Complete Correction of Kommerel's Diverticulum with a Modified Technique Under the Shadow of Covid-19 Pneumonia

Kommerel Divertikülünün Modifiye Teknik ile Düzeltilmesi: Covid-19 Pnömonisinin Gölgesinde Kommerel Divertikülünün Düzeltilmesi

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Öz

Kommerell Divertikülü, aberran sağ subklavyen arterin proksimal kısmının inen aorta da çıkış seviyesinde fokal genişlemesi olarak görülen, aortun nadir bir konjenital anomalisidir. Posterior sol dördüncü aortik arkın embriyolojik bir kalıntısıdır. Kommerell divertikülü doğum anında, emzirme döneminde veya yetişkinlik döneminde ortaya çıkabilen trakea veya özofagus kompresyonuna bağlı semptomlara neden olabilir. Burada disfajilusoria şikayeti ile hastaneye başvuran, Kommerell Divertikülü ile vasküler halka tanısı konulan ve başarılı bir şekilde ameliyat edilen 5 yaşında bir hasta literatür eşliğinde bildirilmiştir.

Anahtar Kelimeler: Covid-19, Kommerel Divertikülü, Vasküler Ring

Abstract

Kommerell's Diverticulum is a rare aortic congenital anomaly, as the focal enlargement of the proximal part of the aberrant right subclavian artery at the level of its exit from the descending aorta. It is an embryological remnant of the posterior left fourth aortic arch. Kommerell`s diverticulum may cause symptoms of tracheal or esophageal compression, which they may arise at the moment of the birth, during the lactation or in the adulthood (dysphagia lusoria). Herein a 5-years-old patient who admitted to hospital with the complaint of dysphagia lusoria, diagnosed as vascular ring with Kommerell`s Diverticulum and operated successfully has been reported in the light of literature.

Keywords: Covid-19, Kommerell's Diverticulum, Vascular Ring

Introduction

Right sided aortic arch and accompanying left ligamentum arteriosum is a rare congenital anomaly seen in 0.05% - 0.1% of healthy population among vascular ring pathologies those occur as the result of congenital abnormal development of the aortic arch (1). The short segment diverticular enlargement of the left subclavian artery at the aortic outlet accompanying to this anomaly is defined as Kommerell's diverticulum (KD) (2). KD may manifest itself with respiratory and/or gastrointestinal symptoms due to tracheal and/or esophageal compression in the neonatal period and in the pediatric age group. It may also be asymptomatic until adulthood and can be detected incidentally (3).

The indications and techniques of surgical intervention for the KD are still controversial, due to the small number of cases. In KD cases detected in childhood, surgical division of the left ligamentum arteriosum, removal of the diverticulum, re-implantation of the left subclavian artery to the aorta and aortopexy are recommended to eliminate residual compression symptoms and to prevent the late aneurysm rupture and/or dissection complications due to KD (4). In adults, through a right thoracotomy, tube graft interposition is performed by removing the descending aorta under cardiopulmonary bypass (CPB), which may cause more complications and even death.

Herein, our surgical technique and results were reported in a pediatric patient who admitted to hospital with the clinic of "dysphagia lusoria" and was diagnosed as Kommerell`s Diverticulum as a result of the examinations.

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Case

5-year-old male patient, who was pre-diagnosed as double aortic arch anomaly by the fetal echocardiography performed in an external center and was followed up by pediatric cardiology until the age of 5 because his asymptomatic progress after birth, was admitted to the cardiovascular surgery department of our hospital with the increasing complaint of dysphagia lusoria in the last 1 year. The patient was examined with transthoracic echocardiography firstly with the preliminary diagnosis of esophageal compression due to double aortic arch by the pediatric cardiology department and an angiographic examination was recommended. MRI angiography revealed right sided aortic arch, left ligamentum arteriosum and KD (figure 1). Surgical correction was decided at the pediatric cardiology and cardiovascular surgery council due to esophageal compression caused by vascular ring anomaly and KD. The patient was undergoing an elective surgery after completing the

preoperative preparations. The thorax was entered through left posterolateral thoracotomy from the third intercostal space in the full right lateral position. Selective lung ventilation was not used. The left lung was retracted anteriorly and caudally, and the mediastinal pleura was opened longitudinally. Exploration was achieved with several fixed sutures on both sides of the pleural incision. The esophagus was dissected and released. The distal aortic arch, left subclavian artery, KD and descending aorta were explored by retracting the esophagus anteriorly. It was observed that the left ligamentum arteriosum originating from the pulmonary artery passed through the anterior of esophagus and poured into the descending aorta posteriorly. It was seen that the trachea and esophagus are compressed by a complete vascular ring structure formed by the right sided aortic arch (anterior/right portion), the base of the left subclavian artery originating from the KD (posterior portion), and the left ligamentum arteriosus (left portion). The ligamentum arteriosum was ligated and divided, by preserving N. Vagus and N. Phrenicus. The patient was heparinized (100 IU/kg). KD was excised by using a side clamp, without the need for CPB. The KD was at the site where the left subclavian artery arises from the aorta, and the anatomical structure was continuing as the subclavian artery after the diverticulum. Therefore, after the excision of KD, the descenden aorta was repaired with continuous double suture technique and the left subclavian artery was re-implanted into the descending aorta. All pressure on the esophagus was removed by performing aortopexy to the lateral wall of the thorax with prolene sutures. Following the bleeding control, a thorax drain was placed and the chest wall was closed in accordance with the anatomy. After the operation, the patient was taken to the cardiovascular surgery intensive care unit and extubated at post-operative 4th hour. The patient's physical examination, blood tests and vital signs were stable in the first 2 days after the operation. On the post-operative 2nd day, chylothorax was occurred in our patient. Somatostatin was added to the treatment. Due to the patient had a fever of 39 °C on the 3rd postoperative day a new Covid-19 PCR test was performed despite of the Covid-19 PCR test results obtained at the time of his hospitalization and 1 day before the operation were negative. Unfortunately, the new Covid-19 PCR test result was positive and in the chest X-ray, there was an infiltrative image compatible with bilateral Covid-19 pneumonia. In his blood tests, despite the other parameters were within the normal range; as which were in a way that supported the diagnosis of Covid-19 infection, WBC was 4930 mcl (neutrophil dominance of 82%), Ferritin was 489 ng/dl, D-dimer was 3612 ng/ml, ProBNP was 827 ng/ml, Procalcitonin was 0.5 ng/ml, CRP was 71.6mg/l and Fibrinogen was 413 mg/dl. Therefore, the patient was transferred to the pediatric Covid-19 intensive care unit for isolation. Paracetamol 15 mg/kg IV. 4 times a day, Lopinavir-Ritonavir 200 mg-50mg PO twice a day, methylprednisolone 2 mg/kg/day IV in 2 doses and acetylsalicylic acid 5 mg/kg PO once a day; was added to the patient's current treatment. In addition, appropriate antibiotic treatment was added to the treatment, in terms of hospital-acquired pneumonia superinfection. Blood, urine and sputum cultures were sent microbiology laboratory. The patient was started on high-flow oxygen therapy (36 l/min, FiO_2 50%) due to the need for oxygen. On the 3rd day of his transfer to the pediatric Covid-19 intensive care unit, the patient's fever regressed, oxygen requirement, CRP and Procalcitonin values decreased; therefore, methylprednisolone treatment was stopped. Chylothorax has completely discontinued on the 6th day of his transfer (postoperative 9th day). Four days after the steroid treatment was stopped, while he was receiving 6-8 l/min oxygen support with a simple mask, the patient had a fever of 38.8 °C again and his oxygen requirement increased but there was no significant increase infiltration image in the chest X-ray. The patient was started on high-flow oxygen therapy again (36 l/min, FiO_2 50%). Colimycin and Amphotericin-B were added to the treatment.

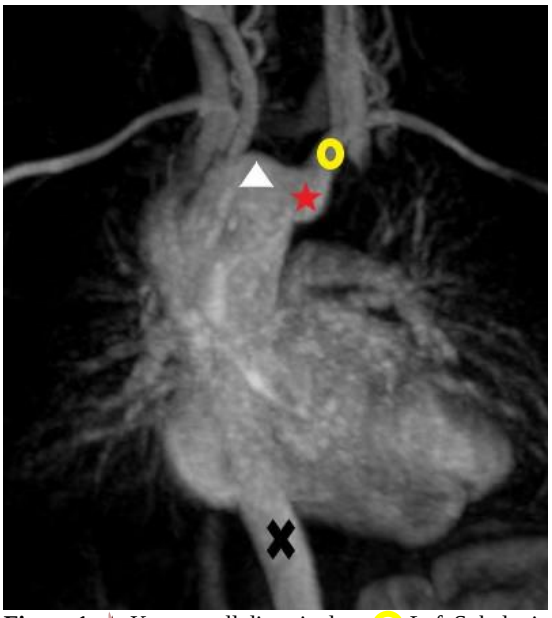

Figure 1. ★:Kommerell diverticulum ○:Left Subclavian Artery, X:Descenden aorta, △ :right sided aortic arch

The patient was consulted again with pediatric cardiology and evaluated with transthoracic echocardiography. In echocardiography; the cardiac functions were normal and no mural thrombus was seen. Pediatric Inflammatory Multisystem Syndrome was considered in the patient whose Ferritin was 911 ng/dl, IL-6 was 8.22 pg/ml and did not respond to antibiotherapy, although CRP and Procalcitonin values did not increase. The patient was started on methylprednisolone treatment again.

Two days after methylprednisolone was started, the patient's fever regressed and his oxygen requirement decreased. However, this time, methylprednisolone was continued for 3 weeks until the Ferritin value decreased. On the post-operative 32nd day, the patient was taken to the pediatric ward, with negative Covid-19 PCR test and clinic recovery. On the post-operative 35th day, he was discharged at the end of an uneventful service follow-up**.**

Discussion

The right sided aortic arch and the accompanying left ligamentum arteriosum, is one of the two most common true vascular rings. The left subclavian artery and left ligamentum arteriosum originate from the KD, the embryological remnant of the 4th dorsal aortic arch. This structure may contribute to the compression of the vascular ring anomaly to trachea and esophagus (1). In 5-10% of these patients, congenital heart diseases such as Ventricular Septal Defect, Tetralogy of Fallot, Pulmonary Stenosis and Patent Ductus Arteriosus may accompany (5). In our case, no accompanying congenital cardiac anomaly was detected in transthoracic echocardiography and MRI angiography examinations.

Pediatric patients with vascular ring pathologies, usually admit to the hospital with the complaints of recurrent respiratory tract infections, stridor, cough, shortness of breath or difficulty in swallowing (6). It is seen that approximately one third of these patients have tracheal compression, and more rarely, 5-15% of them have esophageal compression (7). Our patient also had complaints of difficulty swallowing solid foods and weight loss, but he did not have any respiratory complaints. In symptomatic patients with suspected vascular ring anomaly on X-rays, vascular structures and vascular ring anomalies can be detected by transthoracic echocardiography, but CT or MRI angiography examinations are used for clear evaluation of aortic arch anatomy and surgical planning (8). Because of the advantages of MRI angiography such as the use of iodine-free contrast, the absence of ionizing radiation, and its ability to define the cardiovascular morphology, vascular ring, trachea, and esophagus relationship, it was our first line choice for imaging. In literature, various surgical approaches including right or left thoracotomy, median sternotomy and thoracotomy-sternotomy combination of have been used in pediatric and adult patients those have vascular ring pathologies accompanied with KD. Complex aortic surgery procedures using CPB in adult patients have been reported with positive clinical results (9). However, the procedure of graft placement with excision of the descending aorta under cardiopulmonary bypass in adults may cause more complications than the procedure performed in pediatric patients without CPB and generally results in mortality. In pediatric patients, since the tissues are more elastic, the tissues can come together after KD excision, so there is no need for the use of grafts. In addition, better results are obtained because CPB is not used in the operations. Two different techniques have been described for KD, in pediatric cases. In the first of these techniques, in addition to the division of the left ligamentum arteriosum, aortopexy was performed in order to prevent residual compression symptoms that may develop in the long term after surgery. In the second technique, which Backer et al. described and studied the long-term results, the resection of the KD and re-implantation of the left subclavian artery to the left carotid artery is performed in addition to the division of the left ligamentum arteriosum. With this second technique, long-term dissection and aneurysm rupture complications due to KD can be prevented (4,10). The presence of cystic medial necrosis in the histopathological examination of KD samples, taken from patients even younger than 1 year old reveals the risks of aortic dissection, aneurysm development and its rupture in the long term (11). Therefore, we are of the opinion that KD resection should be performed in these patients. In our case, in addition to the surgical technique suggested by Backer et al., we also applied aortopexy to remove the aorta from the esophagus and trachea as much as possible. In this way, we aimed to minimize the effects of compression that may develop in the long-term after the operation in children growing up, and to eliminate the need for reoperation. Also, we re-implanted the subclavian artery to the descending aorta using a side clamp without increasing the complexity of the intrathoracic surgical procedure in order to avoid neurological complications that may occur during anastomosis of the left subclavian artery to the left carotid artery. No neurological complication occurred in our patient.

All patients who will undergo elective cardiac surgery in our clinic are tested for Covid-19 PCR at the time of first hospitalization and 24 hours before the operation. In the post-operative period, the patients are followed up in the cardiovascular surgery intensive care unit, where Covid-19 positive patients are not taken and consist of isolated boxes. All the necessary antisepsis rules are followed by all health personnel without exception. In the intensive care follow-up, the adult patients` companion is not taken, but in mandatory situations, only the mothers of the pediatric patients are taken intermittently to the intensive care unit, if her Covid-19 PCR test is negative. Although the Covid-19 PCR test performed, 1 day before the surgery was negative, he was followed in an isolated box in the intensive care follow-up and the Covid-19 PCR test of the only companion taken with him was negative; the patient had a fever on the post-operative 3rd day and the result of the Covid-19 PCR test was positive. This unexpected situation led to the fact that the patient who was operated with thoracotomy and who

already had respiratory difficulties due to it, was followed in the intensive care unit for 32 days with high-flow nasal oxygen and intermittent CPAP. This situation has also shown that there are serious problems about the sensitivity of Covid-19 PCR tests. Our patient was discharged with recovery, after the Covid-19 PCR test became negative on the post-operative 35th day.

In vascular ring pathologies developed by right sided aortic arch, left ligamentum arteriosum and KD; division of the left ligamentum arteriosum, excision of the KD and aortopexy completely eliminates the possibility of residual compression of the esophagus and trachea. In order to prevent dissection, aneurysm and rupture complications that may occur due to KD in the long term, KD resection can be performed in pediatric patients with the left thoracotomy approach and without the need for cardiopulmonary bypass. In case of anatomical suitability, anastomosis of the left subclavian artery to the descending aorta will avoid the possible neurological risks of carotic-subclavian bypass.

Written consent: A written patient consent certificate was obtained from the patient (or his legal guardian) that his medical data may be published (20/04/2021).

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