Choroid plexus papilloma presenting as an occipital mass with neck pain

Case report

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Abstract

Rationale: Choroid plexus papillomas are rare benign central nervous system neoplasms arising from choroid plexus epithelium. They are most often located in the lateral ventricle, followed by the fourth and third ventricles and, rarely, in the cerebellopontine angle.

Patient concerns: We report an uncommon case of a 17-year-old boy who presented with neck pain that had lasted for more than 1 month, with accompanying pain and numbness in his upper extremities. His conditions included slight dizziness, nausea, diplopia, paresthesia, and an unsteady gait. Magnetic resonance imaging (MRI) showed huge cerebellopontine angle tumor that extended to the front medulla oblongata.

Diagnosis: Choroid plexus papilloma (WHO I) was diagnosed in this patient.

Interventions: The patient was referred for neurosurgical intervention. The very large neoplasm was subtotally resected.

Outcomes: The symptoms of the patient were gradually alleviated after surgery and subsequent radiotherapy treatment, but unfortunately, follow-up of 2 years later revealed that the disease was recurrent and the young man passed away.

Lessons: Neck pain is related to many factors. The case provided an awareness of the origin of severe intracranial disease. It is mandatory to take a thorough clinical assessment with a holistic approach.

Abbreviations: AICA = anterior inferior cerebellar artery, CPA = cerebellopontine angle, CPP = choroid plexus papilloma, CT = computed tomography, EMG = electromyography, MRI = magnetic resonance imaging, NSAIDs = nonsteroidal anti-inflammatory drugs, PWI = perfusion-weighted imaging, TENS = transcutaneous electrical nerve stimulation, VAS = visual analogue scale, WHO = World Health Organization.

Keywords: case report, cerebellopontine angle, choroid plexus papilloma, neck pain, pediatric

1. Introduction

Choroid plexus papillomas (CPPs) are primary neuroectodermal tumors of the choroid plexus and rarely located in the cerebellopontine angle (CPA).[3] They are usually not malignant and occur in less than 1% of brain tumors in patients of all ages, but more commonly found in pediatric patients.[3] CPPs predominantly arise in supratentorial locations in children, while the most common sites in adults were the fourth ventricle and the CPA.[3][4] On the basis of a series of literature reviews, more common clinical presentations of CPA-CPPs include headache and eighth cranial nerve deficits and unsteady gait.[3][6] The symptoms and signs are usually latent and only slowly progressive. We report an uncommon case of young boy involved in CPA-CPPs with neck pain. To the best of our knowledge, neck pain as a primary complaint is not typical in CPA-CPPs disease and the pathogenesis is still not clear. This condition needs further examination and discussion, as it can easily be confused with cervical spondylosis or cervical radiculopathy that has not previously been reported. Different diagnosis leads to different treatment schemes, and so, favorable outcomes and the avoidance of potential risks are possible through more definitive diagnosis.

2. Case report

The case is based on a 17-year-old boy with neck pain that had lasted for more than 1 month, with numbness in both upper extremities that started 1 week earlier. He complained that symptoms were aggravated after playing computer games for a
longer time than normal. This was accompanied by slight dizziness, nausea, diplopia, paresthesia, and an unsteady gait in supplementary inquiries. In addition, his gastroscopic examination showed gastric ulcer and bile regurgitation in previous medical records. The positive signs of vigilance revealed a stiff neck, limited abduction of left eye, and a positive Romberg test in the patient’s physical examination with no other abnormal symptoms.

A plain film radiograph of cervical vertebrae revealed nonspecific curve and intervertebral space and foramen. In consideration of medical history and alerted signs, the patient was immediately arranged for a craniocerebral magnetic resonance imaging (MRI) examination on the second day of admission. MRI showed huge CPA tumor that extended to the front medulla oblongata (Fig. 1 A–C). The patient was referred for neurosurgical intervention. In surgery, the tumor was presented in the lower left CPA and left side of the medulla oblongata. There was ample blood supply and it had a mass size of $3 \times 4 \times 3 \text{ cm}^3$. The fourth ventricle and left foramen of Luschka were compressed by the tumor that was adhesive to the lower cranial nerves and closely adjacent to accessory nerve and the anterior inferior cerebellar artery (AICA). The large neoplasm was subtotally resected and the tissue adhesive to brain stem was not forcibly removed (Fig. 1 E,F). The pathological diagnosis was CPP (WHO I) (Fig. 1 D). The clinical symptoms of the patient gradually alleviated after surgery and subsequent radiotherapy treatment. Unfortunately, in a follow-up 2 years later, the tumor was recurrent. Considering his overall health and tumor condition, the young man could not receive another intracranial surgery nor was he able to receive chemotherapy or radiotherapy. He then passed away.

3. Discussion

CPPs are of neuroectodermal origin, arising from choroid plexus epithelium. They account for approximately 0.4% to 0.6% of all intracranial tumors, 43% arise in the lateral ventricle on one side, 39% arise in the fourth ventricle, 10% arise in the third ventricle, and 9% arise in the CPA. CPPs occur in the CPA are uncommon and almost found in adults. The tumor of CPA-CPPs was found in young boy in this case, which was extremely rare. CPPs are low-grade tumors classified as grade I in the World Health Organization (WHO) grading scheme. By incorporating genetic analysis, the current histologic grading scheme could be refined to create a more accurate, clinically relevant system for categorizing choroid plexus tumors.

Dysfunction of hearing, headache, dizziness, vomiting, gait ataxia, dysphagia, papilledema, and hydrocephalus are frequently reported symptoms and signs of CPA-CPPs. The clinical manifestations of CPA-CPPs are nonspecific and correlated with increased intracranial pressure and the location of tumor and the nerves or cerebral structures involved with the lesion. Neck pain as a primary complain shown in this case was not typical in CPA-CPP disease and the pathogenesis was still not clear that was not previously reported. More attention should be paid to the antecedents and consequences of CPPs on the basis of logic.
analysis of presenting symptoms and following symptoms and neurological evaluations in clinical practices.

Clinical diagnosis of primary CPA-CPPs is difficult due to both the rarity of this presentation and to nonspecific radiological features that cannot be differentiated easily from other tumors preoperatively. The differential diagnosis commonly used include ependymoma, metastasis, acoustic neuroma, meningioma, vestibular schwannomas, and aneurysm. Cerebral angiogram, MR spectroscopy (MRS), and perfusion-weighted imaging (PWI) may provide more information about CPA-CPPs and play an important role in distinguishing between different CPA lesions to improve the diagnostic power..

Immunohistochemical staining of postoperative pathologic findings would facilitate clear diagnosis. Surgical resection is an important element of treatment modality for CPPs. It is recommended that as much of the tumor as possible should be removed, which was associated with a significant increase in both progression-free survival and overall survival. Literature review support a “wait and see” approach after first tumor resection. The prognosis of patients with CPPs is relatively good, which is related with improvement of surgical and intensive care techniques. For patients with incomplete resection, local radiotherapy could reduce the recurrence rate and prolong the survival time. But the efficacy of irradiation is still uncertain and consensus has not yet been reached. Relapse and malignant transition could occur and long-term follow-up is necessary.

Neck disorder been was recently identified as a significant problem and a common health issue in many countries, with a 1-year prevalence ranging from 16.7% to 75.1% for the entire adult population. Latest studies had shown that the occurrence of neck and shoulder pain was relatively common among adolescents, related to sedentary behavior, lower physical activities, digital device (computer games and social media) overuse, academic burdens of schooling, and psychological pressure. Moreover, the occurrence of neck and shoulder pain in adolescence increases the risk of musculoskeletal disorders in future adults.

The primary symptoms of cervical spondylosis are neck pain, movement disorder, and upper extremities paresthesia. The patient in the case had such symptoms after playing computer games for a longer than normal time, which may have caused a greater load to the neck and proprioception impairment. He was in a high-risk population of neck pain. To a certain extent, the manifestation of the patient was mimicking or similar with cervical spondylosis radiculopathy. But we found some doubtful points in physical examination, such as dizziness, diplopia, neck stiffness, unstable gait, nausea, and among others. The digestive tract symptoms was not relieved after taking oral proton pump inhibitor medicine prescribed in a visit to a previous hospital. We speculated the possibility of posterior cranial fossa disease and were astonished with the radiographic images after brain MRI scan. The exact pathogenesis of neck pain was unknown and probably correlated with the adhesive lower cranial nerves forced, neck muscle continuous tension, the irritation of posterior rami of spinal nerves, and nociceptive receptors of intracranial and extracranial structure that needs further exploration. It is possible that bad posture was a precipitating factor of the onset of neck pain.

It is important to make an explicit diagnosis of cervical spondylosis based on the differentiation of neck pain and physical examinations with a more complex holistic perspective on diagnosis. It is necessary to explore related evidences through the selective use of key radiographic computed tomography (CT) or MRI images and electromyography (EMG) to design treatment scheme. This is especially the case if the patient has accompanied neck rigidity, positive pathologic reflex and abnormal muscular strength and tension, and so on. When such symptoms occur, more urgent examinations and interventions need to be executed. If only routine physical therapies such as cervical traction, manipulation, transcutaneous electrical nerve stimulation (TENS) or treatment with nonsteroidal anti-inflammatory drugs (NSAIDs) based on common clinical practice guideline or therapeutic recommendations are pursued, potential risks could occur.

4. Conclusion

Neck pain was related to many factors. We diagnosed an extra-neck factor in this case presentation and the case provided an awareness of the origin of severe intracranial disease. It is mandatory to take a detailed evaluation and analysis of patients involved in neck dysfunction before rehabilitation intervention with a more complex holistic approach. CPPs in the CPA region are relatively rare and they cannot be differentiated easily from other tumors. Once the patient presents with atypical signs, such as neck rigidity, positive pathologic reflex and abnormal muscular strength and tension, more specialized examinations and interventions are recommended to be executed as soon as possible to avoid occurrence of potential risks and protect the patient.

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Author contributions

DD* was responsible for the oversight of the report and integrity of the entire clinical study. CT was in charge of the study concepts, design, manuscript preparation, and editing. LB analyzed and interpreted the patient data regarding MRI findings. HJ administered and monitored the treatment of the patient. WY collected patient reported outcome measures. All authors have read and approved the final manuscript.

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