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Case Report

Familial lower lip facial paralysis with asymmetric smile: Selective neurectomy of the cervical branch

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1. Introduction

Facial nerve paralysis in children is relatively rare and stems from a wide variety of entities. Congenital, acquired, iatrogenic, idiopathic etiologies all play a role, with Bell's palsy remaining the most common cause [1]. While there is significant literature on the most prevalent causes, familial facial nerve paralysis has only been sporadically described [1]. Prior studies have noted association with multiple facial nerve branches, specific alleles 3q21-22 and autosomal dominant transmission [2]. Congenital muscle hypoplasia resulting in asymmetric smiles have also been described, specifically of the depressor anguli oris [3]. While syndromic causes such as Mobius or Melkersson-Rosenthal syndromes are relatively well known, true hereditary causes are rare and poorly understood. To highlight the difficulty with the diagnosis and treatment of these cases, presented are two sisters with isolated familial unilateral cervical branch palsy. This defect has not been reported and frustratingly mimics marginal mandibular weakness. Careful evaluation with intraoperative facial nerve monitoring was used to identify the aberrant nerve branches and allow for selective neurectomy [4].

2. Report of two cases

Two sisters, 8 and 14 years old, presented with isolated right lower lip weakness and asymmetric smile. Both patients had no history of developmental abnormalities nor facial symmetry at rest. Their family history was noncontributory and both children were the product of full term spontaneous atraumatic births. The parents had decided to wait until the children themselves voiced an interest in having an intervention. When they sought intervention, there were difficulties in accessing care in their home state. A thorough discussion was conducted with the parents and patients regarding the various non-surgical and surgical options. Given the significant loss of depression of the right hemi-lip, the etiology was thought to be dysfunction of the depressor anguli oris due to muscular hypoplasia or marginal mandibular nerve palsy. Contralateral chemodenervation with Botox® (onabotulinumtoxin A) of the lower lip depressors was declined due the patient's inability to tolerate multiple awake injections and their desire for a single definitive procedure. Individual muscle sacrifice was also declined due to unpredictable results. Ultimately, the decision was made with the family to improve symmetry with contralateral selective neurectomy. We hypothesized that the marginal mandibular branch would be sacrificed. Genetic testing was additionally offered but declined by the family. Institutional review board approval was granted for this project. The patients presented for exam under anesthesia, facial nerve monitoring, and selective contralateral neurectomy. Facial nerve monitoring (CMAP - continuous monitoring of the action potential) was performed which allowed for preoperative mapping with identification of branch topography and action potentials by a specialized neurophysiologist [4]. In brief, needle probes were placed in the midline mentalis, orbicularis oris and nasalis and attached to a neuromonitor. As a stimulator probe was placed on the facial skin, the monitor would

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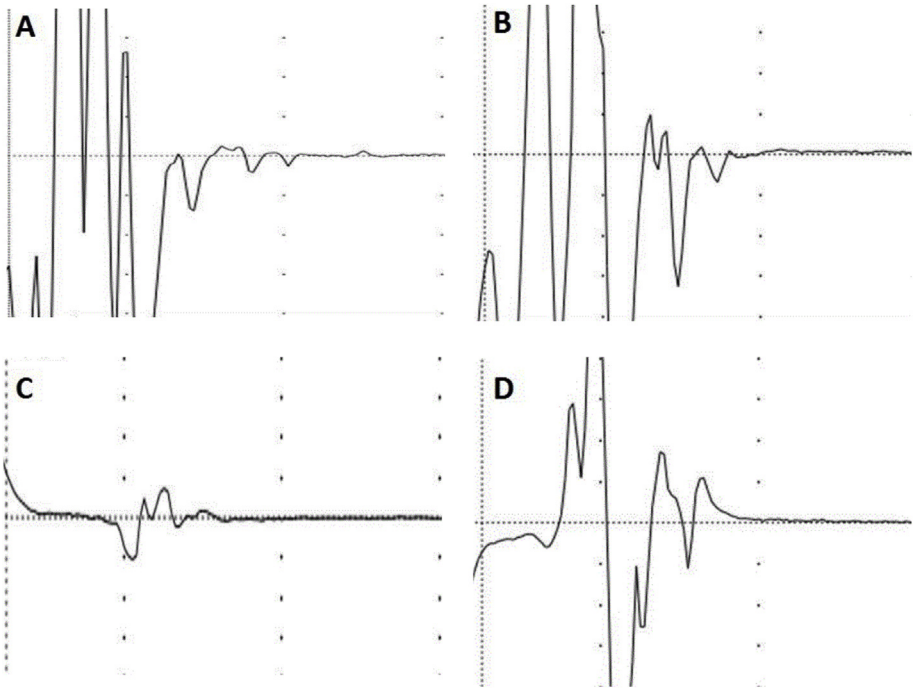

Fig. 1. Electrophysiological facial nerve recording of eight year old sister: (A) Right and (B) left marginal mandibular stimulation elicits normal mentalis muscle CMAP response. (C) Right cervical branch stimulation shows minimal platysma CMAP with no visible clinical contraction. (D) Left cervical branch stimulation shows a normal response.

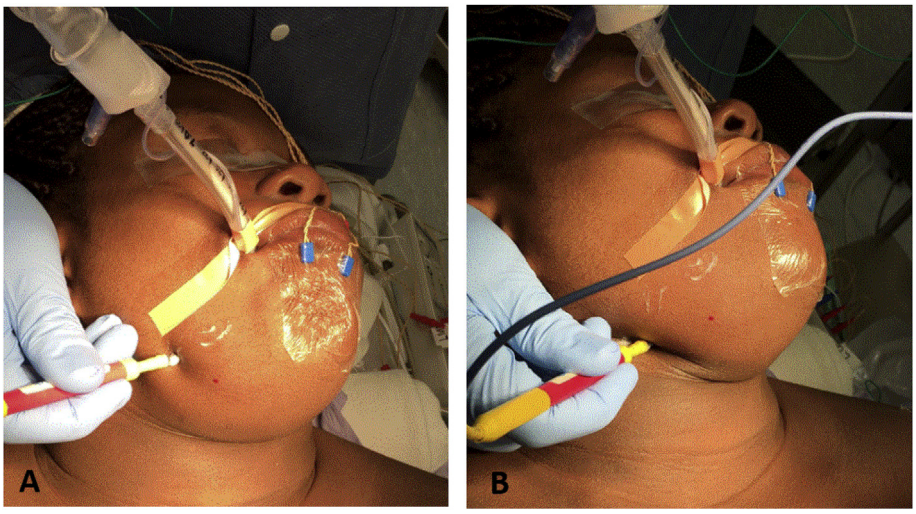

Fig. 2. Intraoperative photos of the older sibling on the paralyzed side showing dimpling with no downward pull upon stimulating the marginal mandibular branch (A) and absence of response to stimulating the cervical branch (B).

detect the action potential of the distal musculature when directly over the nerve. This allowed for a pen to be used to mark the positions of the cervical, marginal and buccal branches on the skin. Once mapped, a bipolar needle probe was placed near the stylomastoid foramen to stimulate the main facial nerve trunk and produce facial musculature tetany as needed through the case. Mapping was performed bilaterally.

The evaluation was performed on the younger sister first. Initial findings were unexpected upon nerve evaluation and revealed symmetric and intact bilateral marginal mandibular branch potentials (Fig. 1), while the right cervical branch was absent with no platysma stimulation. This was ascertained through additional bilateral platysma and cervical branch monitoring. After a thorough intraoperative discussion among the surgical and neuromonitoring staff, it was postulated that there was congenital lack of the cervical branch leading to absence of platysmal lower lip depression masquerading as marginal mandibular palsy. These findings were shared with the family and multiple options were discussed including no intervention, future chemodenervation or surgical neurectomy. They elected to proceed with contralateral cervical branch neurectomy. A 2-cm incision was made in the cervical area over the cervical branch marking. The contralateral branches were identified and stimulation of the cervical branch revealed significant depression of the left lower lip. This branch was identified, doubly tied, and transected with removal of a 1cm intervening segment. Upon awakening, the patient was noted to have a symmetric smile.

The older sister was then brought to the operating room. Neuromonitoring results revealed the same absence of ipsilateral cervical branch function and thus only the contralateral cervical branch was sacrificed. Intraoperative video and photography were taken and illustrate the findings in the two sisters (Figs. 2–4). Postoperatively, both patients had symmetry at rest and with smile without functional deficit at one week and one month (Fig. 5). Further follow-up at one year showed a long-lasting result. Facial photography consent was obtained from the family for both patients.