Original Article

Otologic manifestation of systemic disease

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Abstract

Problem statement: It is well established that numerous systemic dis¬eases have accompanying otologic manifestations. The diseases include syndromic genetic disorders such as Usher syndrome, branchio-oto-renal syn¬drome, Pendred syndrome, Jervell and Lange-Nielsen syndrome, Treacher Collins syndrome, and many others. In addition, other systemic diseases are asso¬ciated with hearing loss (e.g., osteogenesis imper¬fecta, Paget disease of bone, diabetes mellitus, renal disease, hypothyroidism, and others). Over the past 10 years, however, otologic disease has been newly noted to be associated with several inherited and acquired disorders. This chapter briefly discusses the more important of these.

Key Word: Otologic manifestations, Erythematosis, Otosclerosis, Osteogenesis Imperfecta, Arnold-Chiari Malformation

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INTRODUCTION

Acquired immunodeficiency syndrome (AIDS) is caused by infection with a retrovirus called the human immunodeficiency virus (HIV). It is now well recognized that otologic manifestations are common in HIV infections. Further, as our understanding of the infectious disease process and its treatment has progressed, AIDS is increasingly being thought of as a chronic condition as opposed to a fatal one.

Most authors conclude that there are a variety of mechanisms by which HIV infection can result in oto-logic symptoms, and, in many cases, the HIV infection may not have progressed to AIDS. With a typical 10 year window between infection and the onset of symptoms with current treatment, HIV infection may be a possible etiologic factor in hearing and vestibular symptoms with or without other manifestations of HI V infection. The clinician should always keep HIV infection as part of the differential diagnosis of earlyonset hearing loss or vestibular deficit.

Patients with AIDS are subject to systemic infections that may affect the inner and middle ear. Several cases of reactivation of otosyphilis have been documented. In addition, AIDS patients are subject to sudden sensorineural hearing loss (SNHL) due to cryptococcal meningitis. Further, *Pneumocystis carinii,* in addition to causing systemic disease and pneumonia, can cause otitis media. It has been demonstrated that recurrent otitis media is significantly more prevalent in HIV-infected children. Otomycosis has also been documented in this population. More serious infectious.disorders, such as skull base osreomyelitis/malignant otiffs externa caused by organismsIncluding *Pseudomonas aeruginosa* and *Aspergilliislumigatus,* have been described.

There is evidence of a higher incidence of oto-vestibular abnormalities in AIDS patients than in the general population, and - 50% of HIV-positive patients, whether symptomatic or asymptomatic, have abnormalities on auditory and vestibular testing. Some of these abnormalities seem to indicate central auditory and vestibular system abnormalities.

Evaluation

Because contemporary chemotherapy has delayed the onset of AIDS symptoms in patients with HIV infection, otologic manifestations of the infection may occur prior to other symptoms. Thus awareness of the condition is paramount. The evaluation is dependent on the presenting condition, and for the most part, mirrors the evaluation of the condition in the absence of HIV. Audiological and radiological studies are guided by the presenting symptoms. Bloodwork, including CD4 cell counts, will help determine the degree of immunosuppression. For most patients, CD4 counts> *500jmm* will not result in the opportunistic infections that are seen in the more severely immunosuppressed (i.e., < *200jmm).*

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Management

As with the evaluation, management is guided by the specific condition. Treatment of infectious processes includ-es directed antimicrobial therapy, which may be toward an unusual organism. Thus cultures of the infectious process are important.

Immune-Mediated Inner Ear Disease

In recent years it has been suggested that progressive hearing loss with and without vestibular symptoms may be related to the production of autoantibodies directed toward inner ear proteins. This concept has been supported by the fact that some patients with progressive hearing loss respond favorably to treatment with systemic steroids with significant improvements in hearing. Some of these patients may have other evidence of autoimmune disease, such as Cogan syndrome, rheumatoid arthritis, Wegener granulomatosis, Sjogren syndrome, or systemic lupus erythematosis. However, many patients have no other stigmata of autoimmune disease. Although it is widely recognized that some patients with progressive SNHL with and without vestibular symptoms may respond to corticosteroids, the exact mechanism of this syndrome is unclear. Most investigators feel that autoantibodies are produced and directed toward the inner ear. Although early studies identified a 68 kD protein as potentially being associated with a positive response to steroids in suspected cases, its unreliability as a clinical tool has led it to fall out of favor. Studies have demonstrated that elevated antibodies to inner ear structures can lead to SNHL in experimental animals, supporting the presumed autoimmune nature of this syndrome.

Evaluation

A rapid and progressive, and possibly fluctuating hearing loss., with or without associated vertigo, tips off the clinician to suspect an autoimmune etiology. The presence of other autoimmune disorders increases the suspicion. Associated conditions, including Cogan syndrome or relapsing polychondritis, should also be sought. Autoimmune screening laboratories, including antinuclear antibodies, rheumatoid factor, complement C3, and anti-DNA-ds antibodies should be ordered to evaluate for systemic disease. Audiological and radiological (computed
tomography [CT] and/or magnetic resonance imaging [MRI]) studies are also indicated. If one suspects a central nervous system etiology, then a lumbar tap can identify an increased protein count in the cerebrospinal fluid.

Management

Management of autoimmune inner ear disease consists first of a course of steroids (typically prednisone 80 mg/kg/d for 10-14 days) with a slow taper. If symptoms persist despite appropriate doses of steroids, or if symptoms recur after tapering off steroids, options include a more lengthy course of steroids or other immunosuppressive medication, including methotrexate, Imuran, or CellCept (Roche, Branchburg, NJ). Coordinating care with a rheumatologist is highly recommended, with close audiological follow-up to document improvement in hearing. For patients who have lost all hearing, cochlear implantation can be considered.

Controversial Issues

There is ongoing uncertainty regarding the length of steroid administration, and what immunosuppressive medications are the most appropriate for this condition.

Otosclerosis

Until recently, otosclerosis has been regarded as a genetic disease without other systemic abnormalities. Histological otosclerosis is found in the temporal bones of up to 10% of autopsy specimens, and - 0.3% of the population has clinical otosclerosis. Several possible etiologies are now recognized that may contribute to the condition, including genetic, environmental factors (such as the use of fluorides in water), infectious agents (measles virus), as well as possible immune-mediated etiologic factors. Otosclerosis has been shown to be an autosomal dominant disease with variable penetrance in some families. Although generally thought to be a genetic disease, observers have been puzzled by the fact that - 50% of patients with otosclerosis have no family history of the disease. Early studies demonstrated measles antigens in osteoclasts in active otosclerosis lesions (otospongiosis), suggesting a measles etiology, further supported by the finding that several otosclerotic specimens demonstrated measles nucleocapsid gene. Link age analysis studies of large families with otoscle-rosis have shown that T cell receptor b is one gene responsible for familial otosclerosis, implicating the immune system as an underlying etiology. Which of these factors plays the most important role in the genesis of otosclerosis remains to be determined.

Evaluation

Otosclerosis is suspected in a patient with a slowly progressive conductive or mixed hearing loss as doc-umented by audiometry. There may be a characteristic "notch" at 2 kHz in the bone-conduction threshold line (i.e., the Carhart notch). As with other diseases causing a conductive hearing loss, acoustic reflexes are absent; the presence of an acoustic reflex implies a third mobile window syndrome (e.g., superior semicircular canal dehiscence). A CT scan may demonstrate otospongiotic changes surrounding the otic capsule. The disease may be unilateral or bilateral.

Management

There are several treatment options for otosclerosis: (1)

observation, (2) conventional hearing aids, (3) bone-conduction hearing aids (e.g., Baha System, Cochlear Americas, Centennial, CO; Sophono, Sophono, Inc., Boulder, CO; SoundBite, Sonitus Medical, San Mateo, CA), and (4) stapedectomy or stapedotomy. Each of these options is associated with a variety of risks and benefits. There are also some early data suggesting that bisphosphonates can be used to treat SNHL associated with otosclerosis (cochlear otosclerosis). Clinical aspects and management of otosclerosis are further discussed in Chapter 63.

Osteogenesis Imperfecta

Osteogenesis imperfecta (01) in its various forms may manifest with ossicular abnormalities, including otosclerosis with stapes fixation. Osteogenesis imperfecta is a systemic, genetic disease affecting the entire skeleton. A majority of cases are due to mutations in the genes COUAl and CollA2, coding for pro-al and -2 chains of type I collagen. It is inherited in an autosomal dominant pattern, and to date, over 1,500 mutations have been described. The disease consists of at least four subtypes (types I-IV). Type I is the most common; it is a dominantly inherited disorder and usually manifests later in life. Type IV also manifests later in life but is a recessive
characteristic. Types II and III manifest very early in life with multiple fractures and growth retardation. Although 01 is a systemic skeletal disease, it often affects the ossicles of the middle ear, and about half of all patients with 01 will develop some form of hearing loss. The conductive hearing loss seen in 01 is correlated with fractures of the stapes, thinning of the stapes footplate, and fixation around the stapedial annulus. Cochlear degeneration has also been described. In advanced cases, a .progressive SNHL occurs in 01 patients with or without surgical intervention, similar to cochlear otosclerosis.

Evaluation

Evaluation of the patient with 01 includes serial audiograms to document the degree and rate of hearing loss. In most cases genetic testing has been completed by the time the patient sees the otolaryngologist. CT scanning will often demonstrate otospongiotic changes of the otic capsule, causing a lucency surrounding the cochlea, often termed a halo effect.

Management

Management for 01 is similar to otosclerosis, though the results of stapedectomy for 01 have been disappointing. Although stapedectomy may correct many conductive losses, results tend not to be as good as in otosclerosis, with a higher risk of incus fracture with prosthesis placement. Some authors have shown that progressive SNHL occurs in 01 patients with or without surgical intervention. In cases of severe to profound SNHL, cochlear implantation is an option.

Arnold-Chiari Malformation

It is now recognized that vestibular and auditory abnormalities are extremely common in patients with Chiari I malformation. Chiari type I malformation leads to increased intracranial pressure. Unlike Chiari type II malformation, which usually manifests in infancy, Chiari I manifests later in life with recurrent headaches, weakness, vestibular abnormalities, and progressive hearing loss. Studies of a large series of patients with Chiari I malformation showed that eighth nerve symptoms were due to traction on the eighth cranial nerve. Although many of the patients identified with Chiari I malformation had central types of neurotologic findings, many had symptoms suggestive of a peripheral loss.

Evolution

Hearing may be followed with serial audiometry. Patients with central or peripheral otologic symptoms who demonstrate symptoms consistent with Chiari I malformation (headaches, oculomotor deficits, and other cranial nerve deficits) should have an MRI scan. Tonsillar herniation is seen with Chiari malformations with or without hydrocephalus.

Management

Management includes referral to neurosurgery for repair.

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