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Solid tumours of perimeningeal region in children – diagnostic and therapeutic difficulties

Solid tumours primarily localized within head and neck are a serious diagnostic and therapeutic problem. The head and neck site can be divided into three anatomical groups: orbital, nonorbital-nonperimeningeal and cranial perimeningeal. Cranial perimeningeal localization comprises: paranasal sinuses, nasopharynx, middle ear, mastoid process and preauricular-subtemporal region (11). The fact that tumours of the site proliferate in the regions of frequent infections may lead to postponing the proper diagnosis. That localization maintains the biggest therapeutic problem because a radical resection is almost impossible.

Solid tumours developing in the site are soft tissue sarcomas and lymphoepitheliomas. Soft tissue sarcomas originate from prime mesenchymal tissue (2). They account for 7-10 % of all paediatric neoplasms, 50–60 % of them represented by rhabdomyosarcoma (RMS), most frequently localized within the head and neck (35%) (2,7). The peak of incidence occurs between 2 and 5 and between 15 and 19 years of age (7). On the other hand, lymphoepithelioma originates from transitional epithelium of nasopharynx and represents less than 1% of malignant tumours in children, which accounts at the same time for one third of nasopharyngeal cavity tumours. (4) Occurrence of this type of tumour is associated with Epstein-Barr virus infection (6,10). Children between 15 and 18 years of age are affected by the disease.

MATERIAL AND METHODS

The study comprised 13 patients treated for solid tumours primarily localized within perimeningeal region in the Department of Paediatric Hematology and Oncology of Medical University of Lublin between 1992 and 2002. The age of patients ranged from 2 to 17 years (mean 9.5), 4 girls and 9 boys. In the study the character and duration of the symptoms prior to reaching the diagnosis were analyzed. The prognosis and results of treatment were evaluated.

RESULTS

The period from the first symptoms to reaching the diagnosis lasted from 3 to 5 months (mean 4). The symptoms reported by patients were as following: prolonging purulent rhinitis and unilateral occlusion of the nasal meatus – 6 patients, lymph nodes enlargement – 5 patients, swallowing disorders and lockjaw – 3 patients, dysopia, lacrimation – 3 patients, massive epistaxis – 2 patients, earache – 2 patients, dyspnoe – 1 patient, toothache – 1 patient.

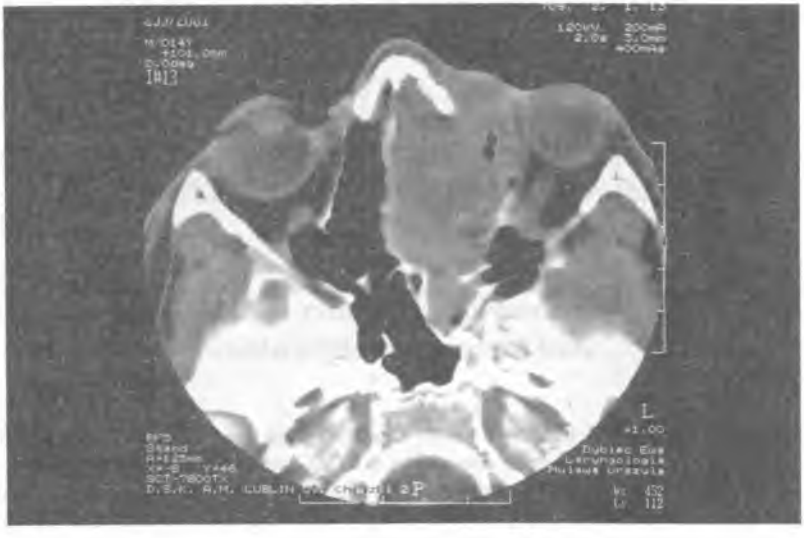

Fig. 1. CT scan of nasopharynx a 15-year-old boy with embryonal RMS before treatment

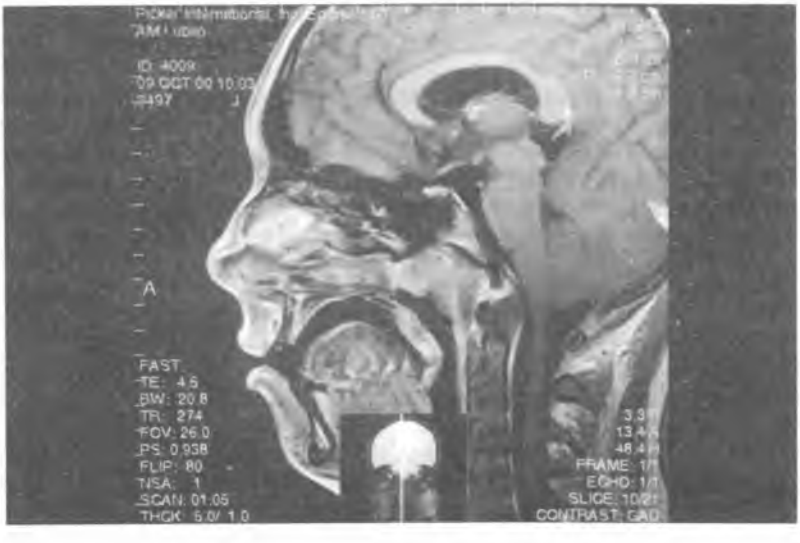

Fig. 2. MRI scan of nasopharynx a 13-year-old boy with lymphoepithelioma before treatment

The diagnosis was based on the result of histopatological examination of the sample of the tumour: embyonal rhabdomyosarcoma – 8 patients, alveolar rhabdomyosarcoma – 1 patient, leiomyosarcoma – 1 patient, lymphoepithelioma – 4 patients (Fig. 1,2). At the moment of making the diagnosis in 2 patients the central nervous system had been already affected, in 4 patients regional lymph nodes had been involved (Fig. 3). Patients with sarcomas were treated according to either IRS III or CWS-96 protocols. Both programmes include multidrug chemotherapy, radiation therapy and resection of the tumour. In the case of lymphoepitheliomas there in no standard management. The main therapeutic procedure is radiation therapy but there are also attempts at chemical treatment and surgical treatment (if required), whose purpose is to remove the residuals of the tumour. The results of treatment: full clinical and radiological remission – 7 patients (2 of them with lymphoepithelioma and 4 with soft tissue sarcomas); in the course of treatment – 1 patient with RMS, in the course of treatment of the first recurrence – 2 patients (1 with sarcoma and 1 with lymphoepithelioma, in both cases the recurrence was diagnosed after 3 months from the termination of treatment), died – 3 patients (1 with lymphoepithelioma and 2 with sarcomas).

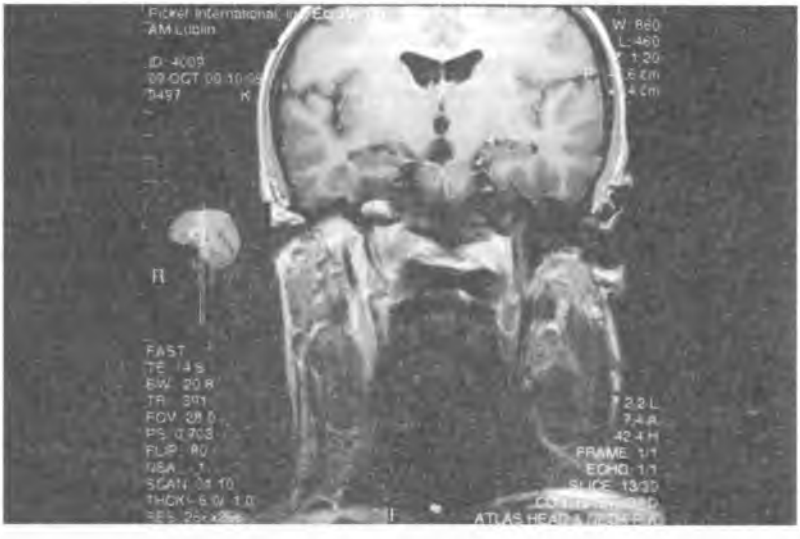

Fig. 3. MRI scan of neck shows metastases of lymph nodes in the patient with lymphoepithelioma

DISCUSSION

Solid tumours primarily localized in the perimeningeal region represent approximately 8 % of all malignancies in children. The very first symptoms are lymph nodes enlargement and/or upper airways infection. In most cases it becomes the reason for the commence of anti-inflammatory treatment by general practitioners. Lack of reaction for the treatment and deterioration of the patients' general state leads to the widening of the diagnostic procedures and reaching the proper diagnosis. Thus, children are admitted into specialized departments at an already advanced stage of the disease. Perimeningeal tumours may spread by continuity giving the metastases to the central nervous system. Distant metastases may involve the bones, lungs, bone marrow and lymph nodes (11). Thus, in the early diagnostics the following imaging

modalities should be performed: computer tomography/ nuclear magnetic resonancel scan of head and neck including the cranial basis, lumbar puncture, CT scan of the chest, ultrasound scan of abdomen, bone scintigraphy and bone marrow aspiration (8,11). Therefore, the treatment of such patients demands multidisciplinary management. It is important to avoid initial surgery because of its non-radical character and the possibility of mutilation of the patient (7). In the patients with soft tissue sarcomas the resection of residual tumour after the inductive chemotherapy should be applied (7,12). Local control may be achieved by simultaneous administering of chemotherapy and radiation therapy after the surgery (2,3). In children with lymphoepithelioma high dose radiation therapy (50-60 Gy depending on the age of the child and the size of the tumour) is crucial (9). Because of insufficient control of the disease process in the case of radiation therapy alone attempts of cytostatic treatment are undertaken (1,5,9). The role of surgery is limited obtaining adequate diagnostic material for histopathological examination and resection of residual tumour following completion of the therapy (13).

In three of the patients with soft tissue sarcomas initial, non-radical surgery was performed. In one child it was complicated by facial nerve interruption, in another patient the tumour capsule was broken during the surgery. In the remaining patients the inductive chemotherapy was applied. The response to treatment was satisfactory. In one child full remission was achieved and the operation was unnecessary. Further chemotherapy and radiation therapy were continued. In two patients the recurrence occurred in the course of postoperative treatment. In one patient cervical and abdominal lymph nodes metastases appeared within 3 months after therapy completion.

In one patient with lymphoepithelioma radiation therapy alone was applied because the patient had been treated 10 years before when there was no experience in chemical treatment. In this case full remission was attained. In the remaining three patients combined chemotherapy radiation therapy and surgery were administered. In one patient the progression of the disease occurred in the course of the treatment, in another patient cervical and mediastinal lymph nodes metastases appeared within 3 months after the completion of the therapy.

CONCLUSIONS

1. Solid tumours of the perimeningeal region cause initial diagnostic problems because of their early stage pseudo-inflammatory manifestation.

2. Because of initial diagnostic problems children admitted into specialized departments are already in the advanced stage of the disease, which creates serious therapeutic problems. Therefore, despite administering intense therapy, the results of treatment are still insufficient.

3. The therapy ought to be complex and it should include chemotherapy, radiation therapy and surgery. The initial operation should be avoided because it is non-radical and often mutilating.

4. General practitioners should pay their attention to the symptoms, which may suggest proliferative process of the perimeningeal region, and particularly to the need of the imaging modalities useful in reaching the correct diagnosis.

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SUMMARY

Thirteen patients, aged 2–17 years, were treated because of primary solid tumours of head and neck location at the Department of Children Hematology and Oncology in Lublin. The authors analyzed clinical symptoms before diagnosis and the duration of these symptoms as well as the kind of tumours. In all cases the tumour was diagnosed on histopathological examination: soft tissue sarcomas – 9 children, lymphoepithelioma – 4 ones. The prognosis and treatment were estimated.

Nowotwory lite okolicy okołooponowej u dzieci – trudności diagnostyczne i terapeutyczne

W Klinice Hematologii i Onkologii Dziecięcej w Lublinie leczono 13 pacjentów z guzami litymi pierwotnie zlokalizowanym w obrębie głowy i szyi. Były to dzieci w wieku od 2 do 17 lat. W pracy przeanalizowano charakter oraz czas trwania objawów klinicznych poprzedzających właściwą diagnozę. We wszystkich przypadkach rozpoznanie postawiono na podstawie wyniku badania histopatologicznego: mięsaki tkanek miękkich – 9 pacjentów, nabłoniak chłonny – 4 pacjentów. Oceniono rokowanie oraz wyniki zastosowanego leczenia.