International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Rare Complication in a Middle Age Child with Pilocytic Astrocytoma: A Case Report

Ander Arranz

Children Teaching Hospital, Košice, Slovakia

Abstract: ***Background*: Brain tumors are the most prevalent type of solid malignancy among paediatric patients. Pilocytic astrocytoma (PCA), often discovered in the posterior fossa, is the most frequently encountered juvenile cerebral tumor. Although malignant progression is uncommon, this gradually developing neoplasia can present with a wide range of complications. Brainstem compression and hydrocephalus, both of which have the potential to be fatal, may be present in these patients and require urgent intervention. However, external ventricular drain (EVD)-associated ventriculitis may present in a minority of these patients and lead to neuroinfection with associated neurologic sequelae. Source control and total surgical resection of the neoplasm along with antibiotic therapy are often considered the cornerstone of therapy. *Case presentation*: We present the uncommon case of a 7-year-old white boy who was admitted to the hospital due to vomiting, severe headache and convulsions. CT scan of the brain showed a large cystic lesion with a bright mural nodule in the posterior fossa of the brain. Likewise, compression of the adjacent structures along with obstructive hydrocephalus and cerebellar herniation were the most worrisome radiographic findings. Following this, an EVD was immediately placed. The therapeutic regimen consisted of dexamethasone, mannitol, sodium chloride, moxastine teoclate, metoclopramide, vancomycin, paracetamol, metamizole, tramadol and phenobarbital. *Conclusion*: Children are most likely to develop this low-grade and often well-circumscribed tumor, which generally has a favorable prognosis. However, prompt detection and early surgical excision can significantly reduce neurological impairment and improve quality of life.***

Keywords: Pilocytic astrocytoma, External ventricular drain, Ventriculitis, Cerebrospinal fluid, Staphylococcus epidermidis

1. Introduction

Astrocytes, a specific type of glial cell that sustains and provides nourishment to the neuronal cells in the brain, constitute the source of the brain neoplasm referred to as PCA. This usually benign and gradually evolving neoplasia represents the most common brain tumor in children. PCA accounts for 15% of all brain tumors in children and comprises 27 to 40% of all pediatric posterior fossa tumors [1, 2]. The incidence rate is estimated at 14 new cases per million in children younger than 15 years of age [3]. Indeed, the majority of these neoplasms occur most commonly within the first two decades of life. Pilocytic astrocytomas (WHO malignancy grade I) are the commonest form of astrocytoma in children and may arise anywhere in the CNS but most commonly occur in the cerebellum [4]. Likewise, the optic nerves, third ventricle, brainstem, hypothalamus and, less commonly, the spinal cord are other noticeable areas where these lesions tend to develop.

Pilocytic astrocytomas occur both sporadically and in association with neurofibromatosis type 1 (NF-1), wherein it particularly affects the optic pathways [5]. Common among many of the NF1-associated tumors is the fact that they have a more favorable course than their sporadic counterparts [6]. The extent and location of the tumor influence its manifestations. However, the majority of the symptoms are caused by elevated intracranial pressure. When the tumor is cerebellar, the symptomatology can be ataxia and signs of increased intracranial pressure, as headache, nausea and vomit [7]. Additionally, visual field loss or several endocrinopathies may result when the lesion is located along the optical pathway or hypothalamus, respectively.

2. Case Presentation

A previously healthy 7-year-old white boy was admitted to the Emergency Department due to several episodes of vomiting, subfebrility, persistent headache and convulsions. The day before the admission, the child had received antiemetic treatment and rehydration support for the same reason. During the physical examination, the child presented with poor oral intake, weakness and lethargy. Neurologic examination revealed bilateral upward eye deviation and tonic spasms of the upper and lower limbs, especially on the right side of the body. Phenobarbital was administered and convulsions ceased briefly. Cardiopulmonary resuscitation (CPR) was successfully performed. Complete blood count showed a significant leukocytosis with neutrophilic predominance and thrombocytosis. Likewise, elevated glycemia (Glu=11.4 mmol/L) and mild hypokalemia were present. Arterial blood gas (ABG) test indicated metabolic acidosis (pH=7.288), hyposaturation (sat02=76%), hypocapnia (pC02=3.99) andnoticeably elevated lactate levels (5.2 mmol/L). Therefore, rehydration support and continuous monitoring were performed. CT scan of the brain revealed convulsive activity and bulbar deviation. Additionally, a cystic tumorous infiltration was detected in the posterior fossa of the brain with compression of the 4th cerebral ventricle and brainstem, along with obstructive hydrocephalus and cerebellar tonsillar herniation. Phenobarbital was re-administered to our patient. Dexamethasone, sodium chloride and mannitol were added to the therapeutic regimen to reduce intracranial pressure. Following this, the patient was urgently transferred to our center for further therapeutic intervention due to obstructive hydrocephalus.

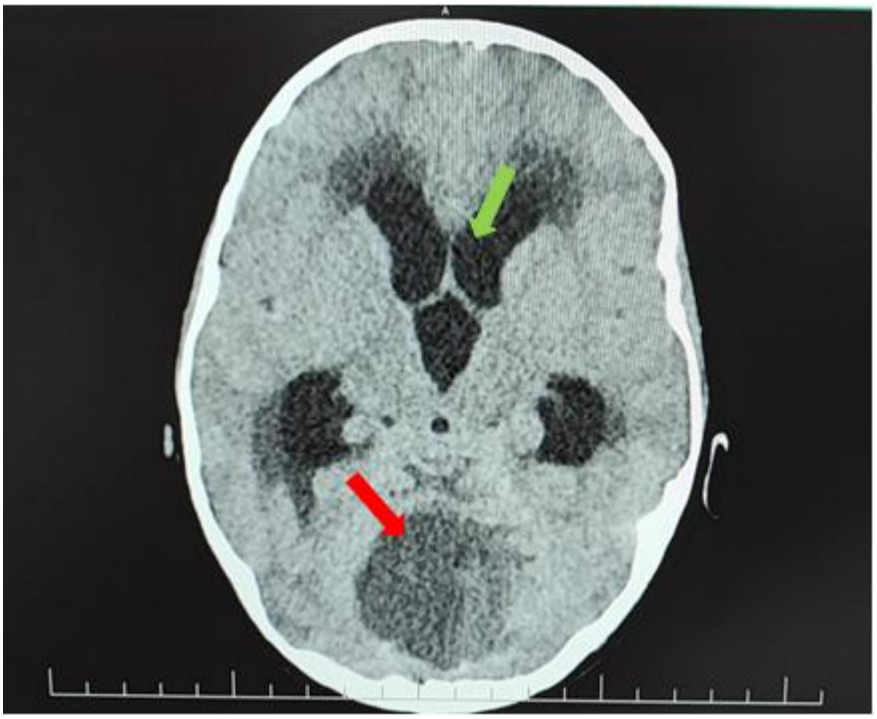

Figure 1: Non-contrast axial CT of the brain. The red arrow indicates tumoral expansion in the posterior fossa between the cerebellar hemispheres with pressure on the adjacent structures including the cerebellum and brainstem along with tonsillar herniation. The green arrow shows severe obstructive lateral ventricle hydrocephalus with signs of brain edema

After the admission, an EVD was immediately placed under general analgosedation. Antiedematous treatment with mannitol was prolonged. However, the ABG test proved an uncompensated metabolic acidosis. CSF analysis suggested hyperglycorrhachia, decreased protein levels and elevated lactate. After consultation with the neurosurgeon, the patient underwent a suboccipital craniotomy and the samples obtained were sent for cytology and histologic examination. The procedure did not include major complications and antibiotic coverage was prescribed. Control MRI after brain tumor resection and preliminary histological findings were evaluated conjunctively as grade I. Pilocytic astrocytoma. Neurologic examination described right-sided neocerebellar symptomatology. A hemogram test showed mild normochromic normocytic anemia and CRP elevation. A new CSF analysis outlined hyperproteinorrhachia and the culture of CSF was positive for Staphylococcus epidermidis. The next day, the patient developed pyrexia (T=38°C) following the course of the infection. The child had polyuria (1980 mL/day) with normal sodium urine levels. Indeed, based on the clinical picture and culture positivity the condition was assessed as ventriculitis, for which vancomycin was subsequently prescribed. The control hemogram test showed improvement with CRP decrease, stationary mild anemia and a mineralogram without significant abnormalities. Additionally, CSF analysis was also within the normal range. Next, the patient was stabilized (HR=78/min; satO2=99%) and tolerated the diet without nausea and vomitus. However, the patient refused mobilization due to persistent dizziness and required further rehabilitation. Following this, the left central venous catheter, permanent urinary catheter and EVD, which contained 71ml of sanguine content, were removed.

3. Discussion

As indicated earlier, depending on the tumor's location and the patient's capacity to report their nervous system complaints, the presenting signs of this condition might not always be immediately apparent. Indeed, hydrocephalus develops when the fragile equilibrium between the CSF generation and absorbance is perturbed and CSF accumulates in the cerebral ventricles. This natural balance becomes disrupted when CSF is excessively generated, improperly absorbed, or obstructed by a blockage like an intraventricular tumor. Nevertheless, EVD is rarely associated with ventriculitis and only 27 case reports to date have confirmed this as the primary source of neuroinfection. Exceptionally, from these, only 11 cases of PCA have been associated with ventricular infection. There is no clear definition for ventriculitis and no accepted diagnostic criteria, which makes the assessment of incidence difficult [8]. EVD-related ventriculitis has consequences not only in terms of mortality but also, more importantly, on the development of severe neurological sequelae [9]. The current research in EVDs has focused on improving the overall safety of the procedure, which includes development of guidance-based systems, virtual reality simulators for training, and antibiotic-impregnated catheters [10].

Additionally, medical imaging of these tumors frequently reveals well-defined, enhancing lesions that are accompanied by cyst development. Only a minority of pilocytic astrocytomas are calcified [11]. Furthermore, as pilocytic astrocytomas often show some infiltration in the surrounding brain tissue, discrimination from a diffuse glioma can be difficult [12]. Management of this condition is not well-defined. Some authors advocate pre-craniotomy ventriculoperitoneal shunt orendoscopic third ventriculostomy or external ventricular drain and subsequent

craniotomy and excision of brain tumor, while others preferinitial tumor excision and subsequent treatment of hydrocephalus when it persists [13-15].

4. Conclusion

Ventriculitis caused by EVD is one of the most serious side effects of this therapy. Cutaneous flora or nosocomial microbes, such as Gram-negative bacteria, are the most commonly related microorganisms. There is a large ongoing research focused on the prevention of EVD-associated ventriculitis by impregnation of the CSF drains with different antibiotics, including rifampicin and minocycline. Other preventive measures include perioperative antibiotic administration, adequate hand hygiene and lessen manipulation to enlarge the drainage duration thereby reducing the risk of infection.

References

[1] AlRayahi, J. *et al.* (2018) 'Pediatric brain tumor genetics: What radiologists need to know', *RadioGraphics*, 38 (7), pp.2102–2122. doi: 10.1148/rg.2018180109.

[2] Burkhard, C. *et al.* (2003) 'A population-based study of the incidence and survival rates in patients with pilocytic astrocytoma', *Journal of Neurosurgery*, 98 (6), pp.1170–1174. doi: 10.3171/jns.2003.98.6.1170.

[3] NORD (National Organization for Rare Disorders). (n. d.). *Juvenile Pilocytic Astrocytoma*. [online] Available at: https: //rarediseases. org/rare-diseases/juvenile-pilocytic-astrocytoma/.

[4] Peter Collins, V. (2013). *Gliomas*. [online] ScienceDirect. Available at: https: //www.sciencedirect. com/science/article/abs/pii/B9780123749840006525 [Accessed 20 Jun.2023].

[5] Lopes, M. B. and Scheithauer, B. W. (2012) 'Histopathology of brain tumors', *Brain Tumors*, pp.138–187. doi: 10.1016/b978-0-443-06967-3.00009-0.

[6] Ullrich, N. (2016). Neurofibromatosis type 1-associated brain tumors. *Journal of Rare Diseases Research & Treatment*, 1 (2), pp.11–16. doi: https: //doi. org/10.29245/2572-9411/2016/2.1017.

[7] Santino, S. F. *et al.* (2023) 'Pathophysiological evaluation of pilocytic astrocytoma in adults: Histopathological and immunohistochemical analysis', *Pathology-Research and Practice*, 248, p.154593. doi: 10.1016/j. prp.2023.154593.

[8] Harris, L. and Munakomi, S. (2020). *Ventriculitis*. [online] PubMed. Available at: https: //www.ncbi. nlm. nih. gov/books/NBK544332/.

[9] Gordon, M., Ramirez, P., Soriano, A., Palomo, M., Lopez-Ferraz, C., Villarreal, E., Meseguer, S., Gomez, M. D., Folgado, C. and Bonastre, J. (2014). Diagnosing external ventricular drain-related ventriculitis by means of local inflammatory response: soluble triggering receptor expressed on myeloid cells-1. *Critical Care*, 18 (5). doi: https: //doi. org/10.1186/s13054-014-0567-0.

[10] Albano, S., Berman, B., Fischberg, G., Siddiqi, J., Liu, B., Khan, Y., Zafar, A., Quadri, S. A. and Farooqui, M. (2018). Retrospective Analysis of Ventriculitis in External Ventricular Drains. *Neurology Research International*, 2018, pp.1–9. doi: https: //doi. org/10.1155/2018/5179356.

[11] Yung, W. K. A. (2003) 'Glial tumors', *Encyclopedia of the Neurological Sciences*, pp.455–464. doi: 10.1016/b0-12-226870-9/00130-1.

[12] Perry, A. and Wesseling, P. (2016) 'Histologic classification of gliomas', *Handbook of Clinical Neurology*, pp.71–95. doi: 10.1016/b978-0-12-802997-8.00005-0.

[13] El-Ghandour, N. M. (2010) 'Endoscopic third ventriculostomy versus ventriculoperitoneal shunt in the treatment of obstructive hydrocephalus due to posterior fossa tumors in children', *Child's Nervous System*, 27 (1), pp.117–126. doi: 10.1007/s00381-010-1263-2.

[14] Rami, A. Q. (2017) 'Neuroendoscopy sway in the treatment of posterior fossa tumours associated hydrocephalus in children', *Journal of Neurology & Stroke*, 6 (3). doi: 10.15406/jnsk.2017.06.00203.

[15] Lee, M. *et al.* (1994) 'Management of hydrocephalus in children with medulloblastoma: Prognostic factors for shunting', *Pediatric Neurosurgery*, 20 (4), pp.240–247. doi: 10.1159/000120797.