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Mucoepidermoid carcinoma of the lung in a 13 year old male: A high grade variant with bilateral supraclavicular lymph node metastasis *

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Summary

High grade mucoepidermoid carcinoma of the lung presenting in childhood is very rare. To date there have been 25 mucoepidermoid carcinoma cases reported in childhood age groups. Except one, all of these were low-grade carcinomas with a benign clinical course. We present a high grade mucoepidermoid carcinoma, in left lower lung of 13 year old male, with bilateral supraclavicular and cervical lymph node metastasis. To our knowledge in this age group this is the second instance of high grade Mucoepidermoid Carcinoma (MEC) associated with metastasis.

Key words: *High grade mucoepidermoid carcinoma, lung, child.*

Introduction

Since Liebow's description of mucoepidermoid carcinoma arising from the tracheobronchial tree in 1952 [5], 25 cases have been reported during childhood [1-10]. This uncommon neoplasm arises from the excretory ducts of the bronchial mucosa. Most children present with a history of a persistent or recurrent cough or pneumonia. The low grade biological behavior characteristic of mucoepidermoid carcinoma in childhood makes conservative pulmonary resection an adequate therapy [1-10].

Case Report

13 year old male was referred to Oncology Hospital Etimesgut Ankara for evaluation of recurrent cough and bilateral supraclavicular-cervical lymphadenomegaly of 3 months duration. There was no history of tuberculosis exposure or foreign body aspiration. He was subfebrile and in respiratory distress. Auscultation of the chest revealed decreased breath sounds in the left middle and lower lobe distribution.

The admission chest roentgenogram showed nearly complete left and partial right lung pleural effusion and mediastinal shift to the right side (Figure 1). Radiologic and scanning examination of the skeletal system were not pathologic. Hemoglobin was 11.5 gm/dl, Leukocyte was 7100/mm^3 with a normal differential count. The sedimentation rate was 42/75 mm/hour. Blood urea nitrogen and uric acid were 26 % mg, 5.3 % mg respectively. A tuberculosis skin test was nonreactive.

Urine sediment and intravenous pyelography was normal. Liver and spleen scanning showed minimal diffuse hepatosplenomegaly. Needle biopsies of pleura and left lung were performed after evacuation of 2500 cc pleural effusion. The histopathological result was chronic pleuritis. Cytological examination revealed Class II. Culture of the effusion

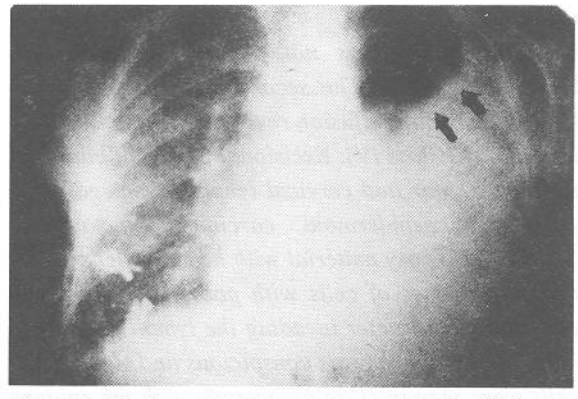

Figure 1. Chest roentgenogram showed pleural effusion on the left and moderate mediastinal shift toward right side.

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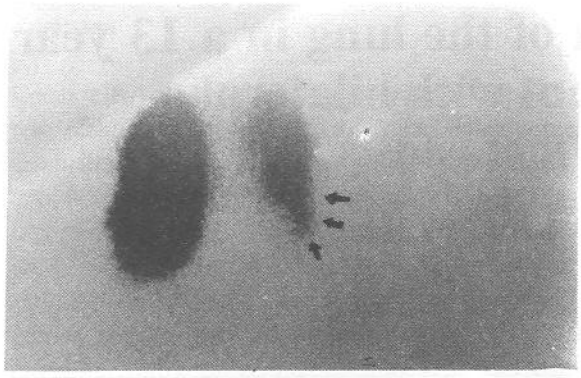

Figure 2. Perfusion scanning of the lungs: The right lung was homogenous and showed mild compensatory expansion. Lower zone of the left lung showed a mass with lack of perfusion (arrow).

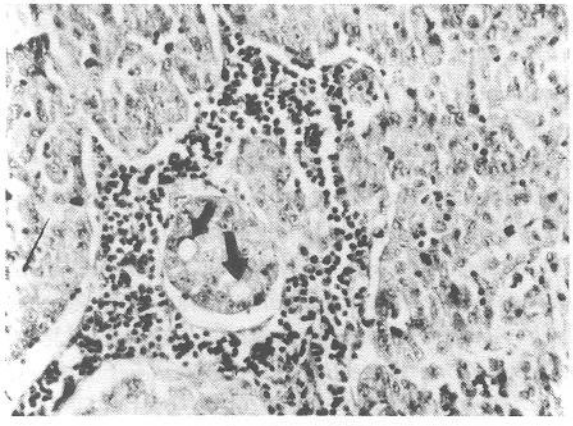

Figure 3. Masses of cells showing poorly differentiated epidermoid character. Nuclear pleomorphism is conspicious and signet-ring type cells can be seen in close association with the epidermoid cells (arrows). HE. 250x.

revealed no bacterial growth. Perfusion Scanning: The right lung was homogenous, and there was hilar enlargement and compensatory lung widening. Lower zone of the left lung showed a mass with lack of perfusion (Fig. 2).

Ultrasonography was not pathologic. Biopsy of testes was normal. The second cytologic examination of the pleural effusion revealed malignant cellular activity (Class IV). Excisional biopsy of bilateral supraclavicular and cervical lynphadenomegaly revealed mucoepidermoid carcinoma metastasis. Staining of biopsy material with hematoxylene eosin revealed masses of cells with poorly differentiated epidermoid character invading the lymph node. Nuclear pleomorphism was conspicious and signet-ring cells were seen in close association with the epidermoid cells (Fig. 3).

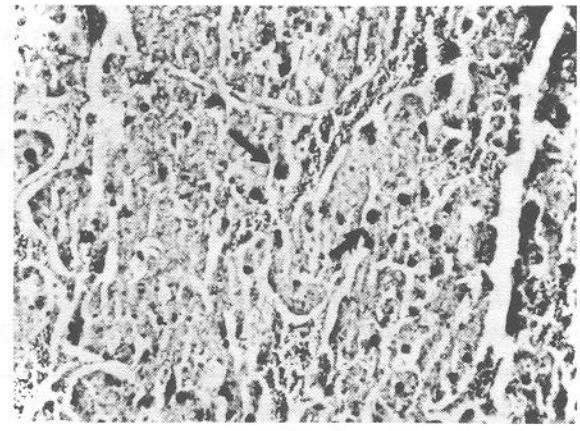

Figure 4. The signet-ring types cells can be seen staining positively with Period Acid Schiff stain (arrows). PAS 100x.

The signet-ring type cells stained positively with periodic acid schiff and mucicarmen stains (Fig. 4). With all these data, the diagnosis was grade 3 highly malignant mucoepidermoid carcinoma with cervical metastasis. Chemotherapy was administered with agressive doses of VACA and Methotrexate. He expired in the 3rd month of the post chemotherapy period.

Discussion

Foreign body aspiration, bronchiectasis, tuberculosis and endobronchial hamartomas should be considered in any child who presents with persistent cough, hemoptysis, dyspnea, wheezing or recurrent pneumonia and/or atelectasis. When symptoms of endobronchial obstruction persist or progress, bronchogenic neoplasms, although rare in this age group, become part of the differential diagnosis.

Chest radiographs are usually normal in cases of tracheal mucoepidermoid carcinoma, but most bronchial lesions present with recurrent or persistent lobar infiltrate, atelectasis, or bronchiectasis due to partial endobronchial obstruction. Cytologic studies of sputum, bronchial washings, or brushings are not helpful in establishing the diagnosis of mucoepidermoid carcinoma, as these lesions are usually covered by intact respiratory epithelium [5,7].

Needle biopsy of lung and cystologic examination of our case were repeated five times. Positive result was only once in cytologic examination.

Needle aspiration shows three cytologic features characteristic of these tumors: **a)** three cell types: spindle cells, epidermoid cells, and mucus secreting cells found in close proximity to each other. **b)** mucus-secreting cells frequently found in close association to more numerous epidermoid cells. **c)** a connective tissue core lined with a layer of spindle cells covered by epidermoid cells [4,7].

Histologic classification into low (grade one), intermediate (grade two), and highly malignant (grade three) tumors on the basis of cellular differentation and mitotic activity is possible. Nuclear hyperchromasia, pleomorphism, and mitotic figures are rarely seen in the lowgrade well-differentiated tumors characteristic of childhood [1]. In the high grade variant, pleomorphism is conspicous, mitoses are present, mucus cells are found in the shape of signet-ring, as is in our case [7].

25 cases of mucoepidermoid carcinoma in childhood have been reported. Mucoepidermoid carcinoma in children appears to remain localized for a long period, exhibiting a low metastatic potential. Only one patient in 25 reported cases was noted to have peribronchial lymph node metastasis. Available follow up of these patients ranges form 8 months to 23 years, and tumor recurrence has not been reported. Radiation therapy has not been employed in any case and seems unnecessary when complete surgical resection is possible [2,4].

Our case is the second highly malignant mucoepidermoid carcinoma with bilateral supraclavicular and cervical lymph node metastasis. He expired in the 3rd month of the post chemotherapy period.

References

1. Breyer RH, Dainauskas JR, Jensik RJ, et al: Mucoepidermoid carcinoma of the trachea and bronchus. The case for conservative resection. Ann Thorac Surg 29:197, 1980
2. Hetta L, Olin C, Rutfors PE: Tracheal adenoma in a 13 year old boy with suspected asthma. Lakartidnigen 77:3532, 1980
3. Kalcsman PG, Olson JI, Eggleston JC: Mucoepidermoid carcinoma of the bronchus: An electron microscpic study of the low grade and the high grade variants. Cancer 43:1720, 1979
4. Lack EE, Harris CGB, Eraklis AJ, Vawter GF: Primary bronchial tumors in childhood. A clinicopathologic study of six cases. Cancer 51:492, 1983
5. Liebow AA: Tumors of the lower respiratory tract, in Atlas of Tumor Pathology. Washington DC, Armed Forces Institie of Pathology, National Research Council Section V, Fascicle 17, 1952
6. Mullins JD, Barnes RP: Childhood bronchial mucoepidermoid tumors. A case report and review of the literature. Cancer 44:315, 1979
7. Seo I, Warren J, Mirkin LD, Weisman SJ, Grosfeld JL: Mucoepidermoid carcinoma of the bronchus in a 4 year old child: A high grade variant with lymph node metastasis. Cancer 53:1600, 1984
8. Stafford JR, Pollock WJ, Wenzel BC: Oncocytic mucoepidermoid tumor of the bronchus. Cancer 54:94, 1984
9. Tepprman BS, Feld R, Fosse RG: Squamous cell carcinoma of bronchus associated with hypercalcemia in a twenty year old. Cancer 47:2690, 1981
10. Torres AM, Ryckman FC: Childhood tracheobronchial mucoepidermoid carcinoma: A case report and review of the literature. J P S 23:367, 1988