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 9 : 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.

Oral involvement manifests as a dry mouth (xerostomia). There is a high incidence of dental caries. Salivary gland enlargement

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.

Artificial saliva sprays, saliva-stimulating Tablets, chewing gum

A trial of systemic

Pilocarpine is worthwhile in early disease to amplify glandular function. 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.

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

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. 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). Periungual nail-fold capillaries are often enlarged and tortuous. Calcinosis, which can be severe, is found mainly in juvenile DM but is occasionally seen in adults.

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.

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.

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 MMF are the first choices of many but azathioprine and ciclosporin are also used as alternatives. 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