Recent Advances in Neurotology

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Introduction

The past decade has seen numerous advances in our understanding, diagnosis and management, both medical and surgical, of patients with inner ear dysfunction and disorders of the cerebellopontine angle and brainstem.

Benign positional vertigo (BPV) remains the most common inner ear disorder seen in neurotology. In this condition patients experience recurrent attacks of episodic vertigo lasting seconds when the head reaches the provocative position (e.g. looking up, bending over or moving from a sitting to a lying position). BPV usually occurs spontaneously but may also result from head injury. BPV following head injury is more likely to be persistent and bilateral, and less likely to respond to physical therapy (i.e. Brant-Daroff exercises, Semont’s and Epley’s manoeuvres).

The discovery of free-floating particles in the posterior semicircular canal (PSCC) of the inner ear at surgery provides the rationale for particle (or canalolith) repositioning in the treatment of BPV. This simple procedure has proven extremely effective in providing patients with relief. However, when BPV does not respond to particle repositioning and proves incapacitating non-ampullary posterior semicircular canal occlusion surgery is usually recommended, providing there is no contraindication to surgery. At the University Health Network this procedure has been universally curative in 32 consecutive patients with incapacitating BPV over the last 15 years with no deterioration in their sensorineural reserve. Autogenous periosteum is typically used to occlude a fenestration made into the posterior canal at surgery in part to prevent movement of the free-floating particles causing BPV. Unlike singular neurectomy (perhaps the most difficult of all procedures in neurotology) the advantage of non-ampullary PSCC occlusion is that it defunctions rather than deafferentates the posterior canal.

The discovery that PSCC occlusion surgery could selectively defunction this canal without ill effect to the rest of the inner ear has led to new surgical approaches through the inner ear that maintain function but allow access to the antero-medial brainstem and cerebellopontine angle (CPA) for treatment of brainstem vascular lesions such as cavernomas or basilar aneurysms that hitherto had been considered untreatable. With this approach, both the posterior and superior semicircular canals are occluded and then resected providing the surgeon direct access to the internal auditory canal and the anterior aspect of the CPA. In nine patients operated on at the University Health Network the sensorineural reserve for hearing was maintained at its preoperative level in seven of nine (77%) patients. Preservation of partial vestibular function (i.e., the lateral semicircular canal and the utricle and sacule) also appears possible.

Other advances have occurred in the systematic analysis of eye movements their interaction with the inner ear. For example, our knowledge of how the vestibulo-ocular reflex (VOR) works in the real world has been significantly enhanced by the introduction of magnetic scleral coil eye movement studies. The subject wears a contact lens with an embedded wire and a similar coil attached to their head, and is placed inside a magnetic frame. Movements of the eyes or head relative to the magnetic field induce a current within the coils that can be measured precisely. This new technology has allowed us to accurately record even the most minute movements of the eyes as they respond to head movements overcoming previous technical problems such as electrode slippage and electrical interference.

Using scleral coil technology we have found that the vestibular portion of the inner ear now appears to have a frequency specific function (i.e. high frequency vs low frequency) in a fashion somewhat analogous to hearing. This explains the phenomenon of oscillopsia (visual blurring with head movement) in patients with normal conventional balance tests and explains why certain individuals cannot adequately compensate following a unilateral vestibular loss despite active vestibular rehabilitation therapy. For the most part caloric testing has been found to be a test of low frequency vestibular function. Although this has not lead to any formal treatment of patients with a high frequency vestibular
loss, the knowledge gained from this test is often invaluable in medicolegal and forensic cases involving workplace and disability issues.

Over the last decade we have witnessed advances in the basic molecular science of how ototoxic agents affect the inner ear. Our increased knowledge of the mechanisms for ototoxicity, both systemic and topical, has tremendous implications for future patient care, prevention of complications and possible treatment of disorders such as Meniere's disease. Intratympanic gentamicin therapy for incapacitating Meniere's disease has successfully relieved vertigo in 80% to 100% of series to date and as a result has replaced the major neurosurgical procedure of vestibular neurectomy as the initial treatment of choice in patients with serviceable hearing.

Somewhat humbling has been the realization that whatever enters the middle ear can ultimately reach the inner ear. Topical aminoglycoside containing drops have now been irrefutably demonstrated to cause clinical ototoxicity in humans. Surgical preparation solutions, especially those that contain alcohol and chlorhexidine, that reach the middle ear and then the inner ear through the round window membrane may also be ototoxic.

We have discovered the genetic basis for the increasingly frequent clinical phenomenon of aminoglycoside antibiotic induced deafness (AAID) and now have a better understanding of cellular injury and apoptosis in individuals exposed to prolonged or toxic doses of aminoglycosides and platinum based chemotherapeutic agents.

Basic research into AAID has demonstrated a mitochondrial pattern of inheritance. There is a mitochondrial DNA 1555 A to G mutation in the 12s ribosomal RNA making it resemble a bacterial 12s ribosome. This increases aminoglycoside binding in the ribosome, which alters protein synthesis leading to the formation of iron binding complexes and free radical formation ultimately resulting in cell death.

Conclusion

The phenomenon of AAID is especially important in Southeast Asia. For example, populations in China appear to have an extraordinary propensity for severe deafness following a single exposure to the aminoglycoside kanamycin. In Shanghai, it has been estimated that over 22% of deaf mutes could attribute their hearing loss to short term aminoglycoside use (often only one dose). Exposure to aminoglycosides (including drops reaching the middle ear) may also be responsible for premature progressive sensorineural hearing loss. We hope that with further advances in molecular biology and genetics we may soon have a way of preventing this type of hearing loss in susceptible individuals and better understand the mechanisms that cause age related changes in hearing and vestibular function.