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Case report

Management of hydrocephalus after cerebellar pilocytic astrocytoma in a pediatric patient: case report and literature review

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ABSTRACT

Background: Pilocytic astrocytoma (PA) is the most common brain malignancy in children. PA is a distinct histological and biological subgroup of gliomas that have a slow growth rate. Most PAs are located infratentorial and in the median cerebral structures such as the optic nerve, brain stem, and hypothalamus. Hydrocephalus is a common finding in patients with PA.

Case presentation: In this report, we describe a case of a patient treated surgically for PA who developed a common complication – hydrocephalus. Pre-operative dilatation of the lateral ventricles with transependymal cerebrospinal fluid flow along with her young age were defined as the risk factors. She required numerous interventions and the findings along with treatment modalities are discussed in detail.

Conclusions: Cases with pre-existing risk factors for post-operative hydrocephalus should be evaluated prophylactic cerebrospinal fluid diversion surgery. If a more conservative approach is employed, the patients' neurological status and ventricle size should be closely monitored. Pediatric patients are negatively affected with repetitive surgeries in terms of physical and mental health. Thus, in order to avoid additional interventions, pre-operative planning of all available options should be discussed with the family.

Key words: pilocytic astrocytoma, hydrocephalus, ventriculo-peritoneal shunt, case report, pediatric

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BACKGROUND

The term *pilocytic* used to describe astrocytoma refers to cells with hairy bipolar processes. Today it is known that pilocytic astrocytomas (PA) are slow growing and well defined tumors. According to the classification of the World Health Organization (WHO), PA are classified as grade I gliomas with low cellularity and mitotic activity. Metastatic spread is extremely rare while some may infrequently undergo malignant transformation [1].

PA can occur anywhere in the central nervous system. However, most PA arise in the cerebellum (42%), followed by the supratentorial lobes (36%), the visual pathway and hypothalamus (9%), brain stem (9%) and spinal cord (2%). In children, the cerebellum is most often affected (67%), and only rarely does PA develop supratentorially [2].

PA constitute 1–5% of all intracranial tumors and 1.7% to 7% of all glial tumors and 70% of cerebellar astrocytomas in children [3]. Note that PA is the most common primary brain tumor in children and accounts for 15.4% of primary brain tumors in children and adolescents and 17.6% of primary brain tumors in children under 14 years of age [4]. PA are potentially curable by surgical removal and are associated with a 10-year survival rate of 90% in children and 63–83% in adults [3]. However, the presence of hydrocephalus can complicate the treatment algorithm [5].

In this report, we describe a case of a patient treated surgically for PA who developed a common complication – hydrocephalus. She required numerous interventions and the details of the management of hydrocephalus after PA surgery are discussed.

CASE PRESENTATION

A 24 month old female patient was referred to our clinic. According to the mother, at the age of 12-months the first operation was performed – a biopsy of a tumor localized in the cerebellum. 2 months later after the pathological examination revealed the lesion to be PA, microsurgical removal of the mass was performed using with the aid of intraoperative USG at the Morozovskaya Hospital (DGKB DZM). After the surgical excision, hemiparesis on the right and concomitant difficulty in crawling remained. After discharge, her condition worsened. A month later she was diagnosed with hydrocephalus and a ventriculo-peritoneal (V-P) shunt was implanted without an anti-siphon. The shunting did reduce most of the symptoms however headache, nausea and vomiting occurred in the upright position. A month later the V-P shunt was removed endoscopic triventriculocisternostomy (ETV) was performed. There was a temporary improvement in the condition, followed by a worsening. 2 weeks later a V-P shunt (Miethke proGAV, pressure 5.0) + Shunt Assistant (20) was re-implanted as the child did not recover from the worsening after ETV. The valve pressure was rearranged to 0 cm H_2O 2 days later. Lethargy and drowsiness improved. She was discharged a week later with weakness of the facial muscles on the left side and a motor strength of 3/5 on the right side. 10 days after her discharge the patient was brought to the emergency department (ED) with lethargy, overall weakness and apathy. The V-P shunt was revised and the anti-siphon was removed. The opening pressure of the valve was set at 5 cm H_2O. The patient was discharged 2 days later only to re-apply to the ED 4 days later with repetitive vomiting and headache.

This time, chronic subdural accumulation in the right parieto-occipital region, subdural accumulation of hemorrhagic density along the outline of the cerebellum (more on the left) and along the convexial surface of the left occipital lobe was found. With the diagnosis of over-drainage, a new V-P shunt with anti-siphon was placed while the previous one was removed (fig. 1, 2).

Figure 1. A, B. Preoperative CT with contrast. Ventricular dilatation with a tumoral formation in the posterior cranial fossa presenting with perivascular edema. Note the dislocation of the amygdala in the posterior fossa and transependymal cerebrospinal fluid flow. C. Early postoperative period CT. Signs of chronic subdural collection bi-frontally.

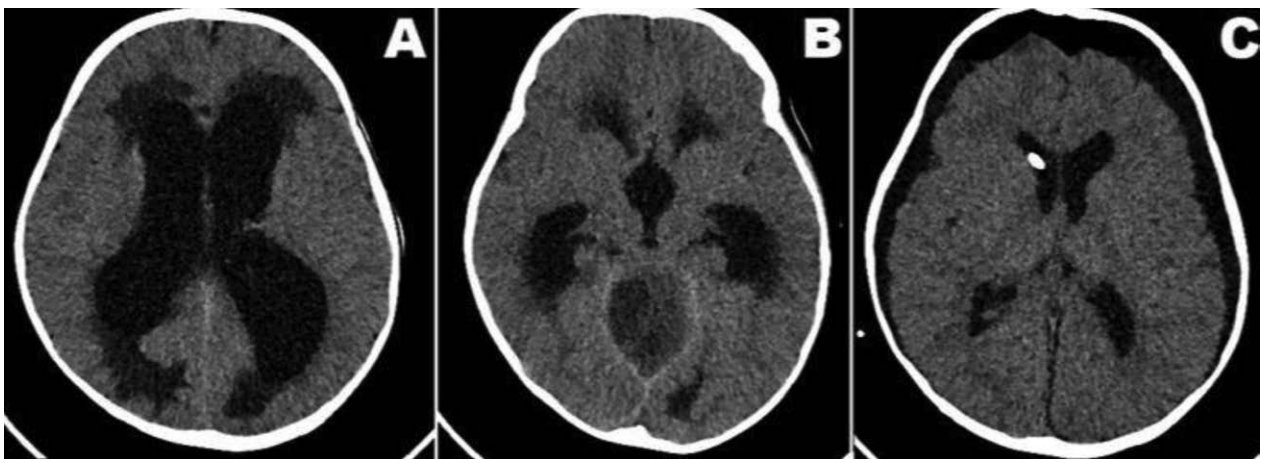
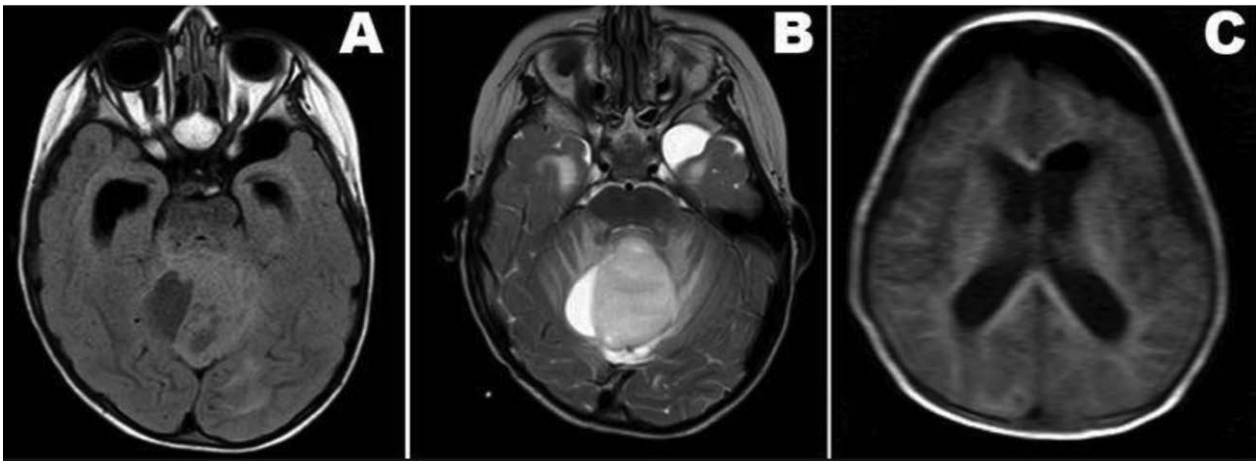


Figure 2. A, B. Preoperative MRI with contrast. Note the perilesional edema and the effaced fourth ventricle. C. Early postoperative period. The postoperative cavity in the posterior cranial fossa was unremarkable. There were no convincing signs of residual tumor tissue. After shunting the total regression of triventricular obstructive hydrocephalus was observed.



DISCUSSION

Hydrocephalus is one of the main complications associated with surgical intervention for lesions of the posterior cranial fossa. The incidence of hydrocephalus after posterior fossa tumor removal ranges from 10% to 40% in the pediatric population [6]. Risk factors for postoperative hydrocephalus are young age (≤2 years), medulloblastoma, and compression of the brainstem. Interestingly, histopathology indicating the presence of astrocytoma ($p = 0.003$) was a negative predictor of hydrocephalus [7]. Moreover, in the adult population, PA was a significant risk factor for the development of postoperative hydrocephalus. A possible pathophysiological explanation may be related to the slow growth of the tumor. Slow growth can lead to the development of chronic hydrocephalus and a decrease in parenchymal compliance, which leads to rigidity of the ventricular walls [8].

The CPPRH (Canadian Preoperative Hydrocephalus Prediction Rule) for pediatric patients with posterior fossa tumors was created to identify individuals at high risk of hydrocephalus. 5 components (age less than 2 years, presence of papilledema, hydrocephalus on preoperative imaging, presumptive tumor diagnosis of medulloblastoma, ependymoma, brainstem glioma, and cerebral metastases) made up a scoring system that ranged from 0 to 10 points [9]. Higher scores correlate with a higher risk of persistent hydrocephalus. In a modified version, the presence of optic disc edema was replaced by radiological evidence of transependymal cerebrospinal fluid (CSF) flow. Our patient had 6 points with a probability of hydrocephalus at 6 months after resection of 0.693. Preoperative risk analysis for similar cases should be treated with caution [10]

Persistent hydrocephalus was more common in children with severe preoperative hydrocephalus ($p = 0.002$) and medulloblastomas ($p = 0.0154$). In addition to clinical characteristics, the molecular subtype can also influence the development of hydrocephalus in pediatric cohorts; however, such data on PA are currently lacking [11]. The main risk factor in our patient was the preoperative dilatation of the lateral ventricles along with transependymal CSF flow. Her young age may have also been a factor in developing postoperative hydrocephalus. The incidence of postoperative hydrocephalus in pediatric patients is about 5 times higher than in adults [12]. With this in mind, it may be prudent to consider the benefits associated with CSF diversion surgery in the pediatric population before or concomitant to the tumor removal. To date, there have been numerous reports of alternatives to V-P shunting, such as prophylactic ETV of the third ventricle, which reduces tumor-associated occlusive hydrocephalus in almost 90% of cases [13]. The efficacy of ETV before surgery for posterior fossa lesions has been described in both adults and children, resulting in a significant reduction in dependence on V-P shunt. However, in patients undergoing ETV, serious complications such as basilar artery injury have been described [14, 15]. The success rate of ETV ranges from 71% to over 90% on longitudinal analysis up to more than 9 years, while the overall complication rate is 9% [14]. Note that the incidence of complications with ETV is lower than with V-P shunting, but it should be recognized that severe complications (intratumorally bleeding, ascending hernia, and death) were recorded in about 1% of patients. Given such data, the question of the prophylactic use of ETV remains controversial [16].

The hydrocephalus in the present case was primarily treated with V-P shunting. However, numerous revisions were required due to shunt dysfunctions, occlusion, and over- or under-drainage. She underwent ETV after V-P shunt dysfunction, which was

also unsuccessful in treating her hydrocephalus. Previous studies have put forth patient age, etiology of hydrocephalus, and prior shunting in ETV success rates. Early failures are attributed to intraoperative technical difficulties such as third ventricular size and shape, thickness, and position of the third ventricular floor, while late failures were attributed to closing of the stoma or cistern scarring [17]. Although cerebellar hemorrhages are not rare in this type of population, they are assumed to be due to head trauma or ruptures of vascular malformations, as well as infections or hematogenous diseases. It should be noted that bleeding in low-grade tumors is related to structural anomalies, and the angioarchitecture is specific to tumor vessels, such as degenerative mural hyalinization and glomeruloid-type endothelial proliferations [18].

Although a study on motor recovery after removal of these tumors in the cerebellum showed a major improvement in the first year postoperatively, the development of the cerebellum being in progress, both in childhood and adolescence, is a factor that is not measured by age at the time of injury but is beneficial for postoperative motor recovery [19].

A comparative analysis was performed in which PA involving the brain stem and those related to neurofibromatosis type I were concluded. We tabulated preoperative volumes and calculated dates from the reports [20]. The median follow-up period was 6 years, 2.07–12.28 years in total [21]. But the MRI examinations were consecutive for at least a 3-month period, showing the lowest negative probability of recurrence of 0.15%; there was no recurrence based on tumor volume [22]. We should avoid any delay in adjuvant therapy due to poor prognosis and avoid any signs of cerebrospinal fluid leak and postoperative CSF drainage to improve wound healing. In the ETV group, postoperative CSF leaks were 0.7%, while in the non-ETV group, they were 13.4% [23].

CONCLUSIONS

Risk factors for postoperative hydrocephalus in pediatric patients with PA are young age (≤2 years), brainstem compression, papilledema and mainly pre-operative hydrocephalus. In patients with known risk factors, it is necessary to consider the use of prophylactic CSF diversion surgery. This may be employed pre or intraoperatively. If the conservative approach is employed, the patients' neurological status and ventricle size should be closely monitored. Pediatric patients are negatively affected with repetitive surgeries in terms of physical and mental health. Thus, in order to avoid additional interventions, pre-operative planning of all available options should be discussed with the family.

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Authors' contributions:
Daniel Encarnación Santos, Bipin Chaurasia, Egor Chmutin: conceptualization, methodology, software.
Gennady Chmutin, Daniel Encarnación Santos: investigation, data curation, surgery.
Gennady Chmutin, Ismail Bozkurt: supervision, visualization, validation.
Daniel Encarnación Santos: writing – original draft, writing – review & editing, formal analysis.

Conflict of interests:
The authors declare no conflict of interest

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Ethics:
This case report is based on retrospective data and does not involve any experimental intervention. As per institutional guidelines, ethics committee approval was not required. Written informed consent was obtained from the patient for publication of clinical information and associated images.