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Case Report

A case of a facial nerve venous malformation presenting with crocodile tear syndrome

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Received : 08 December 19
Accepted : 12 December 19
Published : 03 January 20

DOI
10.25259/SNI_570_2019

Quick Response Code:

ABSTRACT

Background: Crocodile tears syndrome, also known as Bogorad syndrome, is characterized by lacrimation secondary to olfactory and gustatory stimuli and mastication. Crocodile tear syndrome is typically encountered as an uncommon complication of Bell's palsy and usually occurs during the recovery phase of the disease course.

Case Description: We present a case of a 39-year-old male who presented with facial paralysis with ipsilateral crocodile tear syndrome caused by a slow flow venous malformation of the petrous bone and facial nerve.

Conclusion: We present a case of crocodile tear syndrome caused by a facial nerve venous malformation. To the best of our knowledge, this is the only case reported in literature.

Keywords: Bell's palsy, Crocodile tear syndrome, Facial nerve venous malformation

INTRODUCTION

A 39-year-old male presented to the clinic with a complaint of the right hemifacial spasms that progressed to partial facial paralysis. The patient was diagnosed with Bell's palsy and underwent Botox injection with improvement in the spasms. Several months later, he developed partial facial paralysis. The patient was unable to close his eyes and only had lacrimation from the right eye with eating and exercise. On presentation to the neurosurgical service, the patient had a House–Brackmann Grade 4 facial palsy which was noted to be moderate to severe with obvious weakness and disfiguring asymmetry. No other symptoms or relevant history was reported.

The patient underwent both magnetic resonance imaging (MRI) and computed tomography (CT) scan of the brain and temporal bones which demonstrated an enhancing mass lesion in the right petrous bone involving the geniculate ganglion (GG) of the right facial nerve [Figures 1 and 2]. The lesion was expansile, with smooth osseous remodeling. There were no aggressive radiographic features. The lesion was felt to represent either a slow flow venous malformation or a schwannoma of the facial nerve. Due to the close proximity of the lesion to the facial nerve and high risk for facial nerve injury, serial observation was recommended over surgery to the patient. The risks of surgery including complete facial paralysis, hearing loss, and stroke were explained to the patient. In addition, it was made clear that surgery would most likely not result in improved facial function. However, after several months, the patient requested surgery with the hope of improved facial

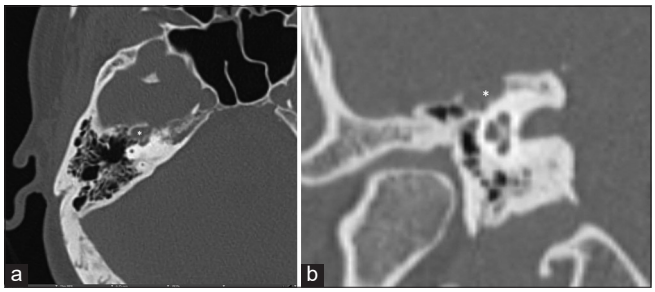

Figure 1: (a) Axial unenhanced computed tomography (CT) demonstrates a lucent expansile lucent lesion in the right petrous bone involving the right facial geniculate segment (white asterisk), (b) magnified coronal unenhanced CT image demonstrating a lucent expansile lucent lesion in the right petrous bone involving the right facial geniculate segment (white asterisk).

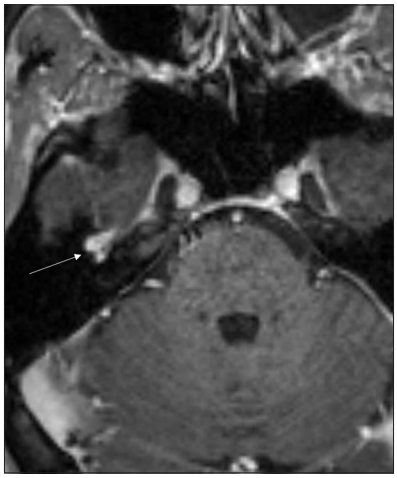

Figure 2: Axial T1 fat-saturated enhanced magnetic resonance imaging demonstrating an enhancing lesion along the right facial nerve geniculate ganglion within the right petrous bone.

nerve function and pathological confirmation. The patient underwent a middle fossa infratemporal craniotomy for biopsy or resection of the lesion [Figure 3]. The patient was placed in supine position with the head turned to the left to expose the right ear. A linear incision overlying the root of the zygoma was made and an extradural dissection to the middle fossa was performed. The lesion was identified with the aid of neuronavigation. There was a bony dehiscence in the location of the lesion, which appeared as a small purple-colored lesion. The lesion was debulked until the facial nerve could be identified. A gross total resection was deemed to be unsafe and would risk a complete facial nerve paralysis. The skull base was then repaired and the wound was closed.

Pathologic examination of the lesion revealed a vascular malformation characterized by a conglomerate of blood vessels of variable caliber ranging from small to large [Figure 4a-d]. The vessel walls were irregular with no obvious elastic lamina, favoring a venous type of vascular malformation.

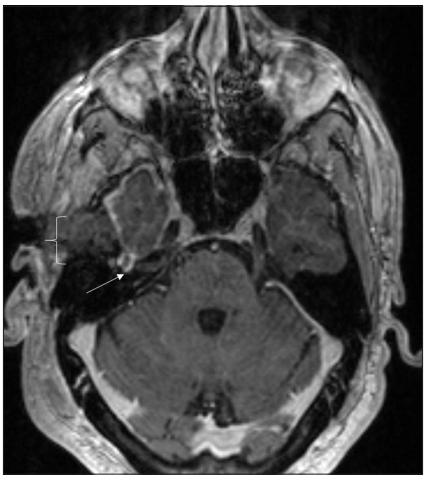

Figure 3: Axial T1-enhanced magnetic resonance imaging demonstrating a defect within the lesion after partial resection (white arrow). Note the right infratemporal craniotomy changes (white bracket).

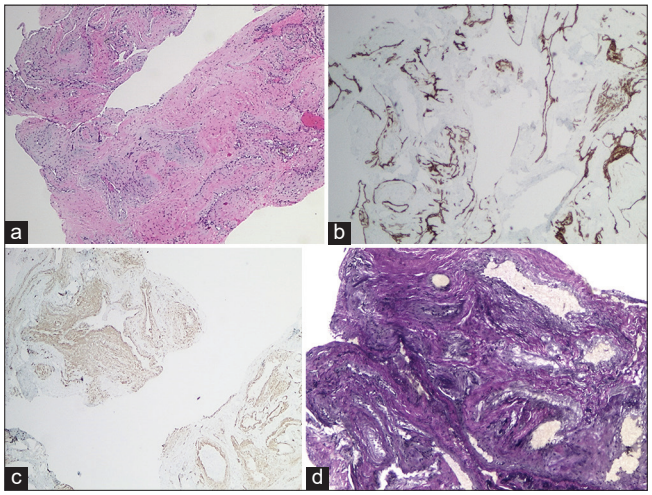

Figure 4: (a) Hematoxylin and eosin stain demonstrating a conglomerate of blood vessels ranging from small to large, (b) CD31 immunostain demonstrating the endothelium lining (brown), (c) smooth muscle actin (SMA) immunostain (SMA positive) demonstrating smooth muscle in the vessel walls, (d) elastin stain demonstrating the absence of elastic lamina, which is typical of venous malformation.

Immediately after resection, the patient regained some function and was able to lift his eyelid, but his facial palsy returned soon after surgery, with overall Grade 4 House–Brackmann palsy. Several months later, the patient underwent facial reanimation surgery and gold weight placement. He had good results postoperatively, but his facial palsy eventually returned. He was noted to have House–Brackmann Grade 4 facial palsy on follow-up appointments. The patient also reports residual symptoms related to crocodile tears syndrome (CTS).

DISCUSSION

Facial nerve venous malformations (FNVMs) are slow-growing, benign vascular lesions that arise from perineural capillary networks. They may involve any segment of the facial nerve and most commonly involve the GG.

FNVMs were previously described as hemangiomas and initially characterized on CT has have irregular ill-defined margins with intralesional bone spicules in a honeycomb pattern.[5]

On MRI, FNVMs are T1 isointense to hypointense, T2 hyperintense relative to adjacent gray matter and avidly enhance.[1,4,11,16,23] Histologically, lesions are characterized by hematoxylin-eosin stains demonstrating dilated vascular spaces lined with endothelial cells that are mitotically quiescent with no internal elastic lamina.[2]

FNVMs account for <1% of temporal bone lesions and 18% of facial nerve tumors.[9] The peak incidence of FNMVs occurs between 30 and 60 years of age slightly more common in females.[6] Most commonly, lesions are reported to be centered on the GG with patients presenting with both progressive and sudden facial paralysis and spasm.[16] Other symptoms include conductive hearing loss, otalgia, pulsatile tinnitus, aural bleeding, and vertigo. Potential explanations for neural injury have included nerve compression, vascular steal, and invasion.[7]

To the best of our knowledge, parasympathetic symptoms related to facial nerve injury have not been reported with FNMV.

CTS, also known as Bogorad syndrome, is characterized by lacrimation caused by olfactory and gustatory stimuli and mastication. The phenomenon is also known as paroxysmal lacrimation or the gustolacrimal reflex.[3,10] This most often occurs in patients recovering from Bell's palsy. The most widely accepted theory is due to injury to the nervus intermedius either due to Bell's palsy or traumatic injury. The nervus intermedius consists of fibers from the superior salivary nucleus which is located in the medulla. The nervus intermedius contains parasympathetic fibers whose stimulation results in secretion of the lacrimal and submandibular glands. Traveling along these fibers is afferent gustatory sensory fibers from the chorda tympani whose cell bodies reside in the solitary nucleus.

During the recovery from Bell's palsy, the parasympathetic fibers undergo regeneration but are misdirected, with fibers previously destined for salivary glands growing to the ipsilateral lacrimal gland. The regenerating nerves grow along the greater superficial petrosal nerve (GSPN) which results in olfactory or gustatory stimulation causing ipsilateral lacrimation.[20] An alternative theory is thought to be the formation of an artificial synapse at the injury site allowing the salivary fibers to jump to the GSPN, leading to lacrimation symptoms.[13]

The incidence of Bell's palsy is 0.08% per year and increases with age.[15] CTS is a relatively uncommon complication of Bell's palsy, occurring in approximately 3.3% of patients with symptoms occurring approximately 6–9 months after the onset of facial paralysis.[21]

Treatment

The most commonly accepted form of the treatment of FNVM is surgical excision.[17] Tumors that are extraneural can be safely separated and resected from the nerve.[18] Preservation of facial function is not possible in cases with direct nerve invasion. Small lesions produce less compression of the facial nerve and are more likely to be safely removed while preserving facial nerve function. Larger lesions are more likely to be closely attached to the facial nerve, making it difficult to separate. The nerve may be sacrificed in these cases and reconstructed with an interposition nerve graft.[22] Recurrence of FNVM is rare after complete or partial excision.[19]

Regarding the clinical management of CTS, different treatment options have been used to stop lacrimation including guanethidine to block adrenergic receptors, propantheline bromide, and homatropine hydrobromide drops; however, the side effects have caused them to fall out of use.[12,20]

Surgical management, including excision of the palpebral lobe of the lacrimal gland, neurolysis of the chorda tympani or vidian nerve, and sphenopalatine nerve block, have been suggested to treat CTS but may be ineffective due to redundant innervation and come with significant morbidity such as vision loss and loss of taste.

The most commonly used treatment of CTS is Botox injection of the lacrimal gland. Botox is an acetylcholine esterase inhibitor and stops transmission along the aberrant parasympathetic nerves to the affected lacrimal gland.[13,14] Botox injection has been shown to cause marked improvement or complete resolution of symptoms, with the treatment effects lasting approximately 6 months. Long-term safety and efficacy, however, has not been established.[14]

Differential diagnosis

The most common imaging differential diagnosis for an FNVM is a schwannoma. Schwannomas more commonly occur at the internal auditory canal (IAC) and present with hearing loss. Hemangiomas occurring at the GG or IAC are typically smaller and more symptomatic.[7] FNVM and schwannoma have overlapping imaging characteristics with avid enhancement and smooth osseous expansion and

remodeling of the facial nerve canal and IAC. The presence of intralesional calcification can be used to distinguish FNVMs from facial nerve schwannoma.[8] It has been suggested that the presence of calcification effectively excludes schwannoma from the diagnosis.[6]

CONCLUSION

We report a case of CTS in a 39-year-old male who presented with facial paralysis related to a venous malformation involving the GG of the right facial nerve. To the best of our knowledge, this is the first case of CTS reported in literature related to a vascular malformation. The diagnosis of CTS is usually made 6–9 months after the onset of Bell's palsy and is characterized by lacrimation occurring with olfactory and gustatory stimulation and mastication. The most common treatment option is the injection of Botox directly into the lacrimal gland with relief of symptoms lasting approximately 6 months.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Rao D, Fiester P, Rahmathulla G, Makary R, Tavanaiepour D. A case of a facial nerve venous malformation presenting with crocodile tear syndrome. Surg Neurol Int 2020;11:3.