A rare case of pediatric cerebellopontine angle meningioma presenting with generalized seizures

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Abstract The cerebellopontine angle (CPA) is a rare location for lesions in children, the most initial symptoms and signs were manifested by cranial nerve palsy and cerebellar dysfunction as well as increased intracranial pressure. However, onset of exclusive generalized seizures caused by CPA meningioma in pediatric patient was not described based on previous literature. Here we reported one rare 3-year-old female patient with a 1-month history of generalized seizures, the presumed diagnosis of the lesion was giant glioma or primitive neuroectodermal tumors (PNETs), postoperative histopathological examination proved it to be a benign meningioma, furthermore, seizure control improved dramatically after removal of the CPA tumor. The authors highlighted the unique clinical characteristics and microsurgical skills in present giant pediatric case. Recognizing of generalized seizures resulted from CPA meningioma and management techniques were also emphasized.
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1. Introduction

Cerebellopontine angle (CPA) tumors are more commonly occurred in adults [1-4], in which accounts for 5-10% of all intracranial lesions[1], However, they are rare in children [5-7], previous literatures demonstrated that the incidence of CPA tumor in pediatric patient is only 1% [1]. In addition, the most initial symptoms and signs in CPA lesions were usually manifested by cranial nerve palsy and cerebellar dysfunction as well as increased intracranial pressure, single generalized seizures is seldom involved in the posterior fossa tumor, on the contrary, seizures as the most common presenting symptom is significantly related to supratentorial locations which implicated in several lobes (28%), parietal lobe (25%), as well as frontal and temporal lobe (18.8% respectively) [8]. In view of onset of sole generalized seizures caused by CPA meningioma in pediatric patient hasn't been reported. Consequently, we reported one exceedingly rare 3-year-old female patient with a 1-month history of generalized seizures, postoperative histopathological examination proved it to be a benign meningioma, furthermore, seizure control improved dramatically after removal of the CPA tumor.

2. Case report

A 3-year-old female patient was admitted to the hospital with a 1-month history of generalized seizures. A generalized tonic clonic seizure mainly occurred in night, there were 4 episodes during one month before admission. Neurological examination on admission showed no significant abnormality. She did not have pulmonary tuberculosis (TB) history, her chest X-ray was normal, the results of routine blood tests were within normal limits. The lesion was iso-intense in T1-weighted MRI sequences and hyper-intense in T2-weighted sequences, the volume of the lesion was 50 × 38 × 46 mm, No surrounding cerebellar edema was also observed, after gadolinium intravenous injection, T1-weighted images demonstrated intense and obvious heterogenous enhancement of the lesion (Fig A-C). "dural tail sign" in the dura adjacent to the tumor was no found in present case. Preoperative electroencephalograph (EEG) was unable to localize the site of the epilepsy.

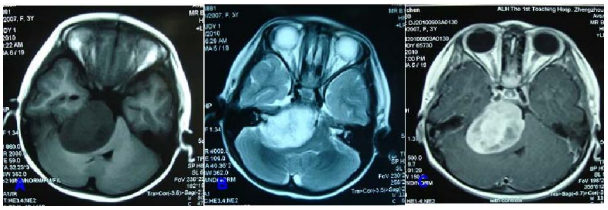

Fig A-B: Preoperative axial T1-weighted and T2-weighted MRI showing iso-intense on T1 and hyper-intense on T2-weighted in right CPA. **Fig C:** Post-contrast T1-weighted MRI showing heterogenous enhancement of the lesion.

Suboccipital retrosigmoidal approach was applied to remove tumor in present case, the cranial nerves of VII, VIII, IX and X were carefully preserved. However, subtotal removal was achieved in present CPA tumor due to the serious adhesion to

the brain stem boundary, the patient's postoperative course was uneventful, and she left the hospital 2 weeks after the operation. Pathological specimens were routinely fixed with formaldehyde and embedded in paraffin and cut for hematoxylin-eosin staining. Postoperative histopathological examination proved it to be a benign fibrous meningioma (Fig D), after treatment with intravenous sodium valproate (SV) followed by oral administration of SV for the total of six months, This patient experienced seizure control dramatically and no complications occurred in this case, small residual tumor adjacent to brain stem was observed in this case on postoperative MRI sacn three months after microsurgery (Fig E-F), adjuvant gamma-knife radiotherapy was performed as subsequent management, no recurrent tumor was observed through three years of following-up.

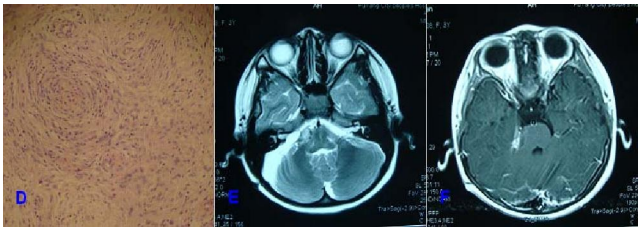

Fig D: Photomicrograph showing the benign meningioma composed of fibrous tissue. (H-E x200)
Fig E-F: Postoperative MRI showing that subtotal removal was achieved on T2-weighted MRI and T1-enhenced MRI respectively.

3. Discussions

The generalized seizures resulted from the posterior fossa lesion is extremely rare, the initial onset of generalized seizures as sole symptom caused by pediatric CPA meningioma have not been reported based on published literatures. Li X [9] reported a series of 34 cases with intracranial meningiomas of childhood and adolescence, generalized seizures was totally involved in the supratentorial convexity, sphenoid ridge lateral 1/3 and cavernous sinus, however, which was no found in the cerebellopontine angle. In addition, other rare initial presentation due to CPA meningioma was described by exceedingly limited literatures. Stein PJ [10] reported one rare case of CPA meningioma with neck and upper extremity pain in 2009. In present case, to our knowledge, this is the first report of the unusual symptom of generalized seizures resulted from CPA meningioma according to previous literatures. Why posterior fossa lesion could lead to generalized seizures? We speculated that the fundamental pathogenesis of seizure may be closely associated with the brain stem compression and increased intracranial pressure owing to giant benign meningioma in posterior fossa. Gan YC [11] described a unique case of one patient with secondary generalized epilepsy caused by a cerebellar quadrigeminal arachnoid cyst, he pointed out the cerebellum may be a source of epileptic activity due to compression by a lesion in the posterior fossa. Consequently, unsuspected CPA meningioma in child patient should be highlighted in the diagnosis of generalized seizure, most importantly, seizure control improved dramatically after removal of the CPA tumor, no generalized seizures were observed after operation through three years of following-up.

Meningiomas constitute 0.4-4.6% of pediatric central nervous system tumors [12-15], which is predominantly located in the cerebral convexity, intraventricular, anterior and middle cranial fossae, falcine and parasagittal regions [16]. Giant meningioma located in the CPA is rare lesion in this pediatric case [17], which is mistaken for giant glioma or primitive neuroectodermal tumors (PNETs). The leading causes of diagnostic errors are as following: (1): The MRI finding is lack of "dural tail sign" in the dura adjacent to the tumor in present case. (2): Gd-DTPA-enhanced T1-weighted images showed a mass with a marked heterogenous enhancement with cystic components. (3): Traditionary conception of malignant tumor especially located in the posterior fossa also plays an important role in the wrong diagnosis in pediatric brain tumor.

The CPA is a rare location for lesions in children with high mortality, Tsai MH [18] showed that two patients died soon after the CPA operation, the whole mortality rate in CPA surgery is accounting for 34.5%. Additionally, tremendous risk during the pediatric brain tumors was observed in numerous previous literatures [19-20], the cause of death may be related to the following factors based on our experiences: (1): child patient is sensitive to hypovolemia due to excessive bleeding during the giant operation, (2): Damaged to vital structures such as brain stem and perforating artery of brain stem, (3): Cranial nerve IX and X injury during the resection of tumor extension into the jugular foramen, (4) postoperative serious complication such as cerebral vasospasm and infarction. Hence, it is our practice that the corresponding measures were taken in every pediatric surgery so as to avoid severe complications: (1) performing excision as rapidly as possible, and administering a blood transfusion at the beginning of surgery. (2) preoperative assessment of resection degree according to the T2-weighted sequences of MRI, brain stem edema and disappear of arachnoid boundary on MRI scan were observed in present patient, this phenomenon means it is impossible to totally resect the tumor from the brain stem. Consequently, it is wise to left small tumor residual in view of significant impact of the patients'

quality of life. (3) Gastric tube was placed on postoperative two days for providing the sufficient nutrition and aspiration prevent if cranial nerve IX and X dysfunction, sometimes, it is necessary to invite otolaryngologist to surgical intervention, (4) suitable hypertension, hypervolemia, and nimodipine was applied to avoid the serious cerebral vasospasm, this exceedingly rare complication such as vasospasm was reported by Lee TT after resection of CPA lesion [21]. (5) Post-operative gamma-knife radiotherapy is an effective and safe treatment for residual CPA tumor with few side effects, this patient with small residual tumor experienced r-knife radiotherapy, she exhibit good health station without severe complication through long-term follow-up, continuous shrinkage of the residual tumor was achieved after the treatment with radiotherapy.

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