Case Report:
Torticollis as a Rare Presentation of Cerebellopontine Angle Choroid Plexus Papilloma in Children

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ABSTRACT

Background and Importance: The rare intracranial neoplasms are Choroid Plexus Papillomas (CPPs), especially in the cerebellopontine angle. The main location of choroid plexus papillomas in adults and children are 4th ventricle and lateral ventricles, respectively.

Case Presentation: We report on a little girl with a cerebellopontine angle CPP who presented with symptoms of torticollis. Assessment of Magnetic Resonance Imaging showed a mass in the right cerebellopontine angle, next to the brain stem. The tumor was completely resected using the right retrosigmoid approach method. A pathological examination determine a typical CPP that this being should be considered an extremely rare cause of a lesion in the posterior fossa.

Conclusion: CPP is usually presented in the atrium of the lateral ventricle in children; however, we reported a rare case in the cerebellopontine angle.

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Highlights

- CPP usually affects in the atrium of the lateral ventricle in children.
- Rarely, CPP may present in the cerebellopontine angle.
- The prognosis of CPP is good, with a 90% survival rate.

Plain Language Summary

A type of tumor named Choroid Plexus Papillomas (CPPs) originates from the lining of tissue in the brain. In fact, these tumors are usually located in the 4th ventricle and in the lateral ventricles in adults and children. However, we reported a rare case in a 2-year-old boy that was presented with the cerebellopontine angle of the brain. This case report is important to provide a better understanding of the tumor presentation in this part and the subsequent management.

1. Background and Importance

The rare intraventricular tumors are choroid plexus tumors that comprise less than 1% of all intracranial tumors and 2%-4% of brain tumors in children [1, 2]. They are most commonly located in the lateral ventricles of children and infants and the 4th ventricle in adults [2, 3]. The cerebellopontine angle is a rare location for the CPP and only a limited number of Choroid Plexus Papillomas (CPPs) have been reported at this location [4-6]. The surgery of this tumor is challenging as patients are typically young, overproduction of Cerebrospinal Fluid (CSF) after surgery, and extreme vascularity of the tumor, which can lead to significant blood loss during surgery and incomplete resection of these tumors [7-9]. Here, we reported on a case of cerebellopontine angle choroid plexus papilloma which was resected by the retrosigmoid approach.

2. Case Presentation

We hospitalized a 2-year-old female patient who presented with torticollis for 15 days. On physical examination, the child was conscious, oriented, and without any neurological deficit. Magnetic Resonance Imaging (MRI) demonstrated a cystic mass without enhancement and restriction in the right posterior fossa next to the brain stem with pressure effect on the fourth ventricle (Figure 1). Based on the radiographic appearances of the lesion, a definite diagnosis of choroid plexus papilloma was reported.

At surgery, a retrosigmoid craniotomy was performed and the tumor was approached through a retrosigmoid approach. A cystic mass was observed in the right cerebellopontine angle next to the brain stem. The tumor was cystic and the gross total resection of the tumor was performed. Fortunately, the patient tolerated the procedure well during the operation. Post-operatively, neurological deficits were observed in the patient. Histopathology was consistent with the diagnosis of choroid plexus papillomas (Figure 2).

3. Discussion

Choroid plexus papilloma is one of the common neoplasms. It occurrence mainly within the ventricular system and involvement of the cerebellopontine angle is rare. During childhood, 80% of CPPs are found in the lateral ventricles, 16% in the 4th ventricle, and 4% in the 3rd ventricle. These tumors are generally more common in males than females [3, 10, 11].

Choroid plexus papillomas are generally classified as benign neoplasms, defined by World Health Organization (WHO) as Grade I and Grade II. Resection of these tumors usually cures the disease and recurrence after gross total resection is rare. As these tumors are highly vascular, they involve young people and may cause post-operative complications such as CSF overproduction, their management is usually challenging [2, 7, 12]. These lesions are highly vascular with several anastomosis of choroidal blood vessels and may affect interventricular space. Thus, adjuvant endovascular embolization can reduce blood loss [9].

Several metastases of Cerebellopontine Angle (CPA) is also probable in this region that are mainly from the neuro-ectodermal origin. Cancers that are reported in the literature to have metastasis to the CPA include

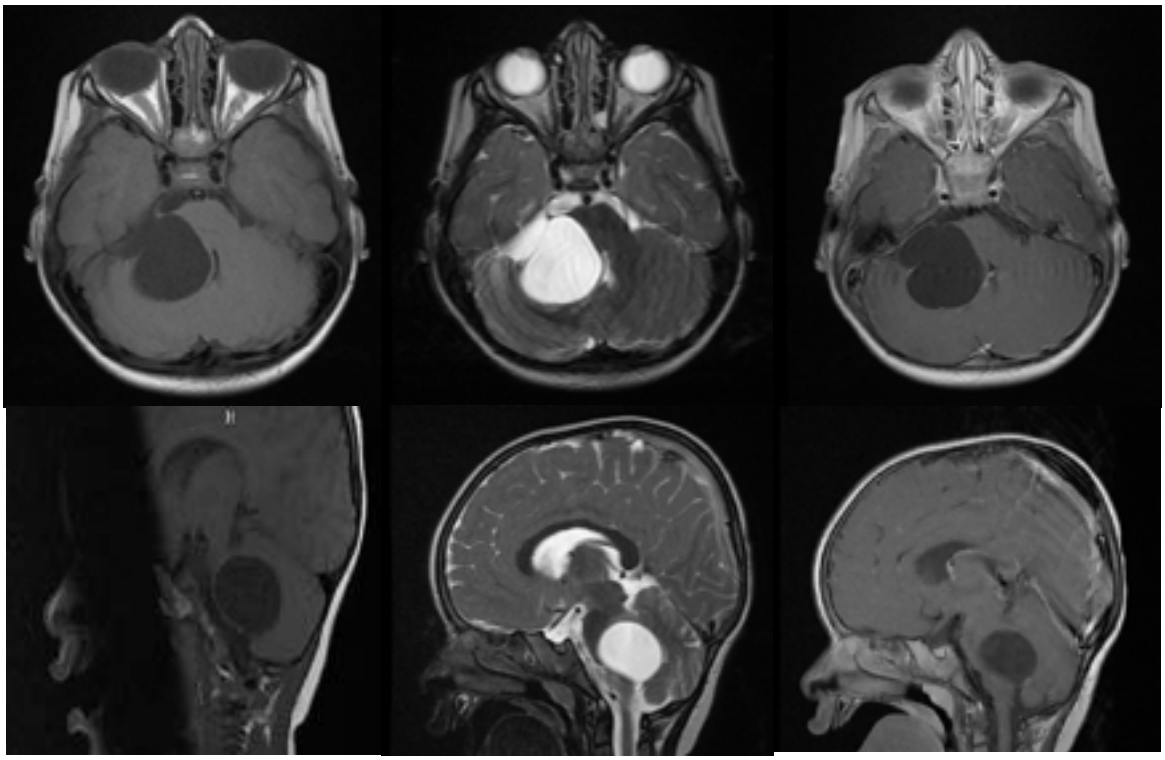

Figure 1. Pre-operative T1, T2, and Gad-MRI showing the tumor without enhancement in the right posterior fossa next to the brain stem with pressure effect on the 4th ventricle

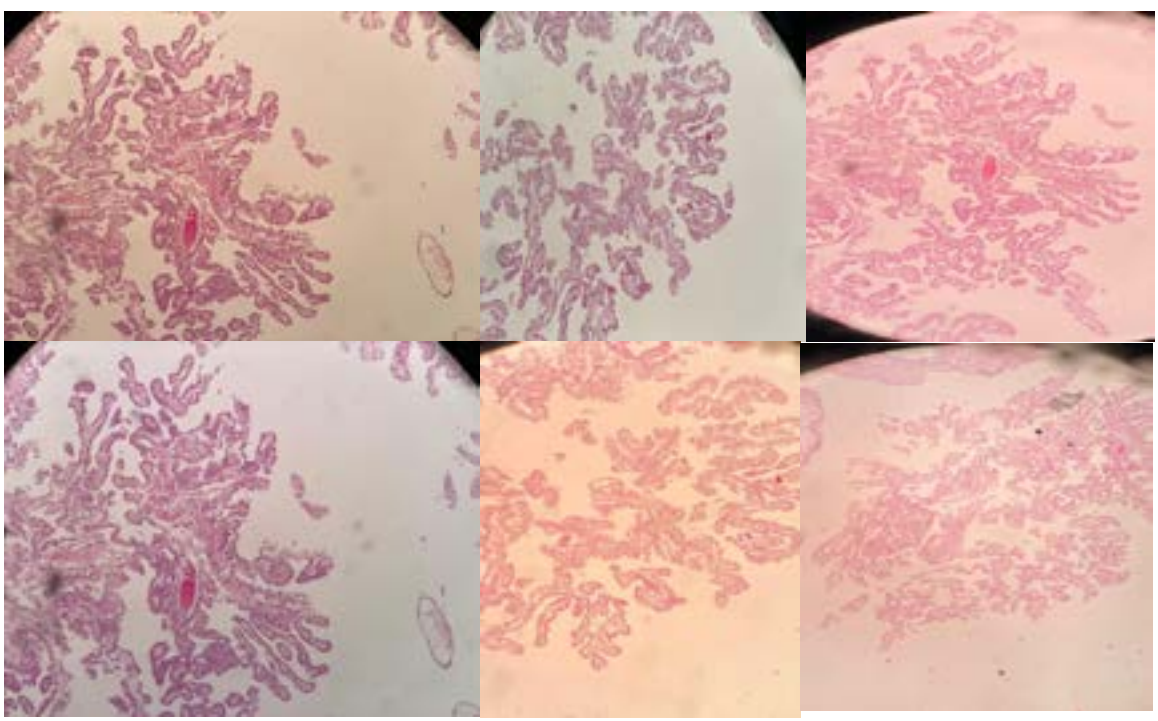

Figure 2. Choroid plexus papilloma showing a single layer of orderly cuboidal to columnar epithelial cells stays on distinct fibrovascular cores. Inset shows a hyalinized blood vessel

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prostate, breast, lung, ocular melanoma, and nasopharynx cancer. Moreover, cases of hematologic cancer with metastasis to CPA also are reported. Therefore, these should be considered in the differential diagnosis of a mass in this region [1].

The treatment of CPP is the surgical excision for the resection of these tumors. The complete tumor should be removed and is usually accessible in as many as 96% of cases. Most of the challenges that neurosurgeons face regarding the management of this type of tumor are related to the heavy blood loss in children, who have a proportionally small circulating blood volume. Although mortalities due to all causes comprise between 0 to 25%, up to 12% of mortalities are reported to be caused by blood loss [2, 7, 12, 13].

Various surgical approaches have been described for cerebellopontine angle tumors. We usually prefer a retrosigmoid approach to a CP angle tumor. The effect of gravity in sitting position and deep venous structures which lie dorsal to the tumor are advantages of using this approach. Yet, the narrow operative corridor which can cause problems in resecting large vascular tumors is disadvantageous. Prevention of bleeding is a major consideration during surgical resection of these lesions in pediatric patients, thus a technique of gentle, repetitive bipolar coagulation of the tumor surface under permanent irrigation (to shrink the tumor and remove it totally) is preferred to its piecemeal resection. Gross total resection of the tumor brings a long-term survival of 90 to 100%. Those malignant tumors or those with extension to leptomeningeal need adjuvant therapy [7, 12].

4. Conclusion

We presented a 2-year-old girl with CPP who presented with symptoms of torticollis. The imaging determined a mass in the right cerebellopontine angle next to the brain stem. We often surgically cure choroid plexus papillomas. The most common location of CPP is in the atrium of the lateral ventricle in children. CPPs have a nearly 90% survival after tumor resection. Although, we reported a rare case of CPP in the cerebellopontine angle.

Ethical Considerations

Compliance with ethical guidelines

The participants were informed of the purpose of the research and its implementation stages. A written consent has been obtained from the subjects. They were also assured about the confidentiality of their information and were free to leave the study whenever they wished, and if desired, the research results would be available to them.

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Authors contributions

Conception and design: Mohammad Faraji-Rad; Data collection: Mohammad Faraji-Rad; Analysis and interpretation: Einoallah Alipour; Drafting the article: Mohammad Faraji-Rad; Critically revising the article: Einoallah Alipour; Reviewing submitted version of manuscript: Einoallah Alipour; Approving the final version of the manuscript: Elnaz Farajirad.

Conflict of interest

The authors have no conflicts of interest in the present study.

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