Case Report

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Thrombus in the Brachiocephalic Trunk in Patient with Bovine Aortic Arch Variation Mimics a Stroke or Multiple Sclerosis

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ABSTRACT

Embolism is mostly associated with arterial rather than venous injury, and with neuronal infarction rather than waste of myelin. Multiple sclerosis (MS) is an autoimmune inflammatory disease of the central nervous system (CNS) with difficult differential diagnosis. MS and stroke have general risk factors. The immune mechanisms of stroke are similar to neurodegenerative diseases and are adjunct to neuroinflammation. In autoimmune diseases the inflammation could increase risk for stroke or amplify the effect of conventional stroke risk factors. In the present case, the young patient was with clinical features susceptible for demyelinating disease. After performed MRI, cerebrospinal fluid examination, evoked potentials and screening for thrombophilia, multiple sclerosis was excluded. Ultrasound doppler examination and computed tomography (CT) angiography was performed because of the acute complaints of numbness and pain in right hand. It was found complete occlusion of the proximal half of the right ulnar artery, occlusion of the distal part of radial artery with its branches forming the superficial and deep palmar arches. The patient was diagnosed with parenchymal thrombus in brachiocephalic trunk, involving more than 80% of its lumen. When there is clinical presentation of relapses with neurological symptoms typical for MS in the differential diagnoses a vascular embolization should be considered and screening for thrombophilia could be useful. Multiple sclerosis could have difficult differential diagnose and a couple of new trials and experimental models show the significance of PAI-1 in etiology of neuroinflammation and neurodegeneration in multiple sclerosis, as well as a risk factor for thrombotic events in young adults.

Keywords: Stroke; Multiple Sclerosis; Thrombophilia

Abbreviations: MS: Multiple Sclerosis; CNS: Central Nervous System; CT: Computed Tomography; PAS: Plasminogen Activation System; MRI; Magnetic Resonance Imaging of the Head; GD: Gadolinium Contrast; CSF: Cerebrospinal Fluid

Introduction

The neurological features of brain vessel embolization are similar to those of MS. (Barnett M, et al. [1,2]). Diagnosing young adults with attacks of acute neurological deficit could be a challenge and testing for thrombophilia should be considered. Thrombophilia is defined as a predisposition for abnormal clot formation. It is a polygenic disorder with variable expressivity. A predisposition to thrombosis may be a result of genetic factors, acquired changes in the clotting mechanism, or more commonly, an interaction between them. Homozygous carrier or the combination of two or more heterozygous abnormal factors can lead to thrombotic disorders in young adults under 45 years. New experimental models show the significance of PAI-1 in etiology of neuroinflammation and neurodegeneration in MS as well as a risk factor for thrombotic events in young adults. (Lebas H, et al. [3,4]) Homozygous carriers of the 4G allele have the highest levels of PAI-1, and 5G homozygous carriers have the lowest levels. (Fay W, et al. [5-8]) In MS, disorder of the plasminogen activation system (PAS) and blood brain barrier are processes that might lead to an ab-

normal fibrin(ogen) extravasation into the parenchyma. Fibrin(ogen) deposits, usually broke down by the PAS, provoked an autoimmune response and demyelination. However, the PAS disruption is not well understood in this type of thrombophilia.

Materials and Methods

Somatic and neurological examination, laboratory tests, magnetic resonance imaging of the head (MRI) and neck with gadolinium contrast (Gd), examination of cerebrospinal fluid (CSF), doppler ultrasound examination, computed tomography (CT) angiography.

Clinical Case

We present a clinical case of a 42year old female admitted to the hospital with complaints of three relapses, couple of months apart. The first attack was six months earlier with numbness in right hand and lasted more than one week. The patient recovered spontaneously without treatment. During the second and third relapses the patient experienced weakness in left hand for 5 days. Multiple sclerosis was discussed as a leading diagnose. MRIs of the head and neck were performed with gadolinium contrast and in the right hemisphere several white matter lesions were found (Figures 1-4), which did not fulfil all revised 2017 McDonald MS criteria. Additionally, the patient was tested for oligoclonal bands in cerebrospinal fluid but no pathological changes were found. Infection disease was also excluded (testing for HIV, syphilis, Lyme disease, hepatitis B virus, hepatitis C virus). To exclude vascular etiology of the lesions genetic testing for thrombophilia was done. The patient was carrier of the 4G allele of PAI-mutation. The patient has conventional risk factors smoking and obesity. Atrial fibrillation was not detected during ECG monitoring and echocardiography did not show any pathological findings.

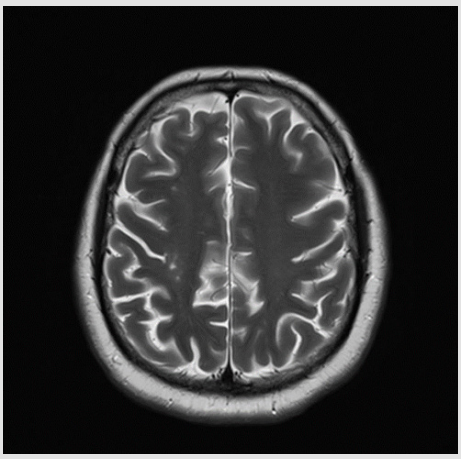
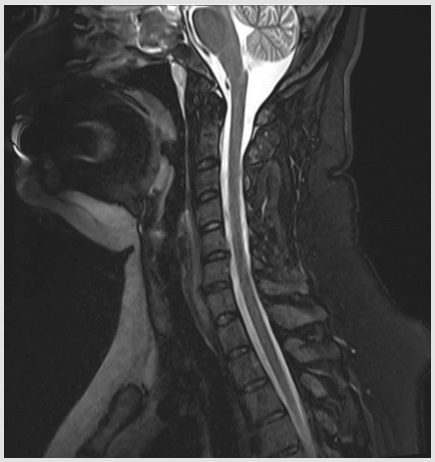


Figure 1.



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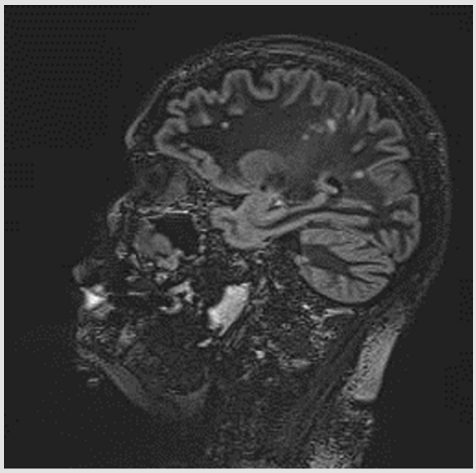

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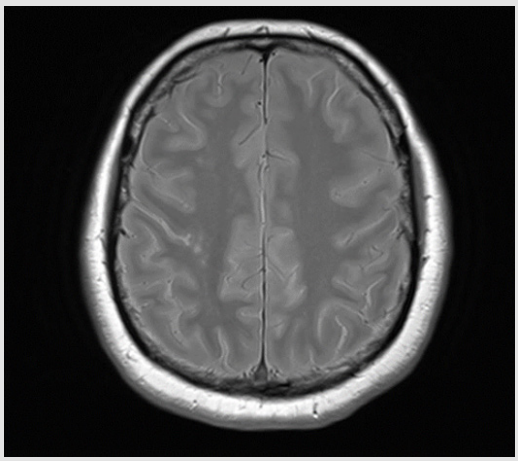

Figure 4.

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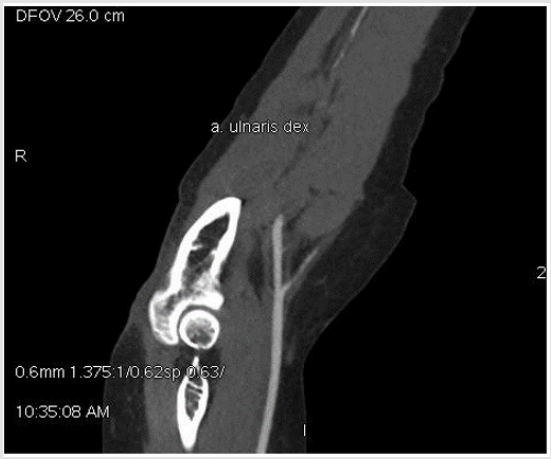
The patient experienced new symptoms during the hospitalization in the clinic. She complained numbness and pain in the right hand. A consultation with vascular surgeon was performed, doppler ultrasound examination and CT angiography of aortic arch and vessels for the right hand. It was found complete occlusion of the proximal half of the right ulnar artery, occlusion of the distal part of radial artery with its branches forming the superficial and deep palmar arches. The patient was diagnosed with parenchymal thrombus in brachiocephalic trunk and bovine aortic arch variation, involving more than 80% of its lumen (Figures 5-10). It was done urgent surgery for thrombectomy and reconstruction of brachiocephalic trunk. Thrombectomia of right ulnar and radial artery were performed with pericardial and right femoral artery patch plastic. The patient had no complications after surgery with complete reverse of the symptoms. Thrombus in the brachiocephalic trunk was discussed as source of embolization and cause for the lesions in right hemisphere. As risk factors for thrombotic event, we defined smoking, obesity, thrombophilia.



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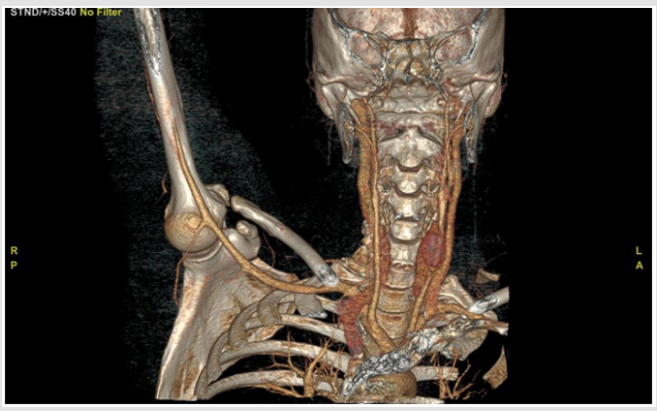

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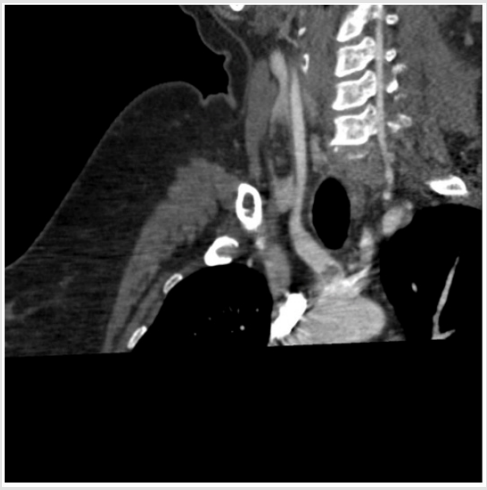

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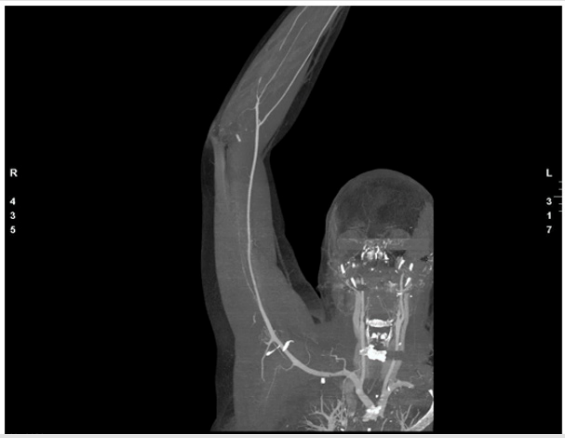

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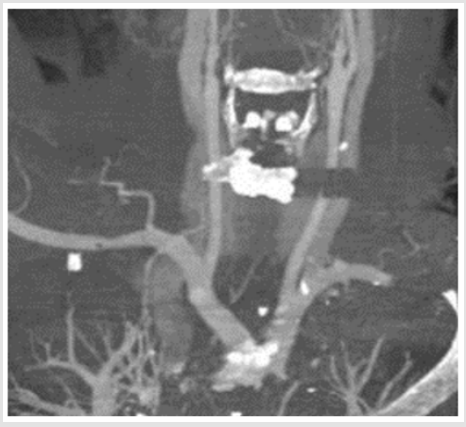

Figure 9.

Figure 10.

Discussion

Multiple sclerosis is a disease of the central nervous system that destroys myelin, oligodendrocytes, neurons and axons. The immune mechanisms of stroke are similar to neurodegenerative diseases and are adjunct to neuroinflammation. The inflammation in autoimmune diseases could increase risk for stroke or amplify the effect of conventional stroke risk factors. PAI mutation 4G/5G is defined as a risk factor for arterial thrombosis. Experimental models show the importance of PAI-1 (overexpressed by reactive astrocytes leading dysfibrinolysis in MS). (Lebas H, et al. [3]) Further studies should be performed to evaluate the role of PAI mutation in MS patients and also for vascular diseases. The thrombotic risk factors associated with MS such as obesity, smoking, endothelial dysfunction, platelet activation, thrombophilia are pro-thrombotic conditions. (D'Haeseleer M, et al. [4]). Distal embolization in the brain and ipsilateral upper extremity could be confusing, because of bilateral neurological signs. Differential diagnosis between multiple sclerosis and ischemic stroke in some of the cases is difficult. We believe that in patients with risk factors for vascular disease and suspicion for MS testing for thrombophilia should be performed.

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