

# Rheumatology

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There are multiple interactions between rheumatology and oral medicine. Three overlapping areas will be covered in this lecture:

- (1) Oral manifestations of rheumatic disease. Several rheumatic diseases have prominent manifestations in the oral cavity, head and neck. Because these manifestations tend to be nonspecific, the underlying condition may go unrecognized unless the oral surgeon suspects the correct diagnosis and implements referral.
- (2) Neck problems that should be of concern to the oral surgeon. Rheumatic diseases may result in neck problems that pose specific risks during the performance of dental procedures.
- (3) Effects of antirheumatic medications. Certain medications used in rheumatology may result in oral lesions and others interfere with hemostasis.

### I. Rheumatic Diseases with Prominent Oral or Head Manifestations

#### A. *Systemic Lupus Erythematosus (SLE)*

Systemic lupus erythematosus is a multisystem condition that affects predominantly women in the childbearing years. Males, children and older people infrequently develop SLE. In addition to its varied clinical manifestations, which may affect any system, multiple immunologic aberrations occur in SLE, in particular the production of autoantibodies that are important in diagnosis. The American College of Rheumatology Classification Criteria for SLE list the salient clinical and laboratory findings in this disease:

#### *ACR Classification Criteria for SLE\**

1. Malar Rash
2. Discoid rash
3. Photosensitivity
4. Oral ulcers
5. Arthritis
6. Serositis (pleuritis or pericarditis)
7. Renal disorder
8. Neurologic disorder (seizures or psychosis)
9. Hematologic disorder (hemolytic anemia, leukopenia, lymphopenia, or thrombocytopenia)
10. Immunologic disorder (anti-DNA or anti-phospholipid antibody production)
11. Antinuclear antibody

SLE may have prominent oral manifestations. Shallow, relatively painless ulcerations are

common. These may occur anywhere in the oral cavity, although the tongue is usually spared. Nasal ulcerations often coexist. Twenty-five percent of SLE patients have sicca syndrome, rarely with the severity seen in primary Sjogren's syndrome. Autoimmune thrombocytopenia may lead to oral petechiae or echymoses as well as gingival bleeding. Thrush is common in SLE patients treated with high-dose corticosteroids. Additional findings that may strike the oral surgeon include a malar rash, sometimes involving the bridge of the nose as well (butterfly rash), palpebral erythema, and alopecia with sparse and fragmented hair at the temples.

#### B. *Scleroderma*

Scleroderma is a condition in which excessive fibrosis (increased collagen production, in excess of degradation) leads to hardening of skin, particularly in the face and distal extremities. Increased vascular reactivity in these patients is expressed as Raynaud's phenomenon, in which, as a response to a cold environment, one or a few digits turn white. Finally, patients with scleroderma exhibit certain autoantibodies which are useful in diagnosis of early cases. Based on the extent of skin involvement, two forms of scleroderma can be distinguished: diffuse scleroderma (dScl) and limited scleroderma (lScl), which is often known as **CREST** (Calcinosis, Raynaud's, Esophageal, Sclerodactyly, Telangiectasis). The diffuse form of the disease has a more severe course, including renal insufficiency in over 10% of patients. Forty percent of patients with dScl have antisclero-70 antibodies. Limited scleroderma is more benign, although individual manifestations of the disease, such as pulmonary hypertension, may be quite serious. Eighty percent of patients with this form of the disease exhibit anticentromeric antibodies.

#### *Types of Scleroderma*

1. Diffuse cutaneous  
Extremities, face and neck, trunk  
Visceral involvement common  
Antisclero-70 antibodies 40%
2. Limited cutaneous  
Extremities face and neck  
Calcinosis common  
Visceral involvement rare  
Anticentromeric antibodies 80%
3. Localized scleroderma

\*An individual fulfilling four or more of the 11 criteria may be classified as having SLE.

Linear scleroderma (may associate with facial hemiatrophy)

Morphea

Because fibrosis is followed by retraction, the oral aperture may be drastically reduced (microstomia). Approximately 25% of patients exhibit dry mouth, usually less severe than in primary Sjogren's syndrome. Because of access difficulties, plus the lack of saliva, dental problems including early decay and gingivitis are extremely common in these patients. In patients with lScl, telangiectases may be quite prominent, predominantly involving the lips, the undersurface of the tongue, and elsewhere in the GI tract. Dysphagia due to esophageal fibrosis is quite frequent and may pose important nutritional problems. The facial skin is often taut, hard-bound, with radial grooves converging to the mouth (pursing). Hardened hand skin, finger contracture and paroxysmal color changes in the digits should make the diagnosis of scleroderma obvious to the dental surgeon.

#### C. *Reiter's Syndrome*

Although we keep here the eponym, an updated designation of this syndrome is reactive arthritis, since the condition usually results from the interaction between an environmental microbial agent (bowel or urogenital infection) and a genetically predisposed individual who carries the HLA-B27 antigen. Somehow, such interaction triggers subacute or chronic inflammatory disease. The classic Reiter's triad includes conjunctivitis, nonspecific urethritis, and arthritis affecting predominantly lower extremity joints, the sacroiliac joints, and tendon insertions. Incomplete cases without conjunctivitis or urethritis are more frequent than classic cases. Shallow and painless oral ulcers are a frequent finding in reactive arthritis. They usually involve the hard palate but may occur anywhere in the oral cavity.

#### D. *Behcet's Syndrome*

This rare syndrome includes painful, deep oral and genital ulcers, acute anterior (with pus in the anterior eye chamber) or posterior uveitis, and a variety of other involvements such as superficial phlebitis, cutaneous vasculitis, arthritis, serositis (pleura, pericardium), meningoencephalitis, colitis, and aortic aneurysm. The condition is chronic and recurrent with asymptomatic period in between. Flares are unpredictable and some manifestations, such as posterior uveitis and meningoencephalitis, may be protracted. Behcet's ulcers involve the mucous membrane of lips, gingiva, cheeks and tongue. Similar lesions occur on the scrotum, and the vulva. Behcet's lesions must be

distinguished from the giant aphthous ulcers syndrome, in which systemic involvement is lacking. Behcet's syndrome is rare in the United States, but it is quite prevalent in north Africa, Turkey, the Middle East, Korea and Japan.

#### E. *Giant Cell Arthritis (GCA)*

Different from the previous conditions, GCA affects predominantly elderly people, women more frequently than men in a ratio of 2 : 1. The classic case exhibits temporal headache and a tender, thickened, often nodular temporal artery. The condition may be unilateral or bilateral. Characteristically, the erythrocyte sedimentation rate (a nonspecific index of inflammation affected mainly by fibrinogen concentration, in itself an acute phase reactant) is markedly elevated, in the vicinity of 100 mm/hour (NI < 30 mm) in 1 hour). GCA may affect other vessels, such as the ophthalmic and posterior ciliary arteries, leading to blindness in about 10% of untreated cases. Thus, early diagnosis is of paramount importance.

Dental surgeons may encounter patients with GCA in several ways. GCA may involve the facial arteries, resulting in pain and swelling near the jaw angle. GCA may affect vessels supplying the masseter muscles, producing intermittent jaw claudication (with cramp-like pain after chewing for a while). It may also involve the lingual arteries, producing paroxysmal color changes in the tongue, paresthesias, pain or necrosis. Finally, oral pain may be prominent, usually at the gum, without local findings. In any older person, persisting gum pain without a clear explanation should raise the suspicion of GCA. A definitive diagnosis of GCA requires superficial temporal artery biopsy. This should be attempted in all cases, even after initiation of corticosteroid therapy, since it takes about one week for the lesion to regress. Treatment with high-dose corticosteroids rapidly corrects the symptoms and prevents blindness.

## II. Rheumatic Diseases that Result in an Unstable or Rigid Neck

#### A. *Rheumatoid Arthritis (RA)*

Rheumatoid arthritis affects women more frequently than men (2:1) and is clinically characterized by symmetric inflammation of distal joints, such as those of digits, wrists, feet, and knees. Shoulders and hips are commonly involved. Interestingly, although the cervical spine is involved in severe cases, the lumbar segment is always spared. Cervical spine changes are prominent at the uncovertebral joints and bursae. Uncovertebral joint synovitis

leads to secondary intervertebral disc deterioration and upper cervical spine instability. Of importance, the synovial bursae surrounding the dens are often affected, leading to lysis and disruption of the transverse ligament of the atlas and atlanto-axial subluxation. In these patients, neck flexion leads to dorsal subluxation of the dens, which thus impinges on the cord.

Symptoms of atlanto-axial subluxation include nuchal headache (due to tissue stretching), and when the cord is pinched, the Lehermitte's sign, i.e., flashes of pain or paresthesiae extending along the four extremities. Diagnosis is based on a cervical spine film obtained with the neck in forward flexion, revealing increased distance between the dens and the anterior arch of the atlas. MRI is helpful to reveal the extent of cord compression. This lesion requires prompt surgical stabilization (wiring together of the C1 and C2 posterior arches).

#### B. *Ankylosing Spondylitis*

In ankylosing spondylitis, a condition that clinically affects men more than women in a ratio of 10:1, inflammatory changes at the vertebral attachment or outermost fibers of the intervertebral discs are followed by ossification. This results in spinal fusion. Lack of motion and osteoporosis make the cervical spine brittle. Interestingly, as in reactive arthritis, most of these patients are HLA-B27 positive. Patients with advanced ankylosing spondylitis are at significant risk when subject to general anesthesia because relatively minor forces applied to

the neck may result in spinal fracture and quadriparesis.

#### C. *Diffuse Idiopathic Skeletal Hyperostosis (DISH, Forestier's Disease)*

DISH is a spinal condition in which four or more vertebrae are welded together by a heavily ossified anterior longitudinal ligament. Cases limited to the dorsal spine are quite frequent (10% of the above-60 population). In extensive cases, the cervical spine is often involved. In these patients, massive, flame-shaped anterior osteophytes typically produce dysphagia. Cord compression may occur, resulting from bulky ossification of the (intra-spinal) posterior longitudinal ligament.

### III. Effect of Antirheumatic Medications

Oral ulcers frequently develop in rheumatoid arthritis patients taking methotrexate. Gold injections often cause metallic taste and various degrees of stomatitis. All of the nonsteroidal anti-inflammatories inhibit platelet adhesiveness, leading to prolonged bleeding time as well as echymoses in predisposed patients. Of the nonsteroidal anti-inflammatory agents, aspirin, because of its acetyl moiety, permanently inhibits platelet cyclooxygenase, altering platelet function until a new crop of platelets develop. Other agents, such as ibuprofen, have a reversible inhibitory effect that lasts only hours. Nonacetylated salicylates, such as choline salicylate and sodium salicylate, lack the acetyl moiety and do not alter the bleeding time nor do the COX-2 inhibitors that are in common use now.