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Primary chest tumours in children

Primary chest tumours in children are rare and appear in 3% of cases, usually 83% of which are malignant. Most common malignant tumours are: neuroblastomas, sarcomas, Hodgkin and non-Hodgkin lymphomas, germ cells tumours and malignant thymomas. Nonmalignant lesions in the chest include cysts, thymomas and teratomas. Early diagnosis of chest tumours is very difficult because of delayed symptoms and highly advanced stage. Symptoms of chest tumours depend on the tumour's mass, localization and progression. These tumours can cause cough, dyspnoea, Horner's syndrome and superior vena cava syndrome (1, 3, 5).

The purpose of this study was to analyze the kind of chest tumours, clinical symptoms before diagnosis, their duration and results of treatment.

MATERIAL AND METHODS

Between 1997-2002, 13 children (6 boys and 7 girls) with primary chest tumours, aged 3 months to 18 years (mean 112.5 months), were treated at the Department of Children Hematology and Oncology. The study included cases of solid tumours of chest and non-malignant lesions. The patients with chest metastases and lymphomas (due to different clinical course) were not analyzed.

RESULTS

The duration of initial symptoms ranged from 2 to 6 months (mean 4 months). Clinical symptoms presented by patients were as follows: • persistent fever – 5 patients, • cough, dyspnoea – 5 patients, • weight loss – 5 patients, • Horner's syndrome – 3 patients, • chest pain – 2 patients, • vomiting – 2 patients.

In one case the tumour was recognized during a routine chest x-ray examination and in another child the diagnosis was connected with chest trauma (Fig.1, 2). All diagnostic procedures were verified with histopathologic outcomes (Tab.1).



Fig.1. Chest X-ray examination of a 12-year-old girl with sa Ewing of ribs VI and VII before treatment



Fig.2. Chest CT scan of a 13-year-old boy with primitive neuroectodermal tumour of ribs I and II before treatment

Table 1. Histopathological results

Tumour	Number of cases
Neuroblastoma	3
Primitive neuroectodermal tumour	3
Sarcoma Ewing	2
Rhabdomyosarcoma	2
Carcinoma embryonale	1
Ganglioneuroma	1
Esophagus cyst	1

Treatment: total surgery was performed in the cases of nonmalignant lesions; early surgery – in 5 cases of malignant tumours, but it was not total; delayed operation – in the other patients. Chemotherapy inductive was given to 6 patients and all patients received chemotherapy postoperative, while 7 patients were subjected to radiotherapy. Results of treatment: all patients with nonmalignant lesions and 4 children with malignant tumours (rhabdomyosarcoma – 1 child, sa Ewing – 1 child, neuroblastoma – 3, primitive neuroectodermal tumour – 1) survived, 3 patients are undergoing treatment (sa Ewing – 1 child, neuroblastoma – 2 children) and 4 children died (PNET – 2 patients, RMS – 1 patient, ca embryonale – 1 patient) because of the disease progression.

DISCUSSION

The majority of tumours developing in the thoracic cavity are asymptomatic in the early stage of the disease. The initial clinical signs, e.g. prolonged fever, lack of appetite, cough, dyspnoea, suggest inflammations of the lower respiratory tract. In such cases, the general practitioners, having performed the physical examination, often diagnose pneumonia and prescribe antibiotics, even without radiographic examinations. The Horner's syndrome is frequently the first symptom of neuroblastoma located in the thoracic cavity (6). The children with this disease are usually directed to a neurologist. After several weeks or even months of ineffective treatment, the X-ray of chest are finally taken. This results in delayed diagnosis and progression of the disease. In patient with benign tumours and non-neoplastic lesions this does not affect the treatment results and remote prognosis. However, in patients with malignant tumours the delay contributes to the dissemination of the neoplastic disease. In 20% of such cases the metastases to the bones, bone marrow, spinal cord or abdominal cavity are detected on diagnosis. The initial diagnostic methods should involve: bone scintigraphy, US examination of the abdominal cavity, bone marrow puncture, lumbar puncture and nuclear MR of spinal cord (2,7). The methods of treatment for malignant tumours include combined chemotherapy, surgery and radiotherapy. In spite of such an intensive treatment, the therapeutic effects are unsatisfactory. In patients with benign tumours and non-neoplastic lesions the management is confined to the surgical removal of the lesion (2,6).

In the patients discussed above, the time from the onset of clinical signs and diagnosis was very long (2–6 months). In 5 patients with malignant tumours pneumonia had been diagnosed and only lack of any improvement following the treatment led to the chest X-ray examination. One of the children sustained the chest injury at school and then the parents noticed chest prominence at the level 6th and 7th rib. The general practitioner diagnosed post-traumatic

haematoma and prescribed an anti-inflammatory oitment. When the treatment results were poor and fever developed he administered an antibiotic. During the next 3 months the patient's condition deteriorated and she underwent a chest X-ray followed by computer tomography. The children with Horner's syndrome were consulted by neurologists several times; however no radiographic examinations were ordered. When the symptoms such as loss of weigh, weakness or fever developed, the diagnostic procedures were widened. The patients were admitted to the Department of Children Oncology with the III or IV stage of proliferative disease. Considering the above mentioned cases, it should be stressed that early routine X-ray examinations are necessary to provide accurate localization and evaluation of the extent of lesions.

CONCLUSIONS

1. The clinical signs of tumour-like lesions located in the thoracic cavity are non-characteristic, which is likely to delay proper diagnosis and lead to therapeutic failure.
2. The general practitioners should thoroughly analyze the cause of signs and symptoms, additionally considering those related to neoplastic dideases.
3. The X-ray examinations play a relevant role in the diagnosis of chest tumours.

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SUMMARY

Primary chest tumours in children are rare and appear in 3% of cases, 83% of which are malignant. Early diagnosis is very difficult because of delayed symptoms and highly advanced stage. Symptoms of chest tumours depend on the tumour mass, localization and time of progression. These tumours can cause cough, dyspnoea, Horner's syndrome and superior vena cava syndrome. The purpose of this study was to analyze the kind of chest tumours, clinical symptoms before diagnosis, their duration and results of treatment.

Pierwotne guzy klatki piersiowej u dzieci

Pierwotne guzy zlokalizowane w klatce piersiowej u dzieci są rzadkie i stanowią 3% przypadków, ale aż w 83% są guzami złośliwymi. Wczesne rozpoznanie jest bardzo trudne, gdyż objawy kliniczne pojawiają się późno i świadczą zwykle o zaawansowaniu choroby. Objawy guzów klatki piersiowej zależą od masy guza, lokalizacji oraz czasu postępu choroby. Do najczęstszych objawów klinicznych zaliczamy: kaszel, duszność, zespół Hornera i zespół żyły głównej górnej. W pracy przedstawiono analizę dzieci ze stwierdzonym guzem klatki piersiowej pod kątem objawów klinicznych poprzedzających rozpoznanie, czasu ich trwania, typu guza oraz wyników leczenia.