CASE REPORT

A complicated course after endovascular repair of an arteria lusoria aneurysm

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Abstract
Arteria lusoria is one of the most common abnormalities of the aortic arch. Usually it is asymptomatic and first noticed as a coincidental finding on imaging studies performed for a different reason. Sometimes, because of symptoms or in rare occasions aneurysmal dilatation, treatment is necessary. Here, we present a case of endovascular repair of an arteria lusoria aneurysm which was complicated by postoperative superior vena cava syndrome and ischaemic stroke.

Introduction
Arteria lusoria or aberrant subclavian artery is one of the most common abnormalities of the aortic arch. Unlike in the normal anatomical situation (figure 1A), the right subclavian artery does not originate from the brachiocephalic artery, but directly from the aorta (figure 1B). Patients are often asymptomatic. However, because of the possible course of the aberrant artery between or along the oesophagus and trachea, symptoms such as dyspnoea or impairment of passage in the oesophagus may occur. There is also a very small risk of aneurysmal dilatation with a chance of rupture. This case report will describe a 77-year-old patient with a complicated disease course after endovascular repair of an arteria lusoria aneurysm.

Case report
A 77-year-old male was referred to our hospital. He had a medical history of prostate cancer, type 2 diabetes mellitus, arterial hypertension, peripheral vascular disease, mild chronic kidney disease and two episodes of deep venous thrombosis. A single-photon emission computed tomography (SPECT) scan was performed for the evaluation of possible metastatic disease.
No metastases were found. However, a large thoracic aneurysm (9 x 14 cm) of a not previously discovered arteria lusoria was seen, causing oesophageal and tracheal compression (figure 2). The patient experienced only minor symptoms (dyspnoea), but because of the size of the aneurysm and the risk of rupture, treatment was needed. Additional diagnostic examinations were performed because coronary artery calcification was seen on the CT scan; the echocardiogram showed normal biventricular function without any valve abnormalities. Coronary angiography showed a significant left main coronary stenosis of 90%. After multidisciplinary consultation, the decision was made to first perform coronary artery bypass grafting (CABG), followed by endovascular repair of the aneurysm at a later stage. The CABG and initial postoperative period were without complications and three days later thoracic endovascular aneurysm repair (TEVAR) was performed. Directly after the procedure, severe progressive oedema of the face, neck and both arms appeared; these are characteristic features of superior vena cava syndrome, which was confirmed by a postoperative CT scan. The underlying cause was not precisely elucidated but it was either caused by mechanical obstruction due to haematoma or to thrombosis in the aneurysm. Treatment with intravenous heparin was started, but given the complexity and potential risks of a second intervention, the decision was made to watch and try to relieve symptoms using diuretics. It was decided not to extubate the patient as long as the upper body swelling was present because of expected upper airway obstruction. Therefore, in order to minimise the risk of dislocation of the endotracheal tube, and the potentially very difficult reintubation in the case of accidental removal of the tube, the patient was sedated for seven days. However, five days after the TEVAR, a neurological exam during a wake-up call (the temporary cessation of sedative medication to be able to monitor the patient’s neurological state) revealed a slight decrease in consciousness and difficulty for the patient to obey commands. In addition, right lower limb paresis and right hemianopsia were seen. A CT scan showed cerebral infarction in multiple vascular territories (frontal, occipital and cerebellar), suggestive of an embolic origin. Since heparin therapy had already been started, there were no clinical consequences. Continuous infusion of diuretics caused the oedema to decrease which made it possible to extubate the patient after 13 days of mechanical ventilation. A few months after discharge, the patient still experienced a slightly paretic right lower limb, but no other neurological deficits. He can walk independently with a walker and is still rehabilitating.

**Discussion**

In the normal anatomical situation, the right subclavian artery and the right carotid artery arise from the brachiocephalic artery. The left carotid artery and left subclavian artery each branch directly from the aortic arch. In the case of arteria lusoria, the right subclavian artery branches directly from the aortic arch, as a fourth branch next to the left subclavian artery. The arteria lusoria then continues behind the trachea and/or oesophagus to the right side of the body and the right arm (figure 1B). This anatomical variation was first described by Hunauld in 1735.[1] It is caused by abnormal development in the embryonic period and it frequently arises from a diverticulum at the proximal descending aorta, which was first described by Kommerell.[2] An aneurysmal diverticulum of the aorta at the origin of an aberrant subclavian artery is called Kommerell’s diverticulum. Patients
are often asymptomatic (in more than 90% of the cases) but compression of the trachea or oesophagus can cause symptoms such as dysphagia (71%), dyspnoea (19%) and coughing (8%).[^3][^4] Impaired passage of food due to compression in these circumstances is called dysphagia lusoria, which was coined by Bayford 50 years after the first description by Hunauld.[^5] It is the most common abnormality of the aortic arch, with an incidence of around 1 to 2%.[^4] It is associated with trisomy 21, with a higher incidence of between 19 and 35%, but is also seen in many other congenital syndromes.[^7] The diagnosis is easily made with a CTA scan or angiography. There are various surgical procedures to correct an arteria lusoria, using sternotomy or lateral thoracotomy with and without the use of cardiopulmonary bypass, as well as endovascular procedures or a combined approach.[^8]

**Superior vena cava syndrome**

Superior vena cava syndrome is the result of a partial or complete obstruction of the superior vena cava, causing venous congestion in the upper body, with characteristic symptoms such as oedema of the face and arms, shortness of breath or even severe respiratory distress due to narrowing of the larynx and pharynx and other symptoms such as headaches, dizziness and ultimately cerebral oedema with possible cerebral herniation. The superior vena cava syndrome is most often seen in patients with tumours in the mediastinum, for example in lung cancer (in approximately 10% of patients with small cell lung carcinoma) or malignant lymphomas (in approximately 3%). Other causes can be, for example, thrombosis in the vessel or vessel wall abnormalities.[[^8][^11]]

When there is a gradual onset, collateral veins have time to develop (often through the azygos and internal thoracic veins), making the symptoms less pronounced. In the case of a rapid onset due to, for example, thrombosis the symptoms are more severe.

Depending on the cause, the treatment may consist of anticoagulation or thrombolysis in case of thrombosis and chemotherapy or radiotherapy for malignant causes. Acute life-threatening symptoms such as upper airway obstruction due to laryngeal oedema should be treated immediately. Cerebral oedema is rare, but possible and should also be treated.
Rhabdomyolysis due to steroids and muscle blocking agents
immediately. Corticosteroids are sometimes used to reduce laryngeal oedema, although the beneficial effect has never been proven.\textsuperscript{[9]} Thrombosis inside the vena cava should be treated with therapeutic anticoagulation. Endovascular stenting is an option if rapid improvement is desired and yields a high success rate, especially in superior vena cava syndrome caused by malignancy. In 80 to 95% of the patients a stent relieves the symptoms.\textsuperscript{[12]}

Considerations
Given the severe oedema of the head and neck with compression of the trachea, it was vital to keep the airway secured with an endotracheal tube until the swelling had decreased. To be sure that the patient would not accidentally extubate himself, we chose to sedate him for a number of days until the oedema of the upper body had decreased. The downside is that any neurological abnormalities are not observed until cessation of sedatives. Given that stroke is one of the most important complications of TEVAR, with incidences ranging from 2 to 8%, it is of great importance to do an early and thorough neurological examination after TEVAR. Stroke is mostly diagnosed within 24 hours after surgery. The mechanism of ischaemic stroke is manipulation of intra-aortal atherosclerotic plaques which can lead to direct embolism. It is thought that the procedure also makes the plaques prone to delayed embolism in the postoperative period.\textsuperscript{[13]}

Conclusion
Arteria lusoria is a relatively common abnormality of the aortic arch. The diagnosis can be easily made with a CTA scan and it may not be discovered until the patient undergoes imaging for a different reason (e.g. undergoes coronary angiography to investigate chest pain). In very few cases symptoms or aneurysmal dilatation are found, both of which could warrant treatment. In this case, there was an aneurysm that required repair due to the risk of rupture. The course of the disease was complicated by superior vena cava syndrome caused by compression of the aneurysm followed by embolic stroke after endovascular repair.

Disclosures
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References