Sjögren's syndrome (SS)

Is characterised by lymphocytic infiltration of salivary and lacrimal glands, leading to glandular fibrosis and exocrine failure.

The typical age of onset is between 40 and 50, with a 10: 1 female-to-male ratio.

Sjögren's syndrome is subdivided into primary and secondary Sjögren's syndrome. Secondary Sjögren's syndrome describes patients with keratoconjunctivitis sicca, xerostomia, or both, in the setting of another connective tissue disease or chronic inflammatory process, such as rheumatoid arthritis, systemic lupus erythematosus

Clinical features

The eye symptoms, termed keratoconjunctivitis sicca, Conjunctivitis and blepharitis are frequent, and may lead to filamentary keratitis. Aqueous tear deficiency produces a dry eye, causing symptoms of grittiness or foreign body sensation, burning, photophobia, and eye fatigue.

Oral involvement manifests as a dry mouth (xerostomia). There is a high incidence of dental caries. Salivary gland enlargement. Patient feel struggles with prolonged speaking.

Patients with xerostomia may have problems wearing dentures.

Often the most disabling symptom is fatigue

Non-erosive arthralgia, Generalised osteoarthritis

Both interstitial lung disease and interstitial nephritis (sometimes complicated by renal tubular acidosis)

Peripheral neuropathy

Lymphadenopathy

Increased lifetime risk of lymphoma,

Investigations

Anaemia, leucopenia, Thrombocytopenia

Rheumatoid factor

• Antinuclear antibody

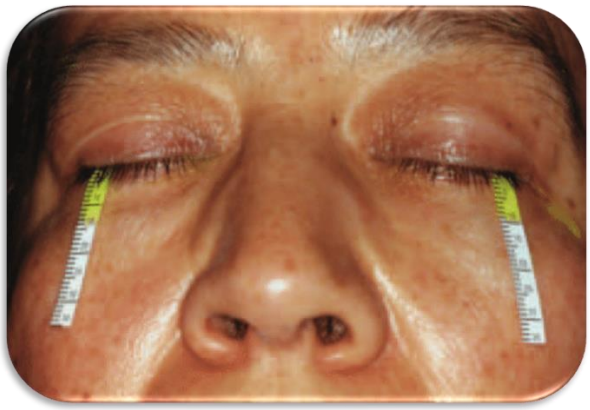
• SS-A (anti-Ro)

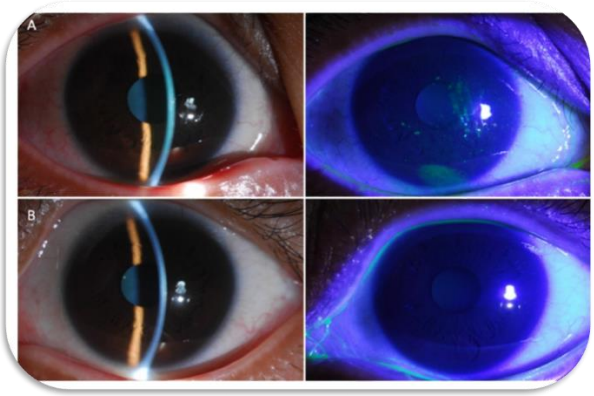
• SS-B (anti-La)

The diagnosis can be established by the Schirmer tear test, which measures tear flow over 5 minutes using absorbent paper strips placed on the lower eyelid

Staining with Rose Bengal may show punctate epithelial abnormalities

It can be confirmed by demonstrating focal lymphocytic infiltrate in a minor salivary gland biopsy





Management

No treatments that have disease-modifying effects have yet been identified and management is symptomatic:

Lacrimal substitutes

Occlusion of the lacrimal ducts is occasionally needed.

A trial of systemic Pilocarpine is worthwhile in early disease to amplify glandular function.

Saliva substitution should be considered the preferred therapeutic approach to alleviate symptoms in patients with no residual glandular function (severe glandular dysfunction), in whom salivary glands cannot be stimulated, either by pharmacological or non-pharmacological interventions. saliva substitutes are available commercially in the form of oral sprays, gels and rinses.

Hydroxychloroquine is often used to address skin and musculoskeletal features and may help fatigue.

For progressive interstitial lung disease and for interstitial nephritis (e.g. glucocorticoids and cyclophosphamide)

Inflammatory Myositis

Polymyositis (PM) and dermatomyositis (DM) are characterized by proximal skeletal and (cardiac and gut) smooth muscle inflammation. Both are notably connected with (either previously diagnosed or undisclosed) malignancy. The increased risk of malignancy associated with DM has been established both at the time of DM diagnosis and more than 10 years after diagnosis. No specific malignancy is associated with DM; rather, the most frequent malignancies are also the most common malignancies in the population.

Idiopathic inflammatory myopathies (IIMs) can occur in any age group, from early childhood to late in adult life. The onset of PM is usually in the late teens or older: the mean patient age at onset is 50 to 60 years. DM shows two peaks: 5 to 15 years and 45 to 65 years. Inclusion body myositis (IBM) is commonly seen in individuals older than 50 years.

A recent report notes a novel association of myositis with hypertension, diabetes, and ischemic heart disease.

Clinical features

The typical presentation of PM and DM is with symmetrical proximal muscle weakness over a few weeks. Patients report difficulty rising from a chair, climbing stairs and lifting, often (though not always) with muscle pain. The onset of muscle weakness is often subacute, occurring during a few weeks, or it can be insidious, developing during several months.

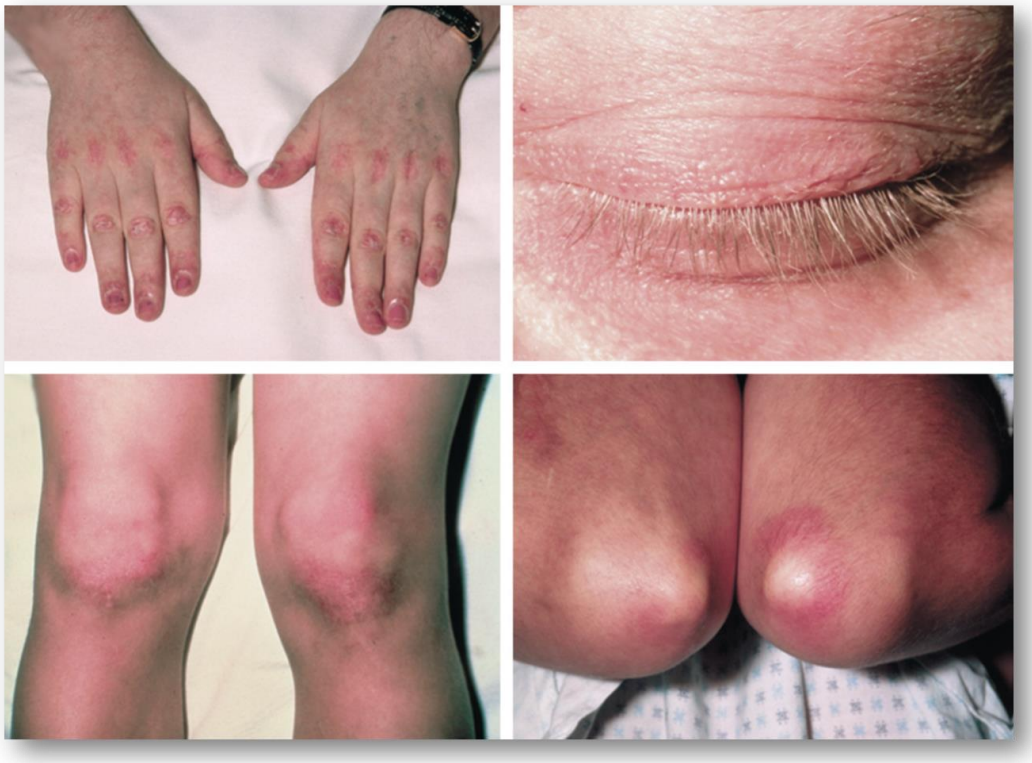
Systemic features of fever, weight loss and fatigue are common. Respiratory or pharyngeal muscle involvement can lead to ventilatory failure or aspiration. Interstitial lung disease occurs in up to 30% of patients.

In DM, the skin lesions include Gottron's papules, occurring over the extensor surfaces of PIP and DIP joints, and a heliotrope rash in the eyelid in combination with periorbital oedema. Similar rashes occur on the upper back, chest and shoulders ('shawl' distribution). Patients with DM often have skin lesions on their fingers, such as periungual erythema, nailfold telangiectasias. Calcinosis, which can be severe, is found mainly in juvenile DM but is occasionally seen in adults.

The skin rash can precede the muscle symptoms by months or even years, and in some patients, the skin manifestations may be the only clinical sign of DM. This condition is often called clinically amyopathic DM or DM sine myositis.

Lung involvement is frequent (30% to 40%) in PM and DM and is a major cause of morbidity and mortality in these disorders. Clinical symptoms such as dyspnea and cough are common. Lung involvement can be caused by weakness of the respiratory muscles or inflammation of the lung tissue (ILD).

IBM is more frequent in men than in women, and it is seen mostly in individuals older than 50 years. The onset is more insidious than that of PM or DM. frequent falls as a result of weakness in the knee extensor muscles. Difficulty swallowing may also be an early clinical feature, reflecting the involvement of the pharyngeal muscles.



Myositis Associated With Malignancies:

The association of cancer is much stronger for DM (20% to 30%) than for PM (10% to 15%). it is imperative to screen for tumors in patients with DM at the time of diagnosis and at relapse, particularly if the symptoms do not respond to conventional immunosuppressive treatment. The types of malignancies vary and include not only hematologic malignancies such as lymphoma but also solid tumors such as lung, ovarian, breast, and colon cancer.

Investigations

Muscle biopsy is the pivotal investigation. Occasionally, however, a biopsy may be normal, particularly if myositis is patchy so, invariably, MRI should be used to identify areas of abnormal muscle for biopsy. Serum levels of creatine kinase are typically raised and are a useful measure of disease activity, although a normal creatine kinase does not exclude the diagnosis. Generally, 80% to 90% of adult myositis patients show an increase in CK during the initial evaluation. However, a certain proportion of patients, especially those in advanced stages of the disease, show normal or relatively modest elevations in CK. AST, ALT, and LDH

Electromyography.

Screening for underlying malignancy should be undertaken routinely (full examination, chest X-ray, serum urine and protein electrophoresis, CT of chest/abdomen/ pelvis; prostate-specific antigen should be included in men, and mammography in women).

Management

Oral glucocorticoids (prednisolone 1 mg/kg daily) are the mainstay of initial treatment of PM and DM but high-dose intravenous methylprednisolone (1 g/day for 3 days) may be required in patients with respiratory or pharyngeal weakness.

Methotrexate and azathioprine are added to steroid. Rituximab appears to show efficacy in a majority of patients. Intravenous immunoglobulin (IVIg) may be effective in refractory cases. hydroxychloroquine has been used for skin predominant disease. IBM is usually resistant to treatment with glucocorticoids and other immunosuppressive agents.

Combining exercise and immunosuppressive therapy is a safe approach and has clear beneficial effects on muscle strength and function.

Emerging evidence suggests that exercise can even decrease muscle and systemic inflammation. Starting approximately 4 weeks after initiation of immunosuppression

Read more in:

Davidson’s Principles and Practice of Medicine, 23rd edition

Kelley & Firestein’s Textbook of Rheumatology, 10th edition