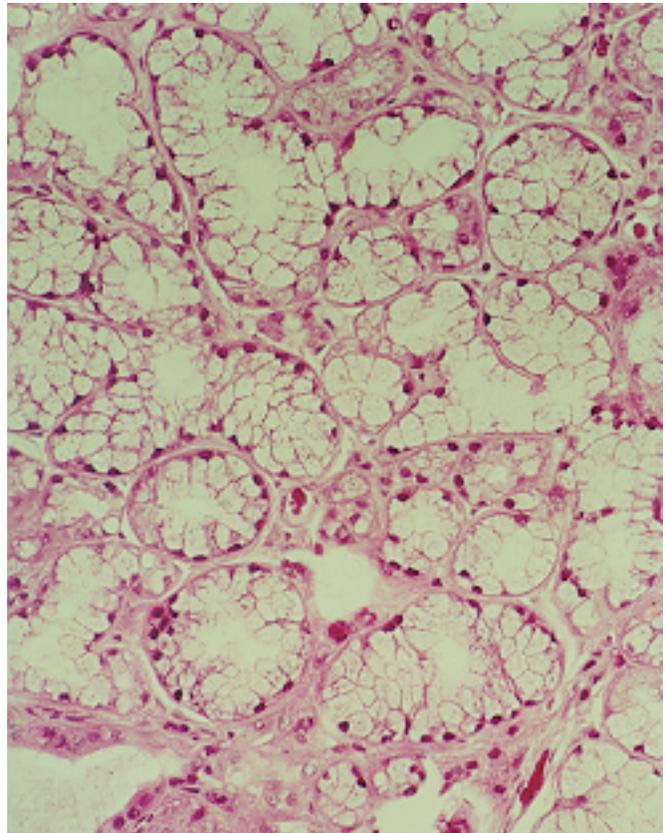


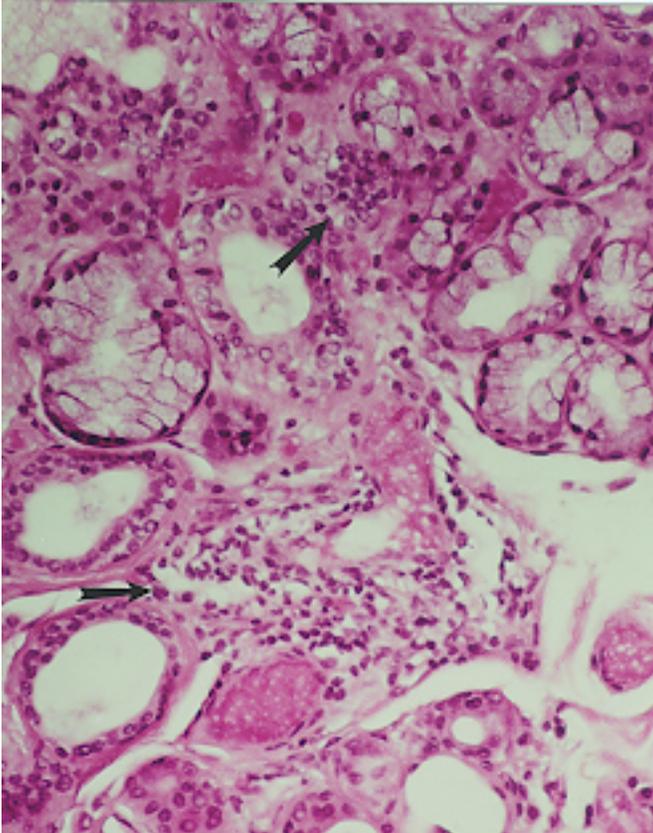
SJOJREN'S SYNDROME

Sjogren's syndrome is an auto-immune disorder of unknown etiology affecting particularly middle aged women. It may be associated with other connective tissue disorders such as rheumatoid arthritis, lupus erythematosus, scleroderma, thyroiditis, and polymyositis. As a primary isolated disorder it is also called "sicca syndrome." Chief head and neck symptoms include dry mouth and dry eyes. The salivary and lacrimal glands may be swollen. The Schirmer test is helpful in diagnosis. The sedimentation rate and serum immunoglobulins, especially, are elevated.

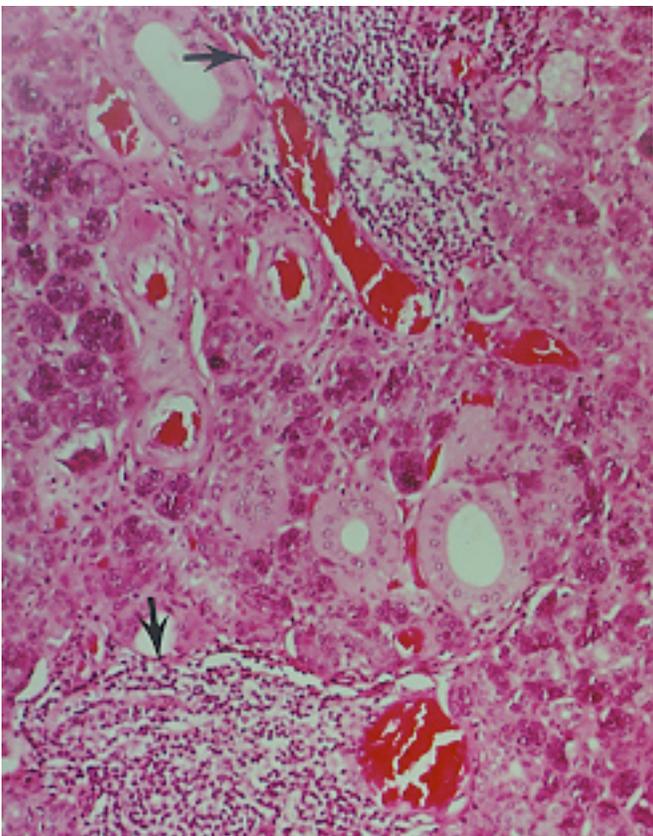
Histologically, in early cases a biopsy of the minor salivary glands of the lower lip shows focal periductal chronic lymphocytic exudate (50 or more lymphocytes and plasma cells) in more than one area with the inflammatory aggregates adjacent to normal acini and ducts. In advanced cases, the glandular acini of major glands disappear leaving largely fat and fibrous tissue along with the ducts of the original gland.

Lip, normal. These are accessory salivary glands of the mucous type. Other areas of the lip contain serous and sero-mucous glands.



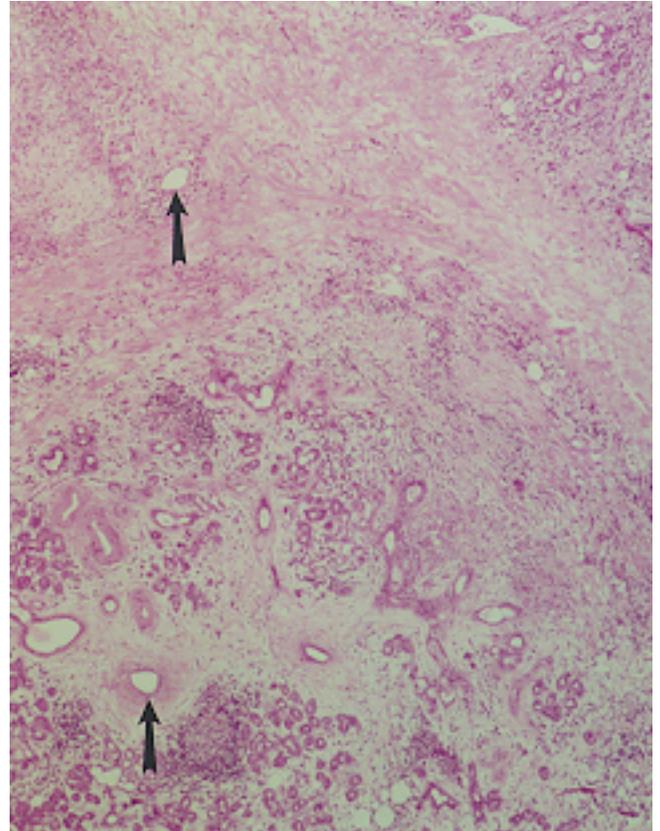


Sjogren's syndrome, lip. Aggregates of lymphocytes and plasma cells (arrows) adjacent to normal acini and ducts.



Sjogren's syndrome, submandibular salivary gland. Periductal aggregates of chronic inflammatory cells (arrows).

Submandibular gland. Marked fibrosis and chronic inflammatory infiltrate in patient with suspected Sjogren's syndrome. Most acini are destroyed leaving chiefly ductal elements (arrows) which may become dilated and filled with debris.



CLINICAL ASPECTS

Since Sjogren's syndrome is a systemic disorder, in addition to xerostomia and xerophthalmia there may be dry skin, dry nose, bronchitis, nosebleed, and otitis media. Sialography, though not diagnostic, may show sialectasia. Biopsy of the parotid gland may be even more useful than lip biopsy but because of risk to the facial nerve, lip biopsy is generally used.

Treatment is largely symptomatic. There is 40 times increased risk of lymphoma in a patient with Sjogren's syndrome. Mikulicz's syndrome is used to include lacrimal and salivary gland enlargement due to additional causes including sarcoidosis, leukemia and lymphoma. This term also is used by some as synonymous with benign lymphoepithelial lesion.