Child-onset Hemifacial Spasm-a case report

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Case report

Keywords: Hemifacial Spasm, Neurovascular Conflict, Carbamazepine, Child-onset

Posted Date: August 17th, 2021

DOI: <https://doi.org/10.21203/rs.3.rs-779146/v1>

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Abstract

Background

Hemifacial spasm (HFS) is a common cranial nerve disease, with peak onset in the forty to sixty years old and child-onset HFS is rare. HFS brings many negative effects on the physical and mental health of patients. Here, we report a 10-year-old case of primary HFS patient.

Case Presentation

Here we report an 11-year-old HFS case last for more than 1 year. An 11-year-old girl complained of frequently involuntary twitch in lower eyelid as the girl was 10 years old, then symptom worsen gradually involved right upper lip in recently months. The girl's daily life was badly influenced. Brain magnetic resonance imaging indicated that AICA compressed right facial nerve. The patient was treated with carbamazepine 200mg per day, and symptom gradually relieve. But bad treatment compliance appeared, and we introduced the advantages and disadvantages of botulinum toxin and MVD to her parents. However, considering the young age, above mentioned options are not considered for the time being. And more observations of medications treatment were needed to guide further treatment.

Conclusion

Child-onset hemifacial spasm is rare and the case we report is one of the youngest known cases of such disease. For such patients, medication is the first choice, but the exact mechanisms remain to be further explored.

Background

HFS is a common cranial nerve disease, mainly manifests as muscle involuntary contractions restricted in the distribution of facial nerve, generally lower eyelid onset, and gradually aggravates to involve the ipsilateral face [1–3]. This disease usually occurs spontaneously and deteriorate as patients are stressful or fatigue [4], but also caused by mini stimulation. Generally speaking, HFS spasm involves only unilateral face[5], mainly the left side[3]. HFS could be divided into primary and secondary based on the potential etiology and primary HFS in majority of cases. Secondary HFS is usually caused by facial nerve injury or compression of nearby primary lesions, such as tumor, inflammatory plaques [6, 7]. Primary HFS is generally caused by neurovascular conflict (NVC), especially root exit zone (REZ) of this nerve [3, 8]. For the compression results in local demyelination, and hyper excitability. Among the compressional arteries, anterior inferior cerebellar artery (AICA) and posterior inferior cerebellar artery (PICA) are the most common, and rarely superior cerebellar artery (SCA), petrosal vein (PV) and vertebrobasilar artery (VBA) [3, 8, 9].

Many researches indicated that HFS is mainly involved middle aged and elderly people [3, 8], it is extremely rare in childhood and adolescence [10, 11]. What's more, more attention should be paid to

childhood and adolescence onset HFS, for many primary lesions usually manifest as HFS, such as cyst[12] and glioma[13, 14]. According to the severity of spasm and its impact on life, Cohen classified HFS into 5 grades[15](Table 1).

Table 1. Cohen Score of Hemifacial Spasm

Grade 0	No spasm
Grade 1	Increased blinking or slight flutter of facial muscle caused by external stimulation;
Grade 2	Spontaneous slight flutter of eyelids and facial muscles without dysfunction;
Grade 3	Significant spasms with slight dysfunction;
Grade 4	Severe spasticity and dysfunction, inability to read or walk for inability to open eyes

In general, medication is the first-line treatment for HFS, such as anti-epileptic drugs and botulinum toxin [16–18], which could alleviate the spasm, but the long-term effect is usually far from satisfactory, and usually lead to some side effects [19–21]. Microvascular decompression (MVD) is a validated option for these patients who are refractory to medications or cannot tolerate the complications [22, 23]. Here we report an 11-year-old HFS case due to compression of the right AICA.

Case Presentation

An 11-year-old girl accompanied by their parents presented to the clinic complained of frequently involuntary twitch in lower eyelid as the girl was 10 years old, then symptom worsen gradually involved right upper lip in recently months even during sleep. For this, the child was unwilling to communicate with classmates or friends and had feelings of inferiority, which seriously impacted her normal life and study. Physical examination of the nervous system: touching can induce spasm, manifested as strong spasm of the right upper lip and lower eyelid, which badly affects vision and walking of the child, but no other positive findings were observed.

Brain magnetic resonance imaging indicated that AICA compressed REZ of the right facial nerve cranial laterally (Fig. 1), and then primary HFS was considered. After comprehensive consideration of the patient's age and family's wishes, the patient was treated with carbamazepine 200mg per day, and symptom gradually relieve, with a Cohen Score of 1 in last outpatient follow-up. But bad treatment compliance in this patient was observed, so we introduced the advantages and disadvantages of botulinum toxin and MVD to her parents. However, considering the young age, the mentioned treatments are not considered for the time being. And more observations of medications treatment were carried on to guiding further treatment.

Discussions And Conclusion

Here, we report an 11-year-old HFS case caused by compression of AICA. This is one of the youngest reported cases of primary HFS in the literature to date.

As we all know, HFS is a common cranial nerve disease, mainly manifested as involuntary convulsion of muscles innervated by facial nerve, the spasm could be provoked by mini stimulus, such as touching and light [4, 18]. Studies have shown that HFS generally involves only unilateral face, mostly on the left side[5, 24], and a few involves bilaterally or accompanied with other cranial nerve diseases, such as trigeminal neuralgia or glossopharyngeal neuralgia[25, 26].

Although HFS generally does not make threat to life, it will affect patients' daily study and life, and cause huge psychological hurt, seriously damage the patient's life quality. Therefore, it is an urgent task for doctors in this field to strengthen the research on diagnosis and treatment of HFS.

Based on different pathogenic factors, HFS can be divided into two types: primary and secondary HFS. Secondary HFS was caused by compression of local lesions near the nerve [6, 7]. The pathogenic mechanism of the primary HFS is not totally made clear, but NVC of the nerve, especially REZ is the most accepted [3, 8]. AICA and PICA is the most common culprit vessels, followed by SCA PV and VBA [3, 8, 9].

At present, medication is the preferred treatment for HFS, such as anti-epileptic drugs and botulinum toxin injection [16–18]. Many patients could achieve partial relieve, but long-term outcome is still far from our expectation and it is easily lead to some serious side effects [27].

For these reasons, MVD has become a validated option for those who are refractory to medications or those who cannot tolerate the side effects[23]. Relevant studies have shown that the long-term efficient could achieve about 90% with little complication rate[28]. For younger patients with poor adherence or severe side effects, MVD is also a feasible option to achieve long-term relief[29].

Generally, primary HFS mainly involve middle to aged people [3, 8], adolescent are less affected, and previously related conditions are mainly case reports [10–14]. Furthermore, for the rarity of youth primary HFS, more attentions should be paid to such patients to exclude secondary HFS, such as cyst[12], glioma[13, 14] and Chiari type 1[30] malformation. As such conditions may lead to more serious outcome, especially in childhood-onset HFS.

In conclusion, child-onset hemifacial spasm is rare and the case we report is one of the youngest known cases of such disease. For such patients, medication is the first choice, but the exact mechanisms remain to be further explored.

Abbreviations

HFS: hemifacial spasm; MVD: microvascular decompression; AICA: anterior inferior cerebellar artery; PICA: posterior inferior cerebellar artery; SCA: superior cerebellar artery; PV: petrosal vein; VBA: vertebrobasilar artery; NVC: neurovascular conflict; REZ: root exit zone;

Declarations

Acknowledgements: Not applicable.

Authors' contributions: CY developed the profile and guided the writing and TS is the main writer of this report, correct the manuscript, follow the patient; NL assisted CY and TS to look for relate paper and conclude the review and follow the patient; JIL played a part in follow the patient and conduct the treatment and diagnosis.

Funding: Funding provided by Science and Technology Department of Guangdong Province (K0214019).

Availability of data and material: Not applicable as this is a case report

Submission statement: This manuscript is original and has not been submitted elsewhere in part or in whole.

Ethics approval and consent to participate: This case report was approved by Ethics Committee of the First Affiliated Hospital of Sun Yat-sen University.

Consent for publication: The authors have obtained the patient's written informed consent for print and electronic publication of this case report.

Conflict of interests: The authors declare that they have no conflict of interest.

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Figures

Figure 1

Brain MRI indicates that the right facial nerve was compressed cranial laterally by an inferior anterior cerebellar arterial loop.

Supplementary Files

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