Oculostapedial Synkinesis post idiopathic facial nerve palsy: Something to listen out for!

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BACKGROUND

Idiopathic facial nerve palsy is a common presentation with variable degrees of both severity and resolution . Patients usually present with facial droop and additional can have issues with eye closure, lack of tearing, change in taste and hyperacusis. Synkinesis is a less common symptom but patients can present with varying involuntary movements accompanying voluntary ones. Oculostapedial synkinesis is one of the rarer forms of synkinesis and can potentially be overlooked, despite the frustrating symptoms for patient. The case highlights the condition and reviews how to appropriately investigate and manage these patients

CASE PRESENTATION

A 56 year old gentleman developed a sudden lower motor neurone facial nerve palsy in October 2015. This was graded as a House-Brackmann classification¹ as a grade VI palsy which was initially treated with high dose prednisolone in the community. He has no other medical co-morbidities of note, no allergies, is a non-smoker and is a retired forge worker. He was referred by his GP to the neurology department as his facial palsy showed no signs of improvement initially, however this gradually improved until he was left with a grade II persistent palsy. After an incidental contralateral meningioma was detected on cross sectional imaging he was referred to the neurosurgeons who managed this conservatively. However, the neurosurgeons referred this gentleman to ENT as despite the improvement of facial nerve paralysis; he has a persistent sensation of crackling in the ear and an inability to hear when eating. On further questioning he complained of a distortion of hearing on eye closure. He had no other associated ontological symptoms; no hearing loss, tinnitus, otalgia or otorrhoea.

DIAGNOSIS AND MANAGEMENT

A diagnosis of oculostapedial synkinesis was made. The incidental meningioma is being followed up in the lateral skulls base clinic with interval imaging and requires no current intervention. The patient was referred to Birmingham for specialist physiotherapy with an expert dealing in facial palsy patients. This conservative management did not improve the patients symptoms and the patient was keen to consider surgical intervention.

A right sided tympanotomy and transection of the stapedial tendon was proposed and the patient was warned to expect an increased sensitivity to loud noises post-operatively and was consented for the potential intra-operative complications of facial nerve injury, change in taste due to chorda tympani damage, tinnitus, vertigo and decrease in hearing. The patient opted for surgery, which was uneventful with no complications. The patient was seen post operatively and had complete resolution of his ontological symptoms immediately.

DISCUSSION

ANATOMY – The facial nerve is complex and has the following branches;

1. motor fibres to the muscles of facial expression and stapedial muscle.
2. Sensory fibres to the ear via the post-auricular nerve.
3. Viscerosecretory fibres to the lacrimal, submandibular and sublingual glands.
4. Gustatory fibres in the form of the chorda tympani (see image).

The role of the stapedial muscle is protective one against loud acoustic stimuli. In the presence of a loud noise, the stapes is pulled posteriorly by the stapedial muscle and tendon, restricting the movement of the ossicular chain and thus limiting conduction of sound energy to the inner ear.

PALSY RESOLUTION – Although the majority of patients who have had a facial palsy improve, upto 30% will continue to have some residual asymmetry as well as having issues with myoclonus, hyperkinesis, synkinesis and spasm. Synkinesis is the presence of involuntary movements alongside voluntary muscle movements. An example of this other than oculostapedial synkinesis is oculo-oris synkinesis or eye closure upon moving the mouth.

WHY DOES SYNKINESIS OCCUR?

There are various theories proposed as to why synkinesis develops³

1. The first is that anomalous regeneration occurs at a site of injury, causing cross-branching of developing axons into inappropriate nerve tubules which results in facial nerve fibres aberrantly innervating muscles of face and causing involuntary action.
2. Ectopic excitation due to lowered depolarisation thresholds induced by injury.
3. Excess intramuscular innervation due to motor end plate axon sprouting, causing anastomoses that connect peripheral nerve branches.
4. There is also a central theory, in which nerve injury causes retrograde changes of the cells in the facial nucleus which cause a hyper-excitable environment for neural activity⁴.

OCULOSTAPEDIAL SYNKINESIS ACTIVITY

The symptoms that are described specifically with oculostapedial synkinesis are dullness to the hearing or a low-pitched tinnitus, which has been described as ‘tympanic noise’⁵, which are associated with facial movement and particularly eye closure. Oculostapedial synkinesis has been described both post-facial palsy^{6, 7} and without facial palsy⁸.

The treatment of the patients involved surgical resection of the stapedial tendon.

INVESTIGATIONS AND MANAGEMENT

Oculostapedial synkinesis is a subjective phenomenon and is difficult to show subjective evidence for on examination. However, variability in tympanic membrane compliance with voluntary facial movement has been identified^{3,6-8} and upto 87% of 47 patients included in a study had changes in middle ear compliance when closing their eyes voluntarily or during orbicularis oculi spasm⁵. In all reported cases, the treatment of patients involved surgical resection of the stapedial tendon.

INVESTIGATIONS

MRI and CT – No facial nerve pathology. Incidental finding of contralateral left-sided cavernous sinus intradural meningioma.

Blood tests – All normal

FBC, TFT, UnE, ESR, Coagulation screen, LFTs, Vit D, Serum ACE level, Serum Borrella burgdorferi,

Repeat MRI w facial nerve sequences

Enhancement of the horizontal portion of the facial nerve bilaterally, which was deemed to be insignificant. No definite enhancement within the right geniculate ganglia. Seventh and eighth cranial nerves normal bilaterally.

AUDIOMETRY

Pure tone audiometry revealed normal hearing thresholds bilaterally. Impedance audiometry showed Type A tympanograms. No notable impedance on facial movement.

CONCLUSION / KEY POINTS

- Synkinesis is the involuntary movement of muscles accompanying voluntary movements and can occur after facial nerve palsy
- Oculostapedial synkinesis is one of the rarer forms of synkinesis but can present with distortion of hearing on facial movement.
- Oculostapedial synkinesis is a clinical diagnosis with subjective symptoms.
- Objective evidence of stapedial synkinesis has been described in the literature and can be shown with variance of tympanic membrane compliance with facial movement.
- Transection of the stapedial tendon has been shown to resolve the symptoms of oculostapedial synkinesis.

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