Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

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CASE SUMMARY

The opinion focuses on a 39-year-old female patient, who was first diagnosed with mild asthma when 7 years old. When she was mature, after her 30th year of life, she developed intensified symptoms of asthma. At that time, she started to be regularly treated for asthma; at approximately the same time, her clinical presentation also included symptoms of dysphagia.

In spite of the initiated therapy, her progressing asthma could not have been controlled. The patient required numerous hospitalizations. Computed tomography performed in 2008 directed the diagnostics process towards a vascular ring formed by an abnormal, aberrant course of the right subclavian artery, which posteriorly compressed the esophagus and indirectly compressed the trachea. Bronchoscopy demonstrated considerable bulging of the posterior tracheal wall when the patient was coughing.

At present, the patient suffers from recurrent ailments - coughing, stridor and dyspnea; recently, she has been on a liquid diet only. She is additionally treated for resistant hypertension, tachycardia, diabetes, hypercholesterolemia, nodular goiter, obesity and degenerative bone disease. She has also had an episode of pulmonary embolism. In her two children, similar vascular rings were successfully released surgically.

DISCUSSION

The vascular ring diagnosed in the patient is an effect of abnormal anatomy of the right subclavian artery (*a. lusoria*), which branches off from the aortic arch as the last vessel and extends extraesophageally to the right, thus affecting the main respiratory tract and the esophagus (*arteria lusoria* means in English "playing" or "merry").

The right *a. lusoria* in various variants (isolated and combined) accounts for approximately 0.5% of congenital heart defects and in some reports it is this artery that is recognized as the most common vascular abnormality involving the mediastinum. Compared to other forms of vascular rings, the aberrant, extraesophageal course of the subclavian artery is more frequently asymptomatic or else the compression symptoms of the esophagus and

trachea are minor and do not require surgical treatment[1]. The onset of clinical symptoms of a vascular ring in an adult patient (both respiratory symptoms and dysphagia) has been described extremely rarely, although it has been noted as late as in the eighth decade of life[2]. As a rule, surgical treatment is employed in symptomatic patients only. If, however, we believe the intensity of compression symptoms to be significant, surgical division of *a. lusoria* is the treatment of choice. The most commonly reported clinical symptoms of a vascular ring in an adult patient are not, however, associated with swallowing disturbances, but rather symptoms involving the respiratory system, such as dyspnea, stridor, recurrent pneumonia, persistent bronchitis and non-specific respiratory symptoms[3]. Various authors emphasize that the onset of dysphagia symptoms may be associated with the patient developing esophageal motility disorders and reflux[4]. In children, reimplantation of the divided artery is not recommended, while such a solution may be considered in adult patients. The release of the vascular ring formed by *a. lusoria* is a simple and safe procedure, which does not require the use of extracorporeal circulation. Left throracotomy through the 4th intercostal space is a good and commonly employed surgical approach. Dividing the aberrantly branching artery is performed proximally to the site of its branching off the aorta. The technique of surgical management of a vascular ring in an adult patient is still subject of discussion and there are advocates of reconstructing the vascular system of the upper extremity following the division of the subclavian artery[5].

While discussing surgical techniques employed in treating vascular rings, one should not forget video-thoracoscopic techniques, which have recently found a permanent place in treatment of numerous vascular abnormalities within the chest; however, such techniques have been used solely in children. To date, no attempts have been made at thoracoscopic division of a vascular ring in an adult patient[6].

EXPERT'S OPINION

The present opinion discusses an adult patient with clinical symptoms of a vascular ring, who additionally suffers from numerous concomitant diseases. One may surmise that asthma diagnosed in childhood was in fact a manifestation of mediastinal compression that facilitated the onset of asthma, if indeed it was asthma. Long-term irritation of the trachea with superimposed inflammatory processes might have facilitated the development of clinical bronchial asthma in its full form[7] or else given pretense of asthma resulting in many years of inappropriate treatment[8]. Such reactions have been observed as predominant symptoms of a vascular ring in adult patients, posing serious diagnostics problems[9].

It appears that the present symptoms of tracheal and esophageal compression predominate and affect the patient's discomfort. Living on a liquid diet for many years is a torment that justifies radical surgical treatment.

Surgical treatment of a vascular ring consists in division of the compressing vessel. Usually, the defect is diagnosed and qualified for treatment in childhood. In case of *a. lusoria* division, if the problem affects a child, there is no danger of ischemia in the extremity supplied by the artery. This is confirmed by the commonly shared belief of cardiac surgeons and pediatric cardiac surgery centers worldwide based on vast experience in surgical treatment of many thousands of patients. Additional support for the belief in a safe division of *a. lusoria*, without upper extremity ischemia and with a minimal risk of subclavian steal syndrome, is found in follow-up of thousands of children, who were for many years subjected

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to the classic Blalock-Taussig shunt with radical division of the subclavian artery. Such procedures were also performed in adult patients in the fifties and sixties of the 20th century.

Nevertheless, theoretically, in case of an adult patients, some concerns about ischemia of the upper extremity are somewhat - though minutely - justified. An adult has lower adaptive reserves in case a large artery is obliterated, while additional aggravating factors (diabetes, hypertension) show a predilection disturbing the blood supply to distal segments of the extremities. The collateral circulation may prove to be less efficient. In this case, support provided by data from the literature is not a reasonable and safe justification of a decision on the type of surgical solution to be employed. The selection of treatment modality may be thus made completely individually, based on the experience of the surgeon and on tests allowing for determining the necessary safety margin.

On the other hand, reimplantation of the subclavian artery would be a much more extensive surgical procedure that would increase the surgical risk, and planning such a procedure a priori is not justified. Reimplantation is associated with a considerably higher traumatization of the mediastinum and the risk of a many-hour-long surgical procedure what would be a great strain for the patient in view of her concomitant diseases.

CONCLUSION

A congenital defect manifested by burdensome clinical symptoms requires surgical treatment as the only causative and rational treatment modality, the more so that the operation is relatively simple and the risk low. Depriving the patient of the possibility of a radical solution would be illogical in this case. What is baffling is not only the very late diagnosis of the defect, but also lack of suggestions on the part of the consulting physicians to perform surgery, the said recommendation having not been voiced for five years after the defect had been diagnosed.

Thus, in view of the absolute clinical justification of the decision to perform radical surgical treatment, for the sake of safety I propose to perform a test in a hemodynamic lab that would answer the question whether obliterating *a. lusoria* may in this particular case result in ischemia of the right upper extremity. A balloon catheter should be introduced to the right subclavian artery. A test balloon obliteration of the artery for a period of 12-24 hours is a procedure that is safe for the patient when prophylactic heparin is employed and the woman is hospitalized in a professional intensive care unit. No ischemic symptoms will be a justification for a surgically simple division of the artery without reimplantation. In the course of the test obliteration of the artery, one may also verify the possibility of blood steal phenomenon to the right upper extremity by the collateral circulation. In case abnormalities of blood supply involving the right upper extremity develop, the vascular ring should be released and the right subclavian artery should be at the same time reimplanted. The blood steal syndrome would also be an indication for artery reimplantation[10]. If the steal symptoms appear at a later time, even after weeks or months[11], reimplantation of the distal end of the subclavian artery to the ascending aorta or brachio-cephalic trunk via a sternotomy is possible.

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