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Parsiyel Pulmoner Venöz Dönüş Anomalisi Anatomisi; Kısa Literatür Araştırması

Partial Anomalous Pulmonary Venous Return Anatomy; A Brief Lıterature Review

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ABSTRACT
Partial anomalous pulmonary venous return (PAPVR) was first described by Winslow in 1739 and is a congenital situation. Two conditions are discussed: Pulmonary veins (PV_S) incorrectly drain into the superior vena cava (SVC) or directly into the right atrium (RA). This anomaly is often seen in the right lung than left lung (2/1 or 10/1). Many radiological methods are preferred to determine the anomalous veins due to more reliable results and these methods help in better identification of the anatomy of the pulmonary vein and its tributaries. In cases requiring surgical intervention and treatment plan or treatment approach, it is important to diagnose this anomaly, to know the accompanying cardiac anomalies, and to have the chance for early treatment, as surgeons, radiologists, and clinicians should keep in mind the possibility of such anatomical variations. As a result, the awareness of the PAPVR anatomy provides to prevent vessel damage. Early surgical repair is a choice of treatment in symptomatic patients. Herein, we aimed to present detailed anatomy and embryology of the pulmonary veins and to explain and present pulmonary venous return anomaly in the light of this information with the latest studies about partial pulmonary vein return anomaly.
Keywords: Partial anomalous pulmonary venous return, cordis dilatation, anatomical variation.

ÖZET
Parsiyel pulmoner venöz dönüş anomalisi (PPVDA) ilk kez 1739 yılında Winslow tarafından tanımlanmış olup konjenital bir durumu ifade eder. İki farklı şekilde olduğu bilinmektedir: Bunlardan birisi, Vv. pulmonalium (Vena) venae cavae superior'a (VCS) direne olmaktadır. İkinci durumda ise Vv. pulmonalium doğrudan sağ atriyum'a (SA) boşalmaktadır. Bu anomali sağ akciğerde sola göre daha sıklıkla görülür (2/1 veya 10/1). Güvenli sonuç vermelerinden dolayı ven anomalilerini göstermek için pek çok radiolojik yöntem kullanılıyor. Bu yöntemler, vv.pulmonalium ve dallarının anatomisinin daha iyi tanımlanmasına yardımcı olmaktadır. Bu bölgede olabilecek anatomik varyasyonların bilinmesi klinisyen, radyolog ve cerrah açısından tedavinin planlanması ve cerrahi yaklaşım durumlarında istenmeyen komplikasyonları azaltmada ve önlemede yardımcı olacaktır. PPVDA anatomisinin bilinmesi damar hasarını önlemeyi sağlar. Ayrıca, PAPVR nadir görülen bir anomalidir ve sıklıkla sessiz seyreder. Bu nedenle Semptomatik hastalarda erken dönemde cerrahi onarım uygun bir tedavi yaklaşımı olmaktadır. Bu yazıda pulmoner venlerin detaylı anatomisi ve embriyolojisini sunmayı ve bu bilgiler ışığında parsiyel pulmoner venöz dönüş anomalisi ile ilgili son çalışmalarla birlikte pulmoner venöz dönüş anomalisini açıklamayı amaçladık.
Anahtar kelimeler: Kısmi pulmoner venöz dönüş anomalisi, kalp dilatasyonu, anatomik varyasyon.

Introduction

Anomalous pulmonary venous drainage was described by Winslow in 1739, firstly and this was the first autopsy description also. Charles T. Dotter diagnosed the PAPVR in two cases of adult males (27 years and 41 years) by angiocardiography and cardiac catheterization and publicated in 1949[1-5]. PAPVR can often be associated with a defect in the atrial septum, and encountered the cases with intact atrial septum[2,5]. Surgical repair was described by Kirklin et al in entitled "Treatment of anomalous pulmonary venous connection in association with interatrial communication" in 1956[5]. The PV either drain into the systemic venous circulation or right atrium in PAPVD[2,6,7]. Also, this anomaly is an uncommon cardiovascular congenital anomalous situation and it occurs due to failure of regression of primitive lung drainage. So, it is discussed partial change in the venous connection of the oxygenated blood of the pulmonary vein to the left atrium. This situation leads to left to right shunt (LRS)[2,4,6,8,9]. Additionally, the PAPVR's drainage place can show

the differences; the most seen type is supracardial (63%)[8]. The prevalence of PAPVR can be estimated as 0.1 or 0.2% in computed tomography (CT) scans and in autopsy cases, it can be reported as 0.4 or 0.7% [2,4,8,9]. 10% of these are of left side. Usually, right upper lobe vein runs out to the right atrium. If this anomaly occurs in the left upper lobe, left upper pulmonary vein flows to a vertical vein and followed by brachiocephalic vein. In adults, PAPVR mostly develops in the left upper lobe followed by the right upper, and drain into the left brachiocephalic vein and the superior vena cava, in these upper lobes, respectively[10,11].

Herein, we aimed to present detailed anatomy and embryology of the PV, and to explain and present PAPVR in the light of this information with the latest studies about partial pulmonary vein return anomaly.

Anatomy and clinical significance

The lung veins, which are the only veins, called the pulmonary veins, are blood vessels. They transfer freshly and higher oxygenated blood from the lungs to the heart's left atria. The veins are part of both the pulmonary circulatory system and respiratory system. The pulmonary veins originate from individual alveoli within the lung as capillary vessels which meet the interlobar pulmonary veins ($ILPV_S$). The $ILPV_S$ create the subsegmental veins which may drain multiple segments of the same lobe of the lung. The subsegmental veins form a confluence to the PV_S. The PV_S also contain peripheral drainage from the deoxygenated blood bronchial veins[12-16]. There are four pulmonary veins. The left superior pulmonary vein, and inferior pulmonary vein drain the left upper lobe-lingula, and the left lower lobe, respectively. The right superior pulmonary vein drains the right upper and middle lobe, while the right inferior pulmonary vein drains the right lower lobe. The PV_S are extremely vascular. Some clinical conditions such as cystic fibrosis, tuberculosis, emphysema, benign tumor, or malignancy require a surgical procedure of the lung, including a segmentectomy, lobectomy, or transplant. If a surgical procedure in this anatomical region it is necessary to be careful not to create damage. The PV_S' innervation is provided by the pulmonary plexus. There is a clinical significance of pulmonary veins due to congestion or anatomical malformation. One of the anatomical malformations is a PAPVR, and the other is a more severe form, total anomalous pulmonary venous return (TAPVR). It may be related to an isolated finding or atrial septal defect (ASD) or Schimitar or Turner syndromes. In TAPVR, all PV_S drain into the right atrium, causing cyanosis. TAPVR needed surgical correction due to a higher mortality prognosis. Another diseases associated with abnormal veins are alveolar-capillary dysplasia with misaligned PV_S and right PV_S'agenesis[12-15].

Schimitar syndrome is a special form of this anomaly. PV_S run out to IVC, right atrium, coranary sinus, azygos vein, hepatic portal vein or hepatic veins in this form. Right lung or right pulmonary artery may be hypoplastic or muscle skeletal anomalies, and same sided diaphragma with congenital heart diseases. Clinical findings'severity are depend on complication such as presence the number and localization of anormal veins, ratio of left to right shunt (LRS), ASD cause or size, and infection. The cases which less than half of the lung volume enters the systemic circulation is asymptomatic. PAPVR usualy is diagnosed in advanced stages. There is no cyanosis. If the anomaly are asymptomatic in childhood, volume overload may develop due to LRS. This situation leads to fatigue, or difficulty in breathing[17-19].

Embryology

During embryonic development, the lung buds arise from the foregut and drain via the splanchnic plexus (PS) into the primordial systemic veins such as cardinal (CV_S) and umbilicovitelline (UVV_S). While the CV_S differentiate into the SVC, and CS, the UVV_S develops into the IVC, venous duct, and portal vein. After, caudal and cranial outpouchings begin to develop in the sinoatrial region of the primitive atrium. The caudal portion eventually regresses, whereas the cranial portion develops into the common pulmonary vein (CPV) and extends toward the lung buds. At approximately day 28 of gestation, the CPV combines the pulmonary portion of the PS and allows to flow of pulmonary blood into the heart. Four primitive PV_S are formed, in association with the CV_S (superior), and UVV_S (inferior) on the two sides. Then, the CPV is included to the left atrial wall with obliteration of pulmonary-splanchnic connections, leaving 4 independent PV_S directly entering the left atrium[3]. The disruptions in lateral pathways or regression in the connections between the pulmonary vascular bed and the systemic venous system result in persistent PAPVR, or TAPVR into the systemic venous circulation[2,7].

Material and Methods

The search was performed in the electronic database PUBMED (from 1949 to January 2023). A search strategy was developed using the keywords in the MeSH index. In searches, the words "partial anomalous pulmonary venous return", "cordis dilatation", "anatomical variation" were searched in English. Inclusion criteria for this review: Case reports, classical anatomy resources, and reviews of partial anomalous pulmonary venous returns were considered. Studies performed on total pulmonary venous return anomaly were excluded. The following steps were performed in order to select the appropriate studies: First, the specified database was searched in English. In the second stage, the headlines were read and the unsuitable ones were excluded. The abstracts were read and those who were not eligible were excluded. At the last stage, full-text articles were read. All steps were performed independently by two evaluators and the proper 19 articles were reached. In the evaluation of the selected articles, the study was planned considering the clarity in the research design and objectives, the definition and rationale of the comparative method, the selected samples, partial venous return anomaly anatomy, embryology, etiology, and prevalence. Case reports were analyzed according to date and especially, current studies of recent years were selected

Discussion

Cardiac Magnetic Rezonans Imaging (MRI) or CT can be preferred for anomalous veins' scans, because of giving reliable results. However CT has some disadvantages such as high radiation. Especially, 3D reconstruction is safe, fast, and suitable for the identification of small branches of the right upper vein and right middle pulmonary vein preservation[3,6,9,10]. These two methods have both advantages and disadvantages compared to each other. MRI is preferred due to no contrast injection, and no ionizing radiation exposure[9,10].

PAPVR is a rare congenital heart defect syndrome and the prevalence is 0.1%-0.4% in (adults)[9,10]. It is defined as the drainage of one or more PV_S to the RA, or systemic circulation[3]. Sometimes, PAPVR may be asymptomatic and recognized in significant pulmonary hypertension's presence[6]. An anomalous vein prefers to the nearest systemic vein, or the RA. On the right side, anomalous veins mostly can prefer to drain into the SVC, IVC, RA, CS, azygos vein, portal vein, or hepatic vein. On the left side, mostly drains into the left brachiocephalic vein, CS, or hemi-azygos vein. In addition, drainage via the subclavian or subdiaphragmatic veins is possible[3,6]. Also, it is usually asymptomatic and detected incidentally. However, cases having a higher degree of left to right shunt can lead to pulmonary hypertension and right ventricular volume overload including dyspnea or arrhythmias. Symptoms and prognosis are directly proportional to shunt severity[9,10]. Pulmonary vein anomalous in the left upper lobe have drainage into the left brachiocephalic vein in adults, whereas these veins in the right upper lobe with drainage into the superior vena cava in children[10]. PAPVR involves drainage of 1 to 3 pulmonary veins into the right-sided circulation. Additionally, PAPVR is often associated with hypoplastic right lung, and ASD and leads to a left to right shunt. Sometimes, PAPVR's presence can be in different lobes, or preserved wrongly. It leads to inadequate resection, increase in the left to right shunt flow, and right-sided heart failure and the postoperative mortality risk[9]. In a case study performed by Tsang et al with an 80year male patient having secundum ASD with right ventricle dilatation and ventricle volume, and progressive dyspnea. Also, there were two anomalous. An anomalous right upper lobe pulmonary veins draining into the SVC with the rest of the PV_S draining normally into the left atrium confirming PAPVR[10].

Okur et al. reported a 34years male with PAPVR performed on CT. It was reported that the pulmonary veins draining the bilateral upper lobe drained into SVC in the right side instead of the left atrium, in the left side drained into the brachiocephalic vein via the vertical vein. Also, there was no cardiac anomalies in CT images[3]. Pendela et al studied the four cases with PAPVR aged 63 year female, 51 year male, 42 year female and 66 year male, respectively. Anamolous veins arise from the right upper lobe, and drain into the SVC, and were seen in ¾ of the cases, whereas, anomalous veins arise from left upper lobe, and drain into left brachiocephalic vein were seen in 51 year male subject case. There were sinus venosus ASD in two cases (a female patient aged 42 year and a male patient aged 66 year). The other cases did not show any defect[6].

In a study in which 37 patients were operated for PAPVR, while anormal pulmonary veins of 13 patients and 23 patients usually drain into superior vena cava and, right atrium, respectively, one patient's anormal left pulmonary vein drain into the coranary sinus. Also, ASD with sinus venosus was seen in 22 patients, whereas fossa ovalis ASD were found 11 patients. On the contrary, ASD was not seen in 4 patients. In these cases, the most seen symptom was tiredness and shorthness of breath. In any of the patients having PAPVR, pulmonary vein obstruction did not develop[5]. Silawal et al studied an 88-year-old female having PAPVR (supracardial type) in combination with the annular pancreas, and a persistent umbilical vein. The pulmonary anomaly comprised the aberrant left superior pulmonary vein connecting the superior lobe of the lung with the left brachiocephalic vein resulting in a left to right shunt. An annular pancreas without any signs causing duodenal compression was additionally found. The author reported that the identification of anomaly or anatomical variations presence, and combination is essential fort he elimination of risks[8].

El Kersh et al performed a study of five case series about a PAPVR: in a female aged 42 year, anomalous vein arises from the right upper and the right middle PV_S and drain into the SVC and the RA junction. Also, sinus venous ASD were present; in a female aged 37 years, anomalous vein arises from left pulmonary vein and drain into left innominate vein at the upper mediastinum. This case had no any other anomalies. In the female patient aged 20 years, sinus venosus ASD and pulmonary arterial hypertension (PAH) were present. Additionally, anomalous veins arise from the posterior segment of (Right upper lobe) RUL, anterior and apical segment of RUL, medial segment of RML, and lateral segment of RML. Furthermore, anomalous veins drain into the SVC at the level of the carina, lateral aspect of SVC just above RA, RA at the cavoatrial junction, and superior vena cava, respectively. The other case was male and age was 21 years. An anomalous vein arises from RUL and drains into the superior vena cava. Interatrial septal defect was PFO. In the female patient aged 66 years, anomalous veins arise from RUL and drain into SVC. The other complications were no seen[2].

In a CT study of Dutta and Zuiderveld's conducted with a male aged 61years who had many complaints such as high cholesterol, impaired fasting glucose, and obesity and difficulty in breathing, chest pain and spreading to the left shoulder. The patient had a left upper lobe pulmonary vein draining into a vertical vein. It then run into the brachiocephalic vein. This is a sign of left upper lobe PAPV. At this stage, cardiac anomaly or anormal pulmonary findings were no noted. For this reason, no surgical intervention was recommended to this patient. Additionally, it was reported in this study that this was a incidental case. Approch of the left sided PAPVR is decided according to severity of disease or findings. If 50% or more of the pulmonary blood flow returns through an incorrect connection, it is considered clinically significant, and surgery is consider for symptomatic patients who has tricuspid regurgitation, or right ventricular enlargement[11].

In summary, the awareness of the PAPVR anatomy provides to prevent vessel damage. This becomes even more essential in identification of anomaly or anatomical variations presence, and combination for the elimination of risks. So, the detailed anatomy or variations of the pulmonary veins are required due to the great topographical and clinical significance in terms of surgical and radiological procedures. Additionally, in cases requiring surgical intervention and treatment plan or treatment approach, it is important to diagnose this anomaly, to know the accompanying cardiac anomalies, and to have the chance for early treatment, as surgeons, radiologists, and clinicians should keep in mind the possibility of such anatomical variations. Management or approach to this anomaly or anotomical variation is decided according to the severity of disease.

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