Could Hemiplegia Vegetativa Alterna be a Cerebral Sign of Heart Valve Disease?

Hemiplegia Vegetativa Alterna, Kalp Kapak Hastalığının Serebral Bir İşareti Olabilir mi?

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Dear Editor;

Hemiplegia Vegetativa Alterna (HVA) is the clinical condition characterized with ipsilateral Horner's syndrome (HS) and contralateral hemi-hyperhidrosis and is mainly used to define the condition at stroke patients with posterior cerebral artery (PCA) or middle cerebral artery (MCA) occlusion (1,2). The cases with involvement of uncrossed excitatory and crossed inhibitory hypothalamo-spinal sympathetic pathways have been also defined as HVA by the researchers (1). However, as the lesions straggle or cover a large area, it was not possible to identify a certain anatomical localization, which would explain the clinical condition. Our case is a combination of the previously presented cases and includes both MCA and PCA occlusions.

A 39-year-old female patient was referred to the neurology intensive care unit (ICU) with loss of consciousness, hemiplegia on the right side of the body, and aphasia. She was somnolent in her neurologic examination. She had no responses to verbal stimuli and localize painful stimulus with her left side. It was impossible to have cooperation with the patient due to the mixed aphasia and her pupils were anisocoric (right>left), and pupillary light reflex can be taken. In the primary position, eyes and head were deviated to the left side. Patient had right central facial paralysis, and muscle strength of right upper and lower extremities were 1/5. Nevertheless, in her daily follow-ups, in addition to anisocoria of her left eye rima oculi was narrow compared to her right rima oculi. Autonomic disturbance was detected in the form of hypohidrosis in the left half of face, and hyperhidrosis in the right half of body; especially in the

right half of face and in the right arm reaching out to the elbow (Figure 1). In the brain CT (BCT), there was a large infarct in the left MCA and PCA area making a distinctive shift effect to the opposite hemisphere (Figure 2). In the cranial MRI taken 15 days later, it was observed that this shift effect had disappeared (Figure 3). Atrial fibrillation with high ventricle response was detected at the ECG. The findings at transthoracic echocardiography were as follows; moderate-intensity mitral stenosis, valve area of 1.1 cm², maximum gradient of the valve was 25 mmHg, mean gradient was 15 mmHg, systolic pulmonary artery pressure was 40 mmHg. In addition to the findings above, Wilkins valve score was 7, there wasn't any thrombosis in the left atrium and left atrial appendix.

Figure 1. Horner syndrome on the left side and on right half of the face partially recognized hyperhidrosis

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Figure 2. Large infarct area in left MCA and PCA with marked shift effect in CT

Figure 3. Shift effect associated with large infarct area disappeared in left MCA and PCA areas in cranial MRI (15 days later)

MCA occlusion, heart valve disease and severe neurological deficit with large infarct area are the similar points of our case with published Uca et al's case (2). Even though contra-lateral hyperhidrosis or central HS can exist in the same anatomical localization, their co-existence would be seen rarely (4,5,6,7). We believe that it would be difficult to localize the lesion in HVA cases, and the syndrome would manifest more often, especially in cases of large lesions and severe neurological deficit situations.

It is possible that HVA is not encountered in all cases with MCA, PCA occlusion, or in all cardio-embolic stroke situations. This clearly shows that ischemic syndromes of specific vessels are not even associated with occlusion area, but can be associated with a certain variations of an artery in the area including previous brain damage, collateral circulation and aberrations in Willis polygon.

We decided to have that title for our letter due to the two HVA syndrome patients with previous heart valve disease. The aim is not to be pretentious but to increase etiological inference and also increase interest in HVA syndrome.

In recent times there is a growing interest to alternate syndromes in cerebrovascular cases (7). Syndromes related to specific vessels do not always describe the occlusion's area or its nature. However, knowing about artery syndromes assists the researcher to localize the lesion and to decide whether the origin is vascular or not. The typical nature of HS but high rate of neglected hyperhidrosis could decrease recognition and the number of HVA case reports, and similarly more careful examinations for sweating dysfunction could extend the series. Thus, it could be possible to make more efficient comments regarding both the anatomical localization and etiological reasons in the future.

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