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Seizure as Initial Manifestation of Aortic Dissection Type A

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Background: Seizure as the initial manifestation of aortic dissection is rare.

Case report: An 88-year-old female experienced a first generalized tonic clonic seizure, which was terminated with midazolam. Acute cerebral magnetic resonance imaging and angiography were non-informative. After awaking she complained about cramping pain in the right upper extremity, which was accompanied by involuntary flexion movements of the right upper extremity. Blood pressure was initially normal. Blood gases revealed metabolic acidosis and blood chemical investigations a markedly increased D-dimer. Consecutively blood pressure declined and transthoracic echocardiography showed pericardial effusion. A computed tomography scan of the thorax revealed an aortic dissection type A. The patient died 16 hours after admission after cardiothoracic surgeons had refused surgical treatment.

Conclusion: This case shows that a generalized tonic-clonic seizure may be the initial manifestation of an aortic dissection type A in the absence of thoracic chest pain and that brachyalgia may not develop earlier than with progression of the dissection. [West J Emerg Med. 2010; 11(5):510-511.]

INTRODUCTION
Seizure as the sole initial manifestation of aortic dissection in the absence of chest pain has been reported only once. Here we report a second patient in whom aortic dissection type A initially manifested exclusively as generalized seizure without chest pain.

CASE REPORT
The patient is an 88-year-old female with a previous history of arterial hypertension, obstructive lung disease and bilateral total hip endoprosthesis. She was not drinking alcohol and was living well on her own in an asylum. While riding a tram on her way to a festival, she experienced a sudden onset generalized seizure with loss of consciousness and cloni of the upper and lower extremity with right-sided predominance. A dose of 15mg midazolam from the emergency physician stopped the seizure and when she arrived at the emergency department she was comatose and quadriplegic and the brain stem reflexes were absent, except for the pupils, which were widened and reacted slowly to light bilaterally. Deep tendon reflexes were generally absent and there was general muscle hypotonia.

Blood pressure was 130/80mmHg. Blood chemical investigations revealed moderate renal insufficiency, elevated liver function parameters, leucocytosis of 12.3/nl (n = 4.0-9.0/ nl), slight anemia and a D-dimer of 20.0microg/ml (n <0.5microg/ml). Blood gas analysis revealed metabolic acidosis with a pH of 7.36, a base excess of -8.5, and a lactate of 2.4mmol/l. Electrocardiogram (ECG) showed sinus rhythm. Magnetic resonance image (MRI) of the cerebrum 45 minutes after the seizure did not show ischemia or intracerebral bleeding. On MR angiography no major abnormality of the vasculature was detected. Her state of consciousness improved from coma to somnolence, and she started to move her extremities.

After awakening she complained about recurrent cramping pain in her right shoulder and right upper arm, which was accompanied by automatic flexion of the right upper limb. Aortic dissection was suspected and a computed tomography (CT) of the thorax initiated. Before carrying
it out, however, she developed low blood pressure to non-measurable values and was transferred to the intensive care unit for suspected cardiac shock where hydroxi-ethyl starch and vasopressin helped to increased blood pressure. However, the right pupil widened and became non-reactive to light, and she developed left-sided hemiplegia. Echocardiography showed normal systolic function, a small right ventricle and a pericardial effusion with an embedded clot. Subsequently, a CT scan of the thorax and the aorta showed aortic dissection type A extending into the left descending aorta, the brachiocephalic trunk with an occlusion of the right common carotid artery. For right-sided pain she received sufentanil with success. Carotid ultrasound eight hours after onset confirmed the complete occlusion of the right carotid artery. After two cardio-thoracic departments had refused surgical intervention the patient died without regaining consciousness, 16 hours after admission.

**DISCUSSION**

The patient is interesting for the epileptic seizure without chest pain as the initial manifestation of an aortic dissection type A. Such a scenario has been previously reported in a single 46-year-old patient, who presented with right hemiconvulsive movements due to ischemic right middle cerebral artery stroke.\(^1\) The only risk factor for aortic dissection in the present patient was arterial hypertension. Whether steroids in the broncholytic spray favored the development of the aortic dissection, as has been previously reported in a patient with systemic lupus erythematosus, remains speculative.\(^2\) Disorders predisposing for aortic dissection, such as X-linked heterotopia or Turner syndrome were excluded.\(^3,4\)

Why seizure was the initial manifestation remains elusive. However, one could speculate that the dissection initially resulted in general cerebral hypoperfusion and thus hypoxia or stenosis of the common carotid artery, as well cerebral ischemia, which did not show up on diffusion-weighted imaging. The B1000 sequence might have been negative because the MRI was carried out too early or because ischemia was initially not intensive enough. Neither her son nor her friend reported previous seizures or syncopes. While it is conceivable that the seizure was not the consequence but the cause of the dissection, arguments against such a scenario are that she did not have a history of epilepsy (her history was negative for a cerebral lesion) nor have seizures been reported as triggers of aortic dissection. One could also speculate that the initial event was a rhythm abnormality with cerebral embolism inducing the seizure but spontaneous resolution of the clot. Worsening after the MRI could be attributed to progression of the dissection resulting in complete occlusion of the right carotid artery leading to a non-reactive right pupil and left-sided hemiplegia. Possibly, hemiplegia already existed at the initial presentation but was masked by the sedation with midazolam. However, because the pupils were reactive, though delayed, at the initial investigation, progression of the dissection during or after the cerebral MRI is more likely. Neurological manifestations are not infrequent at onset of an aortic dissection, but in the majority of the cases they are accompanied by chest pain.\(^5\) Altogether about one third of the patients with aortic dissection and chest pain initially present with neurological manifestations.\(^5\) Seizures occur in 3% of these patients.

This case shows that a generalized tonic-clonic seizure may be the initial manifestation of an aortic dissection type A in the absence of thoracic chest pain and that brachyalgia may not develop earlier than with progression of the dissection.

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