Vestibular Schwannoma - A Dangerous Cause of Sensorineural Hearing Loss in Pilots

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Introduction

Sensorineural Hearing Loss (SNHL) is a common incidental finding in middle aged Pilots reporting for Medical Examination. Often, the individual may not even be aware of the disability as the speech frequencies are mostly unaffected till later. The aetiology of the condition can range from the innocuous (Age related Presbyacusis) to the alarming (Noise or Drug induced) to potentially life threatening (Intracranial tumor). Since the vast majority of aircrew are routinely exposed to noise in the form of aircraft engines, radiotelephony sets, etc, there is a predisposition to attribute all SNHL to occupational noise exposure. However, this may not always hold true. A high index of suspicion should be maintained by the medical examiner when screening fresh cases of SNHL.

Key Words: Sensorineural Hearing Loss, Vestibular Schwannoma, Intracanalicular Tumors

Case Report

Case No 1: A 45 year old transport pilot with over 7600 hours of flying time reported for routine medical examination. He had been diagnosed as a case of triple vessel Coronary Artery Disease six months previously and had undergone Coronary Artery Bypass. He was asymptomatic, on regular medication in LMC A4G4 (T-24). He gave no history of having noticed any hearing loss.

During ENT examination, it was observed that the individual was able to hear Forced Whisper (FW) only at 400 cm in the left ear. Conversational Voice (CV) was heard in this ear at the normal 600 cm. The other ear had normal hearing with both CV & FW at 600 cm. Both tympanic membranes were found to be intact & mobile. Rinne's tuning fork test was positive, Air Conduction (AC) being better than Bone Conduction (BC) on both sides. Weber's test was carried out using 512 Hz tuning fork and revealed lateralization of sound to the right ear (better side). Absolute Bone Conduction (ABC) was reduced on the left side. Otoneurological examination revealed no abnormality. The individual did not have any spontaneous or positional nystagmus. Gait was normal. There were no cerebellar signs. There was no evidence of any other cranial nerve weakness.

Nose and throat were examined and found to be normal. Pure Tone Audiometry (PTA) revealed normal AC hearing thresholds for left ear till 1 KHz, followed by a dip to 60 dB at 2 KHz, 75 dB at 4KHz, 90 dB at 6 KHz and improving thereafter to 65 dB at 8 KHz. BC thresholds were 5- 10 dB less than AC thresholds across all frequencies. The right ear had normal AC thresholds at 15-25 dB across all frequencies, except for a small dip to 35 dB at 6 KHz. BC in the right ear was at 10 – 15 dB across all frequencies, except for 6 KHz where it was at 30 dB. Speech Discrimination Score (SDS) in left ear was abnormally poor (50%) as compared to the right ear (98%). Impedance Audiometry revealed bilateral 'A' type tympanogram. Caloric response of left ear was decreased as compared to the right ear for both hot and cold stimulation. MRI examination was carried out using T2 and T1 weighted images. A well-defined, lobulated, extra axial mass was seen in the cerebello pontine angle cistern on the left side measuring 18x19x21 mm. It was hypointense on T1 and heterogeneously hyperintense on T2 and showed hetergenous post contrast enhancement. It extended into the left internal auditory meatus, which was widened. There was a mass effect in the form of indentation on the left side of pons

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and anterior aspect of left cerebellar hemisphere. On the basis of the examination and investigation results, a diagnosis of Vestibular Schwannoma (Left side) was made.The individual was downgraded to A4G4(P). He was advised Surgery by the Neurosurgeon, but was unwilling. He underwent treatment with Gamma Knife at a higher centre.

He was reviewed again six months after treatment. The tumor had not progressed in size. Hearing of the left ear was well preserved with CV/ FW being 600/ 400 cm for the left ear and 600/ 600 cm for the right ear. His Vestibular system was found to be well compensated. The individual had a normal Gait and Romberg test. There was no deficiency of Cranial nerve function. PTA revealed hearing thresholds for AC better than 25 dB till 1 KHz, followed by a dip to 70 – 80 dB for the higher frequencies. Hearing thresholds for the right ear were within normal limits. SDS was 99% for the right ear and 55% for the left ear.

In view of the successful treatment with preservation of good hearing and lack of any residual cranial nerve deficits, he has beenupgraded to A3G2(T-24) with a view of gradually returning him to flying duties.

Case No 2: A 46 year old civilian pilot reported to his doctor with complaints of gradually progressive right sided hearing lossof two years duration. On questioning, he also admitted to having occasional tinnitus. However, there was no history of vertigo, headache, facial weakness, visual disturbances, altered sensorium, nausea or vomiting

During ENT examination, it was observed that the individual was unable to hear Conversational Voice or Forced Whisper with the right ear when the left ear was adequately masked. The other ear had normal hearing with both CV & FW at 600 cm. Both tympanic membranes were found to be intact & mobile. Rinne's tuning fork test was false negative on the right side. Weber's test was carried out using 512 Hz tuning fork and revealed lateralization of sound to the left ear (only hearing ear). ABC revealed severely reduced hearing on the right side after the left ear was masked. Otoneurological examination revealed no abnormality. The individual did not have any spontaneous or positional nystagmus. Gait was normal. There were no cerebellar signs or evidence of any other cranial nerve weakness. Nose and throat were examined and found to be normal. PTA revealed profound loss of hearing on the right side, with only an island of hearing from 250 Hz – 2000 Hz at 90 – 100 dB. Hearing on the left side was normal, with AC and BC thresholds better than 15 dB across all frequencies. Impedance Audiometry revealed bilateral 'A' type tympanogram. Caloric testing revealed a hypoactive labyrinth on the right side with normal responses on the left side. MRI examination was carried out using FIESTA sequences. A heterogeneously enhancing extra axial lesion was noted in the right cerebello pontine angle measuring 23 x 15 mm. The lesion extended into the internal auditory meatus and was continuous with the eighth cranial nerve. The intracanalicular portion measured 10 x 5 mm.

On the basis of the examination and investigation results, a diagnosis of Vestibular Schwannoma (right side) was made.

The individual underwent treatment with Gamma Knife. He was evaluated six months later. Tumor size had shrunk by 2 -3 mm. His vestibular system had compensated and his Romberg test and Gait test were normal. There were no cerebellar signs. Profound SNHL on the right side remained unchanged with normal hearing on the left side. SDS was absent on the right side but 99% on the left side. An executive report was asked for, which was complimentary.

In view of the above, he was made fit for flying as Pilot-in-Command with independently qualified co-pilot, which was duly approved by DGCA.

Discussion

Sensorineural Hearing loss (SNHL) is a relatively common finding in middle aged and older individuals. 40 - 45% of patients referred for hearing evaluation are found to be having mild - moderate SNHL [1]. This hearing loss may result from damage to the hair cells caused by noise exposure, viral infections, and fractures of the temporal bone, barotrauma, cochlear otosclerosis, Meniere's disease or aging. Drugs such as salicylates, quinine and its analogues, aminoglycoside antibiotics, loop diuretics and cancer chemotherapeutic agents can often

produce SNHL. However, hearing loss may also be due to cerebellopontine angle tumors such as vestibular schwannomas or meningiomas, demyelinating disease such as multiple sclerosis, vascular disease such as stroke, infections such as meningitis, or immune disorders such as Polyarteritisnodosa or HIV [2]. A finding of SNHL, must therefore never be dismissed as incidental.

In the Aviation Field, we have a unique system of regular medical examination for all aircrew. These evaluations offer an unrivalled opportunity for early detection and intervention of a vast spectrum of diseases. The finding of hearing loss should be treated just as seriously as that of an ECG abnormality meriting further investigation and evaluation.

Though less than 1% of cases of SNHL can be ascribed to lesions in the Cerebellopontine angle, Vestibular Schwannoma comprises over 80% of these tumors. 95% of Vestibular Schwannoma occurs in sporadic fashion, being due to recessive mutation of chromosome 22q12 which leads to deficiency of a cytoskeletal protein (Merlin) resulting in over proliferation of Schwann cells of the superior or inferior vestibular nerves at the transition zone (Obersteiner-Redlich zone) of peripheral and central myelin. The remaining 5% cases are familial and occur in association with Neurofibromatosis Type II [2].

Vestibular schwannoma typically presents as a unilateral hearing loss, alongwith occasional tinnitus. As it grows in size, it progressively causes other symptoms such as facial nerve palsy, numbness of midface, and decreased visual acuity, diplopia, headache, nausea, vomiting, vertigo,and dysphagia, hoarseness of voice, altered sensorium and ultimately death due to central respiratory compromise. The growth rate is usually slow, averaging 1.8 mm per year.

The hearing loss is typically retrocochlear in nature. Speech discrimination scores are typically much worse than predicted by the pure tone threshold. Indeed, asymmetric hearing loss, especially at 3 Khz in combination with a poorer SDS should raise a suspicion of possible Vestibular Schwannoma [3] though there is no single 'typical' audiogram [4]. There may also be loss of the acoustic reflex or presence of acoustic reflex decay. Vestibular testing typically shows reduced caloric response in the problematic ear but is neither sensitive nor specific.

Brainstem Evoked Response Audiometry (BERA) has over 90% sensitivity and specificity in detecting Vestibular Schwannoma. There is typically a delay in Wave V latency or an absence in identifiable wave peaks. However, in 30% cases, a small intracanalicular tumors may be missed.

The Gold Standard of diagnosis is Magnetic Resonance Imaging with Gadolinium Contrast. High Resolution Fast Spin Echo T2 weighed image can also be used to obtain scans in less time without the need for contrast [5].

The diagnosis of Vestibular Schwannoma as the cause of mild SNHL in a pilot is a challenging situation as it may adversely affect his occupational ability. Mild SNHL in itself does not necessitate any restriction on flying. However, the finding of intracranial tumor necessitates a ground category. Furthermore, the proposed treatments for Vestibular Schwannoma may adversely affect the hearing and/or balance. So, even after successful treatment, the issue of returning to flying duties needs to be considered seriously.

Vestibular Schwannoma can be treated by invasive neurosurgery or non-invasive Stereotactic Radiation (Gamma Knife) [6]. Neurosurgery entails exposure of the tumor in the CP Angle by a Translabyrinthine, Retrosigmoid or Middle Fossa approach. The translabyrinthine approach results in a total loss of hearing in that ear. Retrosigmoid and Middle Fossa approaches preserve hearing, but have disadvantages of prolonged brain retraction and reduced exposure. The facial nerve may be at risk, especially with a middle fossa approach. Post-operative complications include CSF leak, haemorrhage, stroke and meningitis [7]. Many patients have post-operative vertigo which may take a few months to subside.

Stereotactic Radiation entails treatment using several precisely collimated beams of ionizing radiation which pass through different paths to deliver a high dose to a specific intracranial target.Meta-analysis of several studies has

revealed that Stereotactic Radiation offers significantly better long term hearing preservation as compared to surgery, with similar long term tumor outcome [8, 9]. It is suitable for patients with tumor size 2 – 3 cm who have good hearing, or those in whom surgery is not possible. The aim is to arrest the further growth of the tumor, while preserving hearing, balance and facial nerve function. Control rates of upto 97% with 10 year followups are achievable [10]. Hearing is preserved unchanged in 50 – 77% of cases. Complications such as facial nerve dysfunction, trigeminal nerve neuropathy and hydrocephalus occur in less than 1%. Long term followup with regular MRI scans will be required.

It is noteworthy that when asked to choose a treatment modality for themselves, most Neurosurgeons preferred Stereotactic Radiosurgery even though they had recommended surgery for the patient [11].

After treatment for intracranial tumor, the individual should be given 6 months to 1 year to recover. Subsequent assessment must include a complete evaluation of hearing levels, balance and cranial nerve function. IAP 4303 para 6.11.5 lays down the requisite standards of hearing for military aircrew [12].

A minimum of 450 cm Conversational Voice and 300 cm Forced Whisper will be required for A2 category. If the hearing is less than this but is better than 300 cm CV and 200 cm FW, flying may still be possible in category A3 if the SDS is better than 50%. ICAO para 6.3.4.1 lays down a minimum standard of 35 dB at 500 Hz, 1 kHz or 2kHz and 50 dB at 3 kHz for each ear, separately [13]. Hearing loss greater than this can also be declared fit provided there is a minimum of 50% score on SIT and he can hear CV at 200 cm. The individual should have no major balance or cranial nerve dysfunction.

Clinical Practice guidelines for Acoustic Neuroma developed by the American Society of Aerospace Medicine Specialists [14] stress on the adequate functional performance of the aviator, post treatment. Conservative observation is not deemed consistent with the safe performance of cockpit duties, due to the ever present risk of sudden deterioration and abrupt symptomatic progression of the untreated tumor. However, six months after successful treatment of a unilateral tumor with surgery or Gamma Knife, the aviator may be made fit, provided that there is complete resolution of post treatment vertigo and there is no facial weakness, restricted eye movements, strabismus or tracking deficit. There should be acceptable protective mask sealing, which may not be possible if there is residual facial nerve palsy. Unilateral hearing loss, even total loss, may be waiverable provided adequate hearing remains in the other ear.

In a study of over 120,000 medical examinations conducted on over 40,000 pilots between Sep 2002 and Mar 2010 at the ENT Dept at the National Pilot Expertise Center in Clamart, France, only 10 pilots were diagnosed with Vestibular Schwannoma. Of these, only 2 experienced a negative impact on their fitness to fly [15]. With world class medical facilities now available in India, we should be able to offer similar a standard of care to our pilots.

A diagnosis of Vestibular Schwannoma need not mean the end of a Pilot's career. The tumor can be treated by Gamma Knife, and as long as there is no further progression of the tumor with no worsening of SNHL and no complications such as facial palsy or persistent vertigo, the pilot can be allowed to fly.

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