SJOGREN'S SYNDROME — A CLINICAL STUDY OF 12 LOCAL PATIENTS

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SYNOPSIS

Twelve patients having Sjogren's Syndrome seen over a 5-year period were studied. 2 out of 3 clinical features — Dry eyes (Keratoconjunctivitis sicca), Dry mouth (Xerostomia) and an associated connective tissue disease — were used as inclusion criteria.

The patients were all Chinese, with 11 women and 1 man. Their mean age was 45 years (Range 33 – 68 years). Nine patients had decreased tear secretion (Schirmer's test positive). 60% (7 patients) had parotid gland enlargement. 9 patients had an associated immunological disorder. None had rheumatoid arthritis. Only 1 had Rheumatoid Factor present. 92% of the patients responded to instillation of methylcellulose eyedrops. One had spontaneous remission after 3 years.

A notable feature of this report is the absence of rheumatoid arthritis among local patients with Sjogren's Sydnrome.

INTRODUCTION

Sjogren's Syndrome is a chronic inflammatory autoimmune disorder characterised by a mixed cellular infiltration of exocrine glands. The most notable features were infiltration of the lacrimal and salivary glands resulting in xerophthalmia (dry eyes) and xerostomia (dry mouth) respectively. Historically filamentary keratitis was described by Leber in 1882, xerostomia by Hadden in 1888, and lacrimal and salivary gland enlargement without keratoconjunctivitis sicca or xerostomia by Mikulicz. However, the full description was not forthcoming until Henrik Sjogren, a Swedish ophthalmologist, published his review in 1933 (1).

This study characterises some features of local patients with this disorder.

METHODS

12 patients with Sjogren's Syndrome seen in the Department of Medicine, Singapore General Hospital were studied from 1980 to 1985. The Department of Medicine is a general medicine unit having 107,900 patient attendances over the 5 year period.

Sjogren's Syndrome was diagnosed when patients had at least 2 out of 3 clinical features: (1) Keratoconjunctivitis sicca (Dry eyes), (2) Xerostomia (Dry mouth), (3) An associated connective tissue disease.

Abnormal Schirmer's test results were defined as wetting of less than 5 mm of the filter paper in 5 minutes (2).

Labial or salivary gland biopsies were done by the ENT surgeon. Positive results were those of Grade III or IV by the criteria of Greenspan et al (3).

Hematological investigations and immunological markers were sent to the respective hospital laboratories. The Rheumatoid Factor was assayed by the Latex fixation method and titers greater than or equal to 1:80 were considered positive. Tests for SS A and SS B antibodies were not available.

RESULTS

a) Age

The ages of the patients range from 35 to 68 years with a mean of 45. (Fig. 1)

b) Sex, Ethnic group

Of the 12 patients, 11 (92%) are female and only 1 male. All the 12 patients were chinese.

c) Interval between onset of symptoms to diagnosis

The interval between onset of symptoms to diagnosis of the syndrome is short. 5 of the patients were diagnosed within 1 month of onset of symptoms and a further 6 were diagnosed within the year. The last patient had a lapse of 4 years prior to diagnosis (mainly due to a delay in seeking medical consultation). (Fig. 2)

d) Schirmer's test and salivary gland biopsy

Schirmer's test was performed in 9 patients.

7 patients have decreased tear secretion in both eyes while the remaining 2 have decreased tear secretion in only 1 eye. 5 biopsies were performed, with 3 showing features consistent with Sjogren's Syndrome.

e) Parotid gland enlargement

7 patients had parotid gland enlargement — 7 of them have bilateral involvement and 1 also had associated salivary gland enlargement.

f) Associated immunological disorder

9 patients have this association as shown in Table 1. 2 are associated with primary biliary cirrhosis, 2 with bronchial asthma and 1 each with chronic active hepatitis, mixed connective tissue disease, polymyositis, fibrosing alveolitis and alopecia areata.

g) Hematological investigations

Raised Erythrocyte Sedimentation rate (ie. > 20

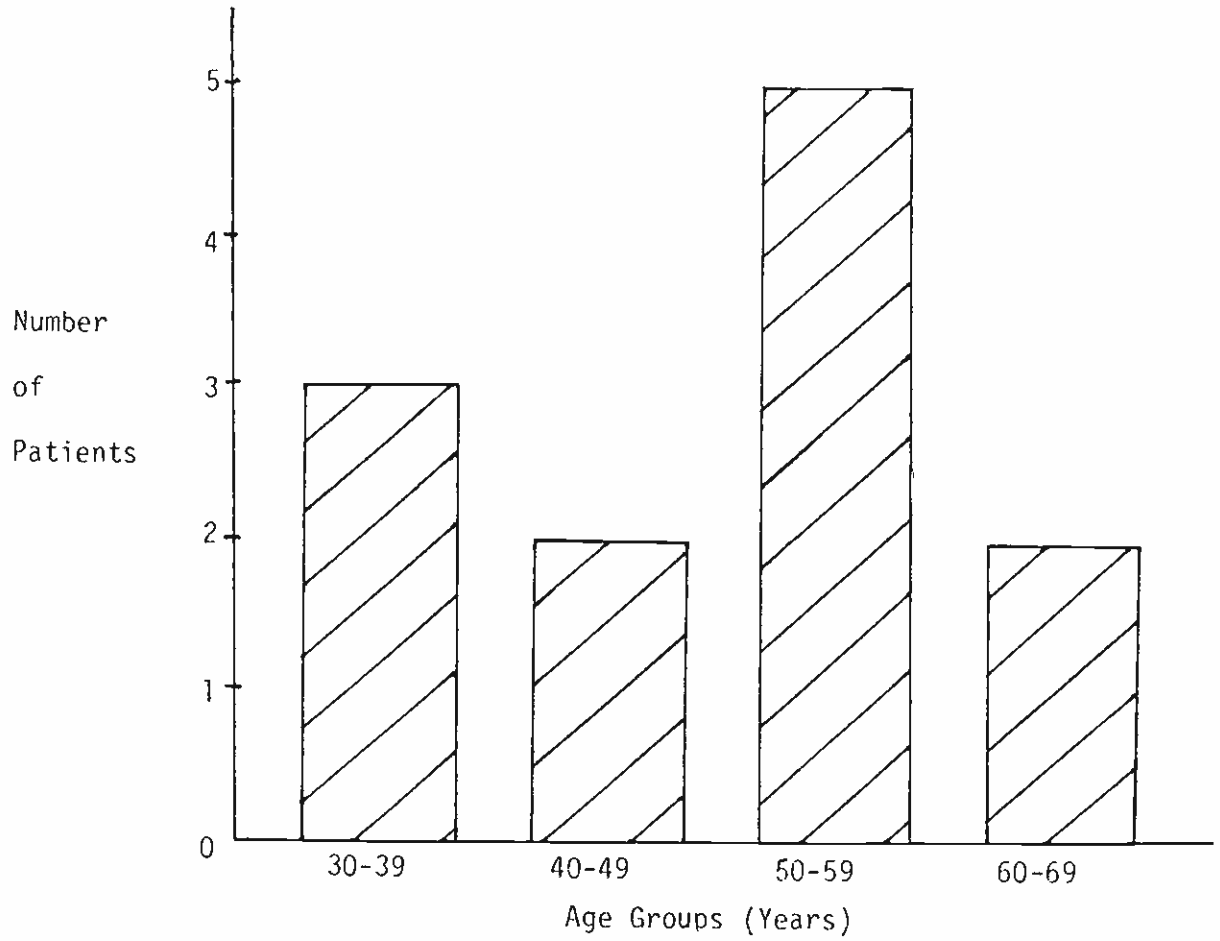

FIGURE 1: AGE DISTRIBUTION OF PATIENTS

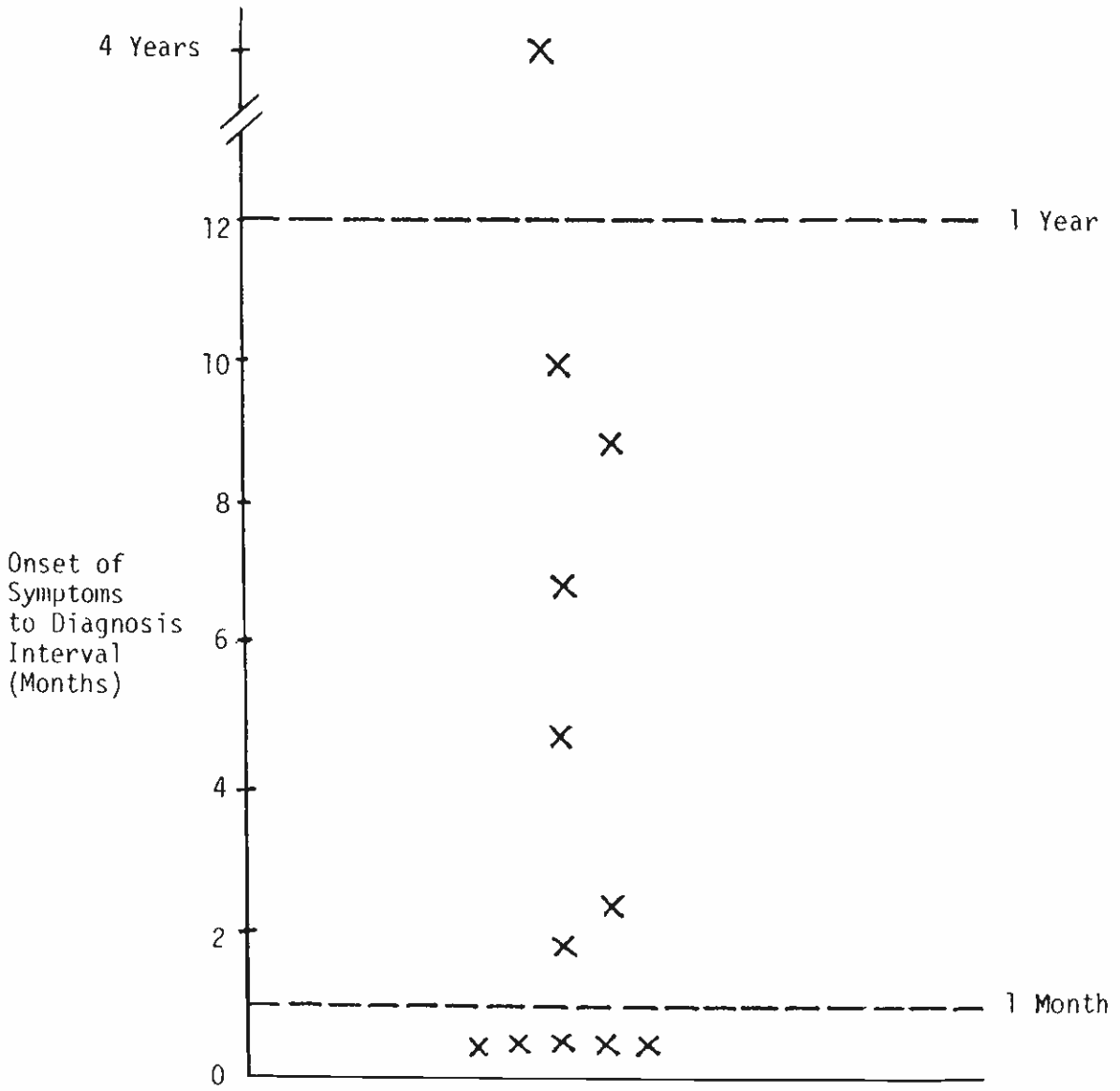

FIGURE 2: ONSET OF SYMPTOMS TO DIAGNOSIS INTERVAL

mm/hr) was present at time of diagnosis in 9 patients. The values ranged from 27 to 93 mm in the first hour. Normal total white cell count as well as lymphocyte count were noted in all patients. 3 patients have haemoglobin level less than 12.5 mg.dl. The only male patient had a mild eosinophilia (8% of the total white count).

TABLE I
ASSOCIATED IMMUNOLOGICAL DISORDER

Primary biliary cirrhosis	2
Bronchial asthma	2
Chronic active hepatitis	1
Mixed connective tissue disease	1
Polymyositis	1
Fibrosing alveolitis	1
Alopecia areata	1

TABLE 2
IMMUNOLOGICAL MARKERS OF SJOGREN'S SYNDROME

6 Patients have associated immunological markers	
Anti-nuclear factor	2
Mitochondrial antibody	1
Smooth muscle antibody	1
Rheumatoid factor and Anti-nuclear factor	1
Thyroid microsomal antibody and anti-nuclear factor	1

h) Immunological Markers

The associated immunological markers are shown in Table 2. 6 patients have associated immunological markers: Antinuclear factor (2 patients), mitochondrial antibody (1), smooth muscle antibody (1), Rheumatoid factor and antinuclear factor (1), Thyroid microsomal antibody and antinuclear factor (1).

DISCUSSION

Systemic autoimmune disorders have a higher prevalence among females e.g SLE (4), Progressive Systemic Sclerosis (5), Rheumatoid Arthritis (6). Female to male ratios range from 3:1 to 9:1. This study shows a similar predominance of the female sex (11:1). In many systemic autoimmune disorders, the syndrome is not well recognised in men. They may be present differently or have different clinical features. A recent study (7) of male patients with Sjogren's Syndrome reveals extraglandular involvement like articular, neurological, inflammatory vascular and lymphoproliferative disorders. The only male patient in our study does not have any extraglandular involvement.

The association of Sjogren's Syndrome with Rheumatoid Arthritis is widely recognised. 15% to 40% of patients were found to have this association (8,9). Surprisingly, none of the 9 patients with an associated immunological disorder had Rheumatoid Arthritis. No special explanation can be given as this general medical department does see a fair number of patients with Rheumatoid Arthritis. In our Rheumatology Clinic, none of our Rheumatoid Arthritic patients were found to have dry eyes or dry mouth.

A recent study (10) showed that Sjogren's Syndrome patients had high levels of SS A and SS B antibodies as compared to normal controls. The association of Rheumatoid Factor, Anti Nuclear Factor or SS A/SS B antibodies has been proposed as one of the criteria for the diagnosis of Sjogren's Syndrome (11). Six patients have associated immunological markers. 3 of the 6 patients with immunological markers have positive antinuclear factors and 1 of them also has an associated rheumatoid factor. The other 3 patients with mitochondrial antibody, smooth muscle antibody and thyroid microsomal antibody are patients with Primary Biliary Cirrhosis and Chronic Active Hepatitis. This relatively large number of such patients reflects the large number of patients with gastroenterological problems seen in this department.

Treatment of the dry eyes ranges from simple instillation of artificial tears, insertion of synthetic plugs into the punctae to transposition of the parotid duct. All our patients responded to artificial tears instillation. 1 had spontaneous remission of symptoms after stopping treatment.

With this study we were able to characterise some of the clinical features of local Sjogren's Syndrome patients. A notable feature is the lack of associated Rheumatoid Arthritis among our patients. Further study with larger numbers of patients will be very helpful to characterise the full clinical and serological features of our local patients.

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