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Case Report

Pilocytic astrocytoma: A rare case report ☆

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ABSTRACT

Pilocytic astrocytoma (PA) is a rare, low-grade glioma predominantly affecting children and young adults, with a favorable prognosis and a high survival rate. Despite its' slow growth, PA often presents with significant symptoms at diagnosis due to its tendency to reach a large size, especially in pediatric patients, where compensatory mechanisms can delay detection. This case report describes a 7-year-old girl with progressive vision loss, headache, and balance disturbance. The imaging result revealed a right cerebellar mass that was in accordance with pilocytic astrocytoma and obstructive hydrocephalus. Histopathologic analysis confirmed the diagnosis with the typical features of PA such as Rosenthal fibers and eosinophilic granular bodies. This case report highlights the critical role of radiologic and histopathologic evaluations for PA diagnosis and management; also the unique presentation and challenges in managing PA, emphasizing the importance of timely intervention to optimize neurological outcomes.

Introduction

Pilocytic astrocytoma (PA) is a histologic and biologic subset of glioma that only accounts for 15% of central nervous system (CNS) tumors [1]. Pilocytic astrocytoma is commonly found in children and young adults; it has a good prognosis with a 10-year survival rate exceeding 95% [2,3]. The age-adjusted incidence of PA is 0.37 per 100,000 people annually, and it only accounts for 0.8% of cases of CNS tumors in young adults [4,5].

Surgical removal is the cornerstone of PA management. Adjuvant therapies such as radiotherapy and chemotherapy should be considered in recurrent tumor and progressive tumors that involves the cerebellar peduncle or brainstem. Patients may have permanent cerebellar dysfunction and cognitive disturbance after surgery although it is the main

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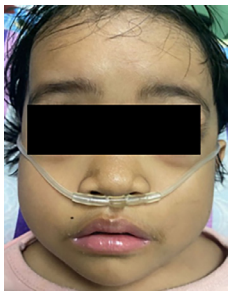

Fig. 1 – Patient's clinical condition.

treatment. There are some ongoing trials to develop the latest PA therapy modalities [5–7]. Pilocytic astrocytoma is commonly diagnosed at a large size due to its slow glowing rate and the ability of the pediatric brain to compensate. Radiologic examinations play important roles in PA diagnosis [7]. We would like to present a case about a 7-year-old girl with pilocytic astrocytoma.

Case report

A 7-year-old girl presented with loss of vision 1 month prior to admission. Initially, she had a headache 6 months prior to admission, which was continuous and not exacerbated by activities. She had blurred vision 4 months prior to admission and postural imbalance 3 months prior to admission. Other symptoms, such as weight loss, seizure, and vomiting were denied. She had a complete immunization history. The history of lumps on the different body parts and developmental disorders was dismissed. She was an elementary school student and had normal activities.

She was moderately ill and compos mentis. The clinical examination was within normal limits except for her anthropometric status. Her body weight and height were 19 kg and 108 cm, respectively. Her nutritional status was normal based on body mass index (BMI)-for-age curve and stunted based on body height-for-age curve. Her visual acuity was no light perception. The funduscopy revealed bilateral papillary atrophy. She had bilateral cranial nerve (CN) III, IV, and VI palsy based on the cranial nerve examination (Fig. 1).

The laboratory examination revealed leukocytosis (10,230 cells/mm^3). The cerebrospinal fluid (CSF) analysis revealed increased glucose and protein levels of 91 mmol/L and 55 mmol/L, respectively. The head computed tomography (CT) scan with contrast showed an inhomogeneous solid mass originating from the right cerebellum, which compressed the fourth ventricle and right cerebellum, suggesting pilocytic astrocytoma with differential diagnoses of sub-ependymoma and posterior fossa ependymoma and obstructive hydrocephalus with trans-ependymal edema (Fig. 2).

Based on the history taking, physical examinations, laboratory examinations, and radiologic examinations, her diagnosis was noncommunicating hydrocephalus due to infratentorial space occupying lesion (SOL) on the right cerebellum, with a suspicion of pilocytic astrocytoma. A surgical resection by craniotomy was planned. The intraoperative finding was a white-brownish soft tumor with an indistinct margin and easily bled. The histopathologic examination revealed pilocytic astrocytoma (Fig. 3).

About 1 week after the craniotomy, she had *ventriculoperitoneal shunt* insertion with a *high-pressure infantile hpbio shunt*. The cerebrospinal fluid was used for further analysis. The cerebrospinal fluid analysis revealed improved glucose and protein levels of 69 mmol/L and 20 mmol/L.

Discussion

Pilocytic astrocytoma is a rare, slow-growing glioma classified as a grade I tumor by the World Health Organization (WHO) [8]. The term "pilocytic" refers to hair-like bipolar cells and has been used to describe astrocytoma variants since the 1930s [9]. The estimated incidence of PA is around 0.839 per 100,000 and begins to decline between the ages of 10 and 14 years. Pilocytic astrocytoma predominantly occurs in individuals aged 5 to 14 years, with a peak incidence between 6 and 8 years, and is somewhat more prevalent in males [7,9–11]. A study showed the male-to-female ratio of PA is 1.1:1 in pediatric cases and 1.8:1 in adult cases [12]. Pediatric PA has a good prognosis with a 5-year survival rate ranging from 80% to 95% and a 10-year survival rate of around 90%. The prognosis of pediatric PA is better than adult PA [3,8]. Our patient was a 7-year-old girl, and her age was included in the most common age group in which PA cases are commonly found.

The most common sites of PA are cerebellum and regions around the third ventricle (42%). The whole neuroaxis might be affected by PA, particularly the optic nerve, chiasm optic, hypothalamus, cerebellum, brainstem, thalamus, basal ganglia, and cerebral hemisphere [8,9,13]. The tumorigenesis of PA is poorly understood. Pilocytic astrocytoma does not have genetic changes such as tumor protein 53 (TP53) mutation, O^6-methylguanine-deoxyribonucleic acid (DNA) methyltransferase (MGMT) methylation, endothelial growth factor receptor (EGFR) amplification, and phosphatase and tensin homolog (PTEN) like in high-grade glioma. The consistent pattern of 1p or 19q loss of heterozygosity (LOH) has not been found and it does not correlate with patients' prognoses. Pediatric PAs may occur *in utero*, and cerebellar PAs reflect the transcriptome of specific embryonic cerebellar cluster cells. Immune cells may have a role in ensuring PA is under control, and hormonal factors, inflammation stimulus, other microenvironment components, and certain events can increase tumor growth at certain time points [7,14]. Genetic alterations involving the mitogen-activated protein kinase (MAPK) signaling pathway are considered a hallmark of PA [12].

The symptoms of PA are insidious due to its slow glowing rate, and early symptom identification depends on the localization and patients' ability to communicate the neurological changes and the discomfort. Most PA cases are diagnosed at a large size [7,9]. The symptoms appear due to the compressed adjacent tissue or increased intracranial pressure [15,16]. Headache is the most common symptom (90% of cases). Children usually complain that the headaches are worse during or just after sleeping. The headache occurs be-

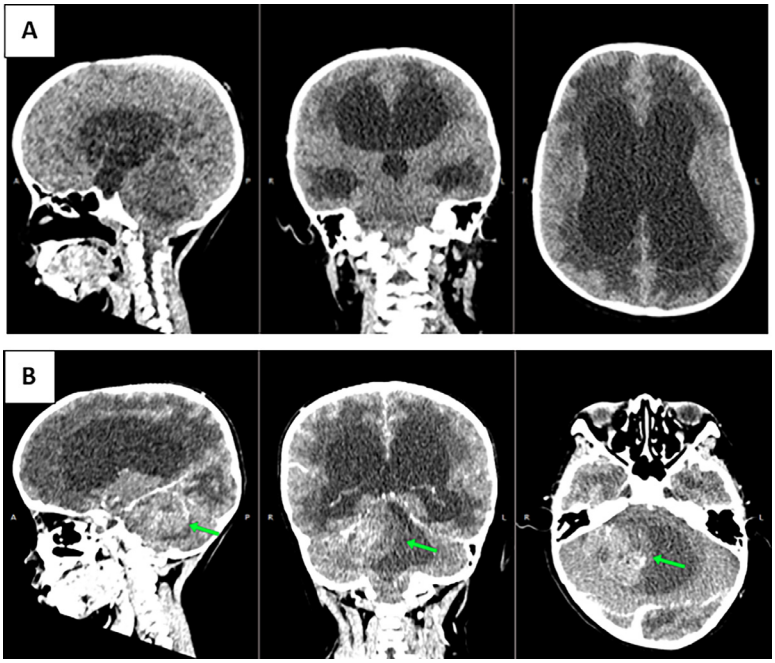

Fig. 2 – The head CT-scan. (A) Axial, coronal, and sagittal noncontrast head CT scans). The cortical sulci, gyri, bilateral sylvian fissure, and interhemispheric fissure were compressed. There were asymmetry and blunted bilateral lateral ventricles. and (B) CT scan of the head with contrast axial, coronal, and sagittal sections). There was a hypodense periventricular lesion on the anterior and posterior *cornu* of the bilateral lateral ventricles. There was a well-circumscribed isodense mass with inner hypodense component and a regular edge originating from the cerebellum measuring 3.60 x 4.55 cm x 4.43 cm, compressing the fourth ventricle and right cerebellum with postcontrast rim enhancement. The basal cisterns and ambience were dilated.

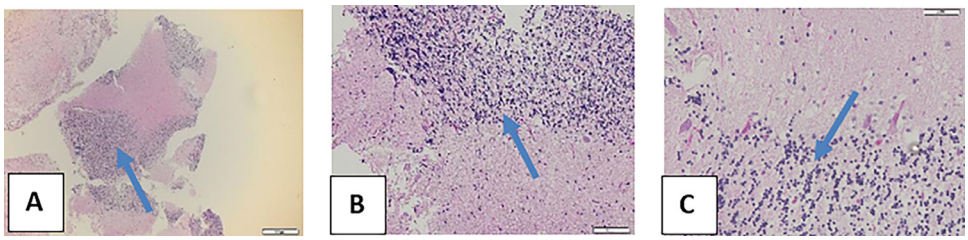

Fig. 3 – The histopathologic examination. (A) 20x magnification, (B) 100x magnification and (C) 200x magnification. (The histopathologic examination revealed loose fibrillar stroma, which partially condensed with round–oval disseminated hyperplastic cells; no elongated hyperchromatic round nuclei and mitosis were found; there were Rosenthal fibers and eosinophilic granular bodies; and ependymal and Purkinje cells of the cerebellum were partially normal).

cause of increased intracranial pressure due to obstructed cerebrospinal fluid circulation in the aqueduct of Sylvius or the fourth ventricle. Vomiting and lethargy are also commonly found in worsened hydrocephalus. The symptoms of cerebellum PA resemble the posterior fossa localization symptoms such as ataxia, horizontal nystagmus, and dysmetria. Cranial nerve defects, particularly cranial nerves III and VI, dysdiadokokinesia, dysarthria, tremors, and signs of increased intracranial pressure can also be found. Patients may experience a visual field defect or loss of sharpness if the tumor is in the optic tract [7,9,15,17].

In this case, our patient initially had hazy vision and a balance disruption before subsequent headache and vision loss. It is consistent with the research in which headaches are the most prevalent symptom. Visual and balance disturbances occur because the tumor is located in the cerebellum. The physical examination revealed cranial nerve III, IV, and VI palsies. The literature also stated that cranial nerve III and VI palsies are the most common cranial nerve palsies in PA.

The preoperative diagnosis of PA is established based on the combination of morphologic and nonmorphologic radiologic findings with a clinical approach based on tumor location. Pilocytic astrocytoma usually has an atypical appearance or symptoms because it is a low-grade glioma [16]. Hydrocephalus is the most common radiologic findings (90% of cases) [7]. The radiological images are evaluated based on the

degree of contrast enhancement, calcification, morphological appearance, size, and tumor distribution. The CT scan result usually revealed well-circumscribed round or oval isodense or slightly hypodense with postcontrast enhancement. The high-grade tumor will reveal heterogeneous densities with diffuse enhancement. Magnetic resonance imaging will show hypointense or isointense (T1 sequence) and hyperintense (T2 or FLAIR sequence) with clear or diffuse enhancement. The masses may have cysts or "Tumor nodule in a cyst" appearance (particularly cerebellar and hemisphere tumors). Pilocytic astrocytoma in the optic tract, optic nerve, and optic chiasm may form fusiform masses. Calcification can be found in radiology examinations. Perilesion edema is rarely found due to its slow-growing rate [9,18,19]. Radiologic examinations can also be used for surveillance in patients who underwent surgery, particularly if the tumor is partially resected, but it is still uncertain [20].

The histopathologic findings of PA are a biphasic growth pattern with fibrillar and microcystic regions, along with periodic acid-Schiff (PAS)-stained eosinophilic granular bodies and elongated eosinophilic Rosenthal fibers. The appearance of glomeruloid vessels may be found due to increased vascularization. Oligodendrocyte-like cells can be found in some cases [21].

The radiological findings in this case are in line with the literature. The head CT scan findings were an isodense mass with an inner hypodense component and postcontrast enhancement that occurred due to increased tumor vascularization [21]. The radiological examination also revealed hydrocephalus, 1 of the most common radiological findings in PA [7]. The histopathologic examination findings were also in line with fibrillar stroma with Rosenthal fiber and eosinophilic granular bodies.

Surgery is the main treatment of PA to achieve gross total resection (GTR) is possible without any neurologic deficits. The GTR rate ranged between 50% and 89%. The solid component should be resected for cystic lesions without tumors in the wall of the cyst. The resection surgery should be repeated if there is a part of tumor left behind. Chemotherapy is applied for certain cases in which the tumor progresses and the surgery is not feasible. The combination of chemotherapy and radiotherapy can be considered if the surgery is not feasible or in the recurrent cases. Adjuvant radiotherapy is commonly avoided in low-grade cerebellar astrocytoma cases (particularly in children) because it can affect neurocognitive function. Hydrocephalus management with *external ventricular drain* (EVD) insertion, *ventriculoperitoneal shunt* (VP *shunt*) insertion, or ventriculostomy can be considered because hydrocephalus is common in PA even after tumor resection. Several studies focused on molecular and cell biology to predict the outcomes and create novel therapies [7,9,18].

Conclusion

Pilocytic astrocytoma (PA) is a rare, slow-growing glioma but quite common in children, particularly in the age group of 5 – 14 years. It is associated with a good prognosis. It has atypical symptoms and was commonly diagnosed at a large size. Therefore, radiologic examinations such as a CT scan or MRI is essential to make the diagnosis supported by histopathologic examination.

Patient consent

Written informed consent for publication of their case was obtained from our patient's parent.

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