Surgical Correction of Vascular Ring in Thai Patients at a Tertiary Hospital

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Background: *Vascular ring is a rare congenital vascular disease. The result of vascular ring surgery in Thailand remains unknown as it lacks serial data. The purpose of the present study was to evaluate the result of vascular ring surgery at Siriraj Hospital and demonstrate the usefulness of the computed tomograph angiographic scanning (CTA) for preoperative anatomical diagnosis of the vascular ring for planning the surgical correction.*
Material and Method: *We reviewed our experiences for all patients that underwent vascular rings surgery at our institute between 1979 and 2009, about preoperative diagnostic imaging, operative technique, and clinical outcome.*
Results: *Twelve medical records of patients that underwent vascular rings surgery were reviewed. The age at time of operation ranged from one month to two years (median 3 months). The surgical approaches were eight median sternotomies, three left thoracotomies, and one right thoracotomy. There were two cases of postoperative complication (residual tracheal and left pulmonary artery stenosis and surgical bleeding). There was one hospital mortality (pneumonia and sepsis).*
Conclusion: *Surgical correction of vascular ring is effective and safe. CTA can give the precise anatomic diagnosis of the vascular rings, lead to good surgical planning, proper surgical approach, and good outcome.*

Keywords: *Vascular ring, Computed tomograph angiographic (CTA)*

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Vascular ring is a rare abnormal anatomical arrangement of the great vessels that leads to compression of the trachea and esophagus. Symptomatic vascular ring is about 1% of all cardiovascular anomalies[1]. Vascular rings can be divided into complete and incomplete rings (Table 1, Fig.1). The Congenital Heart Surgery Nomenclature and Database Project[2] classify vascular rings as double aortic arch, right arch/left ligamentum, pulmonary artery sling, left arch/aberrant right subclavian artery, and innominate artery compression. There are variable clinical presentations and they can be asymptomatic. The most common presenting symptoms are respiratory distress, stridor, cough, and dysphagia. In the past, the diagnosis of vascular rings can be performed by multiple tests such as chest radiography, barium esophagography, bronchoscopy, echocardiography, and cardiac catheterization, but these investigations cannot give precise anatomical diagnosis. Recently, both the computed tomograph angiographic scanning (CTA) and MRI are able to give accuracy that lead to good surgical planning and outcome[3-5]. The indications for repairing vascular rings include all symptomatic patients and complete vascular ring patients. The optimal surgical approach depends on the vascular anatomy. It can be sternotomy or thoracotomy. The result of vascular ring surgery in Thailand remains unknown as it lacks serial data. The purpose of the present study was to report the result of vascular ring surgery at Siriraj Hospital and demonstrate the usefulness of CTA for preoperative anatomical diagnosis of the vascular ring for planning of surgical correction at Siriraj Hospital.

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Material and Method

The study was approved by the Siriraj Ethical Committee. A retrospective study was conducted from

Table 1. Vascular rings

Complete ring	Incomplete ring
Double aortic arch	Innominate artery
Right arch dominant	compression syndrome
Left arch dominant	Pulmonary artery sling
Right arch/left ligamentum	Left arch/aberrant RSCA
Retroesophageal LSCA	
Duct of Kommerell	

LSCA = left subclavian artery; RSCA = right subclavian artery

Siriraj registry database on all patients who underwent vascular rings surgery at our institute between 1979 and 2009. Patients were evaluated for demographic, preoperative, operative finding, and postoperative data. The follow-up was achieved by regular clinic visit.

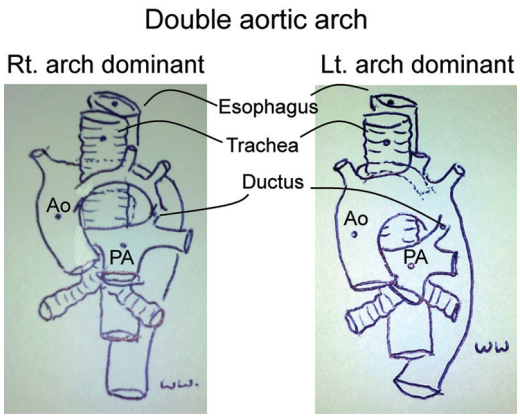

Fig. 1 Double aortic arch.

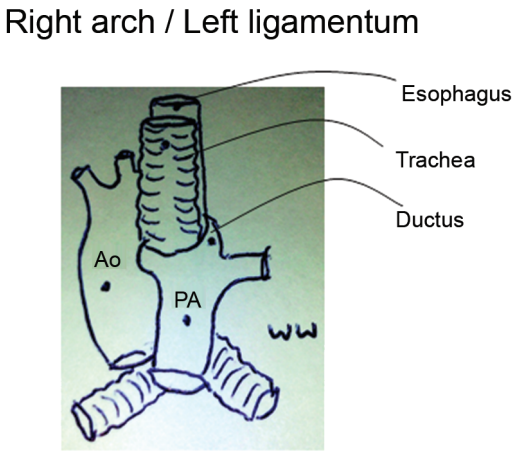

Fig. 2 Right aortic arch/left ligamentum.

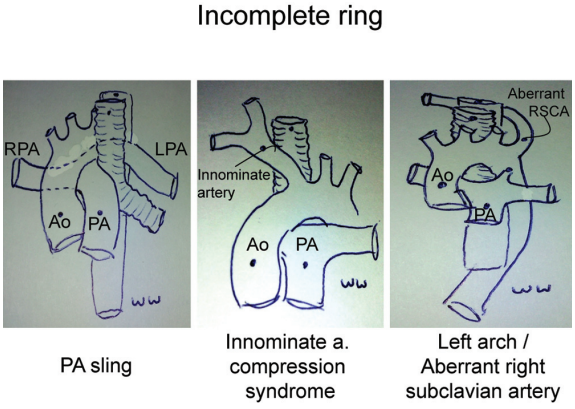

Fig. 3 Incomplete ring.

Results

Between 1979 and 2009 (30 years), we performed 21,346 cardiac surgeries. Of these, 13,006 cases (61%) were acquired heart disease (AHD) and 8,340 cases (39%) were congenital heart disease (CHD). Twelve patients (0.15% of CHD) underwent vascular rings repair by the same surgical team. There were seven boys (58.33%) and five girls (41.66%). The age at time of operation ranged from one month to two years (median three months). The type of vascular rings were two cases of double aortic arch, two cases of right arch/left ligamentum, and eight cases of pulmonary artery sling. The surgical approaches were left thoracotomy in three cases, right thoracotomy in one case, and median sternotomy in eight cases. The length of hospital stay was from nine days to three months. There were two postoperative complications (16.66%), one case of residual tracheal and left pulmonary artery stenosis and one case case of surgical bleeding. There was one hospital mortality (8.33%) due to pneumonia and sepsis (Table 2).

Discussion

CT scan had been used in Thailand since 1981. Prior to this period, the diagnosis of vascular rings could be performed by multiple tests such as chest radiography, barium esophagography, bronchoscopy, echocardiography, and cardiac catheterization but these investigations could not give precise anatomy diagnosis. After 1981, CT scans have the ability to give accuracy that lead to good surgical planning and outcome.

There were two interesting cases in the present study (Table 2). Case number 10 was a four months old girl with respiratory stridor since birth. She had repeated attack of pneumonia and needed ventilator support. With difficulty in endotracheal extubation, bronchoscopy revealed external pulsatile compression of the lower trachea. The CTA was performed and revealed double aortic arch (left arch dominant) with tracheo-esophageal compression (Fig. 4). Then the non-dominant right aortic arch and ligamentum arteriosum were divided through the left thoracotomy (Fig. 5). She was extubated 47 days after surgery.

Case number 11 was a 2-years-old girl with history of dyspnea, dysphagia and recurrent pneumonia since six months old. Esophagography revealed posterior indentation of upper esophagus (Fig. 6). Bronchoscopy revealed narrowing of the lower trachea and left main bronchus. The CTA was performed and revealed complete vascular ring (right

Table 2. Vascular rings cases

No.	Year	Sex	Age	Dx	Sx	LOS	Result
1	1981	M	2 months	DAA	Left thoracotomy, Division of anterior arch and PDA	30 months	Alive
2	1994	F	2 months	Right arch/left ligamentum	Left thoracotomy, Division of ligamentum arteriosum and Kommerell diverticulum, Reimplantation of LSCA	2 weeks	Alive
3	2000	M	6 months	PA sling, tracheal stenosis	Median sternotomy, Reimplantation of LPA, Tracheoplasty under CPB	1 month	Dead
4	2002	F	1 year	PA sling, tracheal stenosis, tracheal bronchus of RUL	Median sternotomy, Reimplantation of LPA, Tracheoplasty under CPB	9 days	Alive
5	2003	M	1 month	PA sling, tracheal stenosis	Median sternotomy, Reimplantation of LPA, Tracheoplasty under CPB	3 months	Alive
6	2004	M	7 months	PA sling, tracheal stenosis	Median sternotomy, Reimplantation of LPA, Tracheoplasty under CPB	16 months	Alive
7	2007	M	2 months	PA sling, tracheal stenosis	Median sternotomy, Reimplantation of LPA, Tracheoplasty under CPB	1 month	Alive
8	2008	M	5 months	PA sling, tracheal stenosis	Median sternotomy, Reimplantation of LPA, Tracheoplasty under CPB	1.5 months	Alive, residual tracheal stenosis, LPA stenosis
9	2008	F	2 months	PA sling, tracheal stenosis, tracheal bronchus of RUL	Median sternotomy, Reimplantation of LPA, Tracheoplasty under CPB	1 month	Alive, post op. bleeding
10	2009	F	4 months	DAA	Left thoracotomy, Division of posterior arch and PDA	3 months	Alive
11	2009	F	2 years	Right arch/left ligamentum	Right thoracotomy, Division of ligamentum arteriosum and Kommerell diverticulum	10 days	Alive
12	2009	M	2 months	PA sling, tracheal stenosis	Median sternotomy, Reimplantation of LPA, Tracheoplasty under CPB	1 month	Alive

M = male; F = female; DAA = double aortic arch; PA = pulmonary artery; RUL = right upper lobe; PDA = patent ductus arteriosus; LPA = left pulmonary artery; CPB = cardiopulmonary bypass; LOS = length of stay

sided aortic arch/left ligamentum with Kommerell's diverticulum) (Fig. 7). The Kommerell's diverticulum and ligamentum arteriosum were divided through the right thoracotomy. In this case, CTA gave the precise vascular anatomy. The right thoracotomy was optimal surgical approach, this patient recovered without any complication.

In conclusion, surgical correction of vascular ring is effective and safe. CTA can give the precise anatomic diagnosis of vascular rings, leads to good surgical planning and proper surgical approach. The pulmonary artery sling should be approached by median sternotomy. The double aortic arch should be approached by left thoracotomy.

Fig. 4 Double aortic arch (left arch dominant) with tracheo-esophageal compression.

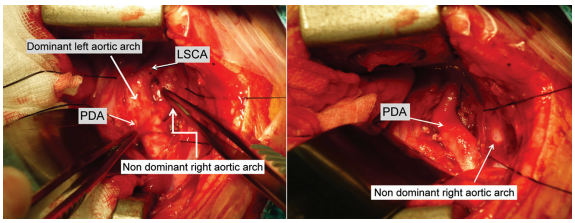

Fig. 5 The non-dominant right aortic arch and ligamentum arteriosum were divided.

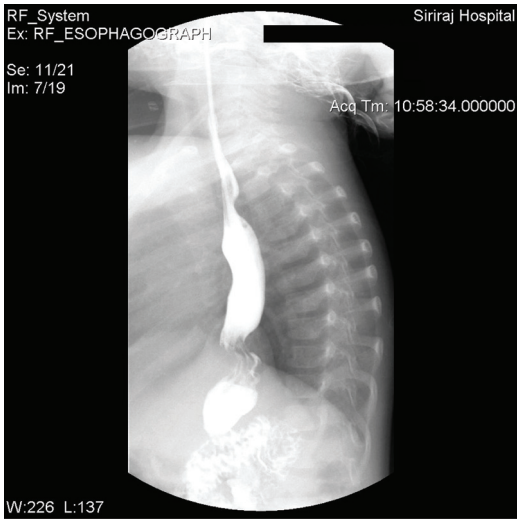

Fig. 6 Esophagography revealed posterior indentation of upper esophagus.

Whereas the right arch/left ligamentum can be approached by left or right thoracotomy depends on the anatomy.

What is already known on this topic?

In the past, the diagnosis of vascular rings can be performed by multiple tests such as chest radiography, barium esophagography, bronchoscopy, echocardiography, and cardiac catheterization, but

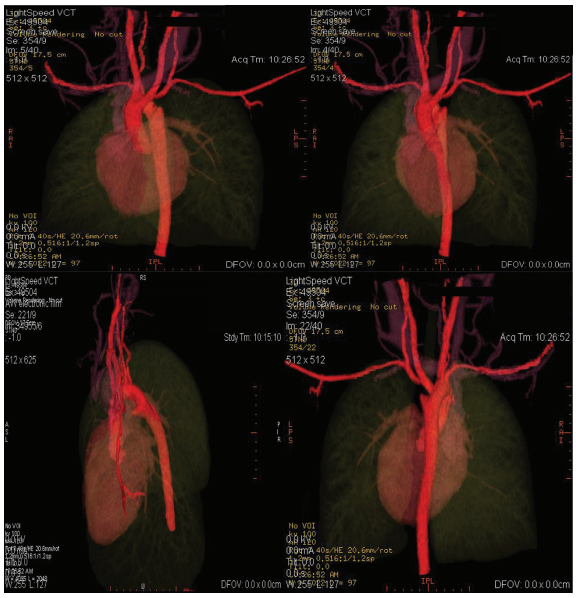

Fig. 7 Complete vascular ring (right sided aortic arch/left ligamentum with Kommerell's diverticulum).

these investigations cannot give precise anatomical diagnosis. Recently, both CTA and MRI have the ability to give accuracy that lead to good surgical planning and outcome.

What this study adds?

The result of vascular ring surgery in Thailand was unknown as it lacked case serial report. The purpose of the present study was to show the result of vascular ring surgery at Siriraj Hospital and demonstrate the usefulness of CTA for preoperative anatomical diagnosis of the vascular ring for planning of surgical correction.

Potential conflicts of interest

None.

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การผ่าตัดแก้ไข vascular ring ที่โรงพยาบาลตติยภูมิในประเทศไทย

วันชัย วงศ์กรรัตน์, สมชาย ศรียศชาติ, วรวงศ์ ศลิษฏ์อรรถกร, ธีรวิทย์ พันธุ์ชัยเพชร

ภูมิหลัง: *โรค vascular ring คือ โรคหลอดเลือดใหญ่ในทรวงอกผิดปกติแต่กำเหนิดแต่พบได้น้อยมาก ซึ่งยังไม่มีรายงานผลการผ่าตัดโรคนี้ในประเทศไทย จุดประสงค์ของการศึกษานี้คือเพื่อประเมินผลการผ่าตัดโรค vascular ring ที่โรงพยาบาลศิริราช และเพื่อแสดงประโยชน์ของเอกซเรย์คอมพิวเตอร์สำหรับการวินิจฉัยโรค vascular ring ก่อนผ่าตัดเพื่อการวางแผนผ่าตัด*

วัสดุและวิธีการ: *ทำการศึกษาผู้ป่วยโรค vascular ring ที่ได้รับการผ่าตัดที่โรงพยาบาลศิริราช ตั้งแต่ พ.ศ. 2522 ถึง พ.ศ. 2552 โดยศึกษาเกี่ยวกับการสืบค้นหาโรค การวินิจฉัย เทคนิคการผ่าตัด และผลการผ่าตัด*

ผลการศึกษา: *มีผู้ป่วยโรค vascular ring ที่ได้รับการผ่าตัด 12 ราย โดยอายุของผู้ป่วยขณะผ่าตัดมีตั้งแต่ 1 เดือน ถึง 2 ปี (ค่ามัธยฐาน 3 เดือน) มีแผลผ่าตัดผ่านทางกระดูกหน้าอก (median sternotomy) 8 ราย ผ่านทางทรวงอก (thoracotomy) ด้านซ้าย 3 ราย ผ่านทางทรวงอกด้านขวา 1 ราย หลังผ่าตัดพบมีภาวะแทรกซ้อน 2 ราย ได้แก่ ท่อหลอดลมและหลอดเลือดแดงปอดตีบ 1 ราย เลือดออกมาก 1 ราย มีผู้ป่วยเสียชีวิต 1 ราย จากปอดอักเสบ ติดเชื้อในกระแสเลือด*

สรุป: *การผ่าตัดรักษาโรค vascular ring นั้นได้ผลดีและมีความปลอดภัย โดยเอกซเรย์คอมพิวเตอร์ช่วยให้รายละเอียดทางกายวิภาคของหลอดเลือดในทรวงอกอย่างแม่นยำ ส่งผลให้สามารถวางแผนผ่าตัดได้ดีและเลือกแผลผ่าตัดได้อย่างเหมาะสม*
