Nephrotic syndrome: first presentation of lymphoepithelioma-like thymic carcinoma

Introduction

Lymphoepithelioma-like carcinoma is an infrequently encountered primary malignancy affecting the epithelial tissues of the thymus gland. This article describes a patient in whom nephrotic syndrome was the initial presentation of lymphoepithelioma-like carcinoma and discusses the clinical course and therapeutic strategy for this disease.

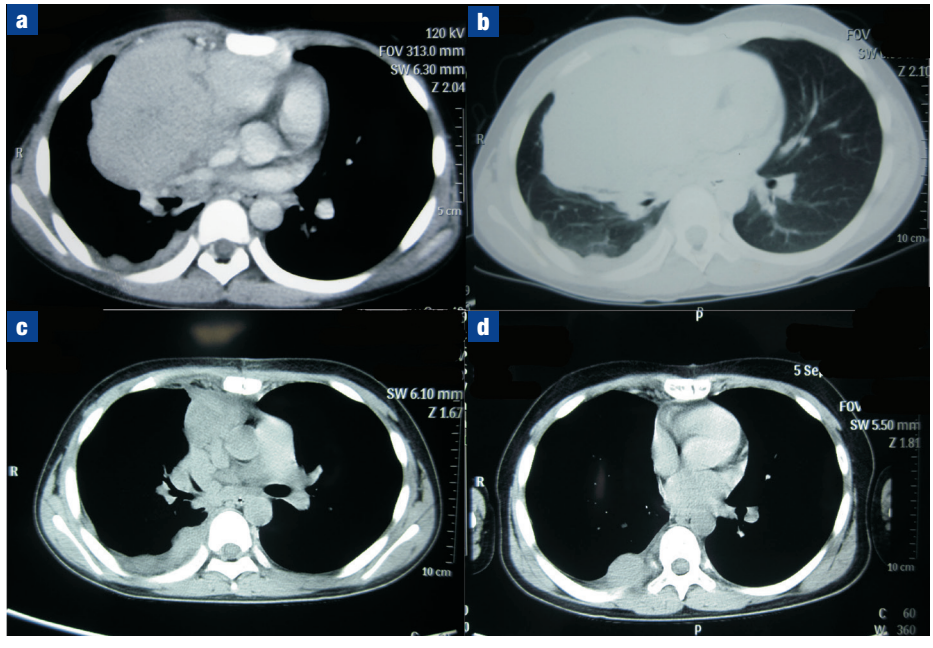
Discussion

Lymphoepithelioma-like carcinoma is a high-grade thymic carcinoma, which behaves more aggressively and exhibits a significantly higher invasive and metastatic potential than low-grade thymic carcinomas. Extrathoracic spread is observed in <10% of cases (kidneys, lymph nodes, liver, brain, adrenals, thyroid, bone) (Arenas et al, 2009). The kidney can become affected before, after or during thymic disease (Karras et al, 2005). Histologically, minimal change is the most frequently seen pathological characteristic in nephropathy (Arenas et al, 2009).

The association between thymic disease and cases of nephrotic syndrome remains unclear, but the thymus is the main organ for immunity, and the production and maturation of lymphocytes is disrupted in patients with thymus carcinoma or thymic hyperplasia before, and even after, thymectomy is performed (Hoffacker et al, 2000). Therefore, thymic malignancies associated with immunological disorders are common.

Treating lymphoepithelioma-like carcinoma is challenging because of the very small number of cases. Cisplatin-based combination chemotherapy is considered an important step in tumour management. Modh et al (2016) concluded that aggressive treatment with chemotherapy, surgical

Figure 1. **a** and **b.** Chest computed tomography scan revealed a large isodense mass (16 cm × 10 cm × 12 cm) encompassing the lung and the pleura. **c.** Chest computed tomography scan 1 month after surgery. **d.** Chest computed tomography scan 4 months after surgery; pleural metastases were enlarged.



CASE REPORT

In January 2009, a 7-year-old Chinese boy was diagnosed with nephrotic syndrome. He received steroid therapy for 1 year and then went into remission.

In May 2013, the patient was readmitted to the authors' hospital with breathlessness at night but no other symptoms. Computed tomography of the chest revealed a large isodense mass (16 cm × 10 cm × 12 cm) encompassing the lung and the pleura (*Figure 1*). A 600 g tumour was located in the anterior mediastinum. The tumour was surgically removed through tumourectomy. Multiple metastases were found in the right diaphragm and the visceral pleura. The tumour stage was IVb. Grossly, the tumour was not well circumscribed (*Figure 2*). The tumour tissue was histopathologically examined, revealing a neoplasm composed of epithelial cells with prominent cytological atypia (*Figure 2*). Immunohistochemical analysis showed that the tumour cells were uniformly positive for cytokeratin (CK), CK5/6, epithelial membrane antigen, CD117 and CD5. Lymphoepithelioma-like carcinoma of the thymus was diagnosed.

The child received three cycles of systemic chemotherapy and local irradiation. He exhibited sensitivity to radiation, but the disease progressed in the pleura. The patient remains under radiation therapy.

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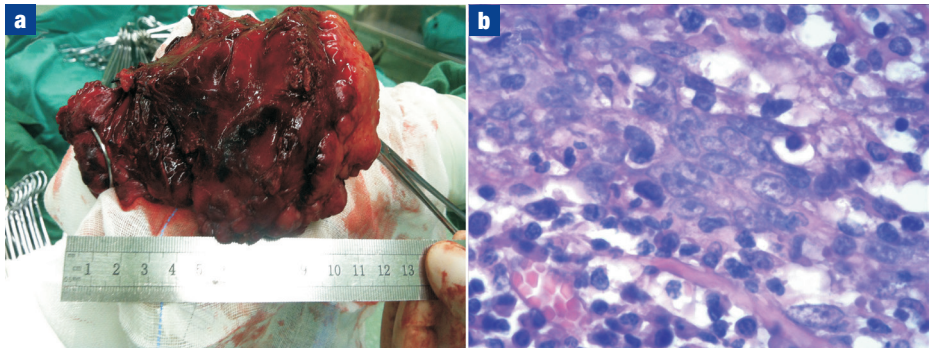
Figure 2. **a.** Gross appearance of the primary tumour. **b.** Pathological slides of lymphoepithelioma-like carcinoma (haematoxylin-eosin staining, 400×).



resection and postoperative radiation therapy can promote the long-term survival of patients with invasive thymic malignancies. In the current case, chemotherapy was ineffective. The child received a total of 54 Gy of radiotherapy which reduced the tumour size, but the disease relapsed in another part of the pleura.

Conclusions

Children presenting with renal diseases should be followed up to detect thymic carcinomas in the early stages. Cellular pathological characteristics and molecular biology linking these two conditions should be further investigated. BJHM

LEARNING POINTS

- Lymphoepithelioma-like carcinoma is usually associated with paraneoplastic syndromes, such as myasthenia gravis, pure red cell aplasia or systemic lupus erythematosus.
- Lymphoepithelioma-like carcinoma is only diagnosed once the neoplasm becomes large enough to infiltrate into the surrounding structure; patients with lymphoepithelioma-like carcinoma can initially be misdiagnosed as having pneumonia.
- Children presenting with renal disease should be followed up to detect thymic carcinomas in the early stages.

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