

CLINICAL AND TACTICAL APPROACHES IN THE DIAGNOSIS OF MALIGNANT TUMORS OF MAXILLOFACIAL AREA IN CHILDREN**TKACHENKO P.I., STARCHENKO I.I., BELOKON S.A.*, GURZHIY Y.V., STARCHENKO O.V.**

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*Received 15/09/2015; accepted for printing 24/03/2016***ABSTRACT**

Among 160 thousand people, who are initially diagnosed with malignant tumors annually in Ukraine, 1.1% is accounted for children. At the same time, unfortunately, isolated data as for the initial and metastatic lesions of maxillofacial area tissues in children are presented in the available modern literature. Currently, identification and treatment of oncostomatological diseases at the early stages of development are faced with considerable difficulties due to greater polymorphism of clinical manifestations of tumors, age peculiarities, dependence of diagnostics of malignant neoplasms from the dynamics of growth, localization, local and general changes. Therefore, the amount of medical errors in pediatric practice is higher than in adults.

The article presents data on the incidence and peculiarities of clinical manifestations of some malignant tumors in maxillofacial area in children.

The resulting data show that 96.6% of patients have been referred to the hospital following the 3-12 months after the first signs of the disease and attendance the medical facility to seek health care; however, in 100% of cases the diagnosis made by the referring medical facility was erroneous.

Malignant neoplasms have been more frequently diagnosed in the age group of 12-15 years (41.4%), and among the nosological forms the most frequent were lymphogranulomatosis (27.6%) and metastatic spread to lymph nodes of the neck (17.2%). During the stages of the diagnosis a tumor process has been detected in 66.5% and 89.7% of cases by the puncture and excisional biopsy, respectively. Informative value of the computer tomography and magnetic resonance imaging in terms of ascertaining the existence of pathological lesions of tissues was 100%.

Despite the comprehensive treatment, provided for the patients in specialized units, 34.5% of sick children died during the first year from the date of making the clinical diagnosis, though cases with 5-year and longer survivability also occurred.

Thus, in clinical practice there is an increasing need in conducting a complex of the state-of-the-art additional methods of examination in children with neoplastic processes, which are not always available for most practical primary health care institutions to date. Physicians of the related specialties should be involved for medical advice in occurrence of the earliest suspicion on malignant growth manifestations. At the same time one should remember that some malignant tumors in children are of embryonic origin, being dysontogenetic, and can be clinically manifested immediately after birth.

KEYWORDS: *children, malignant tumors, maxillofacial area.***INTRODUCTION**

Absence of visible symptoms and “blurring” of clinical manifestations of malignancies at the initial stages of development, late visits to a doctor accompanied by insufficient awareness among

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health care workers about this branch of medicine and lack of oncologic alarm lead to the fact that much of patients come for treatment with advanced forms [Fedyayev I et al., 2000; Vorobyov Yu, Garbuzov M, 2000].

The analysis of the constituents of such disappointing situation shows that in 55% of cases the reason for the late visit to specialized medical facilities is organizational-methodological draw-

backs: incomplete examination of patients, clinical-diagnostic errors (underestimation of clinical data, radiological changes, erroneous interpretation of the results of additional methods of examination), the lack of targeted prevention or obscure determination of sequence of its planning, inadequate preventive examinations, patients' delay at the stages of therapy network [Bernadskiy Yu, 2000; Sokolova N, 2008 a,b]. And, consequently, about 40% of patients have extensive-stage malignant processes in the maxillofacial area and neck due to the medical error, despite the fact that the majority of neoplasms are easy to be visually examined and used for additional methods of study [Fedyayev I et al., 2000].

In pediatric practice, situation is not better: among 160 thousand people, who are initially diagnosed with malignant tumors annually in Ukraine, 1.1% is accounted for children. At the same time, unfortunately, isolated data as for the initial and metastatic lesions of maxillofacial area tissues in children are presented in the available modern literature. Currently, identification and treatment of oncostomatological diseases at the early stages of development are faced with considerable difficulties due to greater polymorphism of clinical manifestations of tumors, age peculiarities [Bernadskiy Yu, 2000; Fedyayev I et al., 2000], dependence of diagnostics of malignant neoplasms from the dynamics of growth, localization, local and general changes [Topolnitskiy O, 2016]. Therefore, the amount of medical errors in pediatric practice is higher than in adults.

This problem is of general medical significance considering the fact that due to topographic and anatomical features of head and neck structures any lesion in maxillofacial area is basically the subject of the diagnosis and treatment, provided by medical professionals of various specialties (dentists, maxillofacial surgeons, otolaryngologists, ophthalmologists, dermatologists, neurologists, etc.) [Bernadskiy Yu, 2000; Topolnitskiy O, 2016].

Previous publications were devoted to the issue of difficulties in identification of malignant tumors in children, having concluded that in their diagnosis one should pay great attention to the detailed collection of complaints, clarification of the history of the disease, do thorough examination, use additional methods of examination more extensive [Tkachenko P et al., 2006; 2011; 2015].

This study presents the accumulated information about the incidence and characteristics of clinical manifestations of some malignant tumors of maxillofacial area in children to a wide medical community.

MATERIAL AND METHODS

The structure of the malignant tumors of maxillofacial area both of primary lesion and metastatic nature was analyzed in 29 children, who were under the observation at the clinic of the Department of Children's Oral Surgery over 15 years (2000-2015). The age of patients ranged from 2 months to 15 years. General clinical methods, computer tomography and MRI have been applied during their examination. In all cases, puncture and excisional biopsy was made by indications, followed up with verification of the biopsy material by the staff of the Department of Pathologic Anatomy with autopsy course, using the standard methods of histological studies [Merkulov A, 1969].

RESULTS AND DISCUSSION

The resulting data showed that out of 29 patients 28 (96.6%) were referred to the clinic following 3-12 months after occurrence of the first signs of disease and attendance to the hospital for medical aid, and in 100% of cases the diagnosis made by the referring medical facility was erroneous and for the majority of cases it was made already at the III-IV stages of diseases' progression.

The table presents the structure of incidence depending on age in children with malignant tumors of maxillofacial area, showing that malignant neoplasms have been more frequently diagnosed in the

Table

The incidence of malignant neoplasms of maxillofacial area in children depending on age

Age groups	Number of observations n (%)
Infant (1 month-1 year)	2 (6.9%)
Babyhood (1-3 years)	2 (6.9%)
Preschool (3-7 years)	6 (20.7%)
Junior school (7-12 years)	7 (24.1%)
Secondary school (12-15 years)	12 (41.4%)



FIGURE 1. Appearance of patient K., 7 years old. Diagnosis: malignant median face granuloma: a – general appearance, b – tomography of bones of the facial skeleton and neurocranium



FIGURE 2. Appearance of patient K., 10 years old. Diagnosis: lymphoblastic lymphoma of submandibular area on the right: a – wound appearance on day 10 after surgical treatment of the “acute purulent lymphadenitis” and before extended biopsy, b – biopsy material, c – sutured wound, d – ultrasonic image of tissues of the upper third of lateral section of neck on the right: two hypoechoic swellings of heterogeneous structure with sharp contours, polygonal shape, size 4.5x2.5 and 2.5x0.8 cm are detected

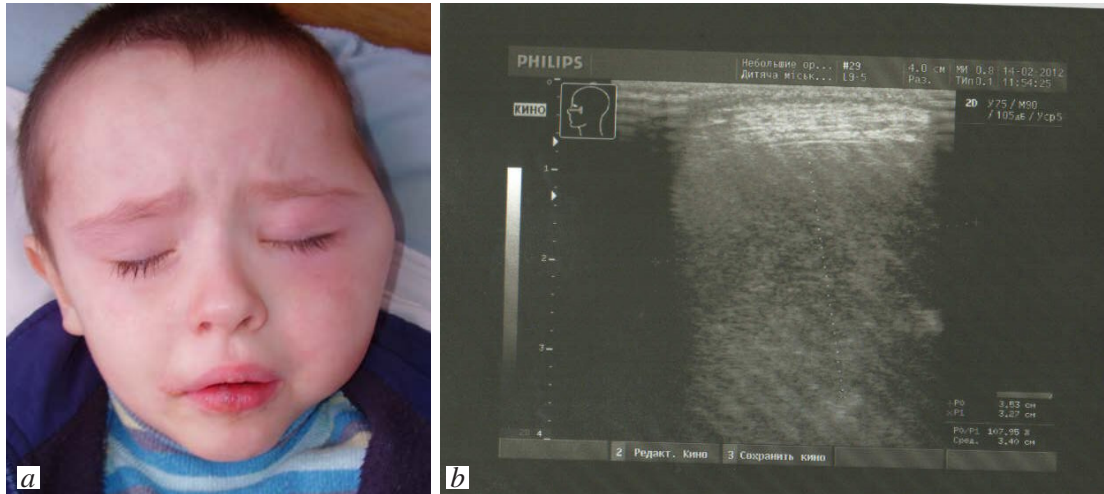


FIGURE 3. Appearance of patient S., 2.5 years old. Diagnosis: rhabdomyosarcoma of parameningeal localization with invasion to maxillofacial area tissues (a). Ultrasonography (b) identified heterogeneity of structure of left cheek tissues with areas of hyper- and hypoechogenicity



FIGURE 4. Appearance of patient O., 8 years old. Diagnosis: rhabdomyosarcoma of parameningeal localization with invasion to peripharyngeal space, submandibular area and retromandibular fossa



FIGURE 5. View of the fragment of lower jaw in patient A., 14 years old. Diagnosis: lytic form of osteosarcoma of lower jaw on the right. Socket did not close up during 2 months after 46 tooth extraction

age group of 12-15 years (12 cases (41.4%)).

Among nosological forms of malignant neoplasms the most frequent were lymphogranulomatosis (27.6%) and metastatic spread to lymph nodes of the neck (17.2%). Malignant histiocytosis (Fig. 1) and lymphoblastoma (Fig. 2) were diagnosed with similar incidence (13.8%). Rhabdomyosarcoma (Fig. 3, 4) and Ewing's sarcoma accounted for 6.9% each, and other primary (Fig. 5) or metastatic lesions of jaws in localization of lymphosarcoma in the abdominal cavity and adenocarcinoma of submandibular gland – 13.8% of cases. The distribution of nosological forms of malignant neoplasms is presented in figure 6.

During the stages of the diagnosis atypical cells have been detected by the puncture in 66.5%, and in 89.7% of cases morphological verification of the di-

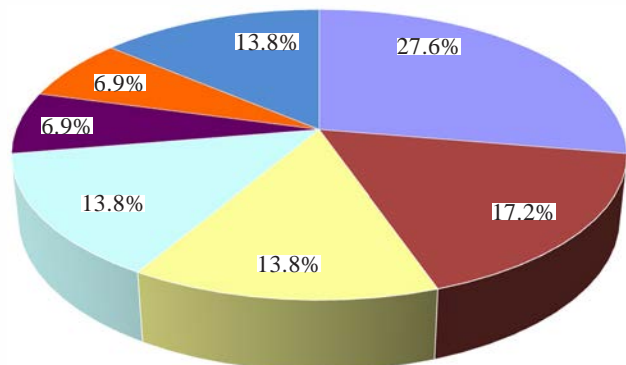


FIGURE 6. Distribution of nosological forms of malignant neoplasms of maxillofacial area in children
 NOTES: lymphogranulomatosis (blue), metastatic spread to lymph nodes of the neck (red), malignant histiocytosis (yellow), lymphoblastoma (cyan), rhabdomyosarcoma (purple), ewing's sarcoma (orange), other lesions (dark blue)



FIGURE 7. Appearance of patient L., 22 years old, who was diagnosed with osteosarcoma of lower jaw on the right at the age of 12: a – the view of neck area after surgery, b – necrosis of hard tooth tissues on the upper jaw, developed as a result of radiation therapy

agnosis was made after excisional biopsy. In 10.3% of patients with suspected metastatic lesion of lymph nodes of the lateral surface of the neck puncture failed to obtain the necessary data on cellular atypia, and the diagnosis was confirmed after their extirpation or direct excision of tumor itself. Informative value of the computer tomography and magnetic resonance imaging in terms of ascertaining the existence of pathological lesions of tissues was 100%.

Unfortunately, despite the comprehensive treatment, provided for the patients in specialized units, 34,5% of sick children died during the first year from the date of making the clinical diagnosis, though cases with 5-year and longer survivability also occurred (Fig. 7).

Below is an example from own clinical practice to illustrate the presented material and clarify the

perception of its content.

The parents of a newborn child contacted the clinic of the Department of Children's Oral Surgery complaining of the presence on child's tongue a swelling, identified immediately after birth. It did not cause any anxiety for the baby; she has been referred for medical advice by the district pediatrician.

Examination showed symmetrical face, coloring of the baby's skin, and visible mucous membranes were natural. Skin turgor was not changed, mouth was opened freely.

On the dorsal surface of the tongue, closer to its root a localized spheroid broad-based swelling, sized 3×4 cm has been visualized; surface and color of mucosa were similar to adjacent tissues. The swelling was of solid- elastic consistency and moved together with the mucous membrane (Fig. 8 a).

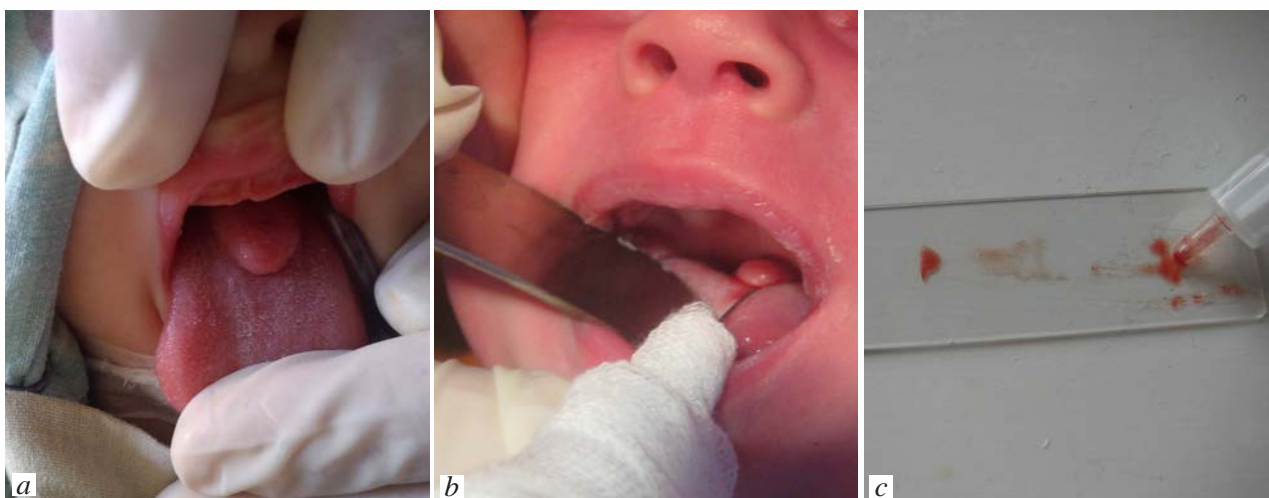


FIGURE 8. Appearance of the newborn child B., 2 months, with neoplasm on the dorsal surface of the tongue (a), performance of puncture of the swelling (b) and view of the obtained material for cytologic screening (c)



FIGURE 9. Stages of surgical treatment of the newborn child B., 2 months, with neoplasm on the dorsal surface of the tongue

Cytologic screening of puncture material allowed detecting only epithelial cells and red blood cells in the punctate (Fig. 8 b, c).

The preliminary diagnosis was rhabdomyoma of the dorsal surface of the tongue. The child was hospitalized for elective surgery and after extensive preoperative examination, involving medical professionals of the allied specialties, the tumor was excised under intubation narcosis within the healthy tissues; the wound was sutured tightly (Fig. 9).

Macroscopically, neoplastic structure resembled muscle tissue both externally and internally. The swelling has not been clearly pronounced as a capsule, but was somewhat more consistent than the tongue that allowed clear identifying of its boundaries. It should be noted that in the area of blind hole the tumor had dense taenia directed deep into the root of tongue, causing the necessity to deepen the wound during its excision.

The course of postoperative period was without complications. Postoperative wound was antiseptitized locally, and antibacterial therapy (medications per kg/body weight) was provided within the framework of the general drug therapy to prevent wound abscess. The suture was removed on 10th day; the wound healed by primary tension.

Surgical material was sent to histological study, the results of which were somewhat unexpected: the swelling, covered with unmodified mucous membrane without clear boundaries grew into the muscles of the tongue. Tumoral tissue was presented with numerous orbicular, oval or polygonal

cells with moderately dense nuclei and relatively large nucleoli. Cellular elements were located closely to each other, forming the solid fields in some areas. Against the background of the cells, described above, cellular elements with pronounced polymorphism, as well as few prolate, spindle cells were detected. The whole cell composition was localized in edematic fibrous stroma with numerous blood microvessels (Fig. 10). Such microscopic picture was consistent with morphological structure of rhabdomyosarcoma [Paltsev M, Anichkov N, 2005].

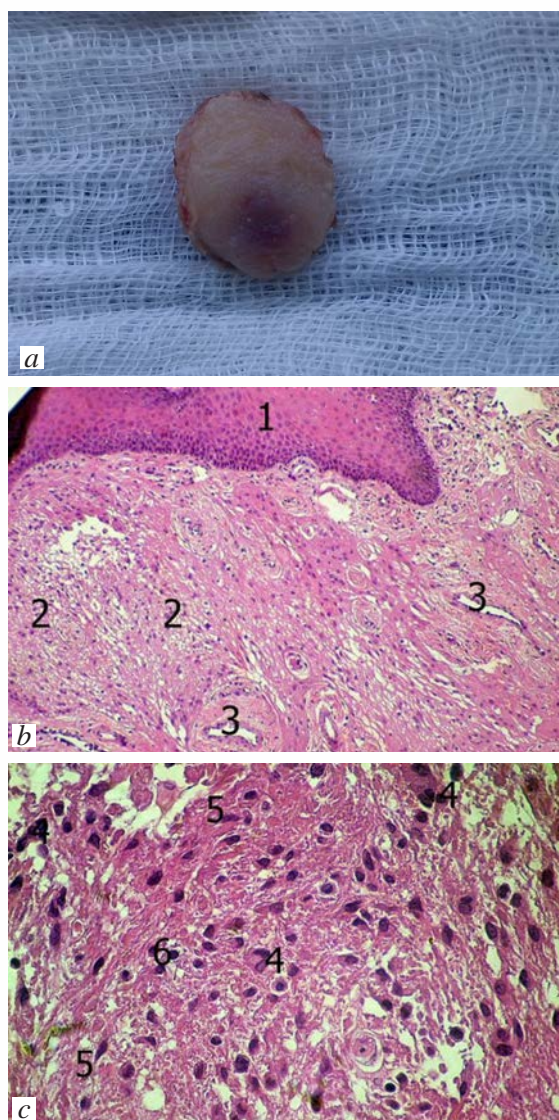


FIGURE 10. Macroscopic (a) and microscopic structure (b, c) of neoplasm of the dorsal surface of the tongue in a newborn child B. Hematoxylin and eosin stain: b – lens 10^x, oc. 10^x, c – lens 40^x, oc. 10^x

NOTES: 1 – Unchanged covering epithelium, 2 – tumor tissue, 3 – blood vessels, 4 – cellular elements with the polymorphism phenomena, 5 – prolate cells, 6 – the most common orbicular cellular elements

On the basis of clinical data and findings of morphological study of the postoperative material, the final clinical diagnosis was rhabdomyosarcoma of the dorsal surface of the tongue.

Once the wound healed the child was referred to the Cancer Institute (Kiev) for the immunohistochemical study of postoperative material to confirm the diagnosis and determine the type and volume of further treatment.

Thus, the peak incidence of malignant neoplasms of maxillofacial area in children occurs at the age of 12-15 years, and lymph nodes are affected most frequently (both initially and metastatically). In clinical practice there is an increasing need in conducting a complex of the state-of-the-art, more informative additional methods of examination of children

with neoplastic processes, which, unfortunately, are not always available for most practical primary health care institutions to date.

Biopsy material sampling for preliminary verification of tumors should be used extensively in occurrence of the earliest suspicion on malignant growth manifestations in the least suspicion on the occurrence of manifestations of the malignant growth, and physicians of the related specialties should be involved for medical advice, that allows making the most rational decision in each individual case.

It should be remembered that some malignant tumors in children are of embryonic origin, being dysontogenetic, and could be clinically manifested immediately after birth, especially in localization on root of the tongue area.

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