LYMPHOEPITHELIOMA – A TUMOUR RARELY OBSERVED IN CHILDREN (3 CASES)

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ABSTRACT

Lymphoepithelioma rarely develops in children. In its early stage it may manifest itself only as enlarged cervical lymph nodes, which is likely to cause difficulties in the initial diagnosis. Radiotherapy is the treatment of choice, however, chemotherapy has also been used. The paper describes three cases of children treated for lymphoepithelioma at the Department of Children Hematology and Oncology in Lublin between 1991 - 2001. in all cases, the diagnosis was based on histopathological examinations of the tumour or lymph node biopsies. The methods of treatment used were different in each case and the longest disease free survival was 3 years. Therefore it seems necessary to design a uniform protocol of treatment for children with lymphoepithelioma.

The first child was treated only to radiotherapy. This choice was based on lack of results of well conducted trials of cytostatic treatment. The good result of our treatment confirmed the data presented by other with the use of radiotherapy alone.

In the second case, in case of large tumour, paralysis of cranial nerves, this patient was refered to combined treatment (radio-chemotherapy). In this case the partial remission was achived.

In the third case, chemotherapy (cisplatin + 5-fluorouracil) resulted in perfect remission of the primary tumour and metastases to the lymph nodes while radiotherapy allow to achive complete control of the primary tumour and metastases to lymph nodes.

Key words: lymphoepithelioma, children, treatment, radiotherapy, chemotherapy.

INTRODUCTION

Lymphoepithelioma is a rare childhood tumour originating from the transitional epithelium of the nasopharynx. Its first clinical sign is the enlargement of the cervical lymph nodes. The other signs and symptoms include: bleeding from the nose, sore throat, difficulties in swallowing, hypoacusia. The cranial basis infiltration results in the paralysis of the cranial nerves and severe pain [1]. Due to high radiosensitivity, radiotherapy is the treatment of choice, however, chemotherapy has also been undertaken [2].

CASE REPORTS

The paper presents three cases of children treated for lymphoepithelioma at our department between 1991 - 2001.

Case report I. A 15-year-old boy was referred to the department in September 1991 because of gradually enlarging cervical lymph nodes on the left side, weight loss and trismus. The laboratory tests revealed: high SR (65/100), mild anaemia, leucocytosis with neutrophilia. The CT scan of the nasopharyngeal cavity showed an abnormal tissue mass filling the left palatopharyngeal space. The histopathological findings of the tumour's biopsy confirmed lymphoepithelioma. Radiotherapy of the nasopharyngeal cavity and enlarged lymph nodes was used (50 Gy), which resulted in complete regression of the proliferative disease. The boy was followed-up in the Oncological Outpatient Clinic for 3 years and showed total remission.

Case report II. An 8-year-old girl was admitted to the Department in December

1995 because of dropping of the left upper eyelid, vision disorders and massive bleeding from the nose. The laboratory tests did not reveal any abnormalities. The CT scan of the head visualized a giant mass filling the whole nasopharyngeal cavity, which penetrated the final conus of the left orbit. The histopathological findings showed the features of lymphoepithelioma. Chemotherapy (Epirubicin + Cisplatin) and radiotherapy (56 Gy) were used. After the fifth cycle, the left eyelid dropping subsided and MRI revealed partial regression of the tumour. The girl was qualified for surgery once the peripheral blood parameters have become normal. However, after two weeks the patient was again admitted to the Department because of a rapidly growing tumour located in the lateral-anterior surface of the right shank. The CT scan showed the 5.7 x 2.3 cm tumour in the soft tissues of the right fibula. The chest X-ray revealed the metastases in the right lung. Chemotherapy was changed to BEC (Bleomycin, Epirubicin, Cisplatin) and then to MEV (Methotrexate, Cyclofosfamid, Vincristine). Moreover, the lung (10 Gy) and right shank (50 Gy) radiotherapy was instituted. The pulmonary lesions regressed while the tumour of the right shank increased. Additionally, dissemination of the inguinal lymph nodes was observed. The girl was subjected to palliative treatment and died due to progression of the disease.

Case report III. A 14-year-old boy was referred to the Department because of bilaterally enlarged cervical lymph nodes. The laboratory tests showed no abnormalities. MRI revealed a tumour of the nasopharyngeal cavity and the histopathological findings confirmed lymphoepithelioma. The patient was subjected to chemotherapy (4 cycles of Cisplatin + 5-Fluorouracil) and radiotherapy of the nasopharyngeal cavity and cervical lymph nodes (56 Gy) and total regression of the tumour and metastatic lesions was achieved. The boy has been followed–up for three months now and total clinical and radiological remission has been observed.

DISCUSSION

Carcinomas of the nasopharyngeal cavity constitute less than 1% of childhood malignant tumours, which is, however, 1/3 of the nasopharyngeal tumours [3]. The majority of nasopharyngeal tumours in children belong to the non-differentiated group and their development is related to the Epstein-Barr (EBV) infection [4,5]. At present, the tumours previously described as lymphoepithelioma are diagnosed as poorly differentiated epithelial carcinomas. The clinical manifestations, ways of spreading and prognosis are similar in children and adults [6].

The primary tumour of the nasopharyngeal cavity may be latent and the disease manifests itself only in enlarged cervical lymph nodes, which leads to initial diagnostic difficulties [1]. Prior to surgery of the nasopharyngeal cavity, the lymph node biopsy is indicated [7]. Spreading of the tumour other than to the lymph nodes is likely to develop as spreading through the continuity to the cranial basis with the accompanying symptoms [8]. The remote metastases on diagnosis are observed in less than 5% of cases and may develop in the lungs, mediastinum, bones and liver [9]. Therefore, the initial evaluation should include CT/MRI of the head, neck and abdominal cavity, bone X-ray and scintigraphy. If the cranial basis infiltration is detected, the lumbar puncture should be performed. The metastases to the bone marrow are rare [10]. The differential diagnosis should consider angiofibroma of the nasopharyngesal cavity and rhabdomyosarcoma [11].

The main treatment method in lymphoepithelioma is radiotherapy [2]. Operative procedures provide only the diagnostic material from the tumour or lymph nodes or enable to resect the residual lesion left after radiotherapy. The recommended doses in primary tumours are 50 Gy (small tumours in younger children) to 60 Gy (big tumours infiltrating the cranial basis in older children) [12]. However, since radiotherapy does not provide perfect control, the studies on the effectiveness of chemotherapeutic agents are carried out. The drugs used include: Cisplatin, Vincristine, Methotrexate, Bleomycin,

Adriamycin, 5-Fluorouracil, Mitomycin C [13,14,15].

The first child described was subjected only to radiotherapy. This choice was based on good results of this management in localized tumours and lack of experience in cytostatic treatment (the boy was treated over 10 years ago). The therapeutic outcome confirmed the data presented by other authors concerning the use of radiotherapy alone.

In the second patient, the features of poor prognosis were already observed on diagnosis (a big tumour, paralysis of the oculomotor nerve). Therefore the patient was subjected to combined treatment (chemo- and radiotherapy) and partial remission of the primary lesion was achieved. However, the disease could not be stopped when remote metastases developed.

In the third child, chemotherapy (Cisplatin + 5-Fluorouracil) resulted in perfect remission of the primary tumour and metastases to the lymph nodes while radiotherapy (55 Gy) provided good local control of the primary tumour and metastatic lesions.

CONCLUSIONS

1. Lymphoepithelioma is a rare childhood malignant tumour, which, in the majority of case is diagnosed when the metastases (mainly to the lymph nodes) occur.
2. Sporadic incidence of lymphoepithelioma in children created difficulties in finding uniform methods of therapeutic management.
3. It seems that combined treatment (chemotherapy, surgery and radiotherapy) gives chances of long survival and recovery, even in advanced stages of the disease.

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