Facial Nerve Neuroma Presenting With Unilateral Conductive Loss And Dryness Of Ipsilateral Eye

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Introduction

Whenever an unexplained unilateral sensorineural hearing loss is encountered all attempts should be made to rule out a possible intracranial space occupying lesion (Kileney, et. al., 1991). We all know this golden rule. But what if an unexplained unilateral conductive hearing loss is encountered. Can it be due to an intracranial space occupying lesion ? Occasionally, yes. An early sign of a facial nerve neoplasm may be a conductive hearing loss, if the mass is within or extends to the tympanum and mastoid (Coker and Fisch, 1984). We had the opportunity to evaluate such a rare patient. After initial description by Schmidt in 1930, only 260 such facial neoplasms have been reported in the literature (Sanna, et. at., 1990).

Clinical Details

This stoic 42 year old Sudanese gentleman living in Abu Dhabi presented to us with a progressive hearing loss and tinnitus in the right ear of 2 years duration. A year after the onset he developed mild left sided headache and recurrent vomiting along with vague pain in the left side of the body. He suffered partial seizures involving right extremities and episodes of unexplained strange smell, perceived by the subject. Notable and interesting was the patient's retrospective observation that his right eye and nostril were getting dried for the past six years. There was no significant neurological deficit on clinical examination except for reduced hearing sensitivity. A very mild right facial weakness was detected later on, prior to surgery. There was no papilledema, objective sensory deficit on the face or body, cerebellar signs, field cuts or long tract signs. There was no difficulty in balance tests.

Skull X-ray did not show any significant findings. C.T. Scan showed a moderately large enhancing mass with rims of calcification involving right temporal lobe (fig.1). MRI scan showed a right middle fossa extra axial with a calcified rim. Carotid and vertebral angiography showed the mass to be avascular, mid-temporal in site and unassociated with any pathological supply.

CT scan from a 42 year old man showing the presence of a large mass (Facial neuroma). Audiological tests indicated a conductive loss in the ipsilateral side and **ABR** indicated a conductive delay as well a prolonged brainstem conduction time.

The patient opted to go to London for surgery which was done by Prof. Lindsay and Prof. Cheeseman at National Hospital, Institute of Neurology, U.K. The

mass was completely removed necessitating dissection into the middle ear. The facial nerve had to be divided just as it emerged from the the internal auditory canal and a dural nerve graft was inserted. Because of these maneuvers, the patient lost his hearing in the right ear. Otherwise he remained well after surgery.

Audiological test environment and equipment:

Tests were conducted in a prefabricated sound treated room. Pure-tone speech audiometry were carried out using a commercially available diagnostic audiometer (Amplaid 455). For speech testing Arabic speech material developed by Alusi et. al. (1974) were used. Immittance audiometry was done with a Hortmann (CIM 85) impedance audiometer. Evoked potential tests were carried out using MK 10 (Amplaid) single channel system. Scalp, disc silver/silver chloride electrodes were used in acquiring potentials. The contralateral mastoid served as the ground. Care was taken to maintain an electrode impedance across the electrodes below 3k ohms. We used a wide spectrum click stimuli generated by a 100 msec rectangular electric wave applied to a TDH 49 ear phones housed in cushions and an anti static cover made of mu metal. Filter range was 150 to 3000 Hz. Click rate was 11 per sec. The tests were done at two different intensity levels, namely at 60 dB SL to compensate for the right air-bone gap, and the other was done at maximum output of the system i.e., 100 dB HL (130 dB pe SPL).

Results

Immittance audiometry showed 'A' type tympanogram in the left ear and pulsatile rounded tympanogram with an extreme negative pressure . Reflexes were absent in the right probe ear, however when the probe was in the left ear, ipsilateral reflexes were observed at normal levels. When the right ear was the phone ear, the reflexes were elevated i.e., greater than 100 dB HL. Pure-tone audiogram in the left ear showed a mild high frequency drop. PTA2 (Average of 1000,2000, 4000 Hz) was 8 dB HL in the left ear and in the right ear it was 43 dB HL. Bone conduction thresholds in the right mastoid were within normal limits, and thus showed a significant air-bone gap, suggesting a mass tilt. Tone decay test done at 20 dB HL (Olsen and Noffsinger, 1974) was negative in both the ears. Speech discrimination scores were excellent. Introduction of competing speech noise in test ear or non-test ear at 0 dB MCR did not influence the performance. Auditory brainstem response audiometry was done with patient in a supine position. The patient was asked to relax and if possible to go to sleep. Peak I-V difference in the left ear was 4.32 msec. Although this value was greater than the normal range it was acceptable because of the high frequency drop in audiogram. However on the right ear the ABR waves were not only delayed, but also prolonged (4.656 msec). Peak V difference between the ears was (ILD) 0.912 msec (although it was contaminated by air-bone gap). It was extended beyond the expected due to a conductive hearing loss. Thus ABR indicated a conductive lesion, as well as neural lesion. Immittance, pure-tone, speech and special tests corroborated with a right conductive disorder. Caloric responses were preserved in both the ears.

Electro-diagnostic study of facial nerve showed mild degenerative changes and a marginal asymmetry of compound muscle potential with mild nuclear or facial nerve involvement.

Discussion

Facial nerve neuroma is a rare slow growing neoplasm. Brackmann and Bartels (1979) observed only 16 such facial neuromas out of 1354 cerebello pontine angle tumors. It can originate along the course of facial nerve any where from the brainstem to the parotid (Pulec, 1981). Most of them arise in the region of the geniculate ganglion and may extend along the facial nerve into the internal auditory meatus or as far as the parotid. The earlier symptom of dryness of ipsilateral eye and nose is explained by involvement of parasympathetic symmetry fibers (Nervous inter medium) that accompany facial nerve from pons to the geniculate ganglion. "Failure to close the eye especially if lacrimation is reduced will add to drying out of the cornea "(Barr and Irvine, 1990). This gentleman did not have difficulty closing the eyes. In fact this seems to be the harbinger, and is not commonly reported in the literature as a sign of facial neuroma. Facial motor function was not disturbed for several years, where as the conductive loss was an earlier disturbance.

This experience alert us to watch for facial nerve neuroma in any unexplained/unresolving conductive loss (with a flat or rounded tympanogram) necessitating a thorough otoneurological evaluation.

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