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Non-Communicating Hydrocephalus Caused by Posterior Fossa Tumor in Children: A Case Report

Dewa Ayu Lidya Dwiani[1], I Putu Ananta Wijaya Sabudi[2], Putu Budi Muliawan[3]

[1]General Practicioner, Klungkung Regional General Hospital, Bali, Indonesia
Email: *lidyadwiani95[at]gmail.com*

[2] Department of Neurosurgeon, Klungkung Regional General Hospital, Bali, Indonesia
Email: *ananta.neurosurgeon[at]gmail.com*

[3]Department of Neurology, Klungkung Regional General Hospital, Bali, Indonesia
Email: *p.budimuliawan[at]yahoo.co.id*

Abstract: ***Non-communicating hydrocephalus caused by blockade in the flow of CSF from ventricles to subarachnoid space. Pediatric brain tumors are the leading cause of death from solid tumors in children and majority of them are located in posterior fossa. A 13-year-old boy had a complaint of a prolonged headache since 2 months ago. The headache felt throbbing followed by spinning. Other complaints include vomiting and loss of appetite was found. The patient experienced a similar complaint for the first time. He denied a history of previous head trauma. While in the womb, routine antenatal care is carried out every month by the midwife, fetal screening has never been performed during pregnancy. On the neurological status, cerebellar signs were found, such as dysdiadochokinesia, and balance disorders. Head CT shown dilated ventricles suggestive of hydrocephalus and hyperdense lesions suggestive of a tumor in fossa posterior are visible. Therefore, the patient had been diagnosed with obstructive hydrocephalus caused by posterior fossa tumor suspect medulloblastoma dd/ ependymoma, space occupying lesion, and cerebral edema. Patient had undergone urgent VP shunt MP Keen D and then followed by tumor resection.***

Keywords: Non-communicating hydrocephalus, Obstructive hydrocephalus, Posterior fossa tumours, VP shunt

1. Introduction

Hydrocephalus (HCP) in pediatric population is having high morbidity and mortality. The prevalence of hydrocephalus in pediatric population varies from 30 to 423 per 100,000 pediatric population in various reported studies with prevalence been higher in developing nations in comparison to developed world.[1] The incidence of HCP in Indonesia occurs between 0.2 – 4 out of 1000 births, which is found around 40 – 50% of the entire medical visits or neurosurgery.[2] HCP is the enlargement of the ventricular system of the brain due to accumulation of cerebrospinal fluid (CSF) inside the cerebral ventricles.[3,4] HCP classified hydrocephalus as communicating and non-communicating. Communicating HCP caused by insufficient absorption of CSF in the subarachnoid space and non-communicating or obstruction HCP due to blockade in the flow of CSF from ventricles to subarachnoid space. Obstructive type is further sub-categorized into a congenital and an acquired type.[3] Acquired hydrocephalus in pediatric population is commonly attributed to intracranial infections, intracranial hemorrhage, especially intraventricular hemorrhage (IVH) and subarachnoid hemorrhage (SAH), benign and neoplastic lesions.[5] Pediatric brain tumors are the leading cause of death from solid tumors in children. The most common posterior fossa tumors are pilocytic astrocytoma, diffuse brain stem glioma, medulloblastoma, and ependymomal.[6] Brain tumors in children present in about 50% of cases at the time of tumor diagnosis and persisting up to 10-40% of cases after surgical resection.[7]

2. Case Reports

A 13-year-old boy was taken by his mother to the hospital with the main complaint of a prolonged headache since 2 months ago. The headache felt throbbing followed by spinning . Complaints feel worse during activity and become better with rest. Other complaints include vomiting and loss of appetite was found, no fever , no tingling, no blurred vision and ear problems. The patient experienced a similar complaint for the first time. He denied a history of previous head trauma. In history taking the patient's parents denied a family history of similar complaints and no history of metabolic diseases such as hypertension and diabetes mellitus. Family history of malignancy and history of allergies were also denied. The author tried to find risk factors related to pregnancy history. While in the womb, routine antenatal care is carried out every month by the midwife, fetal screening has never been performed during pregnancy. There were no complaints at each control. The patient's mother took vitamins and blood-boosting medication during pregnancy. There is no history of infection or use of alcohol or illegal drugs during pregnancy. Complaints of fever, vaginal discharge, bleeding during pregnancy were denied. The patient was born by caesarean section at term gestational age with a birth weight of 3200 grams and was said to have cried immediately. History of immunization is said to be complete. On physical examination, vital signs and general status were within normal limits. No macrocephaly was found with a head circumference of 45 cm, no crack pot sign , no sunset phenomenon, fontanel anterior had closed. No sunset phenomenon was found. On the neurological status, cerebellar signs were found, such as dysdiadochokinesia, balance disorders.

On computerized tomography scan of the head, dilated ventricles suggestive of hydrocephalus (Figure 1) and hyperdense lesions suggestive of a tumor in fossa posterior are visible (Figure 2)

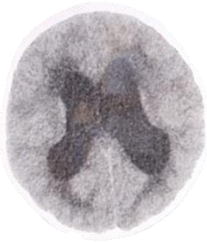

Figure 1: Dilated ventricles suggestive of hydrocephalus

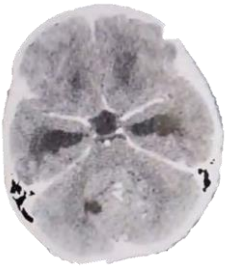

Figure 2: Hyperdense lesions suggestive of a tumor in fossa posterior

3. Discussion

The most common solid tumors in children are central nervous system tumors, and the majority of them are located in the posterior fossa.[8] The most common posterior fossa tumors are pilocytic astrocytoma, diffuse brain stem glioma, medulloblastoma, and ependymoma. Pilocytic astrocytoma are the most common of all with a classic cyst with nodular appearance. Diffuse brainstem glioma usually has epicentres in the pons with the displacement of the basilar artery. Ependymoma is seen to arise from the fourth ventricle. Medulloblastoma has characteristic of showing drop metastasis to the spinal cord through the ventricle system and usually arises from the vermis.[9] Even though this condition is quite common, the optimal treatment algorithm has not yet been determined.[8]

Hydrocephalus can present with many symptoms but often presents in the same pattern. In older children, more common presentations include headaches, visual complaints (blurred or spotted vision), and decreased consciousness. Papilledema is a significant sign in children of any age and may be associated with elevated intracranial. Closure of the fontanelles, along with increased brain volume, makes children much more vulnerable to acute malfunctions with treated hydrocephalus and new presentations of undiagnosed decompensated hydrocephalus. The presentation of hydrocephalus in this population also differs due to the age of the patient.[10]. Patients with posterior fossa tumors may have symptoms depending on location, size, and subsequent increase in intracranial pressure (ICP) due to hydrocephalus. Progressive headache is the most common symptom, followed by nausea, vomiting, and mental changes as symptoms worsening with increasing severity of hydrocephalus. Cerebellar involvement often leads to specific symptoms in the posterior fossa, such as gait ataxia, horizontal nystagmus, tremor, and visual disturbances.[11] Focal neurologic deficits such as bilateral palsies of sixth cranial nerves, which manifest as the inability of the eyes to abduct. Certain clinical signs are considered manifestations of late-stage presentation of hydrocephalus and usually require more urgent intervention. The two most common late-stage presentations are Parinaud syndrome (dorsal midbrain syndrome) and new-onset seizures. Parinaud syndrome is characterized by upgaze palsy; pseudo-Argyll Robertson pupils or pupillary light-near dissociation in which the pupils are able to accommodate but unable to react to light; convergence-retraction nystagmus, in which upon attempted upward gaze the eyes converge and are pulled into the orbit; and abnormal eyelid retraction (Collier sign).[10]

The results of patient's Head CT scan with and without contrast shown masses in fourth ventricle causing constriction of the sulci and enlargement of ventricular system along with contrast enchancement giving impression of medulloblastoma dd/ ependymoma, and brain edema with non-communicating hydrocephalus. Therefore, patient had been diagnosed with obstructive hydrocephalus caused by posterior fossa tumor suspect medulloblastoma dd/ ependymoma, space occupying lesion, and cerebral edema. Medulloblastoma is a highly malignant neoplasm and also the most common malignant brain tumor in children, representing 15% to 20% of all pediatric brain tumors and 30% to 40% of posterior fossa neoplasms. Medulloblastoma mostly found in males, usually before 10 years with a bimodal peak of incidence at 3 years and 9 years of age.[6,9,12] The tumor usually appears in the midline of the vermis and grows into the fourth ventricle. Less typical locations include non-ventricular superior or inferior vermian tumor, cerebellar hemispheric lesions, and extension into the foramina of Magendie and foramina of Luschka to the cerebellopontine angle (CPA). On non-contrast CT, the tumor is often characterized as hyperdense lessions. The tumor typically may appear heterogeneous on imaging, with findings related to cyst and haemorrhage on MR, calcification may also be seen on CT, and shown an enchancement on contrast. Ependymomas are characterized by perivascular arrangement of tumor cells. These tumors are most often seen in children younger than 5 years of age. Ependymomas mostly in the posterior fossa with around 70% of cases, with 90% involving the ventricular ependyma. Ependymomas may also spread through the foramina of Luschka and Magendie. Punctate calcification is demonstrated in 50% of ependymoma cases on CT. Calcification is most often seen in ependymomas than in any other posterior fossa tumor in children. The most important imaging finding in identifying an ependymoma is extension of the tumor through the fourth ventricular outflow foramina.[6]

Children with posterior fossa tumors have hydrocephalus in 60–91% of cases at the time of diagnosis. In most cases of posterior fossa tumors, hydrocephalus is caused by obstruction of cerebrospinal fluid (CSF) flow at the level of the cerebral aqueduct or the fourth ventricle. In these areas, the flow of CSF is stopped, and resulting in triventricular hydrocephalus. Attributing to this pathophysiological mechanism, Morelli et al. in their series of 114 children with hydrocephalus associated with posterior fossa tumor found

that early total tumor removal resolved hydrocephalus in 91% of patients.[8]

Patient had undergone urgent VP shunt MP Keen D and then followed by tumor resection. In the past, most children were diagnosed too late and therefore were usually admitted to hospital in poor neurological state. Neurosurgeons agreed that hydrocephalus should be treated immediately with shunting and only after the child is stable and recovered to continue with the tumor resection. At the time, placement of a shunt before tumor resection was found to significantly reduce morbidity and mortality. However, this also led to shunt complications, such as shunt dysfunction, infection, intraperitoneal tumor seeding, and multiple shunt revisions in a growing child. Nowadays, there are several treatment options for hydrocephalus associated with posterior fossa tumors such as preresection CSF shunting, preresection ETV, temporary EVD placement, postresectional ETV, and postresectional CSF shunting. CSF shunting was an earlier standard approach in treatment of hydrocephalus associated with posterior fossa tumors in children wuth relatively common grave neurological status leading to rapid improvement of conditions. It was also allowed more affordable access and easier tumor resection. This method also decreased a risk of postoperative pseudomeningocele formation and subsequently potential CSF leakage and infection. However, this treatment wasn't ideal. Some authors observed that in preoperatively shunted patients, tumor was displaced closer to the brain stem and made resection more difficult.[8,13]

Lin and Riva-Cambrin in 2015 reported only 10–40% of patients have persistent hydrocephalus after tumor resection. Tumor location in the midline was revealed as a risk factor for development of persistent hydrocephalus in several studies. Culley et al. showed that 40% of patients with midline tumors in their series required shunt placement, in contrast to not one of the hemispheric tumors in their series.[7] Medulloblastoma patients with significant extension into the fourth ventricle defined by Chang stage T3 (tumor >3 cm in diameter with extension into the brain stem) or T4 (tumor >3 cm in diameter with extension up past the aqueduct of Sylvius and/or down past the foramen magnum) had a statistically higher rate of eventual shunt placement.[8]

4. Conclusions

Non-communicating hydrocephalus caused by blockade in the flow of CSF from ventricles to subarachnoid space, which further classified into acquired and congenital. Acquired hydrocephalus in pediatric population is commonly attributed to intracranial infections, intracranial hemorrhage, especially intraventricular hemorrhage (IVH) and subarachnoid hemorrhage (SAH), benign and neoplastic lesions. Pediatric brain tumors are the leading cause of death from solid tumors in children and majority of them are located in posterior fossa. The most common posterior fossa tumors are pilocytic astrocytoma, diffuse brain stem glioma, medulloblastoma, and ependymoma. Children with posterior fossa tumors have hydrocephalus in 60–91% of cases at the time of diagnosis. Most children were diagnosed too late and therefore were usually admitted to hospital in poor neurological state. Neurosurgeons agreed that hydrocephalus should be treated immediately with shunting and only after the child is stable and recovered to continue with the tumor resection.

Ethical Approval
The patient has signed informed consent and agreed for the publication of their data and related imaging as a case report article.

Conflict of Interest
There are no conflict of interest to declare by any of the author of this study.

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Author Profile

Lidya Dwiani received her bachelor's degree of medicine in 2019 from Warmadewa University and she currently working and interning an internship in the Neurology Department of Klungkung Regional Hospital, Bali, Indonesia