

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).

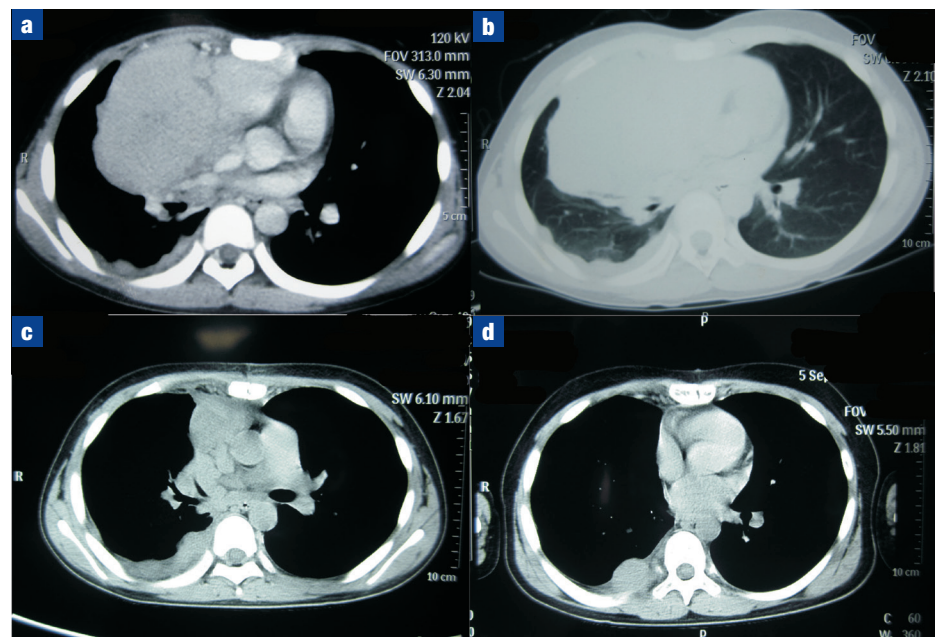
The association between thymic disease and cases of nephrotic syndrome remains unclear, but the thymus is the main organ for immunity, and the production and maturation of lymphocytes is disrupted in patients with thymus carcinoma or thymic hyperplasia before, and even after, thymectomy is

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.

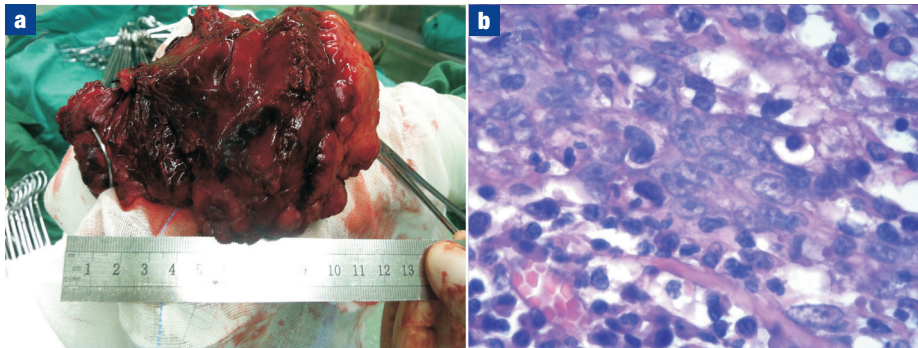
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Figure 2. **a.** Gross appearance of the primary tumour. **b.** Pathological slides of lymphoepithelioma-like carcinoma (haematoxylin-eosin staining, 400×).



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**

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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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Smiths Medical, a leading global medical device manufacturer, has released the CADD®-Solis ambulatory infusion pump version 4.1 that sets the foundation for wireless communication globally. The CADD®-Solis v4.1 pump is UL-marked and has full compliance with the latest product standards, including IEC-60601-1 Edition 3.1. The CADD®-Solis pump maintains the advantages of an ambulatory pump for patient mobility and also provides a single system that effectively delivers IV PCA, epidurals and peripheral nerve blocks from pump to patient.

The CADD®-Solis v4.1 pump enables new features such as:

- Longer drug names to eliminate abbreviating or truncating drug names and concentrations.
- The addition of profiles to categorize drug protocols or represent care areas.
- Wireless two-way communication with the PharmGuard® Server software to help increase efficiencies, reduce costs, and improve patient outcomes.¹

Customers can continue to use previous versions or choose to upgrade any CADD®-Solis model 2110 pump² to CADD®-Solis v4.1 software using SureLink® remote support software, while maintaining their existing drug library. Turning off wireless communications or the lack of wireless access does not impact pump delivery or data retention.

Jeff Hohn, General Manager & Vice President, Infusion Systems, at Smiths Medical, says: "The new CADD®-Solis system is a continuation of Smiths Medical's commitment to advancing patient care and helping to improve outcomes through leading-edge technology. Wireless bi-directional communication sets the foundation for integrating pain management data directly into the patient records in the hospital's Electronic Medical Records (EMR), saving clinician time charting and increasing documentation accuracy. For example, providing PCA data into patient monitoring systems could be used to manage the risk of respiratory depression from the over use of opioids."

For more information on the CADD®-Solis ambulatory infusion pump range, contact Glen Johnson, NordUK Marketing Manager at Smiths Medical 01233 722 100 ● glen.johnson@smiths-medical.com ● www.smiths-medical.com

References
 1 Wireless communication at launch only available for English language pumps
 2 Model 2100 pumps manufactured before 2011 (Serial Numbers <10012365) cannot be upgraded to v4.1 pump software