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Sicca Syndrome Due to Primary Amyloidosis

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Amyloidosis, especially primary amyloidosis, is well known as a mimic of other diseases. Though it has often been described in the cornea, conjunctiva, and eyelid (Brownstein, 1968) and rarely in other parts of the eye and orbit it has not apparently been reported to affect the lacrimal gland. The following report suggests that amyloidosis should be considered in the differential diagnosis of the sicca syndrome.

Case Report

A 71-year-old gardener presented in April 1970 with a short history of breathlessness and swollen ankles. On examination he was noted to have a raised jugular venous pressure, basal crepitations, hepatomegaly, and peripheral oedema. The pulse was regular, blood pressure was 180/90 mm Hg, and heart sounds were normal. There was an ejection systolic murmur but no cardiac enlargement. In addition, it was noted that the mouth was very dry and lacking in saliva. On direct questioning he admitted having had a dry mouth and "gritty eyes" for about one year.

Schirmer's test for lacrimation was positive. A chest x-ray picture was normal; there was no evidence of cardiac enlargement. Electrocardiograms showed left ventricular preponderance but no other abnormality. In addition, he was found to be in moderate renal failure, with a blood urea of 138 mg/100 ml and a serum creatinine of 2.8 mg/100 ml. An intravenous pyelogram showed diminished excretion but no other obvious abnormality. Serum electrolytes, urine microscopy and culture, and serum calcium and phosphorus were all normal. Liver function tests showed a serum alkaline phosphatase of 54 King-Armstrong units and a slightly raised serum alanine aminotransferase of 68 units. Serum bilirubin was 0.9 mg/100 ml. The erythrocyte sedimentation rate (Westergren) was 38 mm in the first hour, haemoglobin 13.8 g/100 ml, and white cell count 9,200/mm³ with a normal differential.

These investigations showed that he had renal, hepatic, and cardiac disease in addition to the "sicca syndrome." A tentative diagnosis of a connective-tissue disease was made but lupus erythematosus cells and antinuclear factor were not found and the plasma protein electrophoresis was normal. A liver biopsy specimen was reported to show only mild non-specific abnormalities. A technetium liver scan showed multiple filling defects suggesting secondary neoplastic deposits; barium studies of the gastrointestinal tract showed no primary site. No definite diagnosis was established and he was treated symptomatically with diuretics and discharged from hospital.

In June 1970 he was noted to have developed bilateral foot-drop; soon after this he went into liver failure, which presented with an obstructive jaundice. Again he was treated symptomatically with neomycin, dietary restriction of protein, and diuretics. The liver at this time appeared to be increasing in size. After recurrent bouts of liver failure he died from a combination of oliguric renal failure and hepatic failure five months after first presenting.

Necropsy.—The body was wasted, jaundiced, and covered with petechiae. Serous effusions were present in the pleural and peritoneal cavities. A pseudocyst of the pancreas was present. The main abnormal finding was gross amyloidosis of liver, spleen, kidneys, and heart. Histological examination confirmed the presence of amyloid in these organs and also in the testes, tongue, pituitary, lymph nodes, oesophagus, pancreas, adrenals, and thyroid. Sections from the left parotid and lacrimal glands showed deposition of amyloid in interlobular septa and between acini which, especially in the parotid, were somewhat atrophic. Apart from a few scattered lymphocytes in the section of the parotid, the lacrimal and salivary glands showed no cellular infiltrate or fibrosis. The histological picture appeared to exclude the possibility of active or healed inflammation. Amyloid was also present in the walls of small arteries and sebaceous glands in the eyelid. Amyloid stains on sections of the previous liver biopsy specimen showed that amyloid was present, but in such small amounts as to be imperceptible without special stains. The cause of death was recorded as renal, hepatic, and cardiac failure due to "primary" amyloidosis.

Comment

The lack of inflammation or fibrosis in lacrimal and salivary glands excludes the common chronic inflammatory form of the sicca syndrome. There seems little doubt in retrospect that in this patient the clinical syndrome was due to amyloidosis. The lack of chronic inflammation here or elsewhere in the body and the distribution of amyloidosis suggests the "primary" form.

This case illustrates once again the varied clinical guises of amyloidosis and raises a further possible diagnosis to be considered in sicca syndrome. Clearly the entity described here is rare, but might occasionally require to be distinguished from Sjögren's syndrome with secondary amyloidosis, which has been recognized previously (Gardner, 1965).

An association of sicca syndrome with hepatic and renal failure, as seen in the present case, will also require to be distinguished from the combination of sicca syndrome with autoimmune liver disease (Golding *et al.*, 1970) and with hyperglobulinaemic renal tubular acidosis (Mason and Golding, 1970).

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