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THE QUALITY OF LIFE OF A PATIENT WITH AN IATROGENIC BRAIN STROKE AFTER ANGIOSURGICAL TREATMENT FOLLOWING THE IMPLANT OF A STENTGRAFT INTO A DISSECTED THORACIC AORTA WITH A CONGENITAL ANATOMICAL ANOMALY OF BRANCHING OF THE RIGHT SUBCLAVIAN ARTERY (ARTERIA LUSORIA)

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SUMMARY

Background:

The aim of this article was the evaluation of the quality of life of patient during long term follow up after the dissection of the descending thoracic aorta and angiosurgical related complications.

Case study:

A 31-year-old hypertensive female patient was admitted with a chest and epigastrium pain, with a 20 months history of asymptomatic DeBekey type III thoracic aortic dissection following CT scanning. A badly conducted Angio-Tomographic examination (Angio-CT) did not reveal the anatomical pathology of branching within the right aorta. The implantation of a stentgraft resulted in the closing of both subclavian arteries, and a iatrogenic brain stroke resulted as a consequence. Successful revascularisation of the right subclavian artery resulted in the disappearance of the neurological symptoms.

Conclusions:

Badly conducted and incorrectly interpreted diagnostic examinations may result in improper treatment resulting in complications as happened in the case of the patient described by us. A close observation of each patient after the performed surgery is, therefore, of the highest importance in order to be able to perform any necessary repair procedures. A long lasting illness of dissection of the thoracic aorta resulted in long, deep rooted stress, connected with a high degree of anxiety which led to a further decrease in the patient's quality of life. Successful secondary repair surgery reduced the stress symptoms and increased her quality of life.

Key words: vascular diseases, anxiety, pain, consciousness

INTRODUCTION

There is a lack of reports on the quality of life for patients with dissection of the thoracic aorta with simultaneous congenital anatomical anomaly of an aberrant right subclavian artery (Arteria Lusoria, AL) (Fig. 1). There are also no works describing the quality of life connected with the postsurgical complications. It should be stressed that any faulty attempt to treat such disorders may decrease the brain blood supply, leading therein to a brain stroke.

It is worth recalling that the aberrant right subclavian artery (Arteria Lusoria, AL) arising from the aortic arch or proximal descending aorta was first described by Hunauld (see: Hunauld 1735). The AL is the most common congenital anomaly of the aortic arch that occurs in about 0.2-2-5% of individuals (Guzman & Eagleton 2012; Huang et al 2009; Myers et al 2010; Vucemilo et al 2014), but is still a rare vascular anomaly. In such a case four vessels arise sequentially from the aortic arch: the Right Common Carotid Artery (RCCA), the Left Common Carotid Artery (LCCA), the Left Subclavian Artery (LSA) and the aberrant Right Subclavian Artery (RSA) (Guzman ED, Eagleton 2012). The AL has to cross to the right and upwards in the posterior mediastinum (Guzman ED, Eagleton 2012). The retropharyngeal course (80-84% of cases) of the AL predisposed to and described by Bayford (Bayford 1794), the so called dysphagia lusoria, is caused by esophageal compression (Guzman & Eagleton 2012; Kommerell 1939; Janssen et al. 2000). The antetracheal course of the AL (4-2-5%) or location between the esophagus and the trachea (12-7-15%) may produce airway obstruction, dyspnea and a chronic cough (Myers et al. 2010; Derbel et al. 2012; Janssen et al. 2000).

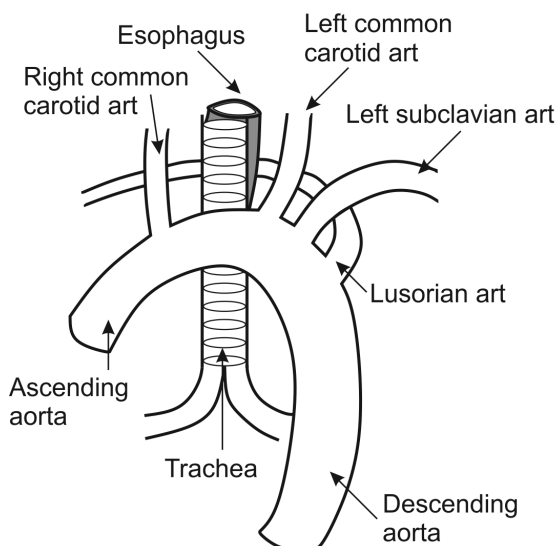


Fig. 1. Schematical path of the aberrant right subclavian artery (arteria lusoria). This artery has its root distal in the left subclavian artery and crosses the spinal column behind the esophagus. Art = artery. Source: Janssen et al. 2000, with own modification

Aneurysmal dilatation of the origin of AL was described first by Kommerell in 1736 (see: Bayford 1794). It occurs in 60% of cases of AL and is known as Kommerell's diverticulum. The dilatation of the AL is associated with the risk of thrombosis, embolisation and rupture (Hunauld 2012; Derbel et al. 2012; Stone et al 2011). The AL remains usually asymptomatic and in 66% of cases is discovered during the examination of other mediastinal anomalies (Stone et al. 2011; Guzman & Eagleton 2012; Baker et al. 2014; Janssen et al. 2000).

Longitudinal cohort studies have noted its frequency in 2.9 patients out of 100,000 persons annually (Mészáros et al. 2000). In the Oxford Vascular Study it was found that AD morbidity was at rate of 6/100 000 person/a year (Erbel et al. 2001). The incidence is higher in men than in women and increases with age (Olsson et al. 2006). Prognosis is worse in women, which is connected with the atypical picture of the illness. The most frequent risk factor in AD is the aortic hyper tension noted in 65–75% patients, which is as a rule badly controlled (Hagan et al. 2000). The mortality rate is high: 21.3% die before reaching the clinic, and 68.2% within the first 2 days after admission (Mészáros et al. 2000).

A lowering of mortality may be made possible only due to rapid angio-tomographic diagnosis and the undertaking of treatment, chiefly angiosurgery. This is connected with the good supply of medical equipment, and a well trained operative team, since such surgery is connected with a high risk of complications and death. This may be concluded from the reports on the implantation of stengrafts into the descending thoracic aorta covering of the left subclavian artery (only one artery) and impairment of the blood flow to the left vertebral aorta. Covering the origin of both subclavian arteries by means of a stentgraft is possible in those cases of congenital anomaly of the right subclavian artery below the left subclavian artery that results in a closing of the blood supply to both vertebral arteries (branches of subclavian arteries) with a full clinical picture of a Steal Syndrome (Subclavian Steal Syndrome - SSS) (Lee et al. 2003).

The aim of this article was an evaluation of the quality of patient life during the long term follow up following dissection of the descending thoracic aorta and angiosurgical related complications.

CASE STUDY

A 31-year-old hypertensive female patient was admitted with a chest and epigastrium pain, with a 20 months history of asymptomatic DeBekey type III thoracic aortic dissection followed in CT scanning. Her medical history included a brain stroke 2 years earlier and suspicion of Marfan syndrome. She did not have a smoking history. Acute coronary syndrome was ruled out. Physical examination revealed no evidence of visceral or peripheral malperfusion with no peritoneal signs.

Computed tomography (CT) was limited to the aorta arch, descending aorta, and to 2-3 centimeters picture of vessels branching from the arch of the descending aorta in the upper part of the exposition (Fig. 2a-c). One can see the broadening of the false channel of dissection of the DeBakey type III and renal arteries,

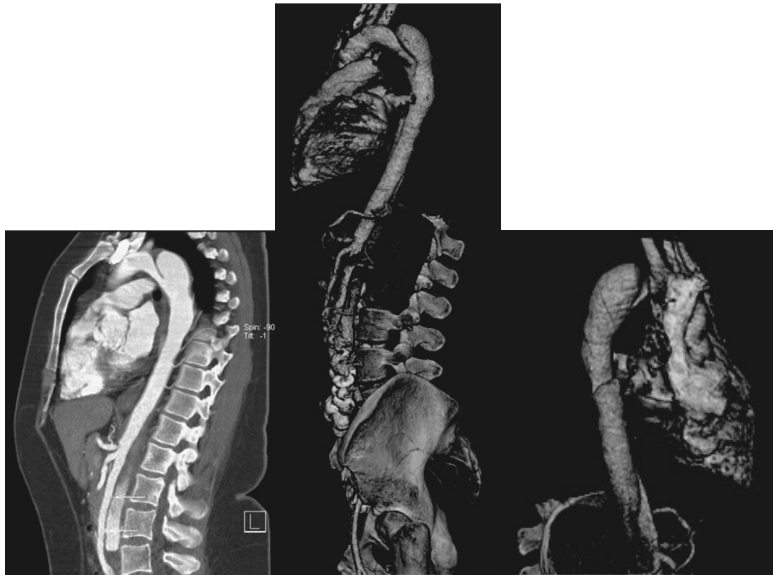


Fig. 2a-c. Preoperative angio-tomography. Spatial reconstruction. The presence of the fourth vessel is difficult to delineate

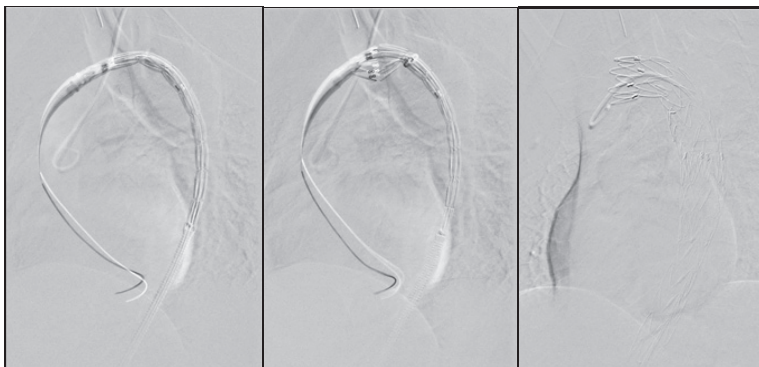


Fig. 3. a-c. Intraoperative angiography. Sequential stages of releasing and implantation of the stentgraft to the descending aorta (a-b). The stentgraft in the descending aorta after fixing it (c). The catheter “pig-tail” (with the bend of its tail) in the left subclavian aorta, which indicated the level of fixing of the upper edge of the stentgraft (a-b)

superior mesenteric artery, coeliac trunc originated from the true lumen was observed in comparison to the earlier computed tomography. Restriction of the upper scans area made impossible aortic arch vessels evaluations. It was the main reason for the lack of diagnosis of the congenital anomaly, which is expressed by the occurrence of AL.

Urgent surgery was performed on the patient, implanting the stentgraft to the descending aorta with a partial covering of the left subclavian artery marked with a catheter “pig-tail” (with bending tip), which was a mark of the level of stentgraft fixing, and enabled one to introduce a contrast to the blood (Fig. 3a-d).



Fig. 4. Angio-tomographic examination performed after the appearance of the neurological symptoms. Only two vessels can be noticed (the right and left common cervical arteries). Subclavian arteries (RSA, LSA) were not exposed

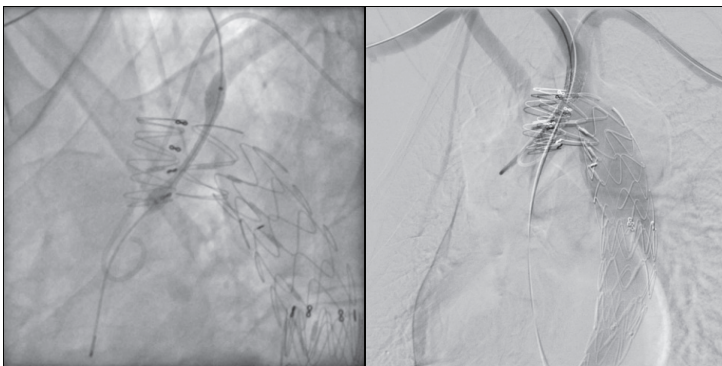


Fig. 5a-b Intraoperative angiography performed during the second surgical procedure. Sequential stages of the stent implantation to the Arteria Lusoria (RSA - AL). (a) The “pig-tail” catheter (with the bending tip as a guide of left subclavian artery and the stent on the balloon catheter indwelled through the right subclavian artery – AL). (b) The view of the expanded stent

After the surgery the patient reported the disappearance of the chest and epigastrum pain, but after a few hours she developed symptoms of a brain stroke, including a loss of consciousness. Computed tomography (CT) revealed two arteries arising from the aorta arch (Fig. 4). It also showed a covering of the entrance to the false channel and an almost complete covering of the left subclavian artery by the stentgraft as well. It was surprising that previous the CT examination had missed the congenital anatomical anomaly of the arising of the right subclavian artery – AL. This explained the reason for these surgical complications.

Ultrasound examination revealed a retrograde flow in the right vertebral artery (RVA) with fully clinically manifested steal syndrome (Subclavian Steal Syndrome, SSS). A successful RSA revascularization was performed next day (Fig.5a-b). Through a right brachial artery and aberrant RSA a 8x39mm balloon-

expandable stent Omnilink Elite was deployed between the wall of the aortic arch and the stentgraft. This restored antegrade flow into both the left and aberrant right subclavian arteries and right and left vertebral arteries as well.

The patient recovered well and a follow-up CT imaging (Fig. 6a-b) revealed stability of the repair, no graft migration, an absence of endoleaks and patient

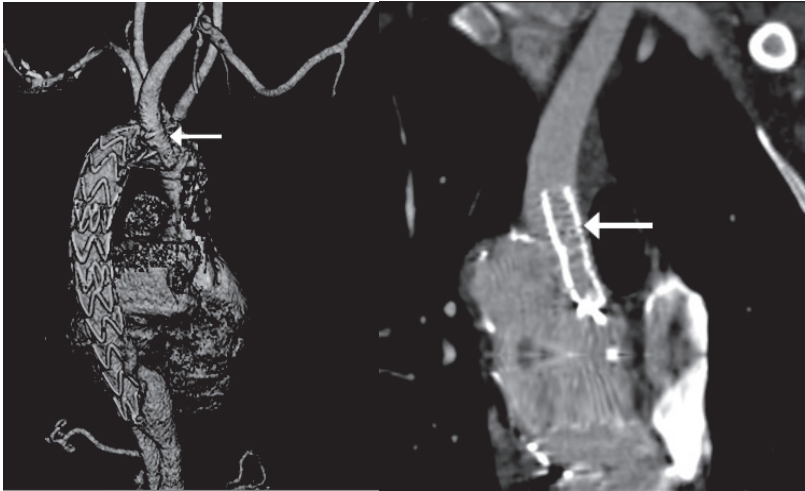


Fig. 6a-b. 3-D reconstruction of CT angi-tomography scans after the second procedure. The arrow points to the stent at the entrance of AL



Fig. 7a-b. 3-D reconstruction of CT angi-tomography scans after the second procedure. The view of the thoracic and abdominal aorta. The dissection of the abdominal aorta below the lower edge of the stentgraft can be well seen

LSA and aberrant RSA origins (RSA with a stent). The patient remained asymptomatic, with no symptoms of arm claudication. Spinal cord ischemia was not observed as well.

NEUROPSYCHOLOGICAL FINDINGS

Examinations were performed before surgery, at a discharge 12 and 24 months after the operation by the same team of neuropsychologists and included standard neuropsychological assessment including: an interview – 20 questions on life satisfaction, Mini-Mental State Examination (Folstein, Folstein and Mc Hugh, 1975), and SF-36 (Tylka i Piotrowicz 2009). The assessments was administered at the baseline before the first operation, between operations, 1 day after the second operation, at periods of 12 and 24 months postoperatively (Tab. 1).

Table 1 shows that the patient manifested mild cognitive impairment in MMSE examination (18/30 points.) manifested by attention deconcentration in working memory disorders. After the first surgery the examination was impossible due to the patient's loss of consciousness. The examination administered a day after the second reconstructive surgery revealed mild improvement in brain functions (20/30 points). This may be connected with the big stress for the organism due to two consecutive operations with the use of general anesthesia as well as the previous brain stroke. The patient recovered and her cognitive and social functioning systematically improved, which was seen in the neuropsychological tests results. Twelve months after surgery the patient scored 28 out of the 30 points possible, and two years later her scores reached the upper level of the norm.

The health related quality of life before the first operation was low (89/171 scores in SF-36). The patient experienced strong thoracic pain radiating between the shoulders and a tremendous death anxiety. She also complained of abdominal pain and abdominal muscles fibrillation. This was due to her dramatically low sense of well-being and low self-esteem. Study after the first surgery was not possible for the reasons already given above.

The second revascularization procedure restored normal blood flow to the brain and resulted in the improvement of cognitive ability. The patient, after regaining consciousness, was informed that the first operation protected her tho-

Table 1. Assessments at baseline before the first operation, between operations, 1 day after the second operation, at 12 and 24 months postoperatively

Test	Before the first operation	Between operations	1 day after the second operation	12 months after the second operation	24 months after the second operation
MMSE	18/30	unconscious	20/30	28/30	30/30
SF-36	89/171	unconscious	136/171	146/171	159/171

Mini-Mental State Examination = MMSE;
SF-36 = Short Form Questionnaire.

racic aorta from rupture with the result of a reduction of thoracic and abdominal pain, and the second recovered the proper functioning of her brain. This has meant that her mood and health related quality of life have dramatically improved.

It was found that in the SF-36 scale results she achieved 136 / 171 pts. Further improvement in the quality of life was parallel with the improvement of her health. 12 months after surgery the patient scored 146/171 pts., and two years later she gained 159/171 pts. Currently, she does not complain about pain nor sleep disorders and nightmares. She also does not feel death anxiety. Moreover, she believes that she independently may perform many everyday life activities (ADL) including sexual ones. She stopped isolating herself from the local society. She feels happy, and, as she put it, her quality of life is almost perfect.

DISCUSSION

The case study presented here has shown that three incorrectly made and interpreted, over a prior period of 20 months, computerized tomography (CT) of the thoracic aorta were not able to detect the presence of genetically related, anatomical abnormalities of the right subclavian artery (Arteria Lusoria, AL).

As a result, the stengraft was deployed too high, which resulted in a closing of the inflow to both subclavian arteries, and vertebral arteries consecutively. Very often, we cover the entrance of the left subclavian artery to obtain complete closure of the entrance to the false channel of aortic dissection.

There is, however the risk of the development of steal syndrome of the left vertebral artery, though this is usually low. Only when the patient manifests severe neurological symptoms and left upper limb ischemia, do we decide to conduct revascularization, usually by-passing vascular prosthesis from the left carotid artery to the left subclavian artery. In the case of closing both subclavian arteries, the symptoms of steal syndrome are usually so severe with a brain stroke that angiosurgical intervention is a necessity.

A young mother of two children with a full awareness of the dissection of her aorta and the possibility of mortal complications had lived under severe stress for 20 months. The stress became stronger with the appearance of abdominal pain after she had been informed about a widening of the dissection and the necessity for an operation which might save her life. After the surgery the patient was happy and did not pay attention to the weakness of her right hand. This was followed by dizziness and a consequent loss of consciousness. Waking up after the second surgery she again manifested her happiness due to the fact that her previous pain had disappeared, and that her hand was functioning again. She was slightly worried with the fact that the aorta was dissected below the deployed stentgraft, and there is no possibility to place a subsequent prosthesis (Fig. 7a-b). She was informed that the risk of complications within that area will be lower if she kept to her doctor's recommendations, such as the systematic control of blood pressure, prohibiting hard and middling-hard work, and the lifting of heavy

objects as well a prophylaxis of constipation.

The long period of pain absence and psychological discomfort over the subsequent two years resulted in an unawareness of the danger connected with the dissection of the aorta below the fixed stentgraft (Fig. 7a-b). In our opinion the high evaluation of the quality of life described by the patient is not compliant with the real state of her health. It may be connected with a pushing out of any awareness of the risk of complications, of her young age and good family relations as well as the need to take care of her two young children.

CONCLUSIONS

1. In the presented case study, badly performed and wrongly interpreted diagnostics examination led to the incorrect treatment, one resulting in complications (a brain stroke).
2. Only careful Intensive Care Unit observation following the first operation allowed one to undertake fast decisions to implement corrective treatment.
3. A long lasting illness such as the dissection of the thoracic aorta resulted in severe stress combined with tremendous anxiety, which all decreased the quality of life.
4. A successful reconstructive surgery reduced the stress and increased to a considerable degree the quality of life.

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