SPONTANEOUS INTERNAL CAROTID ARTERY DISSECTION IN DYSTROPHIC BULLOUS EPIDERMOLYSIS – ASSOCIATION OR DETERMINANT FACTOR?

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ABSTRACT

Pathogenesis of spontaneous carotid artery dissection (sCAD) is still a domain of research. Classical inherited connective tissue disease, primary or systemic arteriopathies and ultrastructural alterations of the arterial wall were reported in sCAD.We report a case of sCAD in a middle-aged woman with dystrophic epidermolysis bullosa, an inherited blistering disease due to a deficiency of type VII collagen at the basement membrane zone (BMZ).

Key words: spontaneous carotid artery dissection, dystrophic bullous epidermolysis

BACKGROUND

Spontaneous carotid artery dissection (sCAD) is an important cause of stroke in the young and middle-aged population(10-25%) **(**1).

A primary or systemic arteriopathy is frequently suspected in patients with spontaneous cerebral or cervical artery dissections (2).

Therefore, all patients with CAD should be examined for signs of connective tissue disorder as skin and joint hiperextensibility, abnormal scars, and retina abnormalities on funduscopy, as angioid streaks (3).

Not only clinically evident, but ultrastructural connective tissue abnormalities were found in sCAD (4).

Epidermolysis bullosa is a heterogenous group of mechano-bullous disorders characterized by defective attachement of the epidermis to the underlying dermis (5). The dystrophic forms of epidermolysis bullosa have been shown to result from distinct mutations in the gene encoding type VII collagen, the major component of the anchoring fibrils (6). We report a patient with such dissection accompanied by dystrophic bullous epidermolysis, a previously unreported association.

CASE REPORT

A 39-year-old woman presented a seizure while shopping in a supermarket and was rushed to the emergency room with altered level of counsciousness. She had had surgery, chemo and radiotherapy for a breast carcinoma 4 years ago and had been on Tamoxifen for 3,5 years. She had been diagnosed with dystrophic form of bullous epidermolysis since the age of new born. General examination on admission showed: BP=130/80 85 mm Hg, HR=80/min, normal body temperature, red discoloration and dystrophic changes of the skin on the back of the hands and feet and dystrophic changes of the nails. Neurological examination showed: altered level of counsciuousness (GCS=9, O_2,V_2, M_5), head and eyes deviation towards right, absent movements

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on the left hemibody with left hypotonia and absent tendon reflexes, left Babinski sign. Laboratory findings were unremarkable (WBC=5.25·10^3/µl, RBC=4·10^3/µl, Hgb=12.8 g/dl, HCT=36,8%, PLT=105·10^3/µl, serum glucose=118 mg/dl, creatinine=0.69 mg/dl, cholesterol=180 mg/dl, TG=140 mg/dl, INR=0.95, APTT=11.7 sec). An emergency CT scan was normal. ECG and chest ultrasound were normal. MRI of the neck and head showed a T2 and Flair hyperintense lesion , with diminished diffusion and increased ADC, in left basal ganglia (head of caudate, upper internal capsule, putamen, lateral globus pallidus) (Fig. 1). MRA studies showed: absent flow on the entire course of the

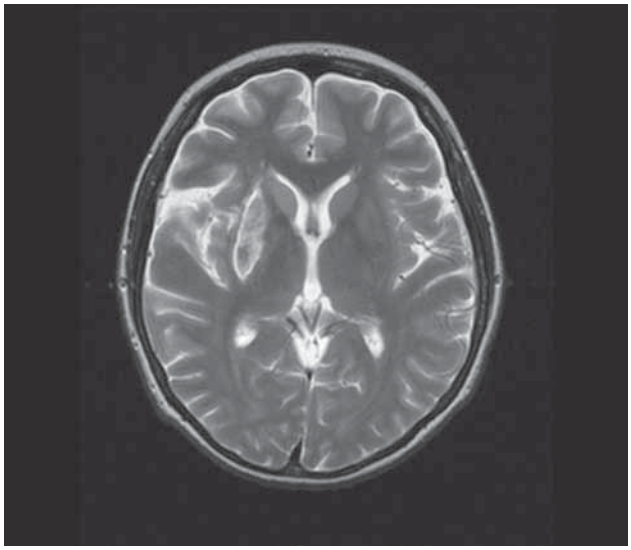

***FIGURE 1**. Cranial T2-MRI-axial sections – The occlusive dissection of ICA resulted in deep MCA teritory infarction (T2 hyperintense lesion in head of caudate, upper internal capsule, putamen, lateral globus pallidus).*

right internal carotid artery beginning from the bifurcation, on the cervical and intracranial segment and continuing on the medium cerebral artery and a hyperintense, crescent-shaped intramural hematoma (Fig. 2 and Fig. 3). The right ACA was supplied from the left ACA through anterior communicating artery (AComA). Dissecting occlusion of the right ICA was diagnosed. Anticoagulation with intravenous heparin was started, followed by oral acenocoumarol. Starting from the second day the patient reported a throbbing pain in the upper laterocervical region that persisted for the next 14 days. Active movements had a good recovery in the leg, while the upper arm regained only a trace of contraction in the proximal region. At 12 months the patient was able to walk alone, but flaccid paralysis persisted in the upper arm. MRA follow-up at 3, 9 and 12 months disclosed the same aspect of occlusion of ICA, while MCA was supplied by collateral flow from ACA and branches from ECA.

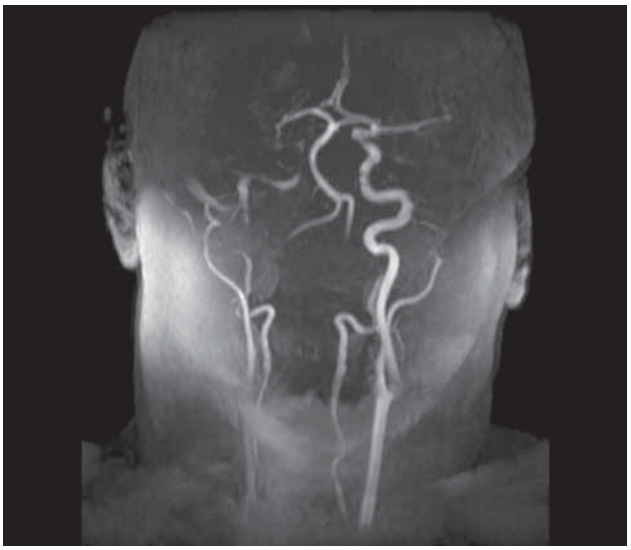

***FIGURE 2**. Cervico-cerebral MRA-3D TOF – Absence of vizualization of right ICA from its origin and during its entire cervical and intracranial course.*

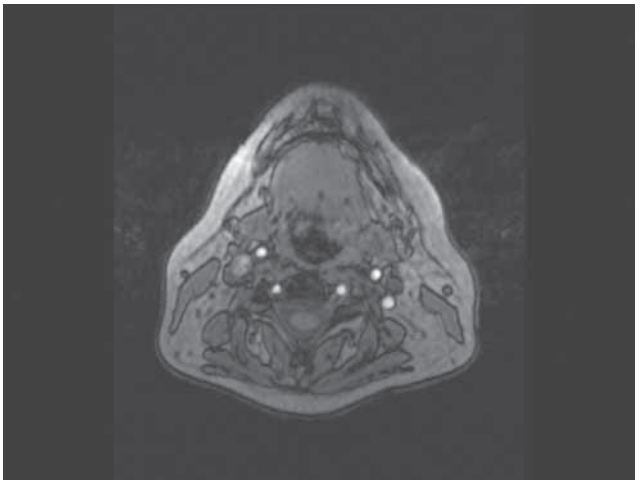

***FIGURE 3.** Cervical axial TOF – MRA demonstrates ICA dissection by a hyperintense, crescent-shaped intramural hematoma and an eccentric flow-void of the patent lumen.*

DISCUSSION

We highlight the association of two disorders based on collagen alteration. Bullous epidermolysis could be considered a marker for presence of ultrastructural alterations of the arterial wall.

Our patient presented seizure and stroke, without any warning signs, as it is reported to happen in one fifth of patients (7).

The affected carotid artery did not recanalize, as patients with initial high grade stenosis or occlusion are less likely to recanalize (8).

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