Introduction

Connective tissue diseases, also referred to in the past as collagen vascular diseases and autoimmune disease, are among the least understood entities in medicine. Great strides toward understanding of the human immune system have occurred in the last 20 years, however, the cause of these diseases is still not understood. These syndromes are difficult to diagnose because many of their symptoms are nonspecific and the signs and symptoms tend to overlap. Additionally, they lack specific laboratory and pathologic tests to confirm the clinical suspicion. The common histologic feature of this group of diseases is widespread inflammatory damage to connective tissues and blood vessels and deposition of fibrinoid material. The location of these inflammatory changes and the pattern of organ involvement leads to the clinical disease designations.

The immune system development requires the maturation and differentiation of a lymphoid stem cell that originates in the bone marrow. This stem cell line gives rise to a variety of subpopulations: the B cell, a lymphocyte maturing under the control of the bone marrow itself; the T cell, a lymphocyte developing under the influence of the thymus; the macrophage or monocyte; and a minor population of lymphocytes called NK cells. These cells can be identified by their specific cell surface characteristics. The immune system is designed to respond to an antigen that is taken up and presented by macrophages to T and B cells that then produce chemical mediators and antibodies in response to the antigen. The connective tissue diseases are believed to develop due to an abnormal interaction of the individuals own immune system to “self” antigens. Three theories exist to explain how this comes about. The antigen may have been sequestered, or hidden, from the immune system during embryogenesis, when the immune system was “learning” what was self and what was foreign. If this antigen is later exposed, it may elicit an immune response as a foreign antigen. Another possibility is that an exogenous antigen may be sufficiently foreign to elicit an immune response, but similar enough to a self-antigen to allow cross reactivity of antibody. A third theory proposes that a self-antigen may be altered by, for example, a viral infection, causing it to be perceived as foreign. Having reviewed some of the basic ideas of immunity, several connective tissue diseases as well as vasulitides will be presented.

Systemic Lupus Erythematosus

General
- Autoimmune multisystem disease.
- Prevalence 1 in 2,000; 9 to 1 female to male; or 1 in 700 women; 1 in 245 black women
- Peak age of onset 15-25
- Unknown cause, pathologic process is immune complex deposition
- Systemic manifestations include photosensitive skin eruptions, serositis, pneumonitis, myocarditis, nephritis, CNS involvement
Specific labs - native(double stranded) DNA, SM antigen, many others often positive
-procainamide, hydralazine, hydantoins, penicillin and sulfonamides may precipitate lupus like reaction
- LE cell is specific

Head and Neck Manifestations
-Skin and mucosal lesions
-Malar or "butterfly" rash first sign of disease in 50%
-Erythematous maculopapular eruption most common head neck and chest may be pruritic and follow sun exposure
-Oral ulcerations- superficial with erythematous halo, may increase to 2 cm, painful
-3-5% ulceration or perforation of nasal septum
-Larynx and trachea involvement rare- TVC thickening, paralysis, cricoarytenoid arthritis, subglottic stenosis
-Acute enlargement of parotid in 10%, may be unilateral, tender confused with acute parotitis
-Xerostomia in some
-Neuropathy cranial nerves affected in 15%
-Discoid lupus-subtype with cutaneous lesions with significant scarring, no visceral involvement

Treatment
-Avoidance of sun exposure use of sunscreens
-NSAIDS
-topical and systemic steroids
-antimalarials
-low-dose methotrexate
-azothioprine, cyclophosphamide restricted to cases with serious visceral involvement
-symptomatic treatment- saliva substitutes, mouthwashes, Klack’s solution, (tetracycline, cortisone, benadryl, nystatin)
-postprandial rinses with H2O2 and H2O 50:50

Diagnostic criteria for SLE

<table>
<thead>
<tr>
<th>Diagnostic criteria*</th>
<th>Percent/incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Malar rash</td>
<td>64</td>
</tr>
<tr>
<td>Discoid rash</td>
<td>17</td>
</tr>
<tr>
<td>Photosensitivity</td>
<td>37</td>
</tr>
<tr>
<td>Oral ulcers</td>
<td>15</td>
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<tr>
<td>Arthritis</td>
<td>50</td>
</tr>
<tr>
<td>Proteinuria (0.5 g/dL) or cellular casts</td>
<td>20</td>
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<tr>
<td>Seizures or psychosis</td>
<td>19</td>
</tr>
<tr>
<td>Pleuritis or pericarditis</td>
<td>19</td>
</tr>
<tr>
<td>Hemolytic anemia, leukopenia, lymphopenia,</td>
<td>11-40</td>
</tr>
<tr>
<td>thrombocytopenia</td>
<td></td>
</tr>
<tr>
<td>Antibody to DNA or Sm antigen, + LE prep,</td>
<td>15-60</td>
</tr>
<tr>
<td>or false ++RPR</td>
<td></td>
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<tr>
<td>Positive fluorescent antinuclear antibody*</td>
<td>95</td>
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</table>

*The diagnosis of SLE requires the presence of four of the 11 criteria (96% sensitivity, 96% specificity).
*Increased antibodies to double-stranded DNA are pathognomonic.

From Bailey’s H&N Surgery Second Edition

Rheumatoid Arthritis

General
-inflammation of synovial tissue with symmetric involvement of peripheral joints, hand, feet, wrists commonly
-can also affect nonarticular muscular structures(tendon, ligaments, fascia)
-occasionally systemic disease such as vasculitis, pulmonary fibrosis
-inflammatory cell infiltrates (lymphocytes), synovial proliferation responsible for pathogenesis
-high incidence of HLA Dw4 locus positive suggesting inherited contribution
-occurs in 1% of the population, women 2-3 times men, 4th – 5th decade, also distinct juvenile form
-onset may be acute, more often insidious, progressive joint involvement
-morning stiffness lasting greater than 30 minutes, or after prolonged inactivity
-subcutaneous rheumatoid nodules
-synovial fluid shows inflammation 2K to 75K WBC’s 50% PMN’s
-diagnosis clinical, rheumatoid factor positive within 12 months of onset in 90% of patients
-other tests RA associated nuclear antigen (RANA), anti-RA-33,
-diagnostic criteria p 195 Baileys

Head and Neck Manifestations
-articular involvement of the ossicles, TM joint, cricoarytenoid joints, cervical spine
-TM joint involvement common 55% symptomatic, up to 70% evident on X-ray
-juvenile RA can cause micrognathia
-RA most common cause of arthritis of cricoarytenoid joint
-30% of RA patients hoarse, 86% show pathologic involvement of joint
-exertional dyspnée ear pain, globus sensation all attributed to C-A joint involvement
- hoarseness also caused by rheumatoid nodules or ischemic recurrent nerve paresis or paralysis
-can present with stridor requiring local and systemic steroids and possible tracheostomy
-possible CHL from ossicular chain involvement, TM may lose stiffness
-also described unexplained SNHL associated especially with rheumatoid nodules
-RA of spinal column predilection for cervical and can lead to subluxation

Treatment
-physical therapy, daily exercise, splinting joint protection,
-salicylates, NSAIDS, gold salts, penicillamine, hydroxychloroquine, immunosuppressive agents, systemic corticosteroids should in general be avoided
-prognosis 10-15 yrs of disease 50% fully employed, 10% completely incapacitated, 10-20% complete remission

Diagnostic Criteria for Rheumatoid Arthritis

1. Morning stiffness (≥1 h)
2. Swelling of three or more joints
3. Swelling of hand joints (proximal interphalangeal, metacarpophalangeal, or wrist)
4. Symmetric joint swelling
5. Subcutaneous nodules
6. Serum rheumatoid factor
7. Radiographic evidence of erosions or periarticular osteopenia in hand or wrist joints

*Criteria 1 to 4 must have been continuous for 6 weeks or longer and must be observed by a physician. A diagnosis of rheumatoid arthritis requires that four of the seven criteria be fulfilled.

From Bailey’s H&N Surgery second edition

Sjogren’s Syndrome
General
-immune-mediated destruction of exocrine glands
-not to be confused with Mikulicz’s disease (swelling due to nonspecific lymphocytic infiltrate
-accompanying nonconnective tissue diseases such as hyperlipoproteinemia, malnutrition, diabetes,
cirrhosis, tuberculosis, and sarcoidosis)
-primary and secondary forms
-primary or sicca syndrome- isolated disorder of lacrimal and salivary glands
-secondary sicca complex when associated with any of the connective tissue diseases
-occurs in 1% general population and 10-15% of RA patients
-9 to 1 female preponderance, onset 40-60 yrs of age
-increased risk of lymphoma, perhaps as high as 44 times risk
-clinical manifestations- xerophthalmia and secondary keratoconjunctivitis, xerostomia,
can also affect skin, vagina, external genitalia, chronic bronchitis, GI tract, renal tubules
-minor salivary gland biopsy demonstrates heavy lymphocyte infiltration
-RF and ANA high in most patients, SJogran’s syndrom A (Ro(SS-A) SS B (La/SS-B) noted in 60% and
30% of patients respectively
-diagnostic criteria p 196 Bailey’s

Head and Neck Manifestations
-80% c/o xerostomia most predominant symptom
-difficulty chewing, dysphagia, changes in taste, fissures of tongue and lips, increased dental caries
-oral candidiasis
-salivary quantification difficult and requires experience, quantitative salivary scintigraphy best
-history of recent salivary gland enlargement, bilateral or unilateral, recurrent not persistent
-eye complaints of dryness, burning, itching, and foreign body sensation
-keratoconjunctivitis sicca- corneal abrasions with rose bengal staining
-Schirmer I test (lacrimal production) 15 mm wetting after 5 minutes normal, 5-15 equivocal, less than 5
abnormal (15% normal subjects less than 5mm)
-Schirmer II – stimulate with 10% ammonia 15 cm below nose, less than 10 mm abnormal
-nasal crusting, epistaxis, hyposmia secondary to loss of nasal glandular secretions
-diagnosis- history- Schirmer I if abnormal then Schirmer II or rose bengal staining abnormal or other
supportive evidence ie. abnormal salivary flow then labial biopsy is indicated

Treatment
-symptomatic treatment increased oral intake, saliva substitutes, artificial tears
-avoid decongestants, antihistamines, diuretics, and other drugs with anitcholinergic side effects
-pilocarpine
-candidiasis treat with clotrimazole or nystatin
-close supervision by a dentist is essential
-surveillance for any developing malignancy

... CONTINUED NEXT PAGE...
Diagnostic criteria for Sjogren’s Syndrome

1. Dry eyes (>3 mo), sensation of sand or gravel in eyes, or use of tear substitutes >3 times a day
2. Dry mouth (>3 mo), recurrent or persistent swollen salivary glands, or frequent drinking of liquids to aid in swallowing dry foods
3. Schirmer-I test (≤5 mm in 5 min) or Rose bengal score ≥4
4. >53 mononuclear cells/4 mm² glandular tissue
5. Abnormal salivary scintigraphy or parotid sialography or unstimulated salivary flow ≤1.5 mL in 15 min
6. Presence of anti-Ro/SS-A, anti-La/SS-B, antinuclear antibodies or rheumatoid factor

*Exclusion criteria: preexisting lymphoma, acquired immunodeficiency syndrome (AIDS), sarcoidosis, or graft-versus-host disease.

*Presence of four or more criteria classifies primary Sjogren’s syndrome with a sensitivity of 94% and a specificity of 94%.

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Scleroderma

General
- also called systemic sclerosis
- increased deposition of collagen in the interstitium and intima of small arteries and connective tissue
- characterized by sclerotic skin changes often accompanied by multisystem disease
- 4-12 new cases per million per year, 3 or 4 to 1 female to male preponderance
- average age of onset between 30 and 50 yrs
- black women worse prognosis than white, men worse than women
- initial presentation includes Raynaud’s phenomenon, edema of fingers and hands, skin thickening
- visceral manifestations seen in the gastrointestinal tract, lung, heart, kidneys and thyroid
- arthralgias and muscle weakness common complaints

- four categories
  1) diffuse cutaneous- worse prognosis earlier and more extensive systemic involvement
  2) limited cutaneous- (formerly CREST-calcinosis, Raynaud’s phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia) ,more benign course fewer renal complications
  3) systemic sclerosis sine scleroderma- visceral manifestations of systemic sclerosis without skin changes.
  4) systemic sclerosis in overlap- concomitant SLE, polymyositis, RA
- diffuse cutaneous- anti-ScL-70, limited cutaneous(CREST) anticientromere
- elevated ESR, mild anemia of chronic disease, ANA, hypegammaglobulinemia, RF, LE cells, abnormal EKG in 50%(low voltage, axis deviation, intraventricular conduction deficits)

Head and Neck Manifestations
- 80% H&N complaints, 30% present with H&N complaint
- tight skin, thin lips, vertical perioral furrows
- skin changes due to underlying dermal and subcutaneous inflammatory process
- edema precedes epidermal atrophy and loss of appendages
- dysphagia is most common initial complaint
- 80% show distal 2/3 of esophagus pathology on barium swallow
- decreased or absent peristalsis with mild to moderate dilation, hiatal hernia is common
- decrease ability to open mouth initial complaint in 19%, 2ndary to skin changes
- gingivitis and periodontal thickening are common, translucent zone around dental roots considered
pathognomonic of scleroderma by some investigators
-25% report xerostomia, xerophthalmia or both
-laryngeal involvement occurs with almost 50% c/o hoarseness
-infrequently Raynaud’s of the tongue presents with blanching and dysarthria
-infrequently trigeminal neuralgia and facial nerve palsy

Treatment
-treatment is symptomatic
-calculator channel blockers in Raynaud’s
-\text{H}_2\text{ blockers for reflux}
-NSAIDS and low dose steroids may relieve arthralgias and myalgias
-hand rehabilitation
-intra-arterial reserpine decreases vasoconstriction promotes healing

Polymyositis and Dermatomyositis

General
-group of disorders characterized by proximal muscle weakness and nonsuppurative inflammation of skeletal muscle
-polymyositis and dermatomyositis subsets
-5 cases per million per year, 2 to 1 female to male, onset 40 to 60 years, pediatric variant 5-15 yrs old
-criteria- proximal muscle weakness, elevated serum creatinine kinase levels, myopathic changes on electromyography, muscle biopsy evidence of inflammation (lymphocytic)
-diagnosis definitive with all four, probable with three, possible with two
dermatomyositis requires characteristic skin rash with above
-anti-tRNA synthetases
-may be associated with other connective tissue diseases
-up to 20% associated with malignancy, lung, ovary, breast, stomach most common parotid, tonsil reported
-reported increase incidence of nasopharyngeal carcinoma in patients with dermatomyositis where endemic

Head and Neck Manifestations
-manifestations reflect proximal muscle weakness
-weakness of neck muscles
-difficulty with phonation and deglutition, nasal regurgitation,
dysphagia secondary to involvement of upper esophagus, cricopharyngeus, pharynx, and sup. Constrictors
dysfuction in swallow can result in aspiration and pneumonia
-skin and mucous membrane lesions of dermatomyositis vary-predilection for eyelids, nose, and cheeks

Treatment
-steroids for symptomatic patients
-methotrexate and other immunosuppressive agents reserved for nonresponders
-\text{H}_2\text{ blockers, metoclopramide}

Relapsing Polychondritis

General
-episodic recurring inflammation of cartilaginous structures, eventually replaced by fibrosis
-women more often than men
-Age of onset 35-45, no racial predilection
-can affect any cartilaginous structure including heart valves and large arteries
-diagnostic criteria- recurrent chondritis of auricles, nonerosive inflammatory polyarthritis, chondritis of nasal cartilages, inflammation of ocular structures, chondritis of laryngeal or tracheal cartilages, cochlear (SNHL, tinnitus) of vestibular damage (vertigo)
-ESR elevated, moderate leukocytosis, mild to moderate anemia
-histologically- loss of basophilic staining of cartilage, perichondral inflammation, cartilage
destruction and replacement by fibrous tissue

**Head and Neck Manifestations**
- auricular chondritis, nonerosive arthritis most common
- sudden onset erythema, pain, sparing EAC feature in 33%, develops in 90%, occas. LAD
- resolution in 5-10 days with or without treatment
- may have concomitant serous otitis or SNHL, 49% inner ear symptoms
- nasal chondritis develops in 75% does not always coincide with auricular symptoms
- nasal and auricular chondritis act similar both causing deformity and dysfunction
- laryngeal involvement nonproductive cough progresses to hoarseness and stridor
- 53% will have airway involvement

**Treatment**
- salicylates, ibuprofen for symptomatic relief
- steroids for life threatening manifestations
- antileprosy drug, dapsone reduces lysozymes decreasing inflammatory response

**Mixed Connective Tissue Disease**

**General**
- coined in 1972
- describes a distinct entity with coexisting features of SLE, scleroderma, and polymyositis
- high titers of anti-U1 RNP, a ribonucleoprotein antibody
- prevalence unknown, 80% female, onset 30-60 yrs.
- diagnostic criteria: elevated titers of anti-U1 RNP plus three of either a) hand edema, b) synovitis, c) myositis, d) Raynaud’s phenomenon, or e) acrosclerosis
- pulmonary involvement is common
- cardiac and renal involvement in 25%

**Head and Neck Manifestations**
- combination of features of other CTD
- mucocutaneous changes, malar rash, discoid lupus, sclerodermatous skin changes, oral mucosal ulceration, nasal septal perforation, esophageal dysfunction

**Treatment**
- steroid therapy for symptomatic relief
- immunosuppressives only after complications involving vital organs develop

**Vasculitides**

Vasculitis is the inflammation and necrosis of blood vessels induced by immunologic mechanisms. Any blood vessel can be affected, therefore, the clinical manifestations of vasculitis may be extremely diverse, and different syndromes may overlap considerably. Confusion of clinical signs and symptoms, difficulties in pathologic categorization and uncertainty as to cause or pathogenesis has made standard classification difficult. The following table presents one classification scheme.
The pathogenesis of most forms of vasculitis is unclear. Multiple mechanisms are probably involved including deposition of antibody-antigen-complement in vessel walls, and antigen deposition triggering lymphocytic reaction and granuloma formation.

Clinically, vasculitis is characterized by multisystem disease. Vasculitis gives rise to symptoms because of obliterative narrowing of vessels resulting in end-organ ischemia and dysfunction. It is unusual for a vasculitic process to affect only one organ system. Specific diagnosis can only rarely be made on clinical grounds alone, and routine or even specialized serologic testing yields a specific diagnosis in only a minority of cases. In most instances, it is necessary to biopsy tissue in order to make a diagnosis.

**Hypersensitivity Vasculitis**

**General**
- collective term for large group of diseases
- result in inflammation of small vessels, arterioles, capillaries, and venules
- most cases related to circulating and deposited immune complexes
- skin clinically involved virtually always, hemorrhage or classic purpura
- involvement of major organ systems is less common

**Head and Neck Manifestations**
- petechiae and purpura of oral and nasal mucosa
- angioedema
- serous otitis media

**Treatment**
- usually self-limiting particularly when only skin is involved
- if systemic involvement present or underlying disease (ie.cryoglobulinemia) more aggressive therapy warranted

**Polyarteritis Nodosa**

**General**
- prototype of vasculitides
-males two to three times more often than females
-50-60 yrs old
-hepatitis B antigen in 30%
-small and medium arteries, aneurysms
-tissues include GI tract, hepatobiliary system, kidney, pancreas, and skeletal muscles
-non-specific multisystem complaints (malaise, weight loss, anorexia, fever)
-progressive arthritis, myopathy, neuropathy, hepatic and renal failure, GI bleeding

Head and Neck Manifestations
-few and primarily involve ear
-sudden bilateral SNHL infrequent presentation, vestibular disturbance
-proposed mechanism is thromboembolic occlusion of inner ear end arteries
-rarely- ulceration of nasal, buccal, soft palate mucosa, cranial nerve palsies VIIth most common

Treatment
-Steroids

Churg-Strauss Syndrome (allergic angiitis and granulomatosis)

General
-resembles polyarteritis nodosa
-triad of systemic vasculitis, asthma, peripheral and tissue eosinophilia
-nasal symptoms also prominent
-unlike PAN, lungs virtually always involved
-malaise, night sweats, fever, weight loss, myalgias
-peripheral eosinophilia as high as 74%, elevated ESR

Head and Neck Manifestations
-nasal obstruction, rhinorrhea, nasal polyps, sinusitis
-25% severe nasal crusting recurring after removal in 24-48 hrs
-occasional septal perforation
-subcutaneous nodule
-history of allergy

Treatment
-corticosteroids

Wegener’s Granulomatosis

General
-necrotizing granulomatous vasculitis of upper airway, lower airway, and kidney
-bilateral pneumonitis (95%), chronic sinusitis (90%), mucosal ulceration of nasopharynx (75%)
evidence of renal disease (80%)
-hallmark pathologic lesion necrotizing granulomatous vasculitis
-antineutrophil cytoplasmic antibody (c-ANCA), sensitivity (65-90%) high specificity
-need tissue to confirm diagnosis, often takes 3-4 biopsies, open pulmonary biopsy occas. necessary
-most commonly involved and accessible tissue is nasopharynx
-untreated mortality of 90% at two years

Head and Neck Manifestations
-nasal symptoms- crusting, epistaxis, rhinorrhea, erosion of septal cartilage, saddle deformity,
-recurrent sinusitis
-oral cavity- hyperplasia of gingiva, gingivitis,
-upper airway- edema, ulceration of larynx (25%), significant subglottic stenosis (8.5%)
-Otologic- (20-25%)- serous otitis media, CHL, suppurative otitis media possibly with grannulation tissue,
SNHL, pinna changes similar to polychondritis, facial nerve palsies

**Treatment**
- meticulous dental and nasal care, removal of crusts from eustachian tube orifices, middle ear drainage
- cyclophosphamide 2 mg/kg plus prednisone 1 mg/kg achieved remission in 93% of patients
- azathioprine or methotrexate alternative to cyclophosphamide
- isolated sinus disease- low dose steroids, topical steroids, saline irrigation, antibiotics as needed
- airway compromise alleviated with systemic steroids,
- subglottic stenosis may warrant tracheotomy

**Giant Cell Arteritis (temporal arteritis)**

**General**
- temporal is form of giant cell with only extracranial vessels affected
- focal granulomatous inflammation of medium and small arteries
- most common vasculitis
- prevalence increases with age to 850/100,000 age 80 and older
- symptoms reflect cranial blood supply
- headache (constant , boring) most common initial complaint, 47%, 90% develop
- ESR usually greater than 50 mm/hr
- confirmation from temporal artery biopsy of symptomatic side, 5-7 cm
- if negative biopsy contralateral side
- false negative rate 5-40%

**Head and Neck Manifestations**
- temporal artery tender and erythematous 50% of time
- scalp often tender
- jaw claudication 50%
- lingual claudication 25%
- vertigo and hearing loss reported
- ascending pharyngeal artery involvement can lead to dysphagia
- cranial nerve deficits, vertebrobasilar insufficiency, psychosis reflect intracranial disease
- blindness occurs in 1/3 of untreated patients- field deficits, amaurosis fugax

**Treatment**
- prednisone
- normalization of ESR and symptoms therapeutic guideline
- treatment may be required for up to two years

**Polymyalgia rheumatica**
- accompanying syndrome in 50% of patients with giant cell arteritis
- clinical syndrome of muscular pain, morning stiffness of proximal muscles, increased ESR without inflammatory joint or muscle disease
- systemic symptoms- low-grade fever, weight loss, malaise
- responds well to low-dose prednisone if isolated
- if with giant cell high dose initially then adjusted

**Behcet’s Disease**
- vasculitis with triad of oral and genital ulcers and uveitis or iritis
- aphthous-like ulcers, characteristically punched out with or without surrounding erythema, covered with pale pseudomembrane
- painful, occur in clusters on lips, gingiva, buccal mucosa, tongue, less often on palate, oropharynx
- genital ulcers similar in appearance
- healing in few days to weeks with some scarring
- symptoms may occur simultaneously or months apart

Other associated findings and treatment
- progressive SNHL, tinnitus, and vertigo have been associated with Behcet’s
- may also have ulceration of nasal, laryngeal, tracheal mucosa
- morbidity from CNS involvement, bowel dysfunction, large vessel arteritis
- azathioprine or methotrexate may be beneficial although not documented

Cogan’s Syndrome
- rare disease of young adults
- vestibulounder dysfunction, interstitial keratitis, and nonreactive syphilis test
- symptoms frequently begin after URI
- vestibulounder symptoms usually bilateral can include fluctuating hearing loss, vertigo, tinnitus, and aural pressure, similar to Meniere’s but bilateral
- ocular symptoms of photophobia, lacrimation, and eye pain
- may resolve spontaneously and reappear months later
- when advanced, hearing loss progressive and severe with decreased or absent vestibular responses on caloric testing
- ear symptoms may precede or follow ocular disease
- eye symptoms easily controlled with topical steroids and atropine
- evidence exists that hearing loss may be avoided if treated with steroids within 2 weeks of onset

Kawasaki Disease
- AKA-mucocutaneous lymph node syndrome
- disease of pediatric age group
- fever, conjunctivitis, red dry lips, erythema of the oral mucosa, polymorphous truncal rash, desquamation of the fingers and toes, and cervical lymphadenopathy
- oral cavity erythema and cervical adenopathy are presenting symptoms
- cardiac abnormalities cause 1-2% mortality rate

Bibliography


