



# Primary pulmonary lymphoepithelioma-like carcinoma in a 12-year-old African child



## Authors:

Jacqueline Goedhals<sup>1,2</sup>   
David Stones<sup>3</sup> 

## Affiliations:

<sup>1</sup>Department of Anatomical Pathology, University of the Free State, South Africa

<sup>2</sup>National Health Laboratory Service, Bloemfontein, South Africa

<sup>3</sup>Department of Paediatrics and Child Health, University of the Free State, South Africa

## Corresponding author:

Jacqueline Goedhals,  
gnmbjg@ufs.ac.za

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Pulmonary lymphoepithelioma-like carcinoma is a rare malignant tumour predominantly occurring in Asian patients. It has identical histological features to nasopharyngeal carcinoma. To date, Epstein-Barr virus (EBV) in pulmonary lymphoepithelioma-like carcinoma has been limited to the Asian population. This manuscript presents an unusual case in a 12-year-old African boy in which the tumour tested positive for EBV.

## Introduction

Lymphoepithelioma-like carcinoma (LELC) can occur in a number of sites including the thymus, salivary gland, stomach, skin, uterine cervix, tonsil, oral cavity, trachea, larynx, urinary bladder and vagina.<sup>1</sup> Primary pulmonary LELC was first described by Bégin et al.<sup>2</sup> in 1987 and is a distinct subtype of non-small cell lung cancer.<sup>3</sup> It occurs predominantly in Asian patients, and only four cases of pulmonary LELC have been documented in black patients.<sup>4</sup> Although LELC tends to affect younger, non-smoking patients, only isolated cases occurring in children have been published to date.

We present an unusual case of primary pulmonary LELC in a 12-year-old African child in which the tumour tested positive for Epstein-Barr virus (EBV).

## Case

A 12-year-old HIV-negative African boy presented with a history of coughing and dyspnoea for a period of 6 months. He was placed on treatment for tuberculosis without microbiological confirmation and his symptoms became more severe over the next month. He experienced difficulty breathing and asthma medications were prescribed. Regardless, his condition continued to deteriorate. On examination at admission, he had clubbing of his fingers and severe respiratory distress with subcostal and intercostal retraction and global wheezing. His trachea was displaced to the left, and the right side of his chest appeared more prominent and hyperinflated. The left chest was dull to percussion. He had decreased breath sounds and air entry of the left hemithorax. His respiratory rate was 28 breaths per minute, with a functional oxygen saturation (SpO<sub>2</sub>) of 81%.

On an X-ray of the chest, we observed a hyperinflated right lung with the mediastinum, trachea and heart all displaced into the left hemithorax (Figure 1a). A chest computed tomography (CT) scan showed a left endobronchial mass that measured 58 mm × 49 mm × 55 mm extending into the trachea, with collapse of the left lung (Figure 1b). A bronchoscopy was performed and the mass was biopsied.

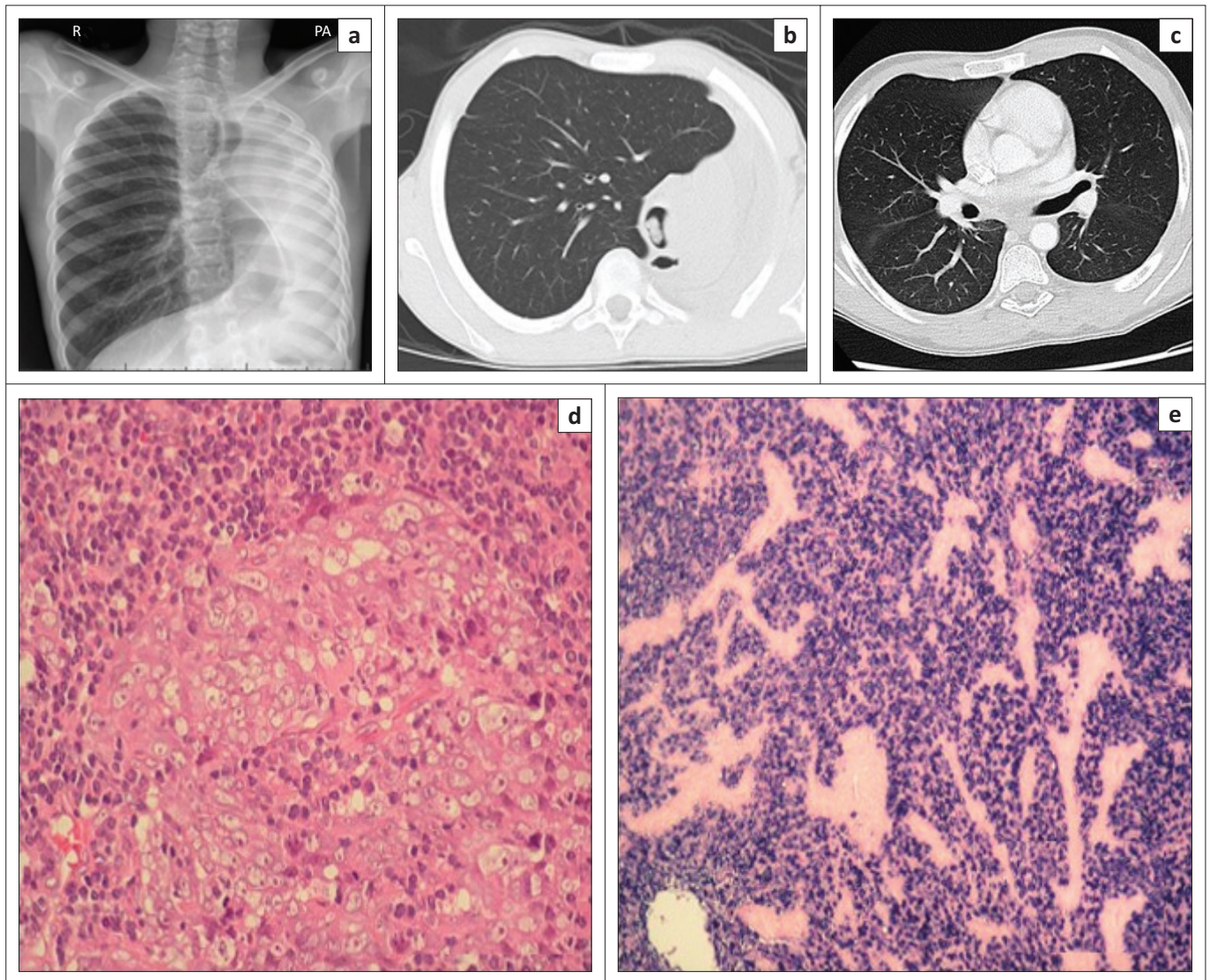
Histology showed nests and sheets of pleomorphic tumour cells with prominent nucleoli and a syncytial arrangement. A marked lymphocytic infiltrate was also present (Figure 1d). Immunohistochemical stains for AE1/AE3, CK5/6 and p63 were positive in the epithelial cells, while a leukocyte common antigen (LCA) was positive in the lymphocytes. A negative placental alkaline phosphatase (PLAP) stain excluded a germ cell tumour and negative chromogranin and synaptophysin stains excluded a neuroendocrine tumour. In addition, stains for CK7, CK20, TTF1 and napsin A were negative. Epstein-Barr encoding region (EBER) *in situ* hybridisation (EBER-ISH) was positive (Figure 1e).

A differential diagnosis of LELC of the lung or metastatic nasopharyngeal carcinoma was made. On further investigation, no nasopharyngeal mass was evident and, consequently, the final diagnosis was that of a primary pulmonary LELC. He was given six cycles of chemotherapy that included cisplatin, 5-fluorouracil and leucovorin. He also received 6 weeks of irradiation with a total dose of 5400 cGy. A follow-up CT scan (Figure 1c) 6 months after completion of treatment showed a small 17 mm × 14 mm mass adjacent to the left main

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EBER, Epstein-Barr encoding region, ISH, *in situ* hybridisation.

**FIGURE 1:** (a) Chest x-ray at presentation shows a collapsed left lung with mediastinal, tracheal and cardiac displacement, (b) CT scan at presentation demonstrates marked hyperinflation of the right lung with a collapsed left lung and a mass in the left main bronchus, (c) CT scan 6 months post-treatment completion shows normal lungs and no evidence of the bronchial mass, (d) Histological features of the tumour. Sheets of pleomorphic tumour cells with a syncytial arrangement infiltrated and surrounded by numerous lymphocytes, (e) EBER-ISH with strong positivity.

bronchus, which was consistent with post-therapy fibrosis, and the lung collapse had resolved. When seen at his 12-month follow-up, the patient was alive and disease-free.

## Ethical considerations

Ethical approval to report the case was obtained from the institutional Ethics Committee (reference number HSREC 176/2017). The child's mother provided written consent to publish the findings and images obtained during work-up of this case.

## Discussion

Pulmonary LELC is a rare tumour most commonly occurring in Asian patients. Patients tend to be younger than those with other types of lung cancer, and there is no sex predilection. They are also more likely to present at an earlier stage. As seen in this case, the majority of patients present with a

history of coughing.<sup>5</sup> Other symptoms include haemoptysis, chest pain and weight loss. The prognosis of patients with pulmonary LELC is significantly better than those with other types of lung carcinoma.<sup>4,6</sup> In a series of 62 patients reported by He et al.<sup>4</sup> the median survival of patients with LELC was 107 months, in contrast to a median survival of 13 months for non-LELC patients. Han et al.<sup>6</sup> showed a significant difference in survival for patients with stage II, III and IV disease. However, there was no significant difference in survival rates among patients with stage I disease. Tumour recurrence and necrosis have been associated with a poor prognosis.<sup>6</sup> Patients over 65 years of age have also been found to have a significantly worse survival outcome.<sup>4</sup>

It is important to distinguish pulmonary LELC from nasopharyngeal carcinoma with metastases to the lungs, as both tumours have an identical histological appearance. Misdiagnosis will lead to incorrect staging and management. In addition, cases with a very prominent lymphoid infiltrate

**TABLE 1. Reported cases of paediatric pulmonary lymphoepithelioma-like carcinoma.**

Author	Age (years)	Gender	Race	Presentation	Outcome	EBER
Liang et al. <sup>3</sup>	9	Female	Chinese	Not stated	Alive at 37.8 months	Not stated
He et al. <sup>4</sup>	15	Not stated	Not stated	Not stated	Not stated	Not stated
Curcio et al. <sup>9</sup>	8	Female	Chinese	Clubbing of fingers and toes	Alive and disease-free at 20 months	Positive
Kundu et al. <sup>10</sup>	13	Female	Indian	Cough, intermittent fever and progressive dyspnoea	Alive and symptom-free after more than 2 years	Not done
Present case	12	Male	African	Cough and dyspnoea	Alive and disease-free at 12 months	Positive

EBER, Epstein–Barr encoding region.

could be mistaken for non-Hodgkin lymphoma, which does not require surgical intervention.<sup>7,8</sup>

Only tumours from the upper aero-digestive tract and thymus have been associated with EBV. This association has been limited to Asians with LELC of the salivary gland and lung.<sup>1,5</sup> This article presents the first documented case of EBV-positive pulmonary LELC in an African patient.

Four cases of pulmonary LELC have been described in children. Curcio et al.<sup>9</sup> reported a case in an 8-year-old girl of Chinese decent, while Kundu et al.<sup>10</sup> described a case of LELC in a 13-year-old girl of Indian decent. A 9-year-old Chinese girl was reported by Liang et al.<sup>3</sup> in a case series of 52 patients, while He et al.<sup>4</sup> found that the youngest person in their series of 62 patients was 15 years old (Table 1). One of the patients reported previously was alive without evidence of recurrence after 20 months<sup>9</sup> and another one after more than 2 years of follow-up.<sup>10</sup> The 9-year-old child reported by Liang et al.<sup>3</sup> was alive after 37.8 months, although no further information was provided. No follow-up data were available for the 15-year-old case reported by He et al.<sup>4</sup> In our case, the patient was a 12-year-old boy of African descent who had responded well to treatment. He was initially misdiagnosed with pulmonary tuberculosis because of the clinical history of coughing and a positive tuberculosis contact. Although pulmonary tumours are rare in children and tuberculosis is extremely common in South Africa,<sup>11</sup> it is important to exclude the possibility of a tumour to avoid delays in treatment.

In summary, we present a rare case of EBV-positive pulmonary LELC in an African child. Metastases from a nasopharyngeal carcinoma should be excluded before making a diagnosis of pulmonary LELC.

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## Competing interests

The authors declare that they have no financial or personal relationships that may have inappropriately influenced them in writing this article.

## Authors' contributions

J.G. was responsible for the histological investigations, provided the histology photographs and wrote the manuscript. D.S. provided the clinical information and radiological images and approved the final version of the manuscript.

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