



On October 23, 2021, Brazilian Society of Pediatric Neurosurgery had the pleasure of holding the **SBNPed GrandRound 2021**. Our Society took the decision to hold an unprecedented event focusing on the presentation of clinical cases. To this end, we selected 15 clinical cases that were presented, most of them by undergraduate students or neurosurgery residents.

ULTRA-DELAYED COMPLICATIONS OF AN IMPERCEPTIBLE SPINAL DORSAL DERMAL SINUS TRACT, MULTIMODAL MANAGEMENT.

Juan Miguel Alemán-Iñiguez, Christian Valencia Padilla

Background: The spinal dermal sinus tract is not exclusive to the gastrulation, since it accumulates both neurulations errors, being a complex malformation and associated with other dysraphisms.

Case Report: The following is a 1 year and 1 month old, male patient from Ecuadorian Amazon, he presents fever and vomiting 4 months earlier and he was admitted with communicating hydrocephalus; a dorsal paraspinal pore of 2x1 mm without production was identified in the skin, the image showed: pachymeningitis, holoespinal abscessesempyemas and an abnormal tract at T6, without apparent lumen; the management was characterized by surgical tract exclusion and proximal pyogenic collections evacuation; caudally lesions were drained with radiointervention guide; a ventriculoperitoneal CSF shunt and long-term antibiotic were indicated.

Discussion: Complications can be: early such as acute meningitis, late like recurrent meningitis or pachymeningitis, ultra-delayed: communicating hydrocephalus, and holoespinal empyemas and abscesses. The discussion points were: ultrasound as a low-cost and easy-to-access screening (sensitivity: 20.0%–100%, specificity: 85.7%-100%), especially in difficult access areas; regarding the management, the dermal sinus tract removal technique is known, however the controversy stemmed from holoespinal abscesses and empyemas, the evacuation with large laminotomies can result in spinal deformities, colletions drainage and irrigation with lavage through a spinal cateter

can be less invasive; the same procedure with tomography or fluoroscopy percutaneous guide can reach the remote lesions.

Key-words: trato espinhal, complicações, infecção.

RESECTION OF A LEFT INSULO-OPERCULAR CAVERNOMA IN A 4-YEAR-OLD CHILD: A CASE REPORT

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Introduction/Background: Cavernomas represent 1.7 to 18% of all vascular malformations in children and 25% are observed in under 18 years of age. The mean age varies from 9.1 to 10.2 years. There is no predominance between gender. In children, cavernoma bleeding is a common manifestation with an incidence varying from 27.3 to 78% versus 8 to 37% in adult patients.

Case Report: RGBB, female, 4-year-old presented a recent onset of increasing headache associated with generalized tonic-clonic seizure. A full neurological examination was performed with no abnomality. Brain Magnetic resonance imaging (MRI) showed a large cavernoma with recent hemorrhage on the inferior suface of frontal operculum and posterior compartment of the left insula. A frontotemporal craniotomy with frontal expansion was performed, followed by transylvian approach. This route required wide splitting of the Sylvian fissure in order to expose the insular cortex and the inferior surface of the frontal operculum. Guided by neuronavigation, a corticotomy was performed in the inferior portion of the frontal operculum, allowing access and complete removal of this lesion without sequelae.







Discussion: Cavernoma's surgical approach aims its radical resection to eliminate the risk of rebleeding. Lateral lesions close to the central core can be removed by transylvian approach. It is mandatory to preserve the lenticulostriate arteries and to know the relationship between the lesion and the internal capsule compartments. Retractors should be avoided. In these deep cavernomas, resection is limited to the cavernoma itself. The surrounding brain infiltrated by hemosiderin must be kept intact.

Key-words: cavernoma in childhood, deep brain cavernoma, insulo-opercular cavernoma

RESECTION OF A SUBEPENDYMAL GIANT CELL ASTROCYTOMA IN A CHILD WITH TUBEROUS SCLEROSIS COMPLEX: A CASE REPORT

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Introduction/Background: 5% to 20% of tuberous sclerosis complex (TSC) patients develop subependymal giant cell astrocytomas (SEGAs), which arise from the subependymal nodules. There is a male predominance with mean age at surgery of 11 years. SEGAs typically arise from subependymal nodules in the area of the foramen of Monro, and can be unilateral or bilateral. They typically project into the ventricle and can produce acute or chronic hydrocephalus.

Case Report: MSS, male, 11-year-old presents mutism perceived in early childhood, around age 2. Since then, he has been followed by pediatrician and geneticist who diagnosed him with TSC. Moreover, the patient presented with a neuropsychomotor developmental delay. In 2019, he presented somnolence, a bilateral abducens nerve palsy and frontotemporal headache. Brain CT and MRI scan showed a tumor in the frontal horn of the right lateral ventricle and hydrocephalus. Ventriculoperitoneal shunt (VPS) and surgical management of the tumor was performed. Anterior interhemispheric approach was performed to tumor resection. Histopathological examination confirmed SEGA and the patient was followed up by pediatric oncologist and pediatric neurosurgeon, with no change in the neurological status.

Discussion: The lack of large population data on the natural history of SEGA means that there is no clear guidance for type and timing of intervention. Current international recommendations propose the use of surgical resection for acutely symptomatic SEGAs. For growing but asymptomatic SEGA, both surgical resection and mTOR inhibitors are potential treatments.

Key-words: Subependymal giant cell astrocytomas, tuberous sclerosis complex, hydrocephalus

LUMBOSACRAL TUMOR IN A PEDIATRIC PATIENT

Camile Cremonese Gobbo, André Bedin, Jorge Wladimir Junqueira Bizzi, Gabriel Frizon Greggianin, Samir Cezimbra dos Santos

Introduction/Background: Chordoma is a rare pathology in the pediatric age group, especially when located in the lumbosacral region. Despite the local infiltrative character, some cases of metastasis are reported. The prognosis of these patients mainly depends on early diagnosis and extensive resection of the tumor.

Case Report: 13 y-o girl transferred with a huge mass lesion in the lumbosacral area. In the primary hospital she was submitted to a biopsy, whose anatomopathological report showed chordoma. The patient started with pain in the lumbosacral region a year ago, which recently evolved to a significant bulging in the area, pain in the posterior region of the left lower limb, urinary retention and difficulty in walking. Surgical approach, both anterior and posterior, was performed with block resection of the tumor. During the procedures there was evidence of metastatic dissemination to ovary and peritoneum. The pathological examination confirmed the diagnosis of conventional chordoma. Due to the late recognition of the condition and the fast evolution of the disease, there was not enough time to perform adjuvant radiotherapy and the patient died.

Discussion: From notochordal remnants, chordoma manifests in less than 5% of patients in the first two decades of life and is rarely found in the lumbosacral region of this patients. These patients open up with manifestation of local pain and, when not diagnosed early, progress with bulging of the lumbosacral region and tumor dissemination. Management is based on a broad primary resection, for complete removal of the lesion, followed by adjuvant therapy.

Keywords: Chordoma, lumbosacral, dissemination, metastasis

PSEUDOTUMORAL FORM OF NEUROCYSTICERCOSIS: A CASE REPORT

Rodolpho Albuquerque Souza, Felipe Gabriel dos Santos Fonseca, Maria Letícia Marques Pinheiro, Alexandre Varella Giannetti

Introduction/Background:Neurocysticercosis is an endemic infectious disease, with high prevalence in developing countries, especially in areas of greater social vulnerability. Its clinical presentation is diverse, according to the cysticercus location and uncommon in pediatric population. We present a case of atypical manifestation of neurocysticercosis simulating, by radiologic aspects, a central nervous system (CNS) tumor (i.e., pseudotumoral form).

Case Report: A 17 year-old boy was admitted at the emergency room with signs of intracranial hypertension. The





imaging exams reveled an obstructive hydrocephalus secondary to an expansive lesion at the right anterior thalamic nucleus. At first, he underwent endoscopic surgery to biopsy and placement of ventriculoperitoneal shunt. After an inconclusive result, a transcalosal microsurgical approach was performed with gross total resection of a well defined lesion. The pathological analysis of the surgical sample was conclusive to a viable cysticercus. At follow-up, the patient shows memory deficits and executive dysfunction, in progressive improvement with rehabilitation.

Discussion: In the life cycle of Taenia solium humans generally acts as the definitive host, carrying the tapeworm in their small intestine. If the patient ingests the eggs shed in the feces of a human tapeworm carrier, it will act as an accidental intermediate host, sheltering the larval stage of the parasite (i.e., cysticercus).Cysticerci can migrate to CNS causing the neurocysticercosis. Despite the generally classic radiologic aspect, we should consider this disease as a differential diagnosis of intracranial masses in endemic regions, since it may manifest as pseudotumoral lesions, like in the case reported, leading to wrong conclusions and consequently inadequate treatment.

Key-words: Neurocysticercosis, pseudotumoral form, cysticercosis, neuroendoscopy, infectious diseases

ENDOSCOPIC AQUEDUCTOPLASTY AND STENT PLACEMENT IN DANDY-WALKER MALFORMATION

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Introduction: The management of Dandy-Walker syndrome is not consensual and surgical options shifted from posterior fossa opening and membrane excision to shunt and neuroendoscopic approaches.1,2 It is important to evaluate clinical and radiological features of each patient to indicate the most appropriate procedure.

Case report: We report a case of a 15-year-old boy who presented with occipital headache and dysphagia started 7 months ago. He was diagnosed with Dandy-Walker submitted syndrome birth and at was to ventriculoperitoneal (VP) shunt at 4-day-old. At the physical examination, he exhibited mild dysarthria and reduced palatal reflexes. The MRI disclosed small supratentorial ventricles; dilatation of the fourth ventricle and aqueduct stenosis. Based on complaints suggesting posterior fossa hypertension and MRI that revealed aqueductal stenosis we proposed an endoscopic aqueductoplasty and stent placement. The patient evolved with remission of the symptoms.

Discussion: VP-shunt is enough to treat hydrocephalus in many patients diagnosed with Dandy-Walker malformation1. However, cystoperitoneal (CP) shunts are favored by authors who advocate placement inside the posterior fossa cyst to maintain downstream flow in the aqueduct and drainage of both ventricles and cyst.5,6 Functional aqueductal stenosis and secondary isolated fourth ventricle are well described conditions and require a combined VP-CP shunt or an endoscopic communication between compartments. Therefore, aqueductal patency is one of the radiological features that dictate the surgical procedure of choice and this analysis is even more important for patients previously treated in order to prevent transtentorial pressure gradient.

Key-words: Endoscopic; aqueductoplasty; Dandy-Walker malformation.

HYDROCEPHALUS SECONDARY TO DENGUE ENCEPHALITIS IN AN INFANT: CASE REPORT

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Background: Dengue is the leading cause of mosquito-borne viral infection. It is responsible for high morbidity and mortality in children living in endemic areas. Nowadays, neurological complications are progressively referred to and include a broad spectrum of symptoms. It can be secondary to metabolic alterations, direct invasion by the virus, and enhanced autoimmune response.

Case presentation: Here, we report a rare case of hydrocephalus secondary to dengue encephalitis. A 13-dayold boy was diagnosed with dengue. He evolved with seizures and impaired consciousness being diagnosed with encephalitis. After clinical treatment, he was discharged fully recovered. Three months later, he presented with signs and symptoms of intracranial hypertension. Brain MRI revealed hydrocephalus secondary to acquired aqueduct stenosis. The patient underwent an endoscopic third ventriculocisternostomy.

Conclusion: Dengue infection is a differential diagnosis for acute febrile neurological impairment in children from endemic areas. Follow-up should be offered after dengue encephalitis to detect possible late-onset complications, as hydrocephalus.

Key-words: Hydrocephalus, Dengue infection, Encephalitis, Third ventriculostomy, Flavivirus, Dengue

MEDIAN SUPRAORBITAL MINICRANIOTOMY FOR BILATERAL FRONTAL EPIDURAL EMPYEMA

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Abstract

Introduction/Background:Intracranial complications of sinusopaties are uncommon complications, even in pediatric population. If left untreated, can lead to poor outcomes.

Case Report: We present a case of a female child, 08 years old, with progressive and persistent bifrontal headache, with fever, decrease in general condition for 01 week, altered consciousness, nausea and vomiting. Recent history of





bacterial sinusitis with incomplete treatment. On examination, sleepy, GCS 14, pupils +3/+3, cranial pairs preserved, no appendicular motor deficits, no meningisms. She underwent CT scan showing bilateral frontal epidural empyema with acute brain hypertension. A right median supraorbital minicraniotomy was made with complete abscess evacuation followed by 08 weeks triple intravenous antibiotic treatment and complete cranial infecction resolution, without other surgical procedures.

Discussion: Frontal epidural empyemas can be treated with minicraniotomies and adapted antibiotic treatment since the extensive irrigation responsible for the ease of contiguity of the frontoethmoidal sinusopathies and the formation of these collections provide a safe route for intravenous antibiotic delivery.

Key-words: empyema, craniotomy, surgical access, Intracranial infection

UNUSUAL COMPLICATION OF INTRACYSTIC INJECTION OF ALPHA-INTERFERON FOR CRANIOPHARYNGIOMA: A CASE REPORT

Felipe Gabriel dos Santos Fonseca, Rodolpho Albuquerque Souza, Luis Augusto Vilela Silveira, Alexandre Varella Giannetti

Introduction: Craniopharyngiomas are tumors whose treatment it's still a challege. Intracystic therapy with interferon alpha is one of the therapeutic modalities, but its use may be followed by complications.

Case Report: We present the case of a patient with craniopharyngioma, submitted to multiple surgical approaches, followed by radiotherapy. Due to a cystic recurrence of the primary lesion in the posterior fossa, the patient underwent implantation of an Ommaya catheter in the cystic portion of the tumor to infusion of interferon alpha, by a retrosigmoid approach in 2020 at the age of 18 years (CT scan with contrast did not show contrast leak). Interferon alpha infusions were performed, during the first cycle, with relatively good tolerance. After the start of the second cycle, however, the patient presented important side effects of worsening motor strength, behavioral changes (depression with suicidal ideation), dizziness e vomiting, in addition to altered image in resonance (hyperintense signal in FLAIR) located in brainstem next to the tumor. In follow up, the patient improved symptoms after a course of corticosteroids and interferon withdrawal and in control MRI showed improvement of the previous alteration.

Discussion: Craniopharyngioma treatment strategies include surgical resection, radiotherapy and intracystic therapies. Interferon acts through its effect on squamous cells, leading to the activation of apoptosis pathways, with control of tumor volume. Despite good results, the use of interferon may be associated with the development of systemic symptoms, psychiatric disorders such as depression, new endocrinological and neurological disorders, in addition to seizures.

Key-words: craniopharyngioma, interferon, Ommaya catheter, complications

FETAL EPIDURAL HEMATOMA SECONDARY TO A MILD BLUNT PRENATAL TRAUMA

Vinícius Gomes de Morais, Bárbara Albuquerque Morais, Cilmária Leite Franco, Paulo Ronaldo Jubé Ribeiro

Introduction/Background: Fetal intracranial hemorrhage affects 1 in every 10,000 pregnancies. In most cases, the etiology of the bleeding is multifactorial, and they can be either related to the mother or the fetus. Blunt prenatal trauma was occasionally associated with these hemorrhages, nevertheless, reports of hematomas secondary to mild traumas are rare.

Case Report: A previously healthy woman, on her second pregnancy with normal prenatal tests, was referred to a routine ultrasound at 35 weeks, which detected an extensive hypoechoic lesion located at the fetus's occipital region. The patient had a mild "domestic accident" 1 week before. She referred that the left side of her belly shocked against the edge of a furniture. The pregnancy continued as planned and a healthy baby was born at term through an elective Csection. A brain MRI at 13 days of age that confirmed an extensive epidural hematoma at the midline occipital region extending inferiorly to the topography of the confluence of sinuses. Blood screening revealed no anemia or coagulation disturbances. At 3 months old, the child was neurologically intact, maintaining the normal growth of the cephalic perimeter. A new brain MRI revealed no more signs of intracranial bleeding, indicating complete reabsorption.

Discussion: Within the prenatal intracranial bleedings, the most frequent are the subarachnoid hematoma and intraparenchymal, scarcely ever the epidural hematoma. Treating these bleedings is challenging due to the ongoing pregnancy. Thus, the prognosis is often reserved, with a mortality rate of 43% and 25% of neurological sequelae.

Key-words: Intracranial hemorrhage, Epidural hematoma, Pregnancy, Blunt trauma, Prenatal

RUPTURED CEREBRAL ARTERIOVENOUS MALFORMATION IN A PEDIATRIC PATIENT: CASE REPORT

Leandro Cândido de Souza, Nicole Silva de Araújo, Matheus Fernando Manzolli Ballestero

Background: Cerebral arteriovenous malformations (AVM) are a rare vascular disorder and even more rare in children, with an incidence of symptomatic cases of 1.1 in 100,000 children/year and a prevalence of 0,02%. Although there is no consensus on the treatment of asymptomatic lesion, symptomatic bleeding lesion are passive of treatment with surgery, endovascular treatment or both.





Case report: We present a case of an 11-years-old child presenting to the emergency room with thunderclap headache and somnolence. The neurological exam showed torpor and no detected focal deficit. A computed tomography (CT) scan evidenced a frontal intraparenchymal bleeding with ventricular invasion. The child was then submitted to an external ventricular shunt and them performed a magnetic resonance imaging (MRI) and a angiography showing an AVM. After discussing with neurovascular team, a surgical intervention was performed draining the bleeding and treating the AVM with complete resection. After six months of follow-up, the child is asymptomatic and without shunt dependence.

Discussion: AVMs are rare lesions in children and must be a differential diagnosis when an intracerebral bleeding is present, especially in a nontraumatic case, as 80-85% of all pediatric AVMs present with hemorrhage. Some findings presented in our case are considered red-flags for AVM, such as severe headache and lethargy, which may indicate increase intracranial pressure (ICP) and/or dural irritation from subarachnoid blood. Although surgery is considered the first-line treatment for urgent intervention, a multidisciplinary approach is essential to increase the breadth of treatment options.

Key-words: arteriovenous malformation, intracerebral hemorrhage, neurovascular disorder, pediatric

TERMINAL MYELOCYSTOCELE ASSOCIATED WITH DERMAL SINUS TRACT AND INTRAMEDULLARY DERMOID CYST-CASE REPORT

Leyzeane Marques do Nascimento, Daniel Dante Cardeal, Bruno Spindola Amaral Garcia de Freita, Hamilton Matushita, Felipe Hada Sanders, Jose Erasmo Dal Col Lucio

Introduction: Terminal myelocystocele (TMC) is a wellrecognized entity. Pang et al. presented imaging, surgical, and histopathological evidence of TMC that argues for its genesis during secondary neurulation and, furthermore, that it arises from failure of apoptosis at a specific point in the late degenerative phase of spinal cord formation. Spinal dermoids, which constitute less than 1% of all pediatric intraspinal tumors, and can be seen in association with dysraphic anomalies, usually in the same anatomical site or even after surgical treatment due to the inclusion of epithelial tissue during the surgical procedure. When congenital, they arise from dislocated ectodermal skin inclusions during neural tube formation between the 3rd and 5th gestational weeks, during primary neurulation and are associated with the congenital dermal sinus. This article reports a case of association between terminal myelocystocele and dermoid cyst in different anatomical sites and time.

Case Report: This is a 1Y9m child, operated on for terminal myelocystocele at birth. She had good neuropsychomotor and sphincter development. It has recently started with

frequent falls, paraparesis, low back pain, constipation and recurrent urinary tract infections. Physical examination showed signs of pyramidal release and MRI thoracolumbar spinal cord anchored in L4 and intramedullary thoracic lesion with dermal sinus. Submitted to surgical exeresis of the same with good evolution.

Discussion: Spinal dysraphisms are complex diseases and may be associated with other spinal column abnormalities. The diagnostic extension of these patients in the search for associated lesions is of fundamental importance.

Key-words: Terminal Myelocystocele; Dermal Sinus Tract, Dysraphism

SOLITARY MYOFIBROMA OF THE SKULL: RESECTION OF INFANT'S FRONTAL BONE LESION

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Background: Infantile myofibromatosis is rare and usually occurs in the first 2 years. Although is a rare disease, the infantile myofibromatosis is the most common fibrous tumor of infancy and has a benign course.

Case presentation: We present a case of solitary myofibroma of the skull. The patient was a 6-month-old male infant with a left frontal bone lesion that had been slowly increasing in size. On clinical examination, he had a mass in the left frontal region of his skull with an area welldefined and nonmobile. The head CT scan showed an osteolytic lesion. The eosinophilic granuloma of the skull was suggested and we proposed a surgical approach. The tumor was removed and the histologic examination revealed a mesenchymal lesion with spindle-shaped cells and eosinophilic cytoplasm. The immunohistochemical examination was positivity for smooth muscle actin, which are associated with myofibroma.

Discussion: This entity is a mesenchymal disorder characterized by formation of nodular tumors in the soft tissue, skeleton and internal organs. Few cases of solitary myofibroma of the skull in infant have been reported in the literature and the etiology of this disease is unknown. It is a lytic lesion with sclerotic rim in most cases and presented in CT scan as an hypodense or isodense lesion. The most commonly differential diagnosis are epidermoid/dermoid cysts and Langerhans cell histiocytosis. The treatment is surgical excision, and chemotherapy is reserved for the disseminated disease. If totally removed, solitary infantile myofibroma usually has a favorable prognosis with a 10% recurrence rate.

Key-words: Solitary myofibroma, bone lesion, skull, juvenile fibromatosis, scalp

COMPLETE RESOLUTION OF GIANT ARACHNOID CYST OF THE MIDDLE FOSSA AFTER SURGERY

Amanda de Oliveira López, João Gabriel Ribeiro gomes





Introdution: The best treatment for intracranial arachnoid cysts (AC) remains controversial. Endoscopic Fenestration and microsurgical approach are currently primary therapy modalities. Cysto-peritoneal shunts has been used after initial therapeutic failure, due to its potential complications.1 We report a patient with AC submitted to microsurgery with complete resolution at follow-up.

Case Report: A 9 months boy was admitted with progressive macrocrania and neurological development delay. Brain MRI showed a giant AC (Galassi III) with mass effect. Microsurgical treatment with communication between cyst and BC and third ventricle was performed. Two weeks later, patient started with irritability and bulging of anterior fontanelle. New scan revealed a Subdural Hygroma (SH) with mass effect. A subduroperitoneal shunt (SDPS) was placed, with symptoms resolution. After 5 months, patient had a traumatic subdural hematoma, and underwent surgery for hematoma evacuation and shunt removal. At 12 month follow up, had normal neurological development and total AC resolution.

Discussion: AC surgery aims to reduce mass effect and signs of intracranial hipertension. Postop SH is more frequent in children under 3 years due to immaturity in CSF absorption.2 Symptomatic and hypertensive SH are treated with subdural-peritoneal shunt. Later, CSF natural dynamics could be re-established and shunt possibly removed.3

Most treated cysts show partial reduction in postop volume.4 Children younger than 2-3 years have greater volume decrease and postoperative cerebral reexpansion.5 Total disappearance of AC is seen in less than 10% of Galassi I/II surgically treated but rarely described in Galassi III. 6

Key-words: Arachnoid cyst, surgery, subdural hygroma, total resolution

ATYPICAL TERATOID-RHABDOID TUMOR IN A 6-YEAR-OLD CHILD

Luiz Fernando Alves Pereira, Ana Maria Mendes Ferreira, Roberto Alexandre Dezena

Introduction / Background: Atypical teratoid-rhabdoid tumor (ATRT) is a rare, highly aggressive, childhood-predominant brain tumor with a very poor prognosis, despite therapy1,2. The scientific literature on this tumor is relatively scarce3.

Case Report: A 6years-old female child with the acute onset of daily recurrent vomiting associated with a bluish vascular lesion on the left eyelid and headache. Neurological examination aBrain MRI showed high grade expansile lesion in left frontal lobe, subfalcine herniation and signs of intracranial hypertension, surrounded by vasogenic edema. Submitted to left frontal craniotomy for microsurgical resection. Total removal was achieved. Patient remained without postoperative deficits. CT 1st postoperative day showed no residual lesion. Histopathological diagnosis of ATRT. Serial MRI scans showed recurrence at the same resection site, and the child underwent other surgeries, with no time for complementary oncologic treatment, evolving to death. right central facial paresis.

Discussion: This is one of the most aggressive brain tumors ever described. Performing several surgeries does not change the prognosis.

Key-words: teratoid-rhabdoid tumor, brain tumor

