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SURGICAL MANAGEMENT OF COMPLETE VASCULAR RINGS: OUR EXPERIENCE

Cardiology

Dr. Vaibhav Chugh Senior Resident, Department of Cardiothoracic Vascular Surgery. ABVIMS & Dr. R.M.L. Hospital, New Delhi, India.

Dr. Rahul Bhushan Senior Resident, Department of Cardiothoracic Vascular Surgery. ABVIMS & Dr. R.M.L. Hospital, New Delhi, India.

Dr. Narender Singh Jhajhria Professor, Department of Cardiothoracic Vascular Surgery. ABVIMS & Dr. R.M.L. Hospital, New Delhi, India.

Dr. Dhananjay Bansal Assistant Professor, Department of Cardiothoracic Vascular Surgery. ABVIMS & Dr. R.M.L. Hospital, New Delhi, India.

Dr. Palash Aiyer Professor, Department of Cardiothoracic Vascular Surgery. ABVIMS & Dr. R.M.L. Hospital, New Delhi, India.

Dr. Vijay Grover* Head & Professor, Department of Cardiothoracic Vascular Surgery. ABVIMS & Dr. R.M.L. Hospital, New Delhi, India. *Corresponding Author

Dr. V.K Gupta Principal Consultant & Professor, Department of Cardiothoracic Vascular Surgery. ABVIMS & Dr. R.M.L. Hospital, New Delhi, India.

ABSTRACT

Vascular rings are rare congenital aortic arch anomalies that may cause trachea-esophageal compressive symptoms in infants and children. It may be asymptomatic in few cases. Due to its rare occurrence and non-specific symptoms, the diagnosis is often missed, thus delaying the treatment. With this case series we share our experience of managing four cases of vascular rings over four years, with age of patients ranging from 3 months to 3 years. All cases presented in pediatric emergency with trachea-esophageal compressive symptoms. In our series we encountered 4 types of complete vascular rings- Left Pulmonary Artery (LPA) sling with Left Ligamentum (LL), Right Aortic Arch with aberrant Left Subclavian Artery from Kommerell diverticulum with LL, Right dominant Double Aortic Arch (DAA) with associated TOF, and DAA with left dominant arch. Two cases had associated cardiac anomalies, which were Atrial Septal Defect (ASD) and Tetralogy Of Fallot (TOF). Median sternotomy was done in two cases with associated cardiac anomalies and left thoracotomy was the approach for rest two cases. Single stage single incision approach was followed in all cases, with no postoperative mortality and no significant morbidity. Early diagnosis and precise planning of surgical intervention provides the best long-term outcomes.

KEYWORDS

Vascular ring, Kommerell diverticulum, LPA sling

BACKGROUND

Vascular rings result from abnormal development of the fourth and sixth paired aortic arches forming anatomically complete or incomplete vascular ring around the trachea, esophagus, or both during fetal life.[1,2] It accounts for 1 to 2 percent of congenital heart disease.[3] They are classified as complete or incomplete rings. The complete Vascular Rings include double aortic arch (DAA), and right aortic arch (RAA) with aberrant left subclavian artery (LSCA) and left ligamentum (LL). The incomplete include innominate artery compression and pulmonary artery sling. Vascular rings, though rare are important causes of trachea-esophageal compression in neonates, infants and paediatric population.[4,5] Vascular rings may remain asymptomatic in few cases.[4] Clinical manifestations of vascular rings are non-specific and incidence is low leading to reduced index of suspicion thus misleading the diagnosis to either pulmonary disorder or gastro-esophageal reflux disease.[6,7] This results in delay in definitive treatment thus increasing the morbidity of patients.

AIM

The main aim of this study is to discuss our experience in managing the cases of trachea-esophageal compression associated with vascular rings. We intend to present the rare cases of complete vascular rings and highlight the importance of early precise diagnosis and planned surgical approach to have the best possible outcomes.

MATERIAL AND METHODS

We encountered four cases of complete vascular ring at our institution between July 2016 to June 2020. Their basic demographic data, type of abnormalities, clinical symptoms and signs, imaging, endoscopic findings and associated other malformations were recorded for further analysis. Diagnostic procedures performed were chest X-ray, transthoracic echocardiography(TTE), barium swallow, cardiac catheterization, bronchoscopy and computerized tomographic (CT) scan. Associated cardiovascular anomalies, treatment, postoperative complications and follow-up were also recorded.

Case 1. A 4 months old male child, weighing 5 kg, a known case of Gastro esophageal reflux disease confirmed on Tc99m sulphur colloid study performed outside our institution, was admitted with complaints of frequent chest infections, dyspnoea and stridor for which he was intubated and mechanically ventilated twice. Pneumonic patch in right upper lobe gradually resolved. Echocardiography revealed 5.5mm ostium secundum Atrial septal defect (ASD) and uncertain pulmonary confluence. Cardiothoracic CT was performed which revealed Left Pulmonary Artery (LPA) sling with Left Ligamentum (LL), with tracheal stenosis [Figure 1]

Operative Procedure: Median sternotomy was done. LPA was arising from Right Pulmonary Artery (RPA) and traversing behind trachea causing compression. Cardiopulmonary bypass was instituted using aortobicaval cannulation and heart arrested. Direct closure of ASD was done along with ductus arteriosus ligation and division. This was followed by LPA to Main Pulmonary Artery (MPA) translocation.
Post-operative recovery was uneventful with complete resolution of respiratory symptoms.

Case 2: A 3 month old male child weighing 3kg presented with cough, dyspnoea and stridor since first week of life with history of repeated hospitalisations. Chest auscultation revealed no murmur. Chest radiography showed RAA with mild tracheal deviation to the left and clear lung fields. Bronchoscopy identified external compression of lower part of trachea from right side with no evidence of stricture. CT Cardiac angiogram revealed prominent RAA with aberrant LSCA arising from Kommerells diverticulum and LL arising from tip of diverticulum joining the LPA origin forming complete vascular ring causing significant esophageal and lower tracheal compression.

Operative Procedure: A left thoracotomy approach was taken. LL was divided. Kommerells diverticulum was found to be away from non vascular structures after the fibrous tissue was dissected, so

diverticulopexy to left rib was done as against the conventional approach of division of the diverticulum and reimplantation of the LSCA. [Figure 2,3]

Case 3. A 3 year old female child presented with tachypnea and central cyanosis. Chest X-ray showed tracheal indentations along with features suggestive of Tetralogy of Fallot (TOF). Echocardiography confirmed the diagnoses of TOF with DAA and bilateral superior vena cavae. CT Angiography showed right dominant DAA with right descending thoracic aorta (DTA) leading to trachea-oesophageal compression. (Figure 4)

Operative Procedure: Intracardiac repair with division of non-dominant left arch was planned. Median sternotomy was done. After looping proximal left arch and proximal left subclavian artery, ductus was divided. Cardiopulmonary bypass was instituted using aortobicaval cannulation. The length of left sided distal arch accessible from this approach for division was short, so we performed LSCA blalocking maneuver. In this technique, after doubly snaring the subclavian artery, the arch clamp was shifted proximal to the left subclavian origin. Then the arch distal to LSCA was divided and ends oversewed with prolene double layer running sutures. Fibrous tissue around trachea and oesophagus was dissected and divided. Left superior vena cava was then cannulated and Intra-cardiac repair for TOF was done in standard manner. (Figure 5,6)

Post-operative recovery was uneventful and after three years of follow-up patient is doing well.

Case 4: A 6 months old male child weighing 4kg, presented with dyspnea, dysphagia and repeated hospitalizations since second week of life. Chest auscultation revealed conducted sounds with no murmur. Bronchoscopy revealed near total lower tracheal narrowing around 1.5cm above carina. Cardiac CT angiogram confirmed left dominant DAA with left DTA and left patent ductus arteriosus (PDA). Right arch was found looping behind the esophagus with Right Common Carotid Artery (RCCA) and Right Subclavian Artery (RSCA) arising from proximal right arch. Thus leading to tracheo-oesophageal compression. (Figure 7)

Operative Procedure: Left postero-lateral thoracotomy was done in view of left ductus and left sided DTA. PDA was looped, ligated and divided. Both arches were looped along with LSCA. Right or the posterior arch was doubly clamped proximal to its confluence with left or anterior arch and divided, Both its ends were over sewed with double layer prolene continuous sutures. Fibrous tissue around trachea and esophagus was dissected and divided. A small iatrogenic rent in esophagus was noted while performing this fibrous tissue dissection and was repaired directly with vicryl 4-0 suture. (Figure 8,9)

Postoperative recovery was uneventful and feeding tube was removed at 9th post-op day after barium study confirmed no leak in esophagus. Patient was discharged on postoperative day 11.

RESULTS

Imaging modalities like CT and echocardiography helped in formulating definitive diagnosis and planning surgical technique. All cases were approached via a single incision. Associated cardiac anomalies with vascular ring can be tackled as one stage procedure via single incision only. All patients were evaluated periodically in postoperative period. Follow-up CT scan revealed marked diminution in tracheo-oesophageal compression. Bronchoscopy confirmed the findings. Symptoms of compression like stridor, dyspnoea and dysphagia nearly resolved. Barium swallow was done in patient with iatrogenic esophageal rent showing no leak. Follow-up echocardiography was done in two patients with associated cardiac anomaly, which revealed adequate repair.

DISCUSSION

Malformations of the aortic arch are rare, but they can be diagnosed early if we keep high degree of clinical suspicion based on clinical manifestations. In the present series, all the four patients presented with respiratory symptoms, which appeared few months after birth in 3 cases and later on in 1 case. Children with symptoms of upper respiratory obstruction must be investigated early, in order to identify the tracheal compression. Early diagnosis of aortic arch anomalies can be achieved if the paediatrician or attending doctor consider these among differential diagnosis.[8] Bronchoscopy is very useful investigation for the diagnosis of vascular rings.[9] For detailed anatomy of the vascular ring and to diagnose associated tracheal pathology, CT scan is best imaging modality.[10,11] Associated cardiac malformations are a known entity and can be detected with the help of echocardiogram.[12] In the present series, one patient presented with the diagnoses of TOF with DAA and bilateral superior vena cavae. Another showed ASD with LPA sling. DAA is the most common type of complete vascular ring encountered in clinical practice.[13] TOF and transposition of great arteries (TGA) are the frequently associated congenital heart lesions with DAA.[14] Therefore, we recommend echocardiogram as the useful adjunct during management of vascular rings.

Symptomatic PA slings causing life-threatening tracheal stenosis may require tracheal surgery, thus increasing the morbidity in post-operative period.[15] In case of a Kommerrells diverticulum it is suggested to completely excise the diverticulum as it may develop aneurysm later in life causing esophageal compression.[16] However, in one of our cases, diverticulopexy to the left rib relieved the symptoms with no complaints even after follow-up of 1 year. Various authors have suggested either staged or single stage dual incision approach for vascular rings with intracardiac lesions. A study by Pankaj B et al. advocated staged repair of TOF with DAA with intracardiac repair by sternotomy first followed by DAA corrective surgery through postero-lateral thoracotomy later.[14] In contrast to this approach, study by Talwar et al. advised the repair of DAA *via* postero-lateral thoracotomy and the intracardiac repair *via* sternotomy in the single setting.[17] We emphasize that vascular rings with intracardiac lesions can be corrected as one stage procedure via single incision approach.[18] The difficulty in division of the non-dominant arch from the midline can be safely undertaken using our LSCA blalocking technique of modified vascular clamp positioning.[18] In the postoperative period, respiratory physiotherapy and antibiotic prophylaxis are of great importance for the long-term treatment of these patients.

CONCLUSION

Early diagnosis, detailed investigations and precise preoperative planning helps formulating the optimal surgical approach for each individual patient and getting the best long term outcomes. Kommerrells diverticulopexy with fibrous tissue dissection may be adequate enough to relieve the compression. One stage procedure via single incision approach is possible for DAA with right dominant arch with associated TOF. Left postero-lateral thoracotomy may be the best approach for DAA with left dominant arch with left ligamentum. Symptomatic LPA sling should be addressed earliest possible with LPA translocation over MPA.

CONFLICT OF INTEREST:
None declared.

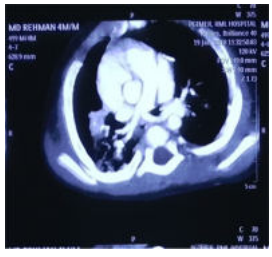

Figure 1: CECT showing LPA sling.

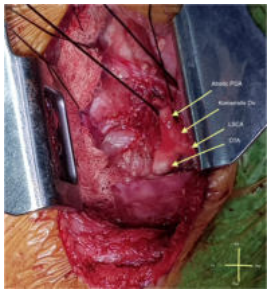

Figure 2: Intra-operative view of Kommerell diverticulum with atretic PDA, LSCA and DTA.

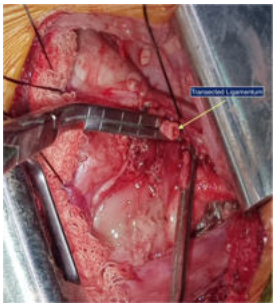

Figure 3: Intra-operative view showing transected Ligamentum

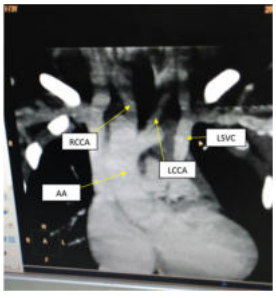

Figure 4: CECT showing the origin of the 2 common carotid arteries from 2 arches and Left superior vena cava.

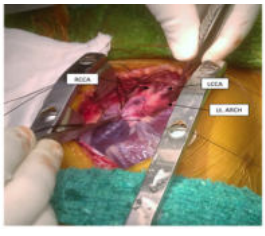

Figure 5: Intra-operative view of the DAA with the respective common carotid arteries .

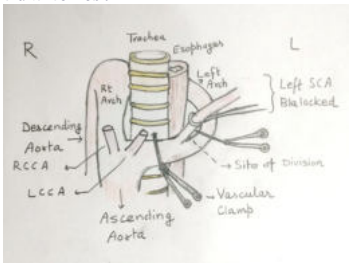

Figure 6: Schematic diagram to show the surgical strategy to clamp and divide the non dominant left arch.

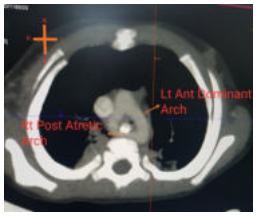

Figure 7: CECT image showing complete vascular ring with atretic right posterior arch looping around trachea and esophagus.

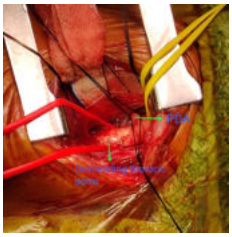

Figure 8: Intraoperative image showing looped ductus before ligation and transection.

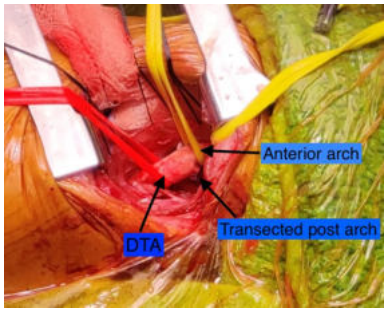

Figure 9: Intraoperative image showing transected posterior arch.

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