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UNILATERAL OCULOMOTOR NERVE PALSY IN PROGRESSIVE SYSTEMIC SCLEROSIS

(oculomotor nerve palsy/progressive systemic sclerosis)

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A case of unilateral oculomotor nerve palsy associated with progressive systemic sclerosis (PSS) is described. Recurrent psychotic symptoms and electroencephalographic abnormalities were also observed. To our knowledge, this is the first case to present oculomotor nerve palsy in PSS. Microangiopathy seems to be a possible cause for the neurological manifestations in PSS.

Neurological disorder in progressive systemic sclerosis (PSS) has been regarded as a rare manifestation (1). Sensory trigeminal neuropathy has been noted in several cases of PSS (2), but the involvement of other cranial nerves is extremely rare. We present herein a case of PSS with unilateral oculomotor nerve palsy.

CASE REPORT

A 72-year-old woman was admitted to the hospital on April 28, 1983, with diplopia. The patient had noticed Raynaud's phenomenon 13 years prior to admission. Dysphagia and substernal discomfort gradually appeared 10 years before admission, and laparotomy was carried out because esophageal cancer was in doubt with an upper G.I. series. However, there was no malignancy in the esophagus. These symptoms persisted even after the operation. The patient consulted a psychiatrist because of

recurrent confusional state starting five years before admission. Mild hypertension was pointed out and antihypertensive therapy was continued from 1980. Three days before admission, she noticed diplopia upon looking to the right.

On general examination, blood pressure was 170/100 mmHg, pulse was 72 beats per minute, regular. Her skin was tight and shiny only over the fingers of both hands (sclerodactyly), but there was no telangiectasia. On neurological examination, she was alert and cooperative. The left eye was deviated to the left on looking straight forward, and its vertical and horizontal gaze to the right were restricted. Marked ptosis of the left side was noted, and the left pupil was dilated and did not react to direct or indirect light stimulation. Other cranial nerves were intact. No abnormalities were detected in the motor and sensory systems in the extremities. The deep tendon reflexes were symmetrical, and the plantar responses were flexor.

Laboratory examinations disclosed that the hematocrit was 34% and the erythrocyte sedimentation rate was 37 mm/hr; other hematological and blood chemistry values were within normal limits. The oral glucose tolerance test disclosed borderline findings. C-reactive protein was ++, but the latex fixation test and VDRL test were negative. Anti-RNP antibody was not detected and none of the viral titers was significantly elevated in the serum. The cerebrospinal fluid was normal. The motor conduction velocity of the tibial nerve was slightly delayed at 39.5 meters per second, while those of the median and the ulnar nerve were within normal limits. A repetitive Tensilon test was negative. An X-ray examination of the esophagus disclosed marked dilatation and diminished peristalsis. The left vertebral and bilateral carotid angiograms and the enhanced CT scan revealed no abnormalities.

She was treated with 30 mg per day of prednisone from the fourth day after admission, but her ocular symptoms did not improve. Thus, prednisone was tapered down. During this tapering down, an acute confusional state developed. An electroencephalogram (EEG) at this time showed diffuse slow waves predominantly in the frontotemporal leads (Fig.1). This disappeared 10 days after the administration of major tranquilizers, but the EEG abnormality continued for three months. The left oculomotor nerve palsy also continued for more than three months and gradually improved.

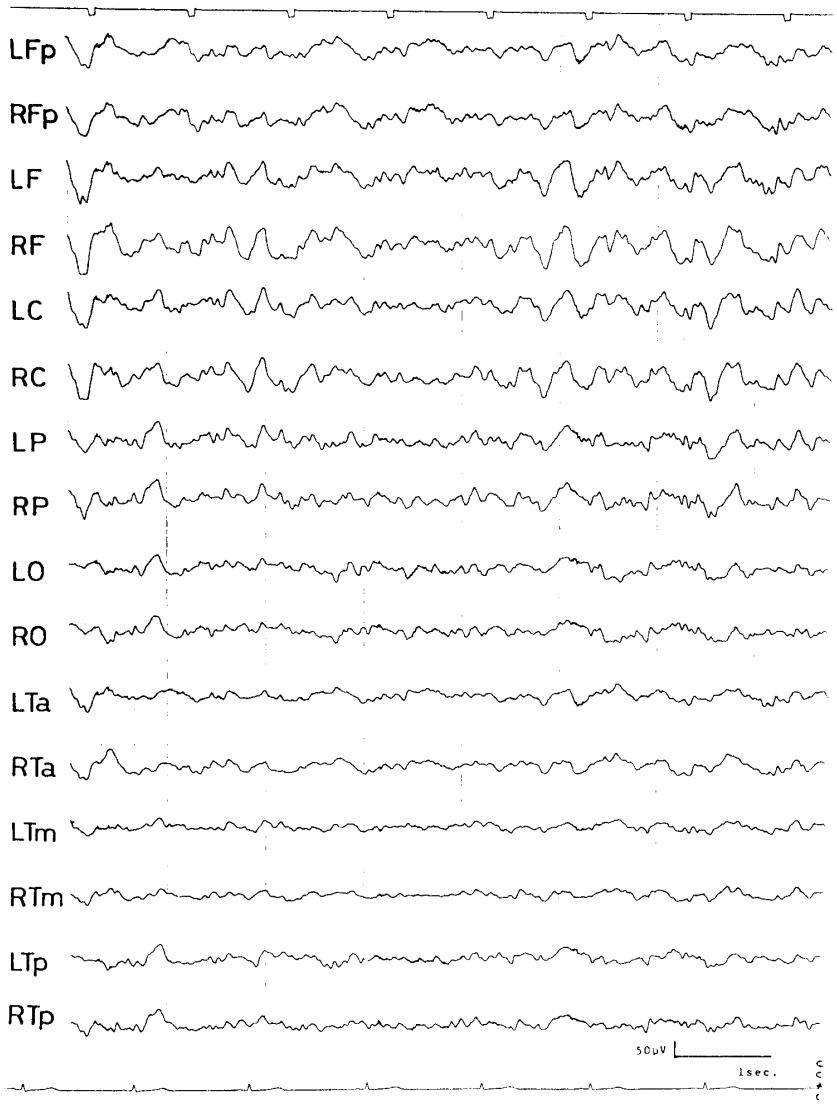

Fig.1. An EEG during the confusional state shows sporadic delta waves in the frontotemporal leads with the basic rhythm of a theta wave.

DISCUSSION

Our patient disclosed a localized sclerosis on the fingers

and no visceral involvement except for the esophagus. Thus, her clinical features were similar to CREST syndrome (calcinosis, Raynaud's phenomenon, esophageal dysfunction, sclerodactyly, and telangiectagia), a benign variant of PSS, although calcinosis and telangiectasia were not observed. As the neurological complication, trigeminal neuropathy has been reported and is said to be more frequent in CREST syndrome than in diffuse sclerosis (2).

The most striking symptom in our patient was a unilateral oculomotor nerve palsy associated with PSS. To our knowledge, the association of an oculomotor nerve palsy with PSS has not been reported previously (3). The differential diagnosis of an oculomotor nerve palsy includes a variety of pathological conditions in addition to connective tissue disease. These conditions usually include aneurysms, tumors, diabetes mellitus, viral infections, and myasthenia gravis. But aneurysms and tumors were not found in the angiograms and the CT scan. Diabetes mellitus, viral infections, and myasthenia gravis were also excluded by the laboratory findings and the Tensilon test. Thus, we suppose that the unilateral oculomotor nerve palsy may have been caused by PSS itself. The mechanism of the involvement of the oculomotor nerve in our patient is obscure. But pathologic changes, that is, thickening of the connective tissue sheaths of the nerves, deposition of mucoid materials around the nerve fibers, and changes in the vasa nervosum have been reported in PSS with peripheral neuropathy (4,5). The pathological changes listed herein, especially microangiopathy followed by fibrosis of the vasa nervosum, seem to be responsible for the oculomotor nerve palsy in our patient.

In addition to the oculomotor nerve palsy, our patient showed psychotic symptoms with EEG abnormalities. This was initially suspected to have been caused by steroid therapy, but prolonged EEG abnormality and a past history of psychotic episodes could not be explained by the side effects of corticosteroid alone. In general, central nervous system involvement in PSS is considered to be quite rare. It has been, however, reported that subclinical abnormalities of EEG in PSS are not infrequent (6), and a case of PSS with cerebral vasculitis has been reported (7). Therefore, it is possible that vasculitis in the central nervous system can cause psychotic disorder and EEG abnormality.

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