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Case Report

Familial Idiopathic Cranial Neuropathy in a Chinese Family

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ABSTRACT

Cranial neuropathy is usually idiopathic and familial cases are uncommon. We describe a family with 5 members with cranial neuropathy over 3 generations. All affected patients were women, indicating an X-linked dominant or an autosomal dominant mode of inheritance. Our cases and a review of the literature suggest that familial idiopathic cranial neuropathy is a rare condition which may be related to autosomal dominant vascular disorders (e.g. vascular tortuosity, sclerosis, elongation or extension), small posterior cranial fossas, anatomical variations of the posterior circulation, hypersensitivity of cranial nerves and other abnormalities. Moreover, microvascular decompression is the treatment of choice because vascular compression is the main factor in the pathogenesis. To the best of our knowledge, this is the first report of familial cranial neuropathy in China.

KEYWORDS: Cranial nerves, Familial, Microsurgical treatment

■ INTRODUCTION

Although familial cases of cranial neuropathy have been reported, most cases occur sporadically. The incidence of familial trigeminal neuralgia (FTN) has ranged from 1 to 6% of patients with trigeminal neuralgia (TN) (5,10). An association between autosomal dominant Charcot–Marie–Tooth disease and FTN has been described (3,11). Moreover, it has also been reported that FTN may be associated with other cranial nerve dysfunctions such as 7th and 8th cranial nerve dysfunctions (2,4,10). Since 1985, more than 9000 procedures of microvascular decompression (MVD) were performed for the treatment of hemifacial spasm (HFS), TN, and glossopharyngeal neuralgia (GN) in our department. In one case of familial idiopathic cranial neuropathy found by the authors, there were 5 female members affected by cranial neuropathy across three generations. This is suggestive of an X-linked dominant or an autosomal dominant mode of inheritance (9).

To our best knowledge this is the first report on familial idiopathic cranial neuropathy which only affects females in China.

■ CASE REPORT

Three microsurgical procedures were performed for the treatment of cranial neuropathy, including TN, HFS and GN, in 5 female members across 3 generations of one family (Figure 1A-D).

Patient A was a 55-year-old woman who suffered right TN (branch 1, 2, 3). Due to the ineffectiveness of conservative treatment, she underwent rhizotomy of right trigeminal peripheral branches 10 years later. Afterwards, her pain was relieved and there was no further serious attack in the follow-up period of 10 years, until her death.

Patient B was a 74-year-old woman who suffered from right TN (branch 1, 2, 3) since she was 50 years old. Due to the ineffectiveness of conservative treatment, she underwent MVD of right trigeminal nerve and selective partial rhizotomy of sensory root 10 years later.

Surgery was performed under general anesthesia. The patient was placed in lateral recumbent position with contralateral side down, head down 15°, rotated 10° to the contralateral side and slight neck flexion. A postauricular skin incision



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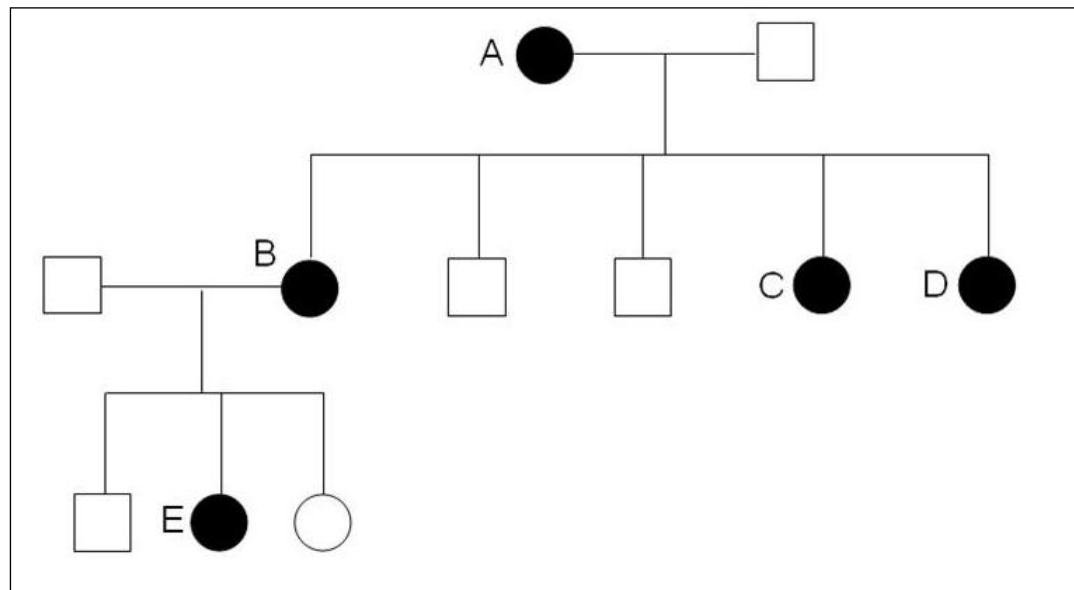


Figure 1: Family tree of patients reported in the present study. Circles and squares indicate men and women, respectively. Black circles indicate affected individuals.

A: right trigeminal neuralgia (TN),
B: bilateral TN,
C: right glossopharyngeal neuralgia and TN,
D: left hemifacial spasm,
E: right TN.

parallel to the hair line was made with a scalpel. A small bone window of 1.5~2.0 cm in diameter was provided between transverse and sigmoid sinuses. The dura mater was opened in inverted T-shape fashion. The cerebellopontine angle (CPA) was exposed by the conventional surgical approach, and the cerebellar hemispheres were lifted with a thin brain spatula. The cerebrospinal fluid was slightly drained and the arachnoid membranes were cut and opened using microscissors to expose the cranial nerves. After cutting the arachnoid membranes between the auditory and trigeminal nerves, the pontine cistern segment of the trigeminal nerve was exposed (12). Maximum care was used to avoid damaging the perforating arteries of the brain stem. The right trigeminal nerve was found to be in contact with a branch of the superior cerebellar artery. The offending artery was removed away from the nerve and a Teflon cushion was inserted into the space between the vessel and brain stem. Selective partial rhizotomy of the sensory root of trigeminal nerve was performed after MVD. Afterwards, the pain resolved and did not recur over the 14-year follow-up. At age 65, she developed left TN (branch 1, 2, 3), and MVD was performed after failed conservative treatment. Intraoperatively, the left trigeminal sensory root was found to be compressed by a branch of the superior cerebellar artery, the anterior inferior cerebellar artery and 2 branches of the superior petrosal vein. The offending arteries were padded off with Teflon, and the veins were ligated after electrocoagulation. Afterwards, the pain was resolved and did not recur over the 8-year follow-up.

Patient C was a 60-year-old woman who suffered from right atypical GN since she was 47 years old. She underwent 3 procedures including MVD of the right glossopharyngeal root, neurotomy of the glossopharyngeal root, and selective partial neurotomy of the upper fila of the vagus nerve over a 5-year period. The former two procedures were not effective but the last was in relieving her pain. Her right TN developed when she was 53 years old and was eliminated by MVD of the right trigeminal nerve. Intraoperatively, the trigeminal sensory root

was found to be compressed by a branch of the superior cerebellar artery. The artery was padded off with Teflon. There was no recurrence of pain over the follow-up period of 7 years.

Patient D was a 56-year-old woman who suffered left HFS since she was 45 years old. She did not undergo any operation.

Patient E was a 44-year-old woman who suffered from right TN (branch 2) since she was 38 years old. She did not undergo any operation.

Except for Patient C, all 5 patients had typical clinical manifestation of idiopathic cranial neuropathy. Additionally, none of the men in this family have cranial neuropathy.

DISCUSSION

Familial idiopathic cranial neuropathy is rare (1,5-8,10). There are few reports demonstrating association of FTN with other cranial nerve disorders in the literature (2,4). In the present study, we report 5 female patients from 3 generations of the same family in China with co-occurrence of FTN, HSF and GN. It is generally accepted that idiopathic cranial neuropathy is non-genetic, since its hereditary form is unclear (7). However, as considered by Coad et al. (2) Carter et al. (1) and Smyth et al.(10), familial idiopathic cranial neuropathy may be subject to autosomal dominant inheritance with low penetrance. Since familial idiopathic cranial neuropathy caused by neurovascular compression (NVC) in the cerebellopontine angle (CPA) could be relieved by MVD, its pathogenesis may be related to patterns of vascularity, and autosomal dominant vascular disorders (e.g. vascular tortuosity, sclerosis, elongation or extension) may be the cause (10). It is possible that the familial development of idiopathic cranial neuropathy may be related to small volume of posterior fossa, anatomic variation in posterior circulation (2), hypersensitivity of cranial nerve and high occurrence of other abnormalities, which may result in NVC of cranial nerve root entry/exit zone. All factors mentioned above may be related to this familial inheritance. Most clinical manifestations

of familial idiopathic cranial neuropathy are typical (6,7). Among familial HFS cases reported in the literature, the left side is more commonly involved (1, 2, 7). This may be related to the frequent dominance of the left vertebral. However, this was not reflected in the reported family (left : right = 2 : 5). In this family, 5 patients from 3 generations were all female. Four FTN patients reported by Kirkpatrick et al. (6) were also all female. In contrast, 3 familial HFS patients reported by Carter et al. (1) were all male. Thus, the relationship between gender and familial inheritance of idiopathic cranial neuropathy is still unclear. The 5 patients in our report suffered 7 cranial nerve lesions, which occurred at ages 55, 50, 65, 47, 53, 45 and 38, respectively. This is consistent with the typical presentation of the disease in middle-age. This phenomenon could be explained by the etiological theory that the cranial nerves become more susceptible to vascular compression due to vascular tortuosity, sclerosis or elongation and downward displacement of the brain stem in the aged. There were few young patients in the familial cases reported by Miwa et al. (7). However, as considered by Carter et al. (1) and Kirkpatrick et al. (6) patients of familial HFS or TN were younger than those of non-familial HFS or TN. According to our experience, nearly half of the young patients who suffered non-familial HFS had small volume of the posterior fossa and/or thickening or adhesion of the arachnoid membrane in the CPA. The familial inheritance of small volume of the posterior fossa may be considered for the cases reported by Carter et al. (1) and Kirkpatrick et al. (6). The neuropathy of multiple cranial nerves (such as bilateral TN) occurs rarely, consisting of about 1% of all cases of cranial neuropathy in our hospital. However, the incidence of multiple cranial neuropathies is higher in familial cases (4,8). In this report, there were 2 patients (Patients B and C) with multiple cranial neuropathies among the 5 patients. Thus the relationship between the multiple cranial neuropathies and familial inheritance requires further study. As in non-familial idiopathic cranial neuropathy, MVD is the preferred surgical procedure for the familial disease; and the results of surgical treatment are satisfactory (7, 10). Further, and possibly, larger studies of familial idiopathic cranial neuropathy will be helpful for more in-depth understanding of the etiology of this disease, and for the assessment of the efficacy and safety of its surgical treatment with MVD.

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