Pontocerebelos angle tumors: diagnostic and tactics of surgical treatment

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Abstract

Tumorile unghiului pontocerebelos: diagnosticul și tactica de tratament chirurgical

Autorii prezintă un caz clinic a unui copil cu tumoră cerebrală în care sunt elucidate particularitățile evoluției clinice și unele aspecte de diagnostic imagistic și tratament, rezultatele obținute fiind confruntate cu datele literaturii. În lucrare se menționează că în cazul tumorilor cerebrale la copii, managementul este bazat exclusiv pe lucrul în echipă în care diferiți membri trebuie să fie familiarizați în amănunte cu toate aspectele patologiei date, precum și prezența unei experiențe de lucru, impunându-se necesitatea unui studiu continuu a problemei abordate. Autorii conchid că recurența tumorilor cerebrale la copii este determinată de rezecția incompletă, subtipul histologic fiind un factor important.

Cuvinte cheie: tumori cerebrale, tumoră pontocerebelară, meningioma, tratament chirurgical, copii

Abstract

The authors present a clinical case of a child with brain tumor in which are related the particularities of the clinical evolution and some aspects of imaging diagnosis and treatment, the results being confronted with literature data. The paper mentions that in the case of brain tumors in children, management is based exclusively on teamwork in which different members need to be familiar with all aspects of the pathology as well as the presence of a work experience, which implies the need for a continuous study of a problem addressed. The authors conclude that the recurrence of childhood brain tumors is due to incomplete resection, the histological subtype being an important factor.

Keywords: brain tumor, cerebello-pontine angle tumor, meningioma, surgical treatment, children

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Introduction

Children brain tumors of the central nervous system (CNS) constitute (2,5 – 3,5 per 100 000 children annually), and therefore contribute to a major part of daily practice in pediatric oncology [3]. Brain tumors are very heterogeneous with regard to tissue, location, pattern of spread, clinical, and age of occurrence from the neonatal period to the adolescence. These tumors also vary in their surgical outcome and carry a different risk of post-operative complications.

At the moment remains the descriptive classification based on the histopathological examination the basic element for the adequate management of child brain tumors.While the localization and cell differentiation is the basis of the diagnostic system,remained the tumor classification "histogenetic. "The last edition

of the "WHO Classification of Tumors of the Nervous System" presents 127 entities, reflecting the wide variety of CNS cellular constituents. Theoretically, all these types of tumors can develop at the child. The number of histological types of brain tumors with particular importance at the child is however significantly lower (table 1) [7]. The majority of pediatric CNS tumors can be classified into five types: meduloblastoma, pilocytic astrocytoma, diffuse astrocytoma, ependymoma and craniopharyngioma.CNS tumors occurring in childhood have important characteristics that differ from those of tumors with other localizations and which deeply influence their behavior:- high degree of invasiveness, even when the histological degree of malignancy is low - heterogeneous structure with mixed tumor areas of different malignancy degree - heterogeneous structure with mixed tumor areas of different malignancy degree - disseminate frequently on the CSF way. Irrespective of the degree of malignancy - tumors can turn, moving from a low degree of malignancy to a high degree.Compared to adult tumors prevails the localization at the posterior brain fossa level: almost half of the pediatric tumors are located at this level [7].

Clinical case:

The 9 years-old patient is hospitalized in the department of neurology with pains of increased intensity in the right hemiface,sensation of benumbness in this part, repeated vomits,double dvisual field.

The detailed anamnesis reveals the appearance of the first clinical signs for about a month.It follows medical treatment for trigeminal neuralgia with transient effect.

The neurological status reveals facial nerve paresis (House-Brackmann III-IV) and abducens on the right part.Trigeminal neuralgia on the right part. Swallowing disorders. Coordination tests with bilateral dysmetria.Unstable in the Romberg test. Pronounced hypotonic in limbs. Taking into consideration the medical discipline and the ineffectiveness of the treatment it is decided to be investigated by cerebral CT (fig.1), by what was detected an expansive process in the region of the pontocerebelos angle on the right part with a mass effect on the cerebral trunk.

Table 1. Brain tumors of special importance for children: the most frequent locations, the according to WHO malignancy grade, and the approximate percentages that they represent reported to all brain tumors at children (modified after Pollack) [7]

Type of tumor	The most frequent locations	Percent of all brain tumors	WHO malignancy grade
Pilocytic astrocytoma	Cerebellum, Hypothalamus, Optic pathways	12-18% 4-8%	
Anaplastic astrocytoma, glioblastoma	Cerebral Hemispheres Brain stem	6-12% 3-9%	III-IV
Pleomorphic xanthoastrocytoma	Brain stem Superficial cerebral Hemispheres	3-6%	II
Oligodendroglioma, mixed glioma	Cerebral Hemispheres anaplastic oligodendroglioma	2-7%	II, III
Ependimoma, anaplastic ependimoma	Lateral and third ventricle Fourth ventricle	2-5% 4-8%	II, III
Choroid plexus papilloma carcinoma	Lateral and Fourth ventricle	2-4%	I, IV
Neuroblastoma	Cerebral Hemispheres		IV
Medulloblastoma	Cerebellum	20-25%	IV
Other primitive neuroectodermal tumors(PNET)	Whole neuroaxis	1-5%	IV
Atypical teratoid/rhabdoid tumor	Infra and supratentorial		IV
Germ cell tumors	Pineal region Hypothalamus	0,5-2 %	I-IV
Craniopharyngioma	(supra) selar	6-9%	I

For the confirmation of the diagnosis, for the discussion of a differential diagnosis as well as for the appreciation of the surgical treatment tactic the patient was investigated by Contrast cerebral MRI-3Tesla.

During the examination in the cerebral MRI is confirmed the presence of a tumor mass located in the posterior cerebral pore, namely in the pontocerebelos right angle with a diameter of about 5 cm,with a moderate capture of the contrast substance and a dislocation of the anatomical structures from the average line. The relatively diffuse contour, T2-hypertense regime (fig. 2). There is a secondary obstructive hydrocephalus expressed by the marked periventricular edema and the obstruction of the adjacent to the tumor licvorian pathways (fig. 3).

The detailed analysis of the clinical and paraclinical data has determinated the establishment of an expansive intracranial process with a pontocerebelous angle. Secondary obstructive hydrocephalus. According to the national and international protocols it was decided to carry out the surgical intervention that had the aim of the tumor ablation, the decompression of cerebral structures, of cranial nerves, of circulatory pathways of the cephalorachidian fluid as well as for histological analysis of the tumor that would determine the subsequent tactic of treatment.

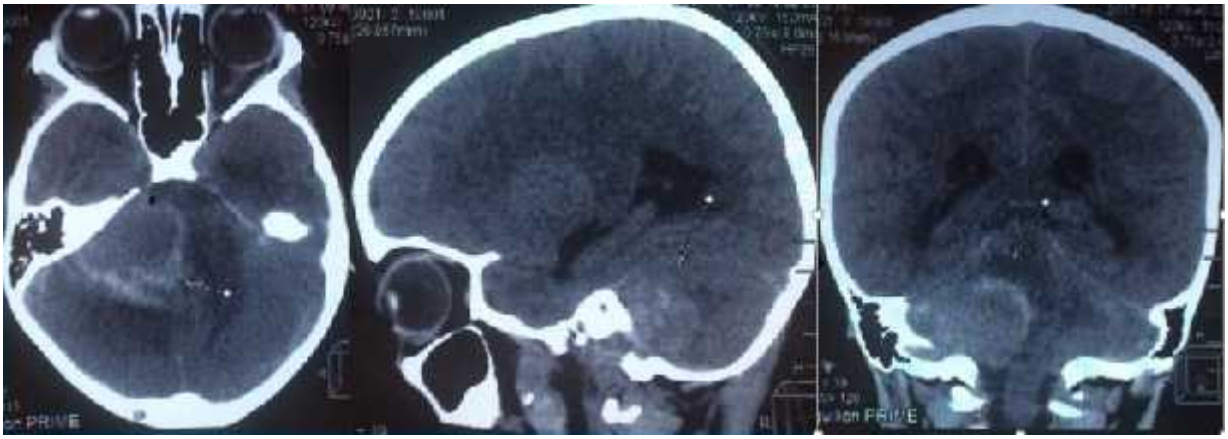

Fig. 1. Pacient L., 9 years. Preoperative CT scan. Explication in text

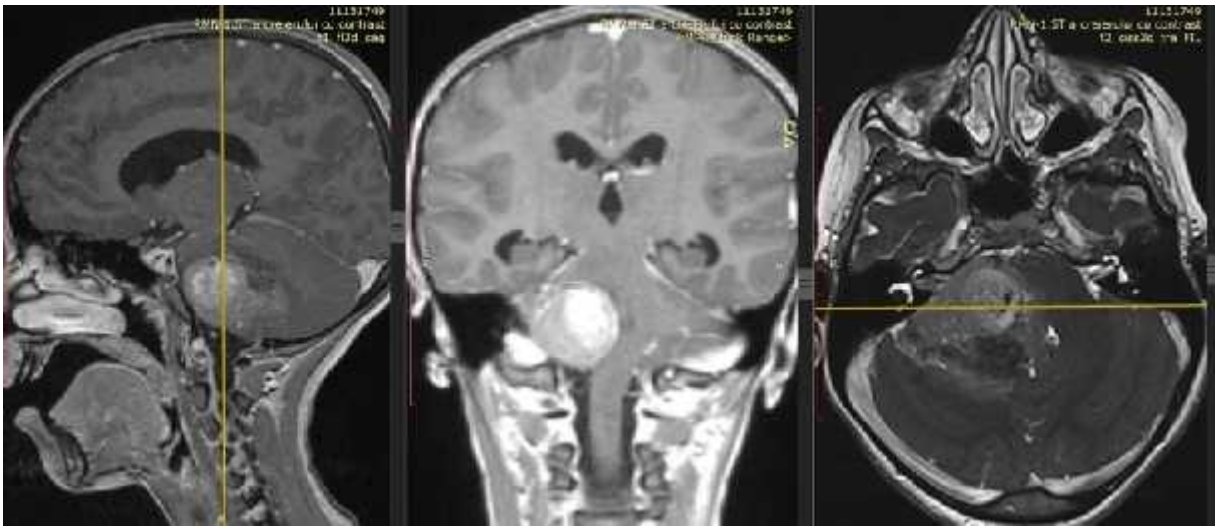

Fig. 2. Pacient L., 9 years. Preoperative MRI: UPC tumor

After a preoperative preparation, the patient was underwent surgery.Surgery with patient positioning in Park bench (patient in ¾ oblique position or almost ventral decubitus, with a roll under the shoulder and the head almost horizontal). Intraoperative the tumor grey-gray color, consecvent tough, weakly vascularized. It was performed the total tumor excision through the retro sigmoid (laterally suboccipital) approach, what was confirmed by the cerebral control CT scan on the second day after the surgery (fig. 4).

It is observed postoperatively a positive dynamic given by the regression of intracranial hypertension syndrome, of trigeminal pain, and the regression of facial nerve paresis (House-Brackmann II).

The histopathological examination denotes the presence of a meningioma meningoteliomatos WHO grade I-II.It was performed the pediatric oncologist consultation with repeated histological examination – it is confirmed the diagnosis and the subsequent treatment tactics that do not require adjunctive therapy.

Two months postoperatively the patient addresses at the emergency department with pronounced headache and repeated vomits that continue for several days. Neurological status aggravated by facial nerve paralysis (House Brackmann IV), abduction nerve paralysis on the right side, bulbar disorders, investigation by cerebral contrast MRI-relapse of expansive process with pontocerebelous angle on the right side with clivus and sellar region invasion (fig. 5).

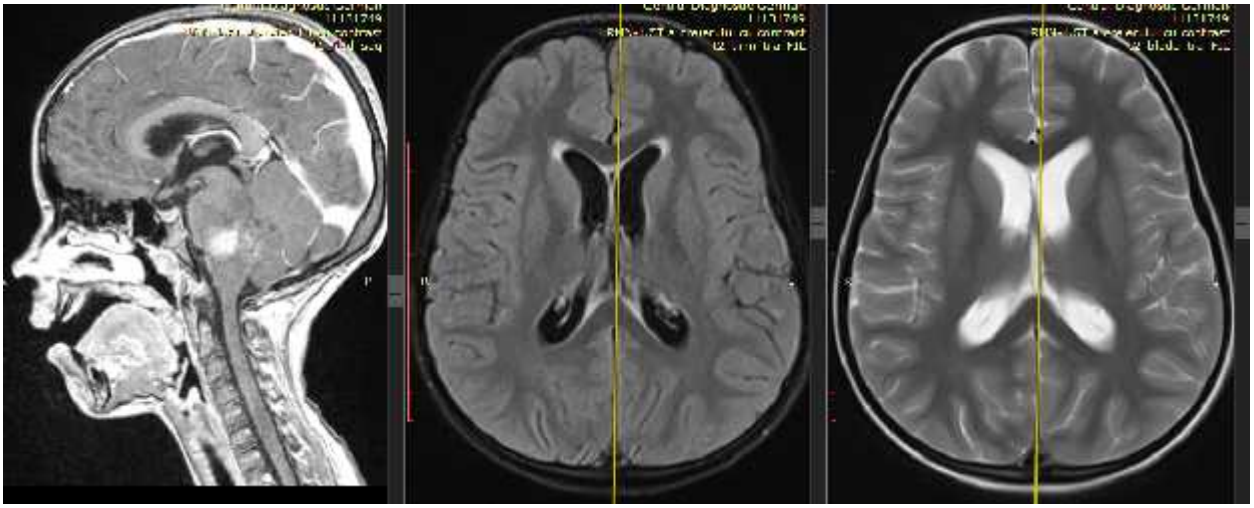

Fig. 3. Pacient L., 9 years. Preoperative MRI: periventricular edema and the obstruction of the adjacent to the tumor licvorian pathways

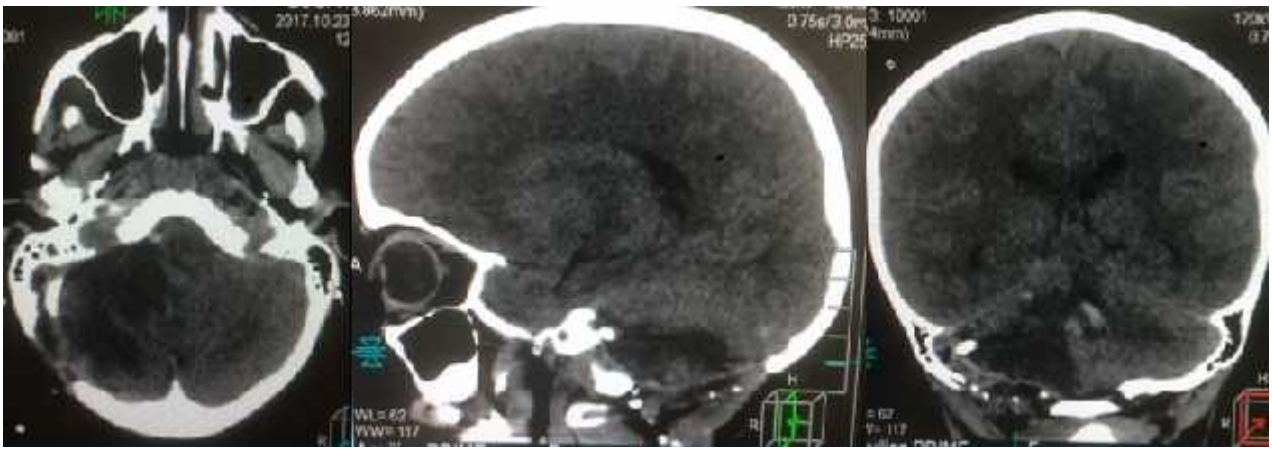

Fig. 4. Pacient L., 9 years. Postoperative CT scan: total tumor excision.

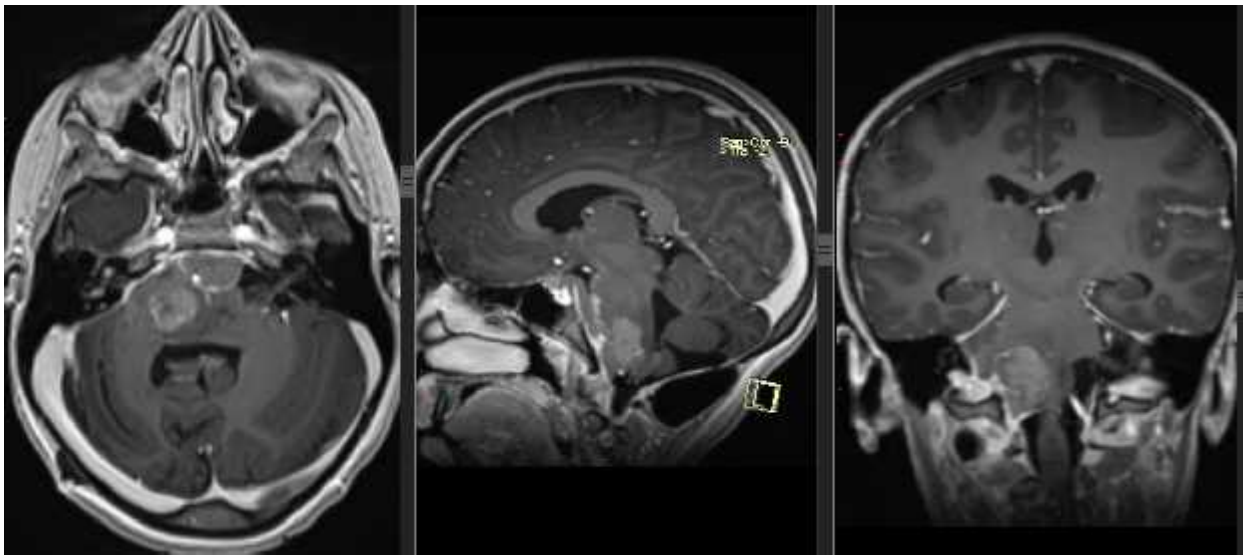

Fig. 5. Pacient L., 9 years. Postoperative MRI scan. Explication in text

Discutions

Meningiomas are uncommon neoplasms in the pediatric age group and differ in various clinical and biological aspects from meningiomas at the adult population [1].

The surgical treatment of posterior pit tumor at children, namely with pontocerebelos angle, has still great difficulty in diagnosis appreciation, performing the surgical step and the histopathological diagnosisas soon as possible.The amount of residual tumor tissue after surgery remains anyway a determining factor for the relapse rate [4, 6].Radical surgery is not possible in most cases due to tumor diffuse boundaries.Extensive tumor resection is useful for thereduction of tumor volume in limited spaces, life quality improving, remission duration increasing and is contributing substantially to the increasing of life expectancy [8]

About 90% of the meningiomas are slow-growing tumors (low-grade tumors), but even they are considered "biologically malignant" because the surgical treatment is often impossible due to localization [5]. Different series in the literature have shown a recurrence rate of approximately 13% [2].

Conclusions

Recurrence seems to be strictly related to incomplete resection and/or histologic subtype of the meningioma. Atypical, aggressive show a higher rate of recurrence and higher incidence of skull base location. Furthermore, there seem to be higher proportion of these tumors in pediatric age with WHO grades II and III.

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