CASE REPORT

A rare cause of occlusion of the brachiocephalic trunk, resulting in fatal stroke: arterial thoracic outlet syndrome

J Heidt1, L de Beer2, GC Admiraal3

1 Department of Intensive Care, Leiden University Medical Center, Leiden, The Netherlands
2 Department of Radiology, Medical Centre Haaglanden, The Hague, The Netherlands
3 Department of Intensive Care, Medical Centre Haaglanden, The Hague, The Netherlands

Abstract - A 48-year old woman was admitted to the intensive care unit with cardiopulmonary failure secondary to a stroke. Computed tomographic angiography showed a complete occlusion of the brachiocephalic trunk (innominate artery). We found bilateral cervical ribs, with anterior dislocation of the subclavian arteries, causing narrowing of the vessel lumen and distal aneurysmal dilatation due to compression by the anterior scalene muscles. The patient died the following day, despite mechanical ventilation and haemodynamic support. We concluded that she had suffered from arterial thoracic outlet syndrome due to bilateral cervical ribs. We presume that the compression caused stenosis of the subclavian arteries and ultimately resulted in a fatal stroke due to the rare complication of retrograde thromboembolic occlusion of the brachiocephalic trunk.

Keywords - Occlusion, brachiocephalic trunk, innominate artery, stroke, arterial thoracic outlet syndrome

Introduction
Occlusion of the brachiocephalic trunk (innominate artery) is rare and seldom a cause of cerebral infarction. In this case report we present a rare cause of brachiocephalic trunk occlusion, which resulted in fatal stroke.

Case
A 48-year old woman was admitted to our emergency department with a respiratory arrest. She was of Hindustan-Surinamese descent, had no significant medical history and used only oral contraceptives. She was a non-smoker and had a normal Body Mass Index. No cardiovascular disease or thrombosis had been reported in her family. She had been having headaches for the last two years. She worked in a childcare facility and had recently been complaining of tired and heavy arms after work.

On the day in question she woke up with a fluctuating numb feeling and white/blue discoloration of the right hand. These symptoms persisted and an ambulance was called, but the paramedics found no abnormalities. During the afternoon the symptoms worsened and she visited an emergency department. Again no abnormalities were found. The report states normal pulsations and colour of the right arm. She was discharged with the diagnosis carpal tunnel syndrome. That evening she complained about severe pain and discoloration of the right hand, and developed nausea, vomiting and loss of sight. She suddenly collapsed and her family tried to provide basic life support.

When the paramedics arrived she had a slow pulseless electrical activity and was in respiratory arrest. After short cardiac resuscitation she spontaneously regained adequate heart rhythm and output.

On arrival at the emergency department the patient had a blood pressure of 110/80 mmHg but no spontaneous respiration. The Glasgow Coma Score was three; her pupils were dilated and non-reactive to light. The patient’s right hand had a dark blue discolouration. Pulsations of the right brachial, radial and ulnar artery were diminished and fluctuating. Doppler ultrasonography showed diminished and fluctuating blood flow. Laboratory results are shown in Table 1. The electrocardiogram showed a sinus rhythm, an incomplete right bundle branch block and no signs of myocardial ischaemia or left ventricular hypertrophy.

We suspected a massive pulmonary thromboembolism or aortic dissection and performed a computed tomographic (CT) angiography. There was neither a pulmonary thromboembolism nor an aortic dissection, but surprisingly we found a complete occlusion of the brachiocephalic trunk (Figure 1). The chest X-ray revealed bilateral cervical ribs (Figure 2). CT angiography showed bilateral cervical ribs with anterior dislocation of the left subclavian artery, causing narrowing of the vessel lumen and distal aneurysmal dilatation due to compression by the left anterior scalene muscle (Figure 3). Because of the lack of intravascular contrast in the right subclavian artery due to the occlusion, the anatomical vascular situation of the right side was less assessable, but seemed to be comparable to the left side. No hypertrophy of the anterior scalene muscles was observed. A CT scan of the cerebrum did not show abnormalities at that point.

The patient was admitted to the intensive care unit. Because of the absence of spontaneous breathing we suspected brainstem
ischaemia with poor prognosis. We therefore renounced surgical and thrombolytic therapy and started treatment with heparin and carbasalate calcium. She developed pulmonary oedema and progressive cardiopulmonary failure, despite inotropic support and mechanical ventilation. Echocardiography ruled out a cardiac source of embolus, and showed impaired left ventricular function without wall motion abnormalities. We suspected heart failure induced by cerebral infarction.

The following morning the brainstem reflexes diminished. A CT scan of the cerebrum showed extensive ischaemic infarction on the right side with severe cerebral oedema (suspect for complete medial and posterior infarction) and compression of the brainstem. The patient died in the afternoon, 24 hours after she had collapsed. The family did not give permission for an autopsy.

**Discussion**

Occlusion of the brachiocephalic trunk is rare. Considerations on differential diagnosis are atherosclerosis [1-4], antiphospholipid syndrome [5,6], dissection [7,8], anatomical variants like a common brachiocephalic trunk [9], and Takayasu’s disease [10-13]. While determining the cause of the occlusion we ruled out these possibilities. Atherosclerosis, dissection, vascular anatomical variants or signs of vasculitis were not observed on the CT scan. Laboratory results (Table 1) showed no signs of systemic inflammation, vasculitis or antiphospholipid syndrome. Moreover, we had strong radiodiagnostic evidence for arterial thoracic outlet syndrome (ATOS). The elevated D-dimer also suggests the presence of a thromboembolic event.

The definition of TOS is “upper extremity symptoms due to compression of the neurovascular bundle by various structures

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**Table 1. Laboratory results**

<table>
<thead>
<tr>
<th>TEST</th>
<th>RESULT</th>
<th>REFERENCE VALUE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Haemoglobin</td>
<td>8.6 mmol/l</td>
<td>7.0 – 9.5 mmol/l</td>
</tr>
<tr>
<td>Erythrocyte sedimentation rate</td>
<td>13 mm in the first hour</td>
<td>2 – 20 mm in the first hour</td>
</tr>
<tr>
<td>Leucocyte count</td>
<td>13.5 x 10⁹/l</td>
<td>4.0 – 10.0 x 10⁹/l</td>
</tr>
<tr>
<td>Platelet count</td>
<td>259 x 10⁹/l</td>
<td>150 – 400 x 10⁹/l</td>
</tr>
<tr>
<td>Sodium</td>
<td>140 mmol/l</td>
<td>135 – 145 mmol/l</td>
</tr>
<tr>
<td>Potassium</td>
<td>3.2 mmol/l</td>
<td>3.2 – 4.7 mmol/l</td>
</tr>
<tr>
<td>Creatinin</td>
<td>86 μmol/l</td>
<td>50 – 95 μmol/l</td>
</tr>
<tr>
<td>ASAT</td>
<td>126 U/l</td>
<td>0 – 31 U/l</td>
</tr>
<tr>
<td>ALAT</td>
<td>109 U/l</td>
<td>0 – 34 U/l</td>
</tr>
<tr>
<td>Alkaline phosphatase</td>
<td>67 U/l</td>
<td>40 – 120 U/l</td>
</tr>
<tr>
<td>γ-GT</td>
<td>13 U/l</td>
<td>0 – 38 U/l</td>
</tr>
<tr>
<td>Bilirubin</td>
<td>8 μmol/l</td>
<td>5 – 19 μmol/l</td>
</tr>
<tr>
<td>C-reactive protein</td>
<td>6 mg/l</td>
<td>0 – 8 mg/l</td>
</tr>
<tr>
<td>D-dimer</td>
<td>&gt; 10.00 mg/l</td>
<td>0 – 0.50 mg/l</td>
</tr>
<tr>
<td>Anti nuclear factor</td>
<td>negative</td>
<td></td>
</tr>
<tr>
<td>Anti neutrophil cytoplasmic antibodies (anti-MPO and anti-PR3)</td>
<td>negative</td>
<td></td>
</tr>
<tr>
<td>Anti cardiolipin antibodies</td>
<td>negative</td>
<td></td>
</tr>
<tr>
<td>Lupus anticoagulans</td>
<td>could not be tested due heparin treatment</td>
<td></td>
</tr>
</tbody>
</table>
in the area just above the first rib and behind the clavicle" [14,15]. This neurovascular bundle contains the brachial plexus, subclavian vein and subclavian artery. Although neurogenic and vascular forms may coexist, these three structures can be individually compressed. Hence, a classification in three types was created: neurogenic TOS (NTOS), venous TOS (VTOS) and arterial TOS (ATOS) [14,15]. The incidence of TOS is not well known due to a variety of diseases that mimic this condition TOS [16], and reported incidences range from 3 to 80 cases per 1,000 population [17,18]. The prevalence of symptomatic TOS has been estimated to be 10 per 100,000 people [19].

The presence of cervical ribs predisposes to the development of TOS, especially NTOS and ATOS. They occur in less than 1% of the general population [14,16-18,20]. Cervical ribs and TOS are more commonly observed in women, with the onset of symptoms usually between 20 and 50 years of age. Cervical ribs occur bilaterally in more than 50%, and are reported in approximately 10% of patients with TOS [16-18,20]. About 30% of cervical ribs are complete ribs fused to the first rib by a true joint or a fibrous attachment. The remaining 70% are incomplete cervical ribs with no direct attachment to the first rib [15]. Only complete ribs have been noted to produce ATOS [20]. Most cervical ribs are asymptomatic, and when symptoms do develop, they are usually neurogenic [15].

ATOS comprises less than 1% of all TOS cases. Symptoms usually develop spontaneously, unrelated to trauma or extensive arm activity. As in our case, ATOS is almost always associated with a cervical or anomalous first rib [14,15,21]. Prolonged compression of the subclavian artery results in fibrotic and inflammatory changes of the arterial wall, causing platelets aggregation resulting in the formation of mural thrombus and macroembolism [22]. This in turn will result in partial or complete thrombosis, poststenotic dilatation, poststenotic aneurysm and digital gangrene [18,23]. Thromboembolic complications are significantly more common in arteries with aneurysmal dilatation [24].

The pathogenesis in our case is anterior dislocation of both the subclavian arteries due to bilateral cervical ribs. This displacement (as shown in Figure 4) resulted in compression by

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**Figure 1.** Coronal CT MIP image: occlusion of the brachiocephalic trunk just above the origo out of the aortic arch, indicated by the arrow (SVC: superior vena cava, LCCA: left common carotid artery, LSA: left subclavian artery).

**Figure 2.** Frontal chest radiograph: bilateral complete cervical ribs, fused with the first ribs by a joint (arrows).
the anterior scalene muscles, and has previously been reported in the literature [25,26]. We presume that in our case, chronic inflammation of the arterial wall resulted in bilateral stenosis and poststenotic dilatation of the subclavian arteries. On the right side a thromboembolism formed, compromising the arterial blood flow of the right arm, resulting in the fluctuating symptoms of claudication, pallor, colour changes, coldness, paresthesia and pain during the day. In the evening ongoing retrograde flow and thrombus propagation resulted in total occlusion of the brachiocephalic trunk, carotid artery and vertebral artery. We presume that the acute occlusion, combined with cerebral embolisms, led to the fatal stroke.

Recognizing ATOS in patients is difficult because they are usually asymptomatic until embolization of the stenoses and aneurysm occurs. The symptoms are related to ischaemia in the affected limb: pain, coldness, colour change or even gangrene [14,15]. An absent radial pulse is common in ATOS on physical examination. Decreased blood pressure greater than 20 mmHg in the affected arm compared with the contralateral arm is an indicator of arterial involvement [18]. There are no sensitive and specific provocative tests [14,15,17,18]. Sometimes a pulsatile mass or a vascular bruit can be found in the supraclavicular region. Diagnostic examinations are X-ray to detect cervical ribs, Doppler ultrasonography to detect decreased arterial flow and CT angiography to detect vascular compression, stenosis and aneurysms [14,15,17,18]. Our case demonstrates the typical ‘non-specific’ and difficult presentation of ATOS. The paramedics and physicians initially missed the diagnosis and failed to recognize the severity of this condition.

In case of proven ATOS, surgical treatment should be considered. Surgical decompression involves cervical rib excision, first rib excision and resection of compressing muscles [15,16,20,27,28]. Additional arterial reconstruction should be performed in the presence of poststenotic dilatation or mural thrombus [15,22]. Vascular reconstruction after decompression presents good results on both short and long term follow-up [26,29,30]. Endovascular treatment with stents has also been described [31], however long term studies showed high chance of restenosis [16]. In our patient we renounced surgical, endovascular and thrombolytic treatment because we suspected a very poor prognosis due to of the poor neurological status after the resuscitation.

To our knowledge retrograde thrombus propagation into the brachiocephalic trunk, resulting in stroke, is very rare and has been described only once [32]. That particular case demonstrated that in the case of less severe neurological damage, and a better prognosis than our patient, surgical treatment can be an appropriate treatment.
Conclusion
After combining the clinical findings, the laboratory results, the chest X-ray and CT-imaging we concluded that our patient had suffered from arterial thoracic outlet syndrome due to bilateral cervical ribs, with complete brachiocephalic trunk occlusion and fatal stroke as a rare complication. Despite its rarity, a sufficient diagnostic approach is crucial when a patient presents with symptoms of arterial occlusion of the upper limbs. A missed diagnosis can result in a severe and potentially fatal medical emergency, as we presented in this case. Therefore, ER and ICU physicians should have sufficient knowledge of ATOS. Early recognition of ischaemic symptoms is essential. Adequate diagnostic measures like chest X-ray to recognize cervical ribs can be very helpful. Timely treatment such as surgery or endovascular intervention is essential and is associated with positive prognosis and outcome.

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References