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

Authors report case of 38 year old women with severe bronchial asthma and vascular ring. The patient's son and daughter were operated in childhood because of symptomatic aberrant retro-esophageal right subclavian artery (ARSA).

The patient has been admitted to hospital several times due to acute dyspnea, stridor and dysphgia. Concomitant diseases included diabetes mellitus, hypertension and obesity. Currently on admission the patient presented high blood pressure, inspiratory and expiratory wheezing stridor. Oxygen saturation was 100% in room air. Echocardiography demonstrated normal left and right ventricular size and function with no significant valve abnormalities. Computed tomography and angiography confirmed diagnosis of left aortic arch with a retro-esophageal right subclavian artery and common origin of the carotid arteries. An aneurysm and dissection of ARSA were excluded. A barium esophagram and gastroscopy revealed compression of the esophagus. On bronchoscopic examination the tracheal lumen was reduced by 1/3 during a cough, no obstruction of the bronchi was found. Pulmonary function test confirmed an asthma diagnosis.

The patient's case has been presented at a multidisciplinary board. Experts have expressed divergent views on further management. They have advised surgery (Roland Hetzer, Department of Cardiothoracic and Vascular Surgery, Deutsches Herzzentrum Berlin), conservative treatment (Bogusław Kapelak, Department of Cardiovascular Surgery and Transplantology, John Paul II Hospital in Krakow), further diagnostic (Tomasz Mroczek, Department of Pediatric Cardiac Surgery, University Children's Hospital, Krakow).

DISCUSSION

A left aortic arch with a aberrant retro-esophageal right subclavian artery was initially described in 1735 at autopsy by Hunauld. ARSA is an anomalous vessel that arises from the aortic arch distal of the left subclavian artery, and runs behind the esophagus to the right. It is a rare congenital anomaly of the aortic arch found in 0.5% of the healthy population and in

30% of fetuses with Down syndrome. Familiar occurrence of ARSA is extremely rare, single case has been described by Arribas. The presence of ARSA is associated with an increased incidence of cardiac defects. Embroyologically, it develops as a result of the persistence of The seventh intersegmental artery caused by abnormal involution of the fourth aortic arch.

Sixty percent of patients with ARSA have Kommerell's diverticulum, that is aortic diverticulum at the origin of ARSA. An aneurysm of the Kommerell' diverticulum have been observed in 3–8% of these patients, dissection of the diverticulum has been also described.

ARSA is usually asymptomatic, but due to compression of the esophagus and trachea may cause dysphagia, cough, stridor, thoracic pain and respiratory infections. Symptoms may develop in adulthood secondary, to the development of atherosclerotic rigidity and tortuosity. Tracheal obstruction may be caused by diverticulum not artery itself**.** ARSA may also become aneurysmal. Rupture and dissection of these aneurysms have been reported and post-rupture mortality rate was approximately 50%.

Kieffer et al. distinguishes four groups of patients with aberrant subclavian artery (aSA): patients with dysphagia caused by esophageal compression by a nonaneurysmal aSA (group 1), with ischemic symptoms caused by occlusive disease of a nonaneurysmal aSA (group 2), with aneurysms of the aSA (group 3) and patients with aSA arising from a diseased thoracic aorta (group 4).

Diagnostic tools include: ultrasonography, barium-contrast examinations of the esophagus, computed tomography, contrast-enhanced magnetic resonance angiography and digital subtraction angiography.

Because of the rareness of the symtomatic ARSA there are no accepted guidelines. Management is individually based. Symptomatic patients or patients with aneurysmal ARSA require surgical intervention. In the treatment various surgical methods are used. First operation by Gross performed in 1946, consisted of ligation of the aberrant vessel. Reports showed that simple division without restoration of flow to ARSA may lead to ischemia of the right arm, and subclavian steal phenomenon. Reconstitution of flow may be obtained by attaching the distal end of the ligated artery to the ascending aorta proximal to the right common carotid artery (first described by Bailey) or to the right common carotid artery (Cooley) either directly or by use of a short interposition graft. The surgical approach depends on ARSA anatomy and center preference, it includes: supraclavicular incision, median sternotomy, left and right thoracotomies. Open surgery of ARSA aneurysms has high perioperative mortality rates. In some cases of ARSA aneurysms endovascular or combined surgical and endovascular interventions have been effective.

Kieffer et al. report surgical treatment in thirty-three adult patient with aberrant subclavian artery (28 with ARSA). Four patients with aneurysms of the aSA or aSA arising from a diseased thoracic aorta died after operation (groups 3 and 4), satisfactory results were obtained in the remaining group. Atay et all. (2006) report uneventful surgery in four patients (3 children and 1 adult).

EXPERT'S OPINION

Presented symptoms may relate to bronchial asthma and compression of trachea by the ring. It is difficult to distinguish the role of asthma and vascular ring in patient's symptomatology.

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Symptomatic ARSA is a strong indication for surgery. Patients without aneurysms of the aSA or aSA arising from a diseased thoracic aorta, have acceptable surgical risk on the basis of Kieffer report. The choice of surgical approach depends on surgeon's experience and the anatomic details.

CONCLUSION

Authors report case of 38 year old women with severe bronchial asthma and familiar occurrence of retro-esophageal right subclavian artery. The presented case is rare, relevant and interesting. Because of the rareness of the disease management must be individually based.

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