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Case Report

Primary lymphoepithelioma-like carcinoma of the lung in an adolescent girl with unusual presentation

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ABSTRACT

Primary lymphoepithelioma-like carcinoma (LELC) of lung is a rare tumour of lung mostly reported from south-east Asia. It occurs in middle aged persons of either sex and presents with the complaint of cough and haemoptysis. We report a case of primary LELC of lung in a young girl with unusual presentation.

Keywords: Primary lymphoepithelioma-like carcinoma, Lung cancer, Clubbing, LELC

INTRODUCTION

Primary LELC is an unusual carcinoma, originally described in nasopharynx.[1] Recently, this tumour has also been described in other sites including lung. According to WHO, primary LELC of lung is classified as a type of large cell carcinoma.[2] This tumour was first reported in 1987, and since then some 200 cases have been reported worldwide.[3] Our knowledge about the tumour is mostly derived from three large cohorts from south-east Asia.[4-7] We report this rare case in a 14-year-old girl who presented with unusual symptoms.

CASE REPORT

An adolescent girl presented to our hospital with the complaint of high grade fever, chest pain and dry cough for the last nine months and generalized bony pains in extremities for the last seven months. The patient was apparently symptom free nine months before when she developed dry persistent cough associated with pleuritic chest pain in the anterior part of chest wall. There was no history of haemoptysis. She developed high grade, intermittent fever associated with sweating which responded to usual doses of antipyretics. After two months, patient developed pain in multiple joints and extremities. Besides, she developed swelling in the distal joints of hands and feet.

With these complaints she visited nearby physician and was treated with empirical antibiotics without much help. She presented to our institution with X ray and two sets of CECT chest that was done five months apart.

At the time of examination, she had high grade fever, moderate pallor and grade IV clubbing. Her vitals were stable and there was no peripheral lymphadenopathy. Besides, she had grade II hirsutism. Respiratory system examination revealed centrally located trachea and dullness on percussion in left infraclavicular, mammary and axillary areas. Air entry was decreased on auscultation in these areas. These findings suggested a mass in the upper part of left lung. Examination of other systems was within normal limits.

Her haematological investigation showed moderate anaemia (haemoglobin 8.4 gm/dl), leucocytosis (TLC 60,000/micro litre) and thrombocytosis (7.7 x 10^5 /microlitre). Her liver and renal function tests were within normal limits. X ray chest showed a well-defined homogenous mass involving left hilum which was invading left upper and middle lobe. CECT chest revealed a mass arising from the left lung encroaching trachea with multiple mediastinal lymph nodes. X ray of both hands showed features suggestive of hypertrophic osteoarthopathy. Based upon the clinical findings, haematological reports and CECT chest findings (Figure 1 and 2) – a provisional diagnosis of lymphoreticular malignancy was made. There was no evidence of an infective focus. Fibro-optic bronchoscopy was done and tissue growth was found occluding left main bronchus and needle biopsy was taken from the same. Bronchoscopic biopsy from mass in the left main bronchus showed fragments of respiratory mucosa with poorly differentiated tumor cells arranged in loose clusters and in sheets in a background of dense lymphocytic infiltration with excess of plasma cells (Figure 3). On immunohistochemical stains these cells showed strong positivity for pancytokeratin (CK), epithelial membrane antigen (EMA) and EBV-LMP 1 protein while negative for leukocyte common antigen (LCA), CD 1a, langerin, desmin, myogenin, CD117, CD30, PLAP, AFP, TTF-1 and neuron specific enolase (NSE). With this histomorphology and immunohistochemical profiles, a final diagnosis of lymphoepithelioma like carcinoma was made. Mutant specific immuno-histochemical analysis for the EGFR mutation in Exon 19 and 21 were negative in tumor cells (Figure 3a).

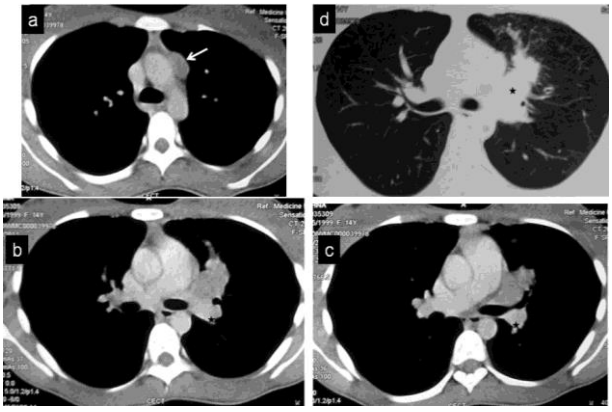

Figure 1a-d: CECT chest showing heterogeneously enhancing irregular shaped mass-like lesion (measuring ~ 3.5 x 2.5 cms) in left perihilar location, causing narrowing of left upper lobe bronchus associated with peri-bronchovascular thickening in lingula. A (2.2 x 1.5 cm) homogeneously enhancing soft tissue lesion in prevascular location, likely enlarged lymph node is also seen.

As LELC of lung closely mimics nasopharygeal carcinoma which can present as occult metastasis in mediastinum, ENT examination with nasal endoscopy and laryngoscopy was done which was normal. FDG PET scan revealed a mass arising from left lung with involvement of multiple mediastinal lymph nodes along with metastasis to left supraclavicular nodes (Figure 4). There was no evidence of any distant metastasis. It was a locally spreading tumour with mediastinal lymph node involvement including left supraclavicular and right paratracheal involvement without distant metastasis. Thus, making the stage of the tumour as III B.

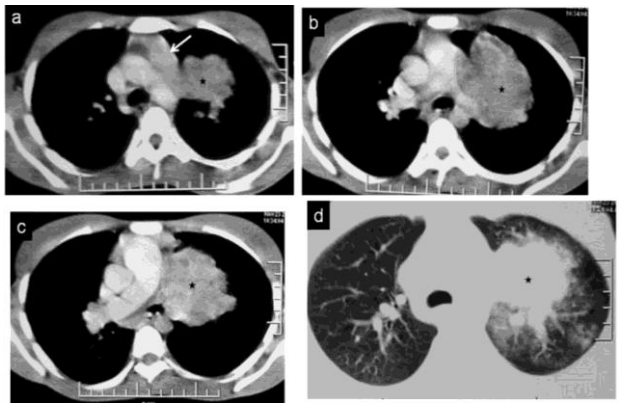

Figure 2a-d: CECT chest showing that the lung mass had dramatically increased in size (measuring 9 x 6 cms), showing heterogeneous enhancement with areas of necrosis within. It is encasing and narrowing the left pulmonary artery, causing obstruction of left main bronchus at its bifurcation with non-visualization of left upper lobe bronchus. There was surrounding ground glass opacity and nodular peri-bronchovascular thickening.

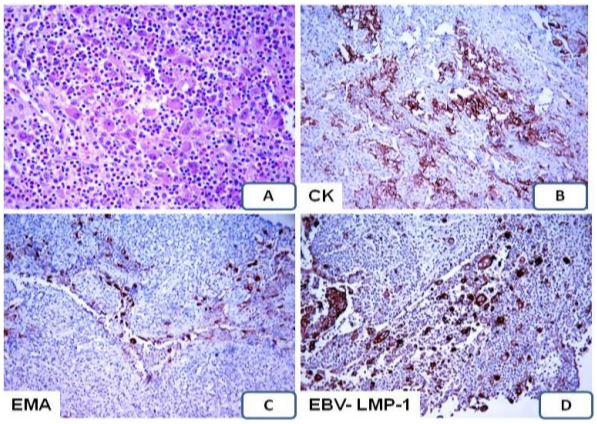

Figure 3: (A) Poorly differentiated carcinoma with prominent lymphocytic infiltration in the background (H&E, x40); (B) The tumor cells are immunopositive for cytokeratin (IHC, CK); (C) epithelial membrane antigen (IHC, EMA); (D) Epstein-barr virus latent membrane protein 1(IHC, EBV LMP-1).

Patient was treated symptomatically for the fever with paracetamol and tepid sponging. For management of pain she was given paracetamol, tramadol and subsequently injection morphine. Surgical oncology opinion was taken

and because of the locally advanced disease and poor performance status she was deemed unfit for surgery. Patient was subsequently referred to the medical oncology team and started on cisplatin based chemotherapy.

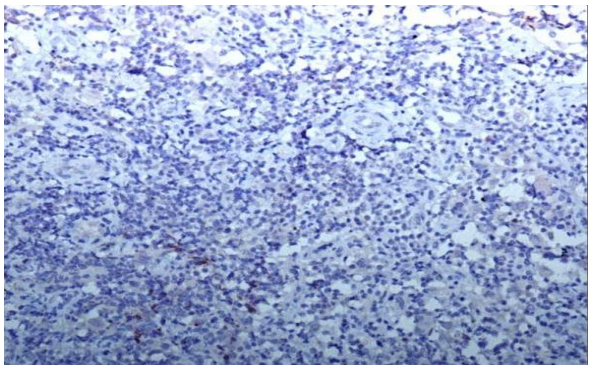

Figure 3E: Photomicrograph shows tumor cells are immuno-negative for mutation specific EGFR antibody.

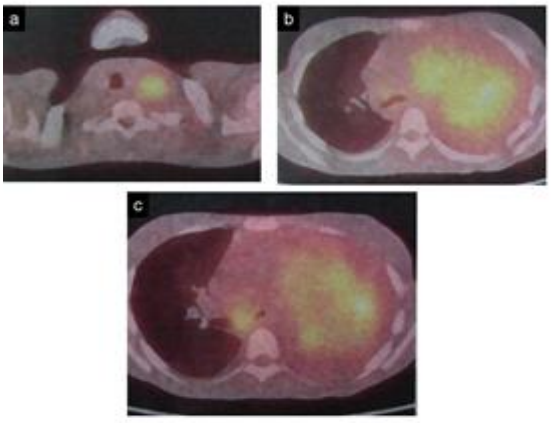

Figure 4a-c: A PET/CT study showed intense uptake in large left pulmonary mass, confluent prevascular and subcarinal LNs, along with metastasiss to left supraclavicular nodes. There was no evidence of any distant metastasis.

DISCUSSION

Primary LELC is a distinct carcinoma of lung which has been reported mostly in the Asian adults of Mongoloid origin.[4-7] It occurs mostly in the middle age without any definite sex predilection.[5] In contrast to the non small cell carcinomas of the lung, primary LELC has been found to have minimal association with cigarette smoking.[5,8,9] Metastasis tends to occur less frequently and late in the course of primary LELC.[4,10]

As per the data of two large case series, about one-third of the patients were asymptomatic at the time of the diagnosis. Most of the symptomatic patients presented with complaints like cough, chest pain or haemoptysis.[4-7] It usually forms unilateral, solitary sub-pleural nodule without affecting major bronchi. However, some cases with bilateral extensive disease at the time of presentation have also been described.[4,11]

Pulmonary LELC is described as a subtype of large cell carcinoma of lungs according to the WHO histological typing of lung and pleural tumours.[12] Histological appearance of the tumour simulates undifferentiated nasopharyngeal carcinoma which is characterized by asyncytial appearance, focal squamous differentiation with prominent lymphocytic infiltration.[10]

Epstein barr virus (EBV) has been associated with lymphoid malignancies like Burkitt's lymphoma. It is also found to be associated with epithelial neoplasms like undifferentiated nasopharyngeal carcinoma and LELC involving lung and other organs.[13] The association between LELC and EBV has been found to be strong in case of Asian population but the same has not been proven in case of white patients.[14] Moreover, the association with EBV is not known with lymphoepithelioma of other sites as cervix, bladder and vagina.[8]

Surgical resection is the major curative method for stage I of the disease whereas patients with stage II or higher stage are treated by combination therapy including post-operative radiotherapy, chemotherapy or both.[11] Cisplatin based combination therapy has shown excellent response and has been used in several studies with good response.[4,5,11] The overall survival of patients with LELC type of carcinoma is better than non-LELC type of carcinoma. Tumour recurrence and necrosis is recognised as poor prognostic factors.[10,15]

The importance of this case report is its rarity. Less than 200 case reports have been described and most of them are from the south East Asian region (of Mongoloid race). Much lesser number of the cases has been reported from Western population (White race) and probably, none reported from Indian sub-continent suggesting increased propensity to certain race. Besides, uncommon age of presentation and symptoms make this case report worth sharing. The age of the patient was 14 years, whereas the tumour is mostly described in middle aged population. In a major case series, the age of patients ranged from 9 to 74 years with the median of 51 years. Most common presentation of the tumour is cough and chest pain whereas this case presented mainly as high grade fever and clubbing, probably due to paraneoplastic manifestation.

CONCLUSION

Primary LELC of lung is a distinct unusual tumour that may have unusual clinical presentation.

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