Chapter 15: Acquired Immunodeficiency Syndrome in Otolaryngology - Head and Neck Surgery

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AIDS (acquired immunodeficiency syndrome) is one of the most significant illnesses to affect the world in the past 100 years. Head and neck manifestations of AIDS are common, and patients may in fact present initially to the otolaryngologist - head and neck surgeon with various complaints. It is the aim of this chapter to familiarize the physician with the various presenting signs and symptoms of AIDS in the head and neck, and to give a perspective on the scope of this disease and its devastating impact on the health care system.

Since 1981, the beginning of the "AIDS decade", there have been more than 150.000 cases of AIDS reported in the USA, and more than 55% of these patients have died. The 5-year mortality rate for AIDS patients from the time of diagnosis now approaches 100%. There are currently estimated to be between 1 and 1.5 million people infected with the AIDS virus in this country; by 1993 the annual incidence may be as high as 100.000 cases per year, with the number of cumulative deaths from this disease approaching 350.000. The disease occurs principally among homosexual or bisexual men and intravenous drug users, but other groups are also involved, including the sexual partners and children of those infected, as well as patients who have received contaminated blood or blood products.

AIDS has generally come to be recognized as a syndrome characterized by the presence of one or more opportunistic diseases indicative of an underlying cellular immunodeficiency without any other known cause of immunodeficiency. The causative agent, the human immunodeficiency virus (HIV), is a retrovirus, namely, a virus containing reverse transcriptase that transcribes viral RNA into DNA. The AIDS virus is lymphotropic, attacking primarily T-helper cells, which are central to the function of cell-mediated immunity. Impairment of this function renders the patient susceptible to a variety of opportunistic infections, including those of viral, fungal, and mycobacterial origin, as well as certain unusual malignancies.

AIDS has been reported in all 50 states although the majority of cases have occurred in urban areas along the two coasts (Centers for Disease Control, 1990a). AIDS is not a uniformly distributed disease. Certain groups are clearly at higher risk according to age, sex, and particularly social and sexual behavior (Volberding and Jacobson, 1990). As of 1990, 91% of affected patients have been male and 9% female, but the relative incidence among females is increasing. Approximately 98% of cases have occurred in adults or adolescents and 17% have occurred in children younger than 13 years old. In the USA 68% of patients have been homosexual or bisexual men and 21% have been intravenous drug users. Between 5% and 10% of cases have been attributed to heterosexual contact; 3% of cases have been attributed to receipt of contaminated blood or blood products. With regard to race, 58% of patients have been white, 27% have been black, and 15% have been from Hispanic and other racial groups.

History

According to the information currently available, it seems likely that the principal entry of the virus into the USA occurred in the mid-1970s from Africa, or possibly from Europe or the Caribbean. In June 1981, the Centers for Disease Control (CDC) reported five cases of *Pneumocystis carinii* pneumonia (PCP in previously healthy homosexual men in the Los Angeles area (Centers for Disease Control, 1981). Over the ensuing months, scores of similar cases were reported, along with numerous cases of Kaposi's sarcoma in patients with similar backgrounds (Gottlieb et al, 1981). Reports of these illnesses in patients with intact immune system was unprecedented, and a deficiency in cellular immunity was postulated. Although these reports marked the beginning of the AIDS epidemic, review of infectious disease data suggests that the disease may have been present in the USA 10 or more years before it was recognized.

In 1981 and 1982 cases of AIDS were reported in hemophiliacs, intravenous drug users, and infants born to intravenous drug users (Rutherford and Werdegar, 1989). Shortly thereafter, the first cases of AIDS associated with blood transfusions were reported. On the basis of information available at that time, there appeared to be three modes of transmission: (1) sexual, especially through receptive anal intercourse, (2) through contaminated blood or blood products, and (3) perinatal transmission. It became increasingly apparent that an infectious agent was responsible. As a result of intensive research efforts over the following few years, by 1984 it was evident that the causative agent was a T-lymphotropic virus, now termed *human immunodeficiency virus type I (HIV-I)*. One year after identification of the virus, a reliable serologic test, the enzyme-linked immunoabsorbent assay (ELISA), was developed. The test was noted to be highly sensitive and, when combined with the highly specific Western blot test, was greater than 99% accurate.

AIDS was first defined by the CDC in 1982 as the presence of one or more opportunistic infections or "opportunistic" malignancies occurring in a patient with no other known cause of immunosuppression. As more information accumulated and tests for the HIV virus became available, the definition was revised. The most important revision occurred in 1987 when the case definition of AIDS was expanded to include a broader spectrum of disease, including such manifestations as enteropathic AIDS (HIV wasting syndrome), HIV dementia, and other disorders (Centers for Disease Control, 1987b).

The term *AIDS-related complex (ARC)* refers to HIV-infected individuals who manifest some symptoms of the illness but do not meet the full criteria for AIDS (Hamburg, 1990). However, it is increasingly evident that this is not a separate entity, but rather one stage in the spectrum of AIDS.

Most data indicate that more than 40% of AIDS patients have presenting lesions in the head and neck (Meiteles and Lucente, 1990). In view of this fact, it is essential for surgeons to be familiar with the various manifestations of AIDS, to understand the primary modes of therapy, and to be informed about the optimal means of protecting themselves and other health care personnel from HIV infection.

Otolaryngologic Manifestations of AIDS

Virtually all medical problems of the head and neck have been encountered in AIDS patients. Most conditions present in the same way as they do in patients who are not HIV positive. However, certain conditions do appear to be more specifically characteristic of this disorder.

Otologic manifestations

Patients with HIV infection can have presenting signs and symptoms in the external, middle, and inner ear (Morris and Prasad, 1990; Lucente et al, 1989). The skin of the external ear may be involved with Kaposi's sarcoma (KS), which presents in a similar manner as KS in other skin areas, that is, as a raised, firm, reddish-blue nodule. Diagnosis is made by biopsy with histologic evidence of atypical spindle cells. Therapy, if necessary, may consist of radiation therapy, chemotherapy, or both.

Several cases of *Pneumocystis* infections in the external ear have been reported. These may present as subcutaneous external canal masses or large aural polyps. Biopsy demonstrates the characteristic protozoan, which appears as a round, multiloculate cyst.

Otitis externa and otitis media have been reported in patients with AIDS. The incidence, causative agents, and response to proper therapy do not appear to differ from those cases seen in the general population. However, AIDS patients with recurrent otitis media and otitis media with effusion should be suspected of having a nasopharyngeal mass; they should be examined and treated accordingly. Kaposi's sarcoma and non-Hodgkin's lymphoma have been reported in the nasopharynx in AIDS patients.

Sensorineural hearing loss has been reported frequently among AIDS patients and has been linked to several causes. The immunocompromised patient is susceptible to hearing loss secondary to meningitic infection from a wide variety of pathogens, including fungal, tuberculous, bacterial, and viral organisms. In addition, the HIV virus itself has been shown to be neurotropic and may be the primary cause for the hearing loss. Finally, various potentially ototoxic agents used to treat the infections encountered in AIDS patients may be responsible for hearing loss in some instances.

Skin manifestations

Among the earliest reported manifestations of AIDS were those occurring in the skin. Kaposi's sarcoma, which manifests as a reddish-blue hemorrhagic nodule, is one of the more common findings (Stafford et al, 1989).

Other reports have documented seborrheic dermatitis, giant molluscum contagiosum, and fungal infections of the skin. A recently reported cutaneous manifestation is the giant herpetic nasal ulcer that extends from the nasal skin onto the upper lip.

Nasal and sinus manifestations

In addition to the usual agents that cause sinusitis in the general population, AIDS patients are also susceptible to unusual sinus infections caused by various uncommon bacteria, viruses, fungi, and parasites. Sinus cultures in AIDS patients with symptoms and radiographic findings characteristic of sinus infection have most frequently demonstrated various aerobic bacteria such as *Hemophilus influenza* and pneumococcus. However, additional reports have shown evidence of sinus infection with such agents as *Cryptococcus neoformans, Alternaria alternata, Acanthameba castellani,* and other unusual pathogens. Routine bacterial infections tend to respond favorably to antibiotics and decongestant therapy. Failure of the patient to respond should lead the physician to suspect an atypical pathogen, and sinus cultures should be obtained promptly so that appropriate antimicrobial or surgical treatment can be initiated. It should be remembered that because of their immunodeficient status, AIDS patients tolerate severe infections poorly. For this reason surgical drainage should often be considered earlier in the course of managing sinus and other infections than it would be in the immunocompetent patient.

Giant herpetic nasal ulcers, not seen previously, have been reported in HIV-positive patients. These ulcers begin in the vestibule and can extend onto the septum and facial skin. Acyclovir is the treatment of choice for these lesions, and response is usually quite good.

Kaposi's sarcoma and non-Hodgkin's lymphoma have been reported in the nasal vestibule, septum, and sinuses of AIDS patients. Diagnosis is made by tissue biopsy, and treatment of both diseases may include radiation and chemotherapy.

Oral and oropharyngeal manifestations

The oral cavity is one of the most common sites of head and neck involvement in AIDS and may often be the initial site of symptoms in an HIV-positive patient. Among the most common conditions noted in this region is Kaposi's sarcoma, which has been reported on the palate, buccal mucosa, gingival mucosa, and posterior pharyngeal wall. The lesion appears, as it does elsewhere, as an elevated or flat reddish-blue nodule (Silverman, 1989).

Another common presenting symptom in patients with AIDS is oral, pharyngeal, or esophageal candidiasis (Brahim and Roberts, 1990). This usually presents as a painful diffuse, thick white membrane (thrush) but may also present in a hyperplastic form and an erythematous (atrophic) form. Another manifestation of oral candidiasis is angular cheilitis, which appears as superficial fissuring and inflammation of the epithelium in the angles between the upper and lower lips. The presence of oral candidiasis in an otherwise healthy but high-risk patient may be an early sign of HIV infection. Diagnosis of candidiasis infection can be made with potassium hydroxide (KOH) smear. Treatment should include topical or systemic antifungal medications.

Although not unique to HIV-positive patients, the herpes simplex virus does present in a more virulent manner in AIDS patients than in the general population. These lesions, which are usually 1 to 3 cm in diameter, may occur anywhere throughout the mouth and lips. They appear as painful ulcers that may persist for weeks. Oral acyclovir is recommended for severe or persistent cases.

Other oral lesions reported in AIDS patients include hairy leukoplakia, non-Hodgkin's lymphoma, acute necrotizing ulcerative gingivitis (ANUG or "trench mouth"), cytomegalovirus infection, and painful aphthous ulcers (Greenspan et al, 1988).

Laryngeal manifestations

Epiglottitis has been reported in several AIDS patients and it appears to be caused by the same organisms as in the general population (Ognibene, 1990). The infection responds well to appropriate antibiotic therapy.

Kaposi's sarcoma involving the upper airway is a common complication of HIV infection. The patient may present with stridor as the initial symptom, and emergency tracheotomy may be necessary as a life-saving procedure (Greenspan et al, 1985). Fiberoptic examination should be used for diagnosis because it is capable of detecting the characteristic nodular red-blue lesion. Biopsy is contraindicated in these cases because of the danger of hemorrhage.

Salivary gland manifestations

Xerostomia is a frequent complaint in many HIV-infected patients. Its precise cause is yet to be elucidated but symptomatic treatment is usually effective.

Cystic parotid enlargement, a phenomenon described relatively recently, occurs commonly in the HIV-positive patient and is usually seen rather early in the clinical course of the disease (Sperling and Lin, 1990; Wenig et al, 1990). As a result, the otolaryngologist is often one of the first physicians to evaluate these patients.

It is not yet known whether these growths represent a primary process of the parotid salivary tissue itself or of the intraparotid lymph nodes. These masses typically present as unilateral or bilateral nontender enlargements. CT imaging reveals multiple cystic masses that on histologic examination are similar to benign lymphoepithelial lesions.

Surgery is generally best avoided because of the refractory nature of this disease and the risks to the facial nerve. However, in some patients in whom the diagnosis is not clear or who are extremely bothered by pain or cosmetic disfigurement, management has consisted of superficial parotidectomy. Increasing experience with this phenomenon has led to the more frequent use of fine-needle aspiration to diagnose and manage these lesions.

Cervical manifestations

One of the earliest reported manifestations of HIV infection was that of persistent generalized lymphadenopathy, which may be present in AIDS and ARC (Davidson et al, 1990). The cervical lymph nodes appear to be among those most frequently involved in this syndrome, which is most likely a result of follicular hyperplasia caused by HIV infection.

Other possible causes of cervical lymph node enlargement in AIDS patients include metastatic Kaposi's sarcoma, non-Hodgkin's lymphoma, and mycobacterial infection. The role of open biopsy and/or fine needle aspiration for persistent lymphadenopathy in HIV-positive

patients is controversial, and future studies will help clarify this issue. For now, it is generally accepted that an open biopsy is not necessary in HIV-positive patients unless there is a high suspicion of malignancy or unusual infection.

Bronchopulmonary manifestations

Pneumocystis carinii pneumonia (PCP) was the earliest reported manifestations of HIV infection. It continues to be the initial manifestation in over 65% of AIDS patients. Initial symptoms may include dyspnea, cough, and low-grade fever. Untreated, PCP is rapidly progressive, and chest roentgenograms may show diffuse infiltration within a matter of days. Diagnosis can usually be made by relatively noninvasive methods such as fiberoptic bronchoscopy and bronchoalveolar lavage. Rarely, open lung biopsy may be necessary.

Other infections that may involve the bronchopulmonary tree in AIDS patients include cytomegalovirus, mycobacteria, and various opportunistic fungi. Neoplasms such as Kaposi's sarcoma and non-Hodgkin's lymphoma involving the chest have also been reported (Ognibene, 1990).

Special Pediatric Problems

Most children with AIDS acquire the illness through maternal transmission. Those who are born with the disease usually do not manifest the illness at birth, but later may develop oral thrush, hepatosplenomegaly, and generalized lymphadenopathy (Chow et al, 1990). The otolaryngologist may be called upon to evaluate these patients because of recurrent ear or sinus infection. These infections are usually caused by most of the same organisms as in the general population (ie, *staphylococcus* and *pneumococcus*) but host failure to respond appropriately renders these patients more vulnerable to these bacteria. Pediatric AIDS patients also appear to be particularly susceptible to gram-negative sepsis from *Pseudomonas* infection. Early and aggressive treatment is indicated to prevent potentially fatal complications.

Occupational Exposure and Transmission

Since the very beginning of the AIDS epidemic there has been a great deal of anxiety among health care workers involved in the treatment of AIDS patients. Considering the lethal nature of the disease and its infectious etiology, some of this anxiety is justified. Fortunately, however, it appears from all existing information that most of this fear is irrational.

The greatest risk of AIDS exposure in the health care setting comes from exposure to contaminated blood, either through needlestick injuries or blood splashes. Polis (1990) reviewed several carefully performed prospective studies that have followed the rate of seroconversion in health care workers with a history of needlestick injury from HIV-positive sources. Patients were followed for a minimum of 6 months. It appears from this information that the risk of seroconversion is less than 0.4% following percutaneous injury. In addition, to date, there have been no documented cases of seroconversion following mucosal blood splashes.

Although the risk to health care workers appears to be quite low, risk does still exist and certain precautionary measures need to be followed to help reduce these risks even

further.

Protective measures for health care personnel

The Centers for Disease Control (1989) has issued a series of guidelines to reduce the risk of HIV infection in health care workers. For many reasons, however, it is not rational to take these specific precautions only in HIV-positive patients. Firstly, AIDS precautions involve little more than practicing infection control measurements that are probably appropriate to all patients in general. Secondly, these precautions can also lessen the incidence of hepatitis, which continues to have a greater morbidity and mortality among health care workers than does AIDS. Most importantly, however, is the fact that the increased prevalence of HIV infection, with or without symptoms, places the health care worker at an increasing risk of potential exposure. The CDC recommends therefore, that *all* patients be treated as potentially infective, and that blood and body fluid precautions be observed consistently for all patients.

General guidelines

The following is a summary of the CDC guidelines for prevention of infection in the health care setting.

1. All health care workers should use appropriate barrier precautions to prevent skin and mucous membrane exposure to all body fluids. Gloves, masks, and gowns should be worn as needed in this regard when contact with blood, mucous membranes, or nonintact skin is anticipated. These gloves, masks, and gowns should, of course, be discarded after contact with each patient.

2. Hands and other skin surfaces should be washed immediately after all patient contact, even if gloves are used.

3. The utmost care must be taken when handling needles, scalpels, and other sharp instruments to prevent injuries. Needles should not be recapped, bent, or broken. Disposable needles and syringes should be placed in puncture-resistant containers immediately after use.

4. Although saliva has not yet been implicated in HIV transmission, it has been found to harbor the AIDS virus; therefore, to minimize the risk during emergency mouth-to-mouth resuscitation, mouthpieces, resuscitation bags, and other ventilating devices should be available in all areas where resuscitative measures may need to be performed.

5. Health care workers with exudative lesions or weeping dermatitis should refrain from direct patient contact and contact with patient care equipment.

Precautions for the otolaryngologist - head and neck surgeon

The otolaryngologist - head and neck surgeon is frequently exposed to body fluids, especially saliva and blood, and therefore should be especially vigilant in taking the appropriate precautions in both the office and operating room. Office procedures such as fiberoptic laryngoscopy, aspiration biopsy, and nasal packing are potentially hazardous in this

regard. Gloves, masks, and gowns should be worn as needed.

There is a high likelihood of secretions and blood being splattered during procedures such as tracheotomy and endoscopy. Masks and goggles should be used during these high-risk procedures; all patients should be treated as potentially HIV infected. Tracheotomy probably has a lower chance of aerosolization if done with the patient under general anesthesia. During otologic surgery such as mastoidectomy, which involves aerosolization of blood, the surgeon is usually protected by the microscope eyepiece. However, assistants and nurses in the area must be duly protected.

Management Following Occupational Exposure

Adherence to CDC guidelines will minimize the risk of exposure to HIV. Yet, despite the observation of strict precautions, numerous exposures to HIV-contaminated fluids have occurred and will continue to occur in the health care setting. The question then is how to best manage these exposures. The CDC (1990b) has issued a set of recommendations to be followed when a health care worker is exposed to a source that is potentially HIV positive:

The worker exposed should report the incident immediately so that the appropriate interventions (eg, hepatitis prophylaxis) may be initiated promptly.

If the source individual has AIDS, is known to be HIV positive, or refuses testing, the worker should be evaluated clinically and serologically for evidence of HIV infection. If the worker is seronegative, testing should be repeated at intervals of 6 weeks, 12 weeks, and 6 months after exposure. During this period the worker should follow public health service recommendations to prevent HIV transmission.

During all phases of the evaluation, confidentiality of the worker should be protected. Recently, the use of Zidovudine (AZT) has been recommended by some experts as a prophylactic measure to be instituted immediately following HIV exposure (Henderson and Gerberding, 1989). AZT is a thymidine analogue that has been shown to inhibit reverse transcriptase, and thus slow or halt the replication of the AIDS virus. Whether the drug offers any real protection after exposure is not known, and never be known because of insufficient data. There are simply not enough patients on whom to conduct an adequate prospective study that will answer this question. In addition, AZT has been reported to cause several side effects, including nausea, vomiting, fatigue, fever, and malaise. The risk of teratogenicity and carcinogenesis from the medication is not known.

Currently, there is considerable debate over this issue. Those who favor the use of AZT prophylaxis do so because of the severity of the illness that may result from HIV infection, the documented antiviral effect of the medication, and the apparent responsibility of the toxicity that may result in persons taking the medication. Those who oppose prophylactic AZT use cite the lack of data demonstrating any benefit and the limited data on the drug's toxicity, teratogenicity, and carcinogenicity.

For now it seems appropriate that exposed workers be provided with all available information on the subject and allowed to make their own decisions in consultation with their personal physicians. If an individual elects to take the drug, informed written consent should

be obtained.

Patient Care Issues

The AIDS epidemic has prompted many physicians to review certain issues related to patient care, including physician-patient communication, patient education, care of the dying patient, and the anxieties that affected patients engender in physicians (Lucente, 1990).

In addition to AIDS, the epidemic of "AIDS phobia" has been widely observed since the disease was first described in 1981. This term applies not to the justified concerns of those who understand the lethal nature of the disease, but to the unfounded fears and anxieties of persons who do not understand the modes of AIDS transmission and the simple measures necessary for protection. These fears have manifested themselves in a variety of disturbing ways: AIDS patients being shunned by co-workers, noninfected homosexuals being fired from jobs, infected children being banned from schools, and so forth. Such fears, anxieties, and phobias are best dealt with by comprehensive educational programs based on the most current information available.

Patient communication

When interviewing patients, identifying risk-group membership can be an uncomfortable task for the physician. This is best addressed in a manner that assures the patient of confidentiality and the physician's genuine desire to help. Most patients are aware of the groups at higher risk, and it may be preferable to begin by asking the patient, "Do you do anything that places you at risk for AIDS?" If the patient does not understand the question or is unaware of the answer, the physician may then explain the various forms of transmission before proceeding with more direct questioning while simultaneously being sure to reassure the patient that the discussion is confidential.

AIDS patients are invariably aware of the seriousness of their disease. Some react with denial, by delaying necessary case and refusing essential treatment. The physician must remind the patient of the need to confront the illness directly and honestly. The physician may wish to seek support from the patient's family and friends, when appropriate. Like patients with other life-threatening illnesses. AIDS patients are under a great deal of emotional stress. Repeated explanations of diagnosis and therapy may be needed in these circumstances. It is helpful to make follow-up appointments, issue written instructions, and emphasize that the physician will be available whenever necessary.

The role of education

The physician plays an important role in educating the patient. Educational programs should include the following:

1. The AIDS patient is taught methods to prevent further spread of the virus.

2. Personal contacts of AIDS patients are instructed to avoid behavior that might transmit the virus, while remaining close to and supportive of the patient.

3. Members of the AIDS risk groups must learn to modify their social behavior to minimize risk.

4. The "worried well" (a term used to describe those who are anxious about the possibility of having AIDS) are encouraged to allay their anxieties by seeking confidential HIV testing.

5. Health care workers are encouraged to provide uniform care to all patients, regardless of known or suspected HIV infection, while protecting themselves from infection by knowing the routes of transmission.

Countertransference

Countertransference is a term that refers to those feelings or emotions that the physician experiences from interacting with patients. Many of the frightening and unpleasant feelings experienced by physicians caring for AIDS patients have been well described by Dunkel and Hatfield (1986). Physicians who are uncomfortable in this regard might discuss these feelings with colleagues or with a psychiatrist or other counselor who can offer advice in dealing with these natural but deleterious perceptions.

Summary

Approximately 40% to 60% of patients with AIDS present with a manifestation involving the head and neck. The otolaryngologist may be the first clinician to evaluate patients with HIV infections and must be knowledgeable about the various possible head and neck presentations.

The otolaryngologist may also be involved in diagnosing and managing the disease and performing biopsies, endoscopies, tracheotomies, and other surgical procedures. In all phases of care, the physician should provide the patient with a medically appropriate and psychologically supportive environment. The patient should be reassured about the continued availability of the physician because this can be of great psychologic support to the patient.

Although universal precautions should be followed in all phases of care, there is no need to avoid such gestures as hand shaking or patting the patient on the shoulder. These can be very reassuring to the patient. The physician should be aware of all sources of information and medical assistance within the community so that appropriate referrals can be made as needed.

The AIDS epidemic presents an enormous challenge to all health care providers. Many practitioners have been forced to reevaluate the patient-physician relationship, including interpersonal communication, the scope of physician responsibility, and the influence of patient care on the physician's own well being. It is often an uncomfortable experience for the physician to manage an illness with such a uniformly fatal outcome, and many may seek to avoid these patients. Failure to do one's best, however, is an abrogation of the physician's primary responsibility to society, to provide his or her skills selflessly to those who seek them.