



The Relationship between Chest Pain and Right Aortic Arch with Left Aberrant Subclavian Artery

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Introduction

Aberrant subclavian artery is a rare anomaly among all anomalies related to aortic arch. In some cases kind of aneurysm named Kommerell diverticulum, originated from right subclavian artery and left aortic arch and the opposite, left subclavian artery association with right aortic arch [1]. Right Aortic Arch(RAA) is a rare congenital anomaly with 0.5 to 1% of normal population. Almost half of the Cases associated with left aberrant subclavian artery and in some, aneurysmal changes in the origin of the artery is inevitable [2]. RAA is categorized in three groups : 1.RAA with left aberrant Subclavian artery 2.RAA in mirror type. 2.RAA with isolated LSA, which RAA with ILSA is a most rare by 0.8 % prevalence [3]. Left aberrant subclavian artery (LASA) is an anatomical variant of right aortic arch which is actually dispart from the RAA as a last branch and usually pass behind esophagus to the left upper limb. However, RASA associated with LAA is more common (0.5_2% of population) the LASA originated from RAA(0.05_0.1)[4]. Kommerells diverticulum or KD, is defined as a aneurysmal form of ASA or descending aorta in proximal part [5] .KD is a rare condition usually accompany RASA more

than LASA [6, 7]. Up to now, few cases has reported. Burckhard F Kommerell was the first one who introduced kommerell in 1936 [8]. Anomalies of aortic arch not necessary influence health condition in childhood and some become symptomatic in adolescence [9]. These aneurysms could be strong threatening for some circumstances like dissection, rupture, emboli to distal parts, compression to adjacent or organs [1]. Rupture and of course mortality rate, is more probable when the diameter is more than 30 mm [10]. Dysphagia also is another symptom which is defensible because of anatomical issues explained above. Most of the patients are symptom free that make the treatment option hard to choose [11]. Some of the technics are transposition or bypass the distal part of aberrant artery to adjust Carotid Artery with or without of ligation the proximal part including excision aneurysm [12-14]. Other options are complete replacement of the arch [15], replacement the descending aorta with transposition aberrant subclavian artery and carotid artery [16], extra anatomical bypass with open standard grafting , ligation of aberrant subclavian artery with descending aorta reconstruction [15].

Case presentation

A 36-year-old woman, admitted to our hospital of Tehran University of Medical Sciences at 2019 years for chest pain and hemoptysis of 3 days evolution and progress. Beside these symptoms, patient had intermittent dysphagia for a long time. After ruling out cardiac problems, for further evaluation not only chest CT angiography was done, but also consult with pulmonologist for flexible bronchoscopy requested (which has no pathologic finding). According to the report of CT angiography, right aortic arch with left aberrant subclavian artery which had a aneurysmal origin was detected, so surgery was planned.

Procedure

After prep and drep in general anesthesia and left lateral position, right posterolateral thoracotomy has performed. Then aortic arch and descending aorta circumferentially was freed. The origin of left subclavian artery aneurysm in proximal behind the descending aorta has been detected, then perfect dissection from the all tissues around carried out to clamp the aorta partially. The aneurysm successfully excised, the exit site of subclavian artery repaired with 4.0 prolens continuously. After that dissection continued to left side of esophagus and subclavian artery ligated. Then position changed to supine, base of the neck opened, left subclavian artery and common carotid

artery explored, with cortex graft, left common carotid bypassed to subclavian artery with 5.0 prolens, distal pulses of superior limb checked, at the end, site of incision repaired and closed.

Discussion

Congenital anomalies for aortic arch usually find incidentally in mediastinal disorder assessments or in autopsy. The aberrant subclavian artery is present in 0.5 to 1.8% of cases [17] which is more common in right than left [11]. During Embryological development, 6-paired aortic arches form in different steps of organogenesis. Descending aorta would be in a left side of vertebral column. The some cases right column will be remained instead, which result right aortic arch. Depending on aortic branches we have some categories that three of them related to right aorta. The incidence of type one, was 59%, in type 2, 39, 5%, in type 3, 0.8%. For example in type 2 (our case) left carotid is a first branch and then right carotid and subclavian artery and the last one is aberrant subclavian (RAA +ALSA). In RAA with mirror type, left innominate artery is a first one then right carotid and subclavian. a. in form of left isolated subclavian artery, left carotid. a is a first then right carotid and subclavian artery, left subclavian without any relation to aorta, communicates with pulmonary. a from left Ductus arteriosus [13, 18-20]. Congenital heart disease include tetralogy Fallot, pulmonary



stenosis with intraventricular defects, tricuspid atresia, truncus arteriosus are seen in 75-85% in RAA type 1 and 3, 5-10% type 2^[13]. Right aberrant subclavian artery is seen in 0.5-1% of normal population. Left aberrant subclavian artery originated from aortic arch is less common. Kommerell diverticulum has a rate of 60% in patient with aberrant subclavian artery.

Aneurysmal formation often happens in the origin of left aberrant subclavian artery and is known as a Kommerell. In previous studies proved that Kommerell has 100% rate in right aortic arch and left aberrant subclavian artery and is diagnosed with CT scan^[5]. Right aortic arch in adults usually presents symptoms related to anomalies due to primary vascular atherosclerosis changes^[7], causes dilation, dissection, and therefore compression effects in adjacent organs. Some of them are dysphagia, dyspnea, stridor, wheezing, cough, choking, recurrent pneumonia, obstructive emphysema, chest pain and so on^[21]. Left aberrant artery can be either in posterior of esophagus (80%), in the middle of trachea and esophagus (15%), and anterior to trachea or main bronchus (5%) so being symptomatic even in the absence of aneurysm is possible^[13, 21, 22]. Left aberrant artery comes along esophagus to get to axillary region^[23]. Most of them are asymptomatic although has compression on esophagus, trachea, laryngeal nerve that causes some symptoms like dysphagia, recurrent aspiration, dyspnea, cough^[24].

First case of LASA introduced in 1735 by Hanauld^[25], but in 1789 Byford was the one who described LASA clinically and invented the word "dysphagia lusoria" as a description^[26]. Kommerell in 1936 described radiologic manifestations. Histopathologic studies recently show intimal cystic necrosis in a Kommerell diverticulum wall which made the diverticulum vulnerable to dissection and rupture^[27]. Clinical importance of this rare

condition is because of high rate mortality and morbidity result of compression effect to adjacent organs and surgical complexity. Other significant complications such as narrowing and stenosis of subclavian artery or pseudo coarctation of aorta even is more rare^[28].

Cases that are asymptomatic and found incidentally, without aneurysm, has no indication for surgery^[14]. In younger patients other associated anomalies such as double aortic arch, right aortic arch with vascular ring ductus arteriosus which encompass esophagus, maybe has a similar symptoms. Respiratory manifestations in infants and children because of flaccidity of trachea are more common^[29]. Dysphagia, caused by pressure on esophagus and can lead to sclerosis and aneurysm. Indication for surgery depends on age, symptoms, in children and infants surgery may be considered earlier because of symptoms^[30]. For adults with mild symptoms can be treated by appropriate nutrition and drugs like PPI^[29]. In 1946, Gross suggested simple isolation and ligation of ASA^[30], although this method is acceptable in children but in adults it causes still syndrome or upper limb ischemia with more probability^[31, 32]. In 1964, Cooley anastomosed right carotid artery to right subclavian so innominate artery has reconstructed^[33]. Instead of traditional procedure which thoracotomy and sternotomy both are done to access the vessels, recently a lonely collar incision with or without synthetic graft is popular^[34]. Endoscopic procedure is an alternative option in patients whom not suitable for open access^[35].

Although rupture and dissection of Kommerell diverticulum is reported but because of rare condition is not completely known. Austin and Wolfe estimate the rupture 19% in 32 patients^[12]. Cina and colleagues reported the rupture and dissection 53% in 32 cases with

LASA and RAA [13]. Nicholas and collegs also report 2 dissection in 10 patients . There is no definite threshold for surgery in cases who are asymptomatic or have minimal symptoms. Cina and colleagues suggested surgery for 3cm or more in size and [13] ota suggested 5cm [7] but overally surgical interventions as soon as possible is recommended [7-37]. Standard method for surgery is Left preferred approach since thoracotomy, resection of a aneurysm replaced by graft, however bypass a distal of subclavian artery to descending aorta is choice in cases which there is no straight alignment between them [1]. This approach created suitable exposure for repair and access to all major vessels. Hypothermic circulatory arrest which is supported by previous study, is a preferred approach since need for clamping aorta, subclavian artery and others is less in result side effects are diminished [1].

Lococo and co-worker presented a case which KA was incidentally found, patient was asymptomatic with size 30 mm, general condition of patient was not appropriate for surgery so follow up choosed. In 12 month follow up patient remained asymptomatic. Thus this conclusion came to mind that if the size of KA less than 30 and incidentally find, beside that patient has poor prognosis maybe closed follow up by radiologic assessment would be helpful [10].

Takeyoshi and co-workers had their first patient in sternotomy incision instead of thoracotomy, although they had to extend the incision to antrolateral to thoracotomy to achieve appropriate exposure to aortic arch and proximal of descending aorta [7, 16].

Conclusion

There is a relationship between chest pain and right aortic arch with left subcellular artery inactivity.

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The Vascular & Endovascular Surgeon of affiliated to Tehran University of Medical Sciences (TUMS) approved this study.

Authors ' contribution

M N, and SS, jZ, F.P. will perform the study and will collect the data for analysis. He also analyse the data.

M N, and SS, F.P, MHD will supervise implementation of whole study.

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