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Foix-Chavany-Marie Opercular Syndrome

Foix-Chavany-Marie Operküler Sendromu

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Dear Editor,

Foix-Chavany-Marie opercular syndrome is a severe form of pseudobulbar palsy caused by bilateral anterior opercular lesions (1,2). It is characterized by a loss of voluntary control of facial, lingual, pharyngeal and masticatory muscles in the presence of preserved reflexive and automatic functions of the same muscles (3,4).

A 35-year-old male with diabetes had history of right leg deep vein thrombosis (DVT), which was treated since the age of 2 years. His sisters also had a history of DVT, and one of his sisters had had two abortions.

The condition started one month ago as an inability to swallow, along with drooling of the saliva, which started suddenly when the patient developed the inability to speak with right side heaviness, mainly in the upper limb. An improvement in the weakness occurred five days later but he still could not speak; the patient then suddenly developed drooling and even an inability to open his mouth and move his tongue. The patient denied having history of hypertension.

An examination of the central nervous system revealed a conscious patient with bilateral facial palsy, bulbar palsy, and normal power.

Laboratory tests showed thrombocytopenia, neutrophilia, erythrocytosis, hypochromic microcytic anemia, and elevated erythrocyte sedimentation rate.

Cerebrospinal fluid analysis showed elevated white blood cells, mostly consisting of lymphocytes (70%); protein was slightly elevated (47 mg/dL), glucose was normal (64 mg/dL), and the fluid was colorless. Cardiac echo showed normal left ventricular size, wall thickness and contractility, an ejection fraction of 66%, mild mitral regurgitation, and no pericardial effusion.

Serology showed positive anti-cardiolipin antibodies [immunoglobulin G (IgG)], anti-double strand DNA antibody, weakly positive anti-neutrophil cytoplasmic antibodies, and anti-phospholipid antibodies (IgG and IgM); anti-nuclear antibodies and anti-p-ANCA were normal. Total serum protein (76 g/L), albumin (45 g/L), and globulin (31 g/L) were normal; alpha-2 (3.4 g/L), beta globin (5.6 g/L) were low, and gamma protein was elevated (19 g/L).

The patient responded to anti-ischemic drugs. Magnetic resonance imaging of the brain with fluid-attenuated inversion recovery sequences showed bilateral, multiple ill-defined foci of abnormal signal intensity involving the parietal and insular regions, some showing cystic centers. There was no mass effect, and no recent hemorrhagic changes, as shown in Figure 1.

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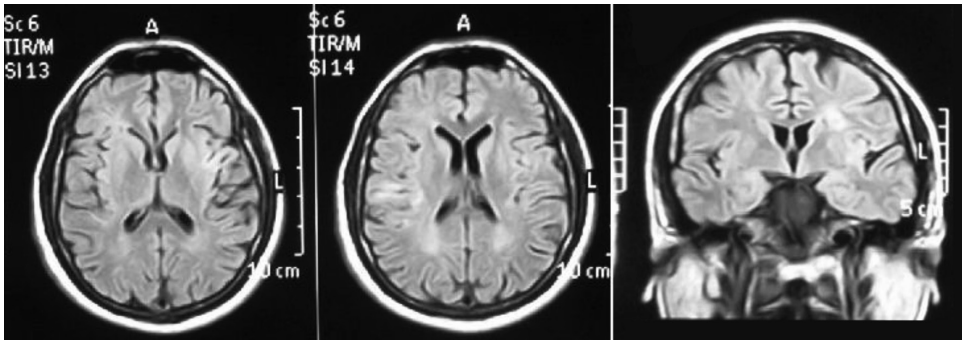

Figure 1. Fluid-attenuated inversion recovery image showing peri-insular hyperintensities (opercula)

Ethics

Informed Consent: Consent form was filled out by all participants.

Peer-review: Internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: A.S.H., F.I.G., Q.A.F., H.A.F., Concept: A.S.H., F.I.G., Design: H.A.F., A.S.H., F.I.G., Data Collection or Processing: A.S.H., F.I.G., H.A.F., Analysis or Interpretation: Q.A.F., A.S.H., F.I.G., Literature Search: H.A.F., A.S.H., Writing: A.S.H., F.I.G., Q.A.F., H.A.F.

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