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Misdiagnosis of Facial Nerve Palsy: A Case Report of Sporadic Vestibular Schwannomas (Non-NFM2) Causing Facial Nerve Palsy Be Misled by Bell's Palsy in a Child

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Abstract

We report a case of a rare sporadic Vestibular Schwannoma of a 9-month-old girl who had a right-sided lower motor type facial nerve palsy. The patient was initially diagnosed with Bell's palsy and received steroid treatment accordingly, two months later the patient's condition deteriorated, and further evaluation of CT and MRI brain was conducted that showed a mass lesion in the posterior fossa causing compression on the facial nerve. Misdiagnosis of facial nerve paralysis is common among children due to multiple related etiologies and varying rates of incidence in comparison to adults. The authors hope to address this issue in this report. **Background:** Facial nerve paralysis has been a matter of concern for many researchers to understand its nature, causes and presentation according to different age groups. In adults, Bell's palsy (BP), the idiopathic form of facial nerve paralysis, is more common compared to children where most cases are due to secondary etiologies. Therefore, pediatricians are in an important position to identify these patients early in order to launch the most effective diagnostic and treatment approaches.

Keywords

Emergency Medicine, Facial Never Palsy, Bell's Palsy, Infants, the Pediatric Vestibular Schwannoma

1. Introduction

The facial nerve, which is responsible for controlling facial expression, lacrimation, and hearing, is considered as the most often paralyzed nerve in the body

[1]. Estimates of the incidence of facial nerve palsy in population aging from 15 to 40 per 100,000 per year [2] [3], while Children's prevalence is much lower, with estimations of 2.7 per 100,000 for children under 10, with 10.1 per 100,000 for those between 10 and 20 years old [4] [5]. Causes of facial nerve palsy in children are either congenital or acquired (infection, inflammation, tumors) [6]. Some tumors such as schwannomas or hemangiomas are the least common cause of facial nerve palsy with 2% among pediatric cases [1] [7]. On the other hand, Bell's palsy (BP) is caused idiopathically and rarely affects children at an estimated rate of 6.1 cases per 100,000 per year for the age group between 1 and 15 years old [8].

Vestibular schwannomas (VSs) are very rare and benign tumors arise from the myelin-producing Schwann cells that line the vestibular cochlea or the eighth cranial nerve [9]. Compared to adults, children are less likely to develop VSs, which often appear as large tumors [10] grow in CPA of the internal auditory canal [11] and start to generate pressure on the brainstem, cerebellum, and adjacent vestibulocochlear nerves [10]. In children, VSs are often associated with neurofibromatosis type 2 (NF2) [11], However, rarely appear sporadically [12] [13]. Only 6% of VSs are diagnosed and treated in the second decade, and even less in the first decade [14].

2. Case Presentation

A 9-month-old girl was brought to the emergency department (ER) by her parents due to facial right-sided deviation with the inability to close the left eye, patient was active with stable vital signs and no recorded trauma or any past medical or family history of neurofibromatosis, her clinical examination was unremarkable other than lower facial nerve palsy. The patient was thriving well with normal development, so no further investigations were held. Accordingly, she was diagnosed with idiopathic Bell's palsy, and she was treated with steroids.

Two months later (the patient now is 11 months of age) the patient returned to the ER with frequent vomiting, low-grade fever, and decreased activity for the past four days. The patient showed signs of psychomotor regression: not rolling, not sitting, decreased interaction with her mother, and irritability. Her vital signs: Temperature 37.2˚C, Heart rate 110 BPM, Blood pressure 130/65mmHg, Respiratory rate 34/min, Oxygen saturation 96%.

Clinical examination revealed hypoactivity, altered mental status, and poor reaction to the environment along with obvious lower facial palsy. Other systems were unremarkable.

One hour later the patient developed a generalized tonic-clonic seizure which responded to benzodiazepine, phenytoin, and levetiracetam.

The initial impression was a brain space-occupying lesion, therefore, an unenhanced axial brain CT scan was conducted which showed a left cerebellar mass with a rightward midline shift obstructing the fourth ventricle and causing obstructive hydrocephaly.

The patient was referred to a higher specialized hospital where she underwent

partial tumor reduction brain surgery to reduce intracranial pressure. The biopsy was obtained and revealed a vestibular schwannoma resulting in a mass lesion in the posterior fossa. Unfortunately, the patient expired two days after surgery due to severe brain edema resulted in brainstem herniation and cardiac arrest.

Radiological findings:

On CT Figure 1:

Left cerebella hemorrhagic lesion measuring 4.2 × 3 cm with mild surrounding vasogenic edema and rightward midline shift obstructing the fourth ventricle causing obstructive hydrocephaly. Dilation of the third and lateral ventricles. No definite tonsillar herniation. Opacification of the right mastoid air cell and right middle ear cavity. Paranasal sinuses are well aerated. The visualized osseous structures are unremarkable.

On MRI Figure 2:

A low signal intensity on T1 W1 and high heterogeneous high signal intensity containing multiple foci of high signal intensity on T2 due to cystic changes and showed heterogeneous enhancement after IV contrast. The mass is delineated posteriorly and laterally by a linear hemorrhage and associated with a smell left subdural hematoma in the subacute phase. The mass showed restriction on D W1 brainstem, it is causing moderate obstructive hydrocephalus of the third and lateral ventricle with mild periventricular edema. Normal appearances of the cerebral arteries and dural venous sinuses. No seeding metastasis seen.

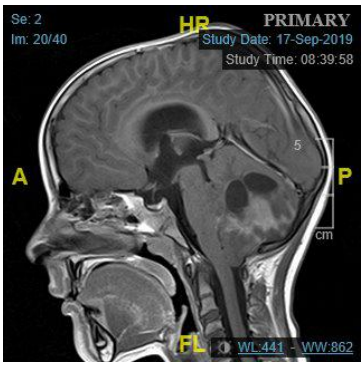

Figure 1. CT brain lateral view.

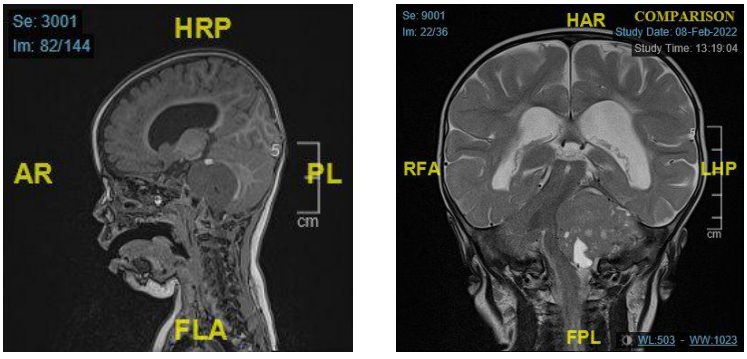

Figure 2. Left: MRI brain with contrast lateral view; Right: MRI brain with contrast sagittal view.

3. Discussion

Facial nerve palsy in children may result from various causes classified as congenital or acquired (infectious, inflammatory, neoplastic, and trauma).

Developmental defects or traumatic causes may lead to congenital facial neuralgia [15]. Only 0.23% to 1.8% of live births result in neonatal facial paralysis, thus it is atypical in neonates, where 78% to 90% of these cases are associated with birth trauma [15] [16].

Lyme disease and varicella zoster (Ramsey Hunt syndrome) are common viral infectious diseases that cause mild otitis media and may lead to facial nerve paralysis in children. Other recognized agents include cytomegalovirus, coxsackievirus, herpes simplex virus, rubella, and mumps virus [1] [15].

Facial nerve palsy can coexist with inflammatory conditions such as vasculitis, Henoch-Schönlein purpura, or Kawasaki syndrome [3] [8]. In Guillain-Barre syndrome facial nerve palsy typically occurs along with muscle weakness and paralysis. In some cases, the facial nerve palsy is the only symptom [3].

Tumors also can compress the facial nerve resulting in facial nerve palsy, for example, neuroma and hemangioma [1]. Cholesteatoma is one of the most common tumors that cause the gradual development of facial nerve paralysis [15]. Other common tumors include head and neck tumors, rhabdomyosarcoma, Burkitt's lymphoma, and primary middle ear lymphoma [3]. On the contrary, facial nerve schwannoma and cerebellar angle tumors are rare causes of facial nerve palsy in children [15].

Bell's palsy (BP)

Bell's palsy is a rapidly developing facial nerve paralysis of unknown etiology but with a very good prognosis as it is a self-healing disease [1]. BP is responsible for most of the recorded cases of facial paralysis with a rate of 40% to 70%, and it may occur in any age group, but it is most common among people between the ages of 15 and 45 years [17]. In other smaller case series, the cause of facial nerve paralysis was BP in 16% to 50% of the children studied [18]. In general, the diagnosis of BP is based on clinical rather than laboratory skills. There are no standard tests necessary to diagnose BP if the patient presents normally and a thorough clinical examination does not reveal any obvious cause [1].

Vestibular schwannomas (VSs)

It is a benign, slow-growing tumor of the vestibular section of cranial nerve VIII [12] accounts for 60% of lesions localized in the cerebellar angle (CPA) [19]. With 0.8% incidence rate in overall pediatric tumors [20], VSs can occur sporadically or accompany a genetic syndrome such as Neurofibromatosis type 2 (NF2). In NF2, vestibular schwannomas appear bilaterally and co-occurring with other brain tumors, whereas sporadic schwannoma is often unilateral, and does not coexist with other tumors [21]. Unlike adults, sporadic forms are extremely rare in children [12] [13], and tend to be more aggressive with higher rates of growth and recurrence [12].

According to Holman *et al.* study [19] on pediatric CPA tumors, only 6% of kids with schwannoma had an NF2 characteristic. In different research, 20% of

patients had NF2-related vestibular schwannoma [22], this numerical difference may be due to a significant portion of NF2 individuals showing symptoms in adulthood rather than infancy [22].

Symptoms are caused by pressure on the vestibulocochlear nerve, the adjacent cerebellum, and the brainstem [10]. The major presenting symptoms of VSs tumors signs are hearing loss, tinnitus, headache, imbalance, hemiparesis and facial numbness [22]. In a study aimed at examining the rare presence of sporadic vestibular schwannoma in pediatrics, of 96 pediatric cases, 24 reported facial paralysis due to the effect of the mass lesion along with other common complications such as cerebrospinal fluid leak (6%) and hemorrhage. (4%) headache (3%) and infection (2%) [23].

4. Results

Misdiagnosis between secondary facial nerve paralysis and Bell's palsy (BP), the idiopathic form of facial nerve palsy, in the emergency department is rare in adults but is high among young ages [24] [25]. Misdiagnosis of the underlying causes of facial nerve paralysis results in a wrong treatment approach, which can lead to poor outcomes, for example, preceding steroid therapy to treat BP for children with leukemias and lymphomas-related facial palsy has poor therapeutic consequences [26].

Some suggestion behind the facial palsy misdiagnosis is the fact that BP, as it is essentially idiopathic, can only be diagnosed by ruling out other etiologies [6] [8], and does not have a specific examination confirming its presence then the possibility of misdiagnosis is very high, especially in diseases with late personations such as CPA tumors [24] [25]. In a 10-year cohort study of 12,272 patients ranging in age from 6 months to 17 years, 41 cases initially diagnosed with BP obtained a new diagnosis with brain tumor within 60 days of the first emergency department visit [26]. Furthermore, poor understanding of the upper and lower neural pathways may play a role in the misdiagnosis dilemma, as cortical and subcortical damage to upper motor neurons causes contralateral facial paralysis involving the lower part of the face (orbicularis oris), in contrast to lower motor facial paralysis, in which paralysis occurs Similar complete flaccidity affecting the muscles of the upper (anterior and circular) and lower muscles of the face (orbicularis auris) [27], for further illustration, BP Paralysis is defined as facial paralysis of lower motor neurons that begins with an acute form without evidence of an auditory, neurological, or local cause. It is usually unilateral and may be complete or partial [1] while CPA fascial schwannomas, like in the discussed case, are benign and slow-growing tumors [28] cause space-occupying lesions [29] as a part of the upper neural pathway, thus complete understating of the differences among neural pathways is essential to conduct the proper diagnosis of facial nerve palsy.

5. Conclusions

Despite the low incidence of tumor-induced facial nerve palsy compared to the

high rate of Bell's palsy, it should take into consideration this rare etiology, especially in children less than 2 years of age where the probability of misdiagnosis is very high. Therefore, it is recommended that establishing the diagnosis of Bell's palsy in this age group is by the exclusion of all other etiologies.

Pediatric vestibular schwannomas present similarly to those in adults, but symptoms of brain mass effect are more common due to the very slow tumor growth with a high incidence of misdiagnosis similar to what has happened with our patient. moreover, children who are diagnosed early can be successfully treated surgically with minimal complications.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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Abbreviations and Acronyms

Vestibular Schwannomas (VSs)

Cerebellar Angle (CPA)

Bell's Palsy (BP)

Neurofibromatosis Type 2 (NF2)