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The Crow-Fukase (POEMS) Syndrome with Vasospastic Angina

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The Crow-Fukase syndrome (CFS) [polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome] is a rare multisystem disorder characterized by polyneuropathy, anasarca, skin changes, endocrinopathy, dys-globulinemia, organomegaly, and polycythemia/thrombocytosis, frequently associated with myeloma [1, 2]. Pathomechanisms underlying the syndrome remain unclear. We describe here a patient suffering from the syndrome associated with an unusual complication, vasospastic angina.

A 36-year-old male dentist was admitted to the hospital with a half-year history of numbness and weakness of extremities. He also complained of diarrhea, back pain, and impotence. Physical examination revealed slight fever, pigmentation of the skin, hypertrichosis, swelling of a cervical lymph node, hepatomegaly, leg edema, and sensorimotor polyneuropathy with greater involvement of distal legs. Laboratory studies demonstrated thrombocytosis (86.0x 10⁴/ul), serum M protein (IgG X type), impaired glucose tolerance, low serum levels of T3, T4 and testosterone, hyperprolactinemia, elevated serum interleukin (IL)-6 levels (12.7 pg/ml; normal <4.0), a solitary osteolytic myeloma in the 10th thoracic vertebra, elevated CSF protein levels (HOMg/dl), decreases in the motor and sensory nerve conduction velocities, and demyelinating changes in the biopsied sural nerve. After the diagnosis of the CFS, the patient was treated with irradiation for the myeloma, prednisolone, and melphalan, resulting in gradual improvements of the clinical and laboratory findings.

Four months after the diagnosis, he suddenly developed chest pain at rest. Electrocardiogram (ECG) disclosed ST elevation in I, aVL, and V3,6 leads. The chest pain was severe and nonresponsive to nitroglycerin. Morphine hydrochloride was intravenously administered, resulting in relief of the chest pain with normalization of the ECG. There was no elevation of serum creatine kinase. To elucidate a pathomechanism underlying the angina attack, coronary arteriography (CAG) was performed. There was no significant organic stenosis of the coronary artery. However, intracoronary injection of acetylcholine induced coronary vasospasm with chest pain and the same ECG changes as found in the angina attack. Nifedipine, iso-sorbide dinitrate, and aspirin were added to the medication. Thereafter, no angina attack has developed with no exacerbation of the CFS.

Association of angina pectoris with the CFS has not as yet been described in the literature. The CAG studies with intracoronary infusion of acetylcholine indicated that the angina was caused by coronary vasospasm [3], although the thrombocytosis, which was probably induced by increased serum IL-6 [4], was a risk factor for angina. Supersensitivity of the coronary artery to spasm induced by acetylcholine may suggest the presence of a functional disturbance of endothelial cells of the coronary artery [3]. Anasarca, one of the major manifestations of the CFS, indicates the presence of vascular permeability changes which may be caused by endothelial dysfunction. A recent study of the CFS indicated a frequent increase of serum IL-6 levels as found in our case, and suggested the abnormality of vascular endothelial cells [5]. Very recently, a similar case of the CFS with vasospastic angina was presented in a local meeting in Japan [6]. The pathomechanism underlying coronary vasospasm in the CFS remains unclear. However, such involvement of the coronary artery would be an important manifestation of the CFS.

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