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TEMPORAL ARTERITIS WITH SJÖGREN'S SYNDROME

(temporal arteritis/sjögren's syndrome/peripheral neuropathy)

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A 79-year-old Japanese woman with temporal arteritis accompanied by Sjögren's syndrome proven upon histological examination was reported. A biopsy of the temporal artery showed typical findings of giant cell arteritis with deposition of IgG and C3 by and immuno-fluorescence study. A complication with latent peripheral polyneuropathy was also obseved. It is known that Sjögren's syndrome is often associated with connective tissue diseases, but a complication with temporal arteritis has rarely been reported.

Giant-cell arteritis includes temporal arteritis and Takayasu's aortitis. Temporal arteritis is common among elderly Europeans and Americans, but is very rare in Japan. On the other hand, most patients with Takayasu's aortitis are young Japanese females (1). It is known that Sjögren's syndrome is often associated with connective tissue diseases, e.g., rheumatoid arthritis, but a complication with temporal arteritis has rarely been mentioned (2). It is for these reasons that the following case is thought to be worth recording.

CASE REPORT

A 79 year old woman developed mild headache in the right temporal region one month prior to admission. At the same time, she noted thickening and tenderness of the bilateral temporal arteries. She had the sensation of a dry mouth and generalized weakness with moderate loss of weight starting seven months

before admission. She had no visual complaints. Physical examination revealed a obese, well developed-woman. Body temperature was 36.1°C, blood pressure was 128/70 mmHg bilaterally and pulse rate was 72 regular beats per minute. The bulbar conjunctiva and palpebrae were normal. The tongue was slightly dry with foamy saliva. The temporal arteries were thickened and elongated with tenderness, and pulsation was not palpable. Bruit was not audible in the neck, orbita or abdomen. The chest and abdomen were normal. Neurological examinations revealed that the cranial nerves including those involved with visual acuity, visual field and optic fundi were normal. Deep tendon reflexes were diminished in the bilateral knees and ankles. Vibration sense was moderately diminished in the ankles, but no muscle weakness was noted. Romberg's sign was negative. The remainder of the physical and neurologic examination was normal.

Results of laboratory studies were as follows: hemoglobin value, 13.7 g/dl; leucocyte count, 9,600 per cubic millimeter, with 75% neutrophils, 20% lymphocytes, 2% monocytes and 1% eosinophils; platelet count, 213,000 per cubic millimeter; erythrocyte sedimentation rate (ESR), 29 mm per hour; and total serum protein, 8.1 g/dl with 4.9 g/dl of albumin, 0.3 g/dl of alpha-1 globulin, 0.8 g/dl of alpha-2 globulin, 0.8 g/dl of beta globulin, and 1.3 g/dl of gamma globulin. The liver and renal functions were within normal range, and electrolytes were normal. Total cholesterol, 257 mg/dl; triglyceride, 329 mg/dl; high-density lipoprotein (HDL), 34 mg/dl; amylase, 139 IU/l; creatin phosphokinase, 37 IU/l. The prothrombin time was normal. The Venereal Disease Research Laboratories test for syphilis and clot test for LE cells were negative. The flocculation test for rheumatoid factor was positive (+). The thyroid and microsome tests were negative, and antinuclear antibody was 10* with a homogeneous pattern. Anti-DNA antibody was 22 IU/l. CH50 was 50 mg/dl and immune complex was negative. Immunoglobulin G (IgG), 1570 mg/dl; IgA, 135 mg/dl; IgM, 50 mg/dl. C-reactive protein (CRP) was (+). Urinalysis was normal. Creatinine clearance was 54.6 ml/min, the Fishberg test was normal, and the PSP test showed 15% for 15 minutes value. Roentgenograms of the chest were normal, and that of cervical spine showed changes of degenerative arthritis, but no vascular calcification. An electrocardiogram and electroencephalogram were normal. As for

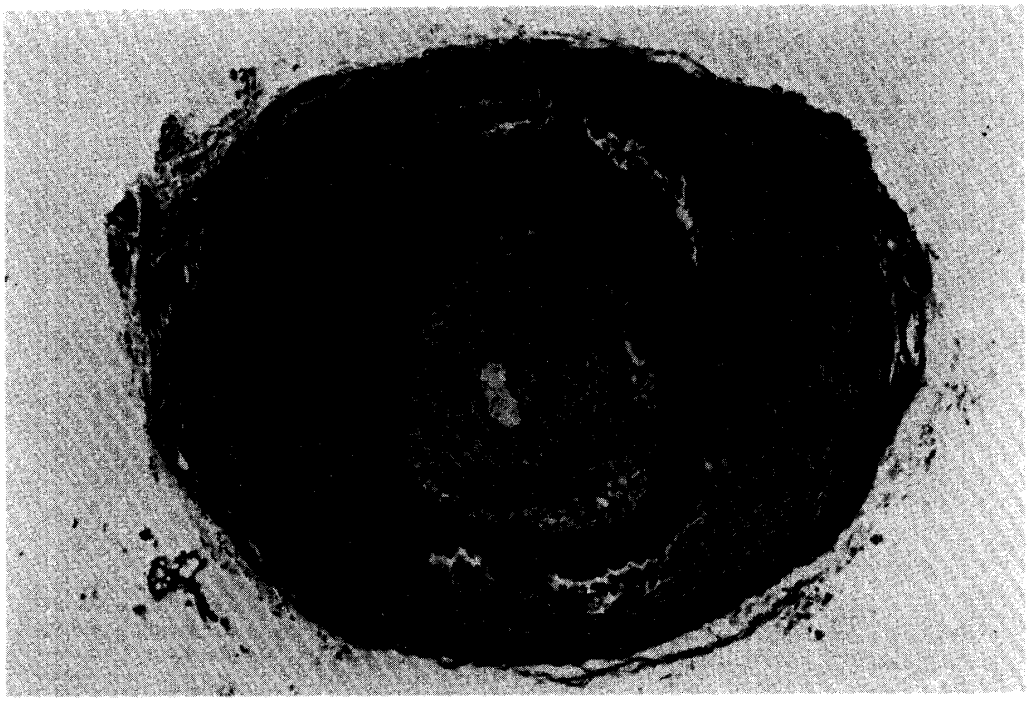

Fig. 1. Right temporal artery: Occlusion of the lumen by intimal proliferation containing moderate numbers of cells with disruption of the internal elastic lamina. Diffuse mononuclear cell infiltration with giant cells in the media. (Hematoxylin and eosin.)

the nerve conduction velocities, the distal latency of the median and ulnar nerves was prolonged, and delay of the sensory conduction velocities of the sural nerve was observed.

The biopsy of the right temporal artery showed almost occluded vascular lumen with marked intimal thickening. Disruption of the internal and external lamina elastica and prominent infiltration of the mononuclear cells with giant cells were observed (Fig.1). These findings were compatible with typical giant-cell arteritis. The immunofluorescence study of the vessel showed prominent deposition of IgG in the mononuclear cells as the cytoplasmic pattern, and C3 deposited in the sarcolemma of the media as the linear pattern.

A Schirmer's test revealed 9 mm for the right and 10 mm for the left and salivary production decreased by 1 ml in 10 minutes. A rose-bengal test showed a dyeing of the bulbar conjunctiva. Sialography revealed mild dilation of the main salivary duct. A biopsy of the labial accessory salivary glands showed mild atrophy of the minor salivary glands with fibrosis and focal mononuclear cell infiltration in clusters, as shown in Fig.2. These findings were compatible with definite Sjögren's syndrome.

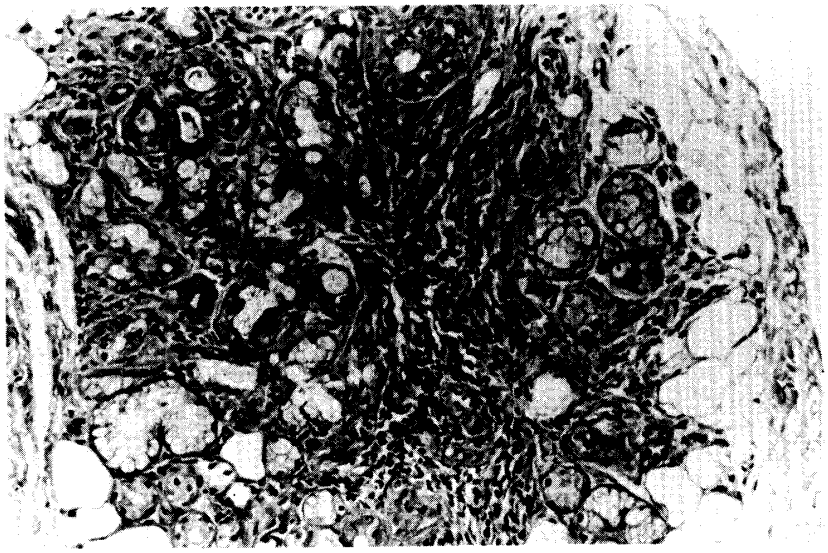

Fig. 2. Labial accessory salivary glands: Mild atrophy of minor salivary glands with fibrosis and focal mononuclear cell infiltration in clusters. (Hematoxylin and eosin. 100*.)

The patient was given by 40 mg per day of prednisolone (PSL) from two weeks after admission (November, 16, 1983), and her headache disappeared within three days. The temporal arteries became soft and palpable, and tenderness disappeared the following week. CRP and ESR normalized two weeks after the administration of PSL. She has been well controlled up to the present by 10 mg of PSL per day.

DISCUSSION

Temporal arteritis is now a well-recognized entity with which most physicians and pathologists in America and Europe are familiar. However, it is considerably rare in Japan, in contrast to the high incidence of Takayasu's aortitis, which is also classified as giant-cell arteritis. Only 72 cases of definite temporal arteritis have been reported in Japan since Kameyama et al. (1966) (3) described the first one.

These two types of giant cell arteritis show quite different clinical pictures. Temporal arteritis occurs in elderly people, in contrast to Takayasu's aortitis which is seen in young females. In addition, the former mainly involves the arteries above the neck, especially the temporal arteries, while the major site of the latter is the aortic arch. It has been known that

the involvement of the peripheral arteries, such as the temporal artery, is very rare in Takayasu's aortitis (1). Polymyalgia rheumatica often accompanies temporal arteritis (4), it's complicating rate is 37.5% (27/72) even in Japan. Alestig et al. (5) have reported a 64-year-old female case of polymyalgia rheumatica associated with Takayasu's aortitis. Such a case, however, has not been reported in Japan, although Takayasu's aortitis is far more common in this country than in Europe or America. There is some doubt concerning the diagnosis of Alestig et al.'s case, because the patient is considered too old for Takayasu's aortitis, and Hunder et al. (6) have reported a case of temporal arteritis producing an aortic arch syndrome.

Therefore, temporal arteritis should be distinguished from Takayasu's aortitis despite the same pathological entity of giant-cell arteritis. It is known that Sjögren's syndrome is associated with connective tissue disease, e.g., rheumatoid arthritis, but its complication with temporal arteritis has rarely been mentioned (7). We could find only one case of this complicaion, namely, that reported by Chagnon et al. (1983) (2). Recently, primary Sjögren's syndrome has come to be noticed with regard to neurological complications due to vasculitis (8). We have experienced more than 20 cases of primary Sjögren's syndrome with neurological symptoms in the past three years. This syndrome may be one of the important causes of peripheral neuropathy of CNS symptoms in elderly people. Temporal arteritis often involves the optic nerve or other cranial nerves, but peripheral neuropathy is a rare complication (9). Therefore, latent peripheral neuropathy in this case may be related with Sjögren's syndrome.

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