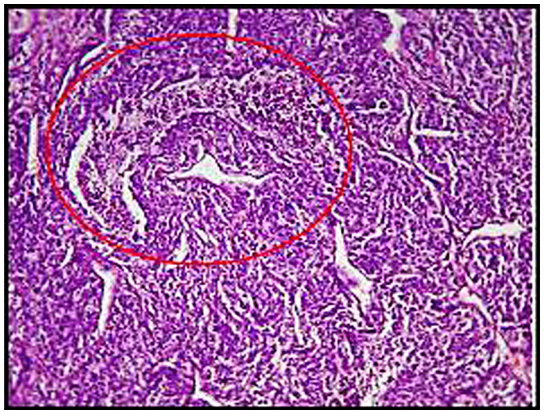

**Figure 17** Microscopically, areas of necrosis and organoid arrangement of tumour tissue(H&E –100×).

It is a very rare tumour with a low incidence rate. In a series reported by Jiang et al. Ref. [28] of 766 patients with resected primary lung cancer, only 22 were classified as LCNET (2.87%). In another study, Takei et al. reported 87 cases out of 2790 (3.1%) [29].

Smoking appears to be a primary cause of LCNET. Several studies reported a strong correlation between LCNET and habitual cigarette smoking [29–32]. Mean age of patients ranges from 62 to 68 years with median age of 65.8 years. Males are the usual sufferers [29,33,35].

The tumour cells are shown to contain a neurosecretory granule. On IHC, the tumour stains for neurone specific enolase, carcinoembryonic antigen, chromogranin, Leu-7 and synaptophysin [29]. These are also described by Wick et al. [37]. To identify a neuro endocrine tumour we need at least one

of the 3 markers – neural cell adhesion molecule, chromogranin A and synaptophysin [29]. They most commonly present as a peripheral tumour [30,34]. Symptoms like cough, haemoptysis, and pneumonia are less likely [35]. Paraneoplastic syndrome is infrequent [29–31].

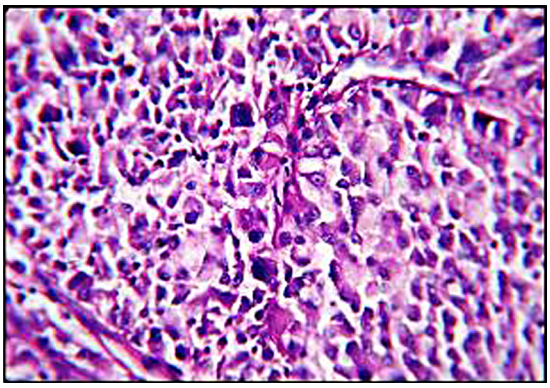

**Figure 18** Large polygonal cells with low N:C ratio, abundant eosinophilic cytoplasm, coarse nuclear chromatin and prominent nucleoli (H&E – 400×).

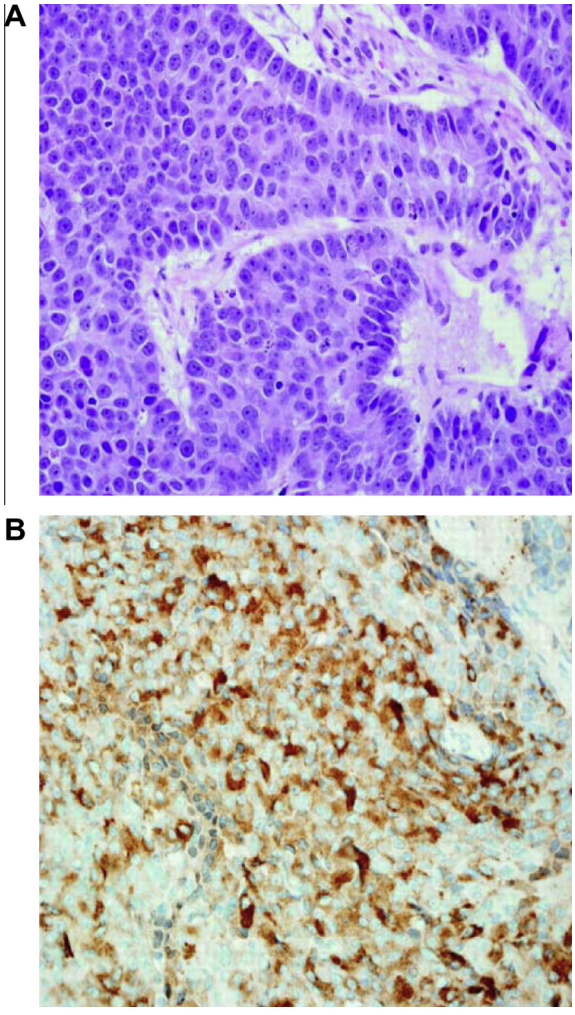


**Figure 19** IHC staining, chromogranin positivity.

In our case, the patient was a 47 year old male, habitual smoker who presented with fever and shortness of breath with involvement of the lower lobe of the right lung. X-ray chest and CT thorax showed mass in the lower lobe of the right lung (Figs. 13 and 14). FNAC showed a pleomorphic large cell with abundant cytoplasm and diagnosed as Non small cell carcinoma (Fig. 15). Right lower lobectomy was done. Mass involved whole of the cut surface (Fig. 16). Microscopy showed a growth with organoid pattern and areas of necrosis consisting of large cells with abundant cytoplasm (Figs. 17 and 18). IHC was done which showed chromogranin positivity (Fig. 19).

The diagnosis of LCNEC needs to be done carefully because there is a significant overlap in the nuclear dimension between LCNEC and SCC [36]. IHC should be done as confirmatory. The five year survival rate in resected LCNEC was only 35% which was worse than the survival rate of 71.3% of LCC with no evidence of neuroendocrine morphology. LCNEC is an uncommon, aggressive neoplasm with poor prognosis. Because of poorer prognosis, all LCC should be carefully searched for the presence of occult neuro endocrine features.

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