Sjogren's Syndrome

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Abstract

The syndrome may present alone (primary) or in association with an underlying connective tissue disorder (secondary). In elderly people Sjogren's syndrome (SS) is subclinical, relatively common and benign. In the elderly Sjogren's syndrome should be considered when sicca symptoms occur with systemic manifestations. Approximately 40% of xerostomia in the elderly is due to Sjorgren's syndrome and accounts up to 20% of Sjorgren's syndrome; arthralgias, Raynaud's phenomenon and purpura were common extraglandular manifestations. It is important to distinguish age-related gland pathology, drug-induced ocular and oral dryness from that due to SS.

Keywords

Sjogren's syndrome · Xerostomia · Raynaud's phenomenon · Purpura

Introduction

The prevalence of SS is about 3% in people above the age of 50 years, and the usual age of onset is at age 40–60 years [1]. It is common in middle-age women and the mean age is 52.7 years [2]. Its prevalence increases with age [3]. The syndrome may present alone (primary) or in association with an underlying connective tissue disorder (secondary) [4]. Approximately 40% of xerostomia in the elderly is due to

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Sjorgren's syndrome, and the elderly account in up to 20% of Sjorgren's syndrome [5]. In a study of 336 consecutive primary SS patients, in 21 (6%) the disease onset was after the age of 65 years [6]. In elderly people SS is subclinical, relatively common and benign [7].

Clinical Profile

SS at presentation is characterised by sicca symptoms of dry eyes (xeropthalmia) and/or dry mouth (xerostomia) due to a lymphocytic infiltration of the lacrimal or salivary glands or both [3, 8]. About 40% of the xerostomia in the elderly is due to SS [5]. In patients with disease onset after the age of 65 years, dry mouth and dry eye were the commonest occurrence. Arthralgias, Raynaud's phenomenon and purpura were common extraglandular manifestations [6]. However, Bostios et al. [6] found no statistical differences in relation to gender, disease duration and ocular and oral symptoms between the elderly onset and the young/adult onset. In the elderly there is often a delay between clinical onset and diagnosis, and this has been attributed to shared features of SS and old age [9].

Diagnosis

In the elderly the diagnosis of SS should be considered when systemic manifestations are associated with sicca symptoms [3]. The revised version of the American European Consensus Group (AECG) classification for diagnosis required the following, namely, the signs and symptoms of oral and ocular dryness, a positive salivary gland biopsy or autoantibodies against SSA/Ro and SSB/La antigens [10]. There is often a delay in the diagnosis in the late onset for the sicca symptoms have been frequently attributed to ageing and or medications [5], and it is important to distinguish age-related gland pathology and drug-induced ocular and oral dryness from that due to SS [9].

Treatment

Oral symptoms: Oral hygiene, salivary stimulation (sugar free chewing gum) and prevention of oral infection (antimicrobial mouth rinses) [4] and systemic stimulation of salivary secretion. Ocular symptoms: Topical (topical tear replacement) and followed by increased tear production. Two muscarine agonists, pilocarpine and cevimerline, have been shown to be effective [4, 11, 12]. Oral cevimeline has been shown to relieve subjective eye symptoms [12]. Systemic symptoms: An anti-CD20 monoclonal antibody (rituximab) that depletes B lymphocytes is a new potential therapy showing promise for severe inflammatory manifestations [4, 13].

Impact

Complications such as dental caries, corneal ulcerations, chronic oral infections and sialadenitis are preventable with early diagnosis [14]. Basic daily functioning such as eating, speaking and sleeping may be affected by the dryness in the mouth thus affecting the quality of life [15]. In SS there is a 20- to 40-fold increase in the incidence of lymphoma [16]. In the elderly, polypharmacy and increased rates of adverse events to medications make treatment complicated in the elderly [17]. Poor prognosis is associated with older age, delayed or inadequate treatment and malignancy (Box 1).

Box 1 Key Points: Sjogren's Syndrome

- Sjogren's syndrome (SS) is an autoimmune disorder characterised by lymphocytic infiltration and destruction of the salivary and lacrimal glands [8].
- The syndrome may present alone (primary) or associated with another connective tissue disorder [4].
- The usual age of onset is between 40 and 50 years and the late onset >65 years [1].

(*continued*)

Box 1 Key Points: Sjogren's Syndrome
(continued)

- The presentation is characterised by sicca symptoms, dry eyes and dry mouth [1].
- In the elderly SS should be considered when sicca symptoms occur with systemic manifestations [3].
- In the late onset the sicca symptoms are often attributed to ageing or to medications and hence a delay in the diagnosis [5].

Multiple Choice Questions

1. The following are true with Sjogren's syndrome (SS), except:
   A. It is an autoimmune disorder characterised by lymphocytic infiltration and destruction of the salivary and lacrimal glands.
   B. The syndrome may present alone (primary) or associated with another connective tissue disorder (secondary).
   C. Primary SS in the elderly is often clinical, severe and relatively less common.
   D. In the elderly sicca symptoms are often attributed to ageing and/or medications.

MCQ Answers

1 = C

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