About a lymphoepithelioma-like carcinoma of the lung with an endotracheal localization

A propos d'un carcinome lympho-epithélioma like pulmonaire de localisation endo-trachéale

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RÉSUMÉ

Introduction: Le carcinome lympho-epithelioma like est une tumeur pulmonaire rare représentant moins de 1% des carcinomes non à petites cellules pulmonaires. Il est classé parmi les entités spéciales dans la classification de l'OMS 2015.

Objectif: Notre objectif était de décrire un nouveau cas de carcinome lympho-epithélioma like pulmonaire et de mettre en évidence les difficultés diagnostiques.

Méthodes : Nous décrivons un nouveau cas colligé dans notre service d'Anatomie Pathologique

Présentation du cas: Les auteurs décrivent le cas d'une patiente âgée de 22 ans sans antécédent particulier qui a consulté pour une symptomatologie respiratoire non spécifique. Les moyens d'imagerie ont mis en évidence une tumeur trachéale associée à des adénopathies médiastinales. Une première biopsie a été réalisée et l'examen anatomopathologique avait conclu à une tumeur maligne avec une différenciation malpighienne focale mise en évidence par les études immunohistochimiques. Une biopsie chirurgicale a été réalisée afin d'asseoir le diagnostic et avait conclu à un cacinome lympho-epithélioma like pulmonaire. Le diagnostic a été retenu après avoir eliminé une éventuelle localisation métastatique d'un carcinome nasopharyngé

Discussion et conclusion: Ce cas illusre la rareté de la tumeur décrite, spécialement chez une patiente jeune et caucasienne. De plus, il met en evidence les principaux challenges diagnostics sur les prélèvements biopsiques.

Mots-clés

Diagnostic, carcinome lymphoepithelioma-like, poumon, pronostic.

SUMMARY

Introduction : Lympho-epithelioma like carcinoma is a rare lung tumour that acounts for less than 1% of non small cell lung carcinomas. It is defined as a special entity among the 2015 World Health Organization.

Aim : Our aim was to describe a completely illustrated new case of lymhpo-epithelioma like carcinoma.

Methods : We describe a new case diagnosed in our Department of Pathology.

Case presentation : The authors describe a case of a 22-year-old woman without a particular past medical history who presented non specific respiratory symptoms. Radiologic investigations revealed a tracheal tumor with enlarged mediastinal lumph nodes. A first biopsy was performed revealing a malignant tumor with a squamous differentiation highlighted by immunohischemistry. A surgical biopsy was performed and the final microscopic diagnosis revealed a lymphoepithelioma-like carcinoma of the lung. This diagnosis was retained after ruling out a possible metastasis of a nasopharyngeal carcinoma whose microscopic features are similar to this subtype of lung tumor.

Discussion and conclusion : This case points out the rarity of this diagnosis, especially in a young and caucasian patient and highlights the diagnostic dilemma caused by this kind of tumor.

Key-words

Diagnosis, lymphoepithelioma-like carcinoma, lung, prognosis.

Lymphoepithelioma-like carcinomas of the lung are rare tumours accounting for less than 1% of lung cancers. In the 2004 WHO classification, these tumours were considered as a particular subtype of large cell carcinomas which are mainly observed in Asia. In the 2015 WHO classification, these tumours are considered as special entities independant form large cell carcinomas. They are usually asymptomatic because of their peripheral localization and present a good response to chemotherapy. We describe a new case of lymphoepithelioma-like carcinoma observed in a caucasian patient. This case is particular by the young age of the patient and the particular proximal localization of the tumour causing diagnostic dilemma especially on biopsy samples.

CASE PRESENTATION

We describe the case of a 21-year-old patient without a particular past medical history, who presented a long lasting cough, dyspnea with recurrent bronchitis. Chest-x-ray showed a mediastinal enlargement. The bronchial endoscopy revealed an endotracheal tumour. A bronchial biopsy was performed. Microscopic examination revealed a malignant tumour made of nests of round cells expressing the P63 antibody. This positivity highlighted a squamous differentiation but the diagnosis of a squamous cell carcinoma was almost improbable because of the young age of the patient and the endotracheal presentation. According to these arguments, the diagnosis of a non small cell carcinoma probably corresponding to a mucoepidermoid carcinoma was retained. A thoracic CT-scan was performed and showed an endotracheal mass with many mediastinal lymph nodes (figure 1). A thoracotomy was performed and an extemporaneous exam of the mediastinal lymph nodes was performed.

Gross examination revealed 3 lymph nodes of 2 cm. Microscopic examination revealed a lymph node infiltration by a non small cell carcinoma process. The definite microscopic examination after fixation and inclusion in paraffin blocks revealed syncytial tumour cells of medium size which were intimately connected to a lymphoid stroma. Tumour cells were characterized by atypical nuclei (Figure 2). An immunohistochemical study using cytokeratin (clone AE1/AE2, Leica, dilution 1 :100), Epithelial Membrane antigen (clone GP1.4, Leica, dilution 1 :100), P63 (clone 7JUL, Leica, ready-to-use), P40 (clone BC28, Ventana, ready-to-use) and EBNA2 (clone PE2, Leica, dilution 1 :25) antibodies was performed. It revealed the positivity of the tumour cells with the cytokeratin and Epithelial Membrane Antigen (Figure 3a). A focal expression of P63 and P40 was also noticed (Figure 3b). Tumor cells didn't express the EBNA2 and LMP antigens (DMAB12153, Creative diagnostics, dilution 1 :100). According to these findings, the diagnosis of a lymphoepithelioma-like carcinoma was retained. Its pulmonary orgin couldn't be retained unless a nasopharyngeal carcinoma was ruled out. A cerebral MRI was performed and was normal.

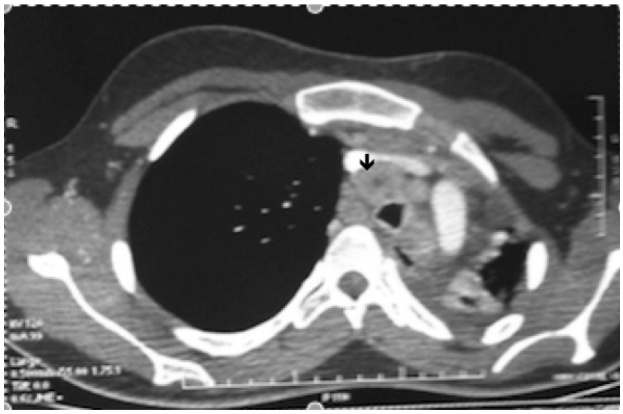

Figure 1 : Chest CT-scan showing paratracheal lymph nodes and an endotracheal process

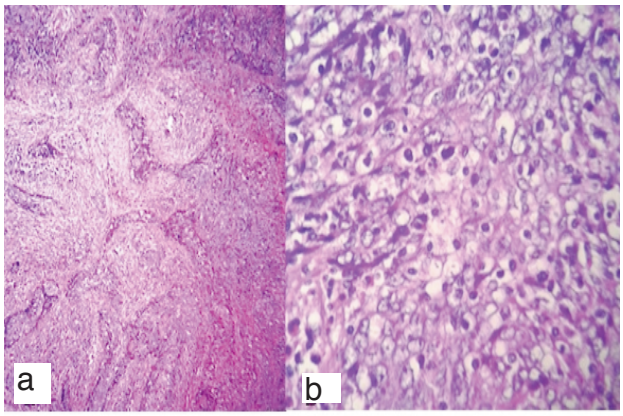

Figure 2: Microscopic examination showing nests of tumour cells intimately connected to an abundant lymphoid stroma (HE x 250). Inset: Tumour cells were ovoid with atypical nuclei (HE x 400).

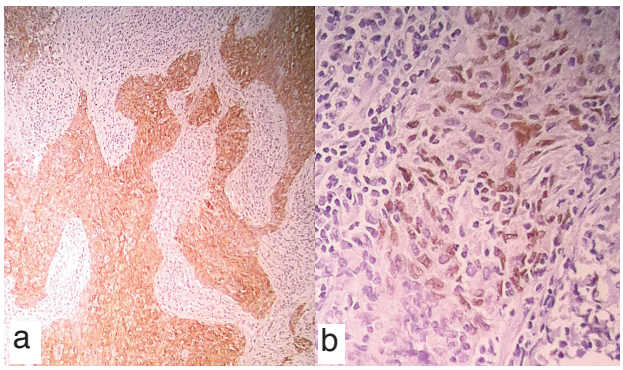

Figure 3: a/ Immunohistochemical study revealing a diffuse expression of the cytokeratin by the tumour cells (HE x 400). b/ Immunohistochemical study revealing a focal expression of P40 by tumour cells (HE x 400)

DISCUSSION

We describe a rare presentation of a lymphoepithelioma-like carcinoma of the lung. In fact, the patient presented a mediastinal process with an endo-bronchial component evoking a salivary gland type tumour or a mediastinal lymphoma involving the trachea. This kind of tumour is generally peripheral and an endobronchial presentation has been rarely reported in the literature (1, 2). Besides, these tumours were mainly reported in Asian patients. Gross findings consist in ovoid tumours ranging from 1 to 11 cm. Microscopic examination reveals the same features observed in nasopharyngeal carcinoma and this diagnosis has to be ruled out before retaining the lung origin. In our observation, the cerebral MRI revealed no abnormalities (1). Microsocpic examinaion shows in almost all cases focal keranizing maturation explaining the positivity of P63 and P40 in the immunohistochemical study. In some cases, tumour cells could express LMP-1 suggesting a potential role of the Epstein Barr Virus (3-5). This expression wasn't revealed in our case.

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