scheme of lymphocytes and plasma cells per 4 mm^2 was established and has been described by numerous authors since its original description (Greenspan et al. 1974). Grade 0 referred to the absence of these cells, grade 1 showed a slight infiltrate, grade 2 showed a moderate infiltrate or less than one focus per 4 mm^2, grade 3 showed one focus per 4 mm^2, and grade 4 showed more than one focus per 4 mm^2. It has been noted that grade 4 (more than one focus of 50 or more lymphocytes per 4 mm^2 area of gland) is seen only in patients with Sjogren's syndrome and was not seen in postmortem specimens. Due to the strong association with the presence of Sjogren's syndrome, focal sialadenitis in a labial minor salivary gland incisional biopsy specimen with a focus score of more than one focus/4 mm^2 has been proposed as the diagnostic criterion for the salivary component of this disease (Daniels, Silverman, and Michalski et al. 1975). It has been pointed out that the focus score cannot separate early from late disease as chronicity of symptoms and focus score did not show a relationship (Greenspan et al. 1974). The highest focus score, however, was seen in patients with the sicca components of Sjogren's syndrome without associated connective tissue disease. Finally, since variation of disease apparently exists from minor salivary gland lobe to lobe, at least 4–7 lobes of minor salivary gland tissue should be removed and examined microscopically (Greenspan et al. 1974).

Incisional biopsy of the parotid gland has at least theoretical benefit and justification in the diagnosis of Sjogren's syndrome. Previous recommendations for major salivary gland biopsy reported potential complications of facial nerve damage, cutaneous fistula, and scarring of the facial skin when utilizing a parotid biopsy to establish or confirm a diagnosis of Sjogren's syndrome. Incisional parotid biopsy may be performed without assuming any of these complications, except in very rare circumstances (Marx, Hartman, and Rethman 1988). Recent studies, in fact, point to a higher yield of diagnosis when using the parotid biopsy (Marx 1995) (Figure 6.6). In Marx's series of 54 patients with Sjogren's syndrome, 31 (58%) had a positive labial biopsy, while 54 (100%) had a positive parotid biopsy (Marx 1995). He concluded his study by stating that incisional parotid biopsy will confirm and definitively document the diagnosis of Sjogren's syndrome (Figure 6.7). The incisional parotid biopsy will also serve to rule out the presence of lymphoma, which is observed to develop in approximately 5–10% of patients with Sjogren's syndrome (Daniels 1991; Talal and Bunim 1964). Patients with Sjogren's syndrome are felt to have 47 times greater incidence of lymphoma than that of an age-controlled population (Marx, Hartman, and Rethman 1988). Ten such lymphomas were reported in Marx's study. They developed 4–12 years after the diagnosis of Sjogren's syndrome was made, with a mean of 7.2 years. In 8 of the 10 cases, a rapid change in the size of the parotid enlargement was noted, and all of the patients exhibited a darkening of the skin overlying the enlarged parotid gland. These changes dictated biopsy of the parotid gland in the background of the systemic disease, with the knowledge that lymphoma does not develop in the lower lip in patients with Sjogren's syndrome.

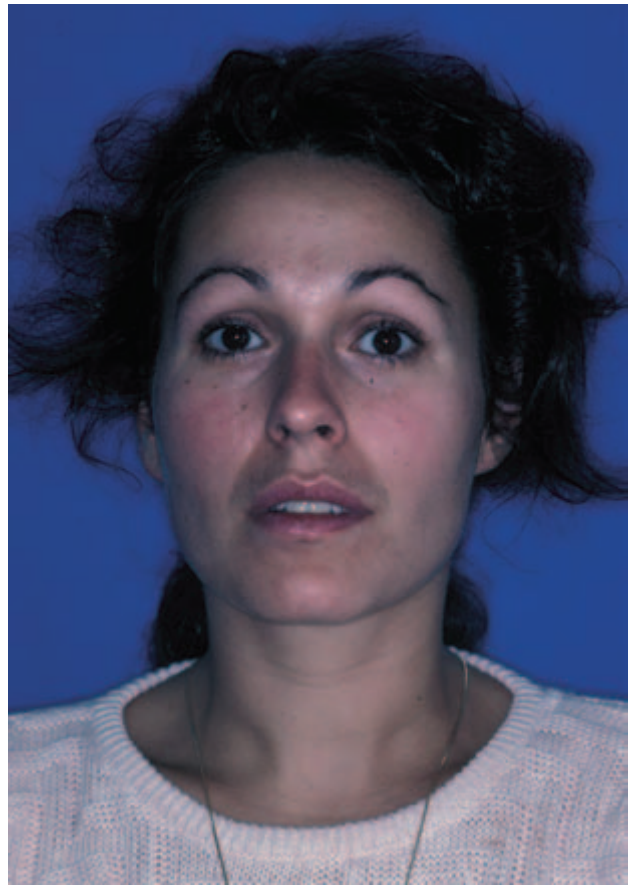

Figure 6.6a. A 32-year-old woman with the recent development of dry eyes and mouth, possibly suggestive of Sjogren's syndrome.