Table 6.1. Frequency of extraglandular findings in primary Sjogren's syndrome.

Clinical Involvement	Percent
Arthritis	60
Kidney	9
Liver	6
Lung	14
Lymphadenopathy	14
Lymphoma	6
Myositis	1
Peripheral neuropathy	5
Raynaud's phenomenon	35
Splenomegaly	3

mune diseases in families of patients with primary Sjogren's syndrome (Anaya et al. 2006). The suggestion is that autoimmune diseases in general may aggregate as a trait favoring a common immunogenetic origin for diverse autoimmune phenotypes, such that a risk factor exists for the development of primary Sjogren's syndrome and other autoimmune diseases. Secondary Sjogren's syndrome is defined when the disease is associated with other clinically expressed autoimmune processes, specifically rheumatoid arthritis, systemic lupus erythematosus, myositis, biliary cirrhosis, systemic sclerosis, chronic hepatitis, cryoglobulinemia, thyroiditis, and vasculitis. Following rheumatoid arthritis, Sjogren's syndrome is the second most common autoimmune rheumatic disorder (Moutsopoulos 1993). Eight to 10 years are generally required for the disorder to progress from initial symptoms to the development of the syndrome. Although typically seen in middle-aged women, Sjogren's syndrome can occur in all ages and in males. It has been estimated that 80–90% of patients are women, and that the mean age at diagnosis is 50 years (Daniels 1991).

CLINICAL MANIFESTATIONS OF SJOGREN'S SYNDROME

Most patients with Sjogren's syndrome develop symptoms related to decreased salivary gland and lacrimal gland function. Primary Sjogren's syndrome patients generally complain of dry eyes, often described as a sandy or gritty feeling under the eyelids. Other symptoms such as itching of the eyes, eye fatigue, and increased sensitivity to light can accompany the primary symptoms. Many of these symptoms are due to the destruction of corneal and bulbar conjunctival epithelium and come under the diagnosis of keratoconjunctivitis sicca. This disorder is assessed by tear flow and composition. Tear flow is measured using the Schirmer test, while tear composition can be determined by tear break-up time or tear lysozyme content. The Schirmer test is considered positive when filter paper wetting of less than 5 mm occurs in 5 minutes, and suggests clinically significant keratoconjunctivitis sicca (Moutsopoulos 1993). There are, nonetheless, numerous false positive and negative results, such that the predictive value is limited. The integrity of the corneal and bulbar conjunctiva may be assessed using the Rose Bengal staining procedure and slip lamp examination. Punctate corneal ulcerations and attached filaments of corneal epithelium indicative of corneal and bulbar conjunctival epithelial destruction are noted on slip lamp examination in Sjogren's patients.

Xerostomia is the second principal symptom of Sjogren's syndrome. Xerostomia can be documented by salivary flow measurements, parotid sialography, and salivary scintigraphy. Salivary flow measurements must be adjusted for age, time of day, gender, and concomitant medications. Patients with dry mouths complain of a burning oral discomfort and difficulty in chewing and swallowing dry foods. Xerostomia is commonly associated with changes in taste and the inability to speak continuously for longer than several minutes.

Salivary gland enlargement occurs in as many as 30% of patients with Sjogren's syndrome during the course of their illness, with the parotid gland being most often enlarged (Figure 6.1) (Kulkarni 2005). Bilateral painful submandibular glands have been described as a presenting symptom of this syndrome (Kulkarni 2005). While the parotid glands are most commonly enlarged, they may be the last glands to be affected in patients with Sjogren's syndrome from the standpoint of decreased saliva production (Pijpe, Kalk, and Bootsma et al. 2007). The parotid glands have a longer-lasting secretory capacity in patients with Sjogren's syndrome, and therefore are the last glands to manifest hyposalivation during the disease. In the more advanced stages of the disease, both unstimulated and stimulated submandibular, sublingual, and parotid func-