Anomalous left superior pulmonary vein draining into the left brachiocephalic trunk: case report

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SUMMARY

Partial anomalous pulmonary venous connection characterizes a direct connection between the pulmonary veins and the right atrium or venous system. This condition is physiologically explained in the recirculation of oxygenated blood through pulmonary vasculature to the arterial system. Patients who have this condition exclusively are usually asymptomatic. During a routine human anatomy dissection, a vein which presented a variable path near the left lung hilum was observed. Following the dissection to observe its path, it was identified as the left upper pulmonary vein. However, its drainage was in the left brachiocephalic vein, which described a pattern of anatomical variation. In the present case, as there is only one abnormality present in the left upper pulmonary vein, it was suggested that the patient was asymptomatic, as the characteristic had only been observed post mortem by chance, with no correction performed. Raising awareness of this condition can help medical students around the world to diagnose it.

Key words: Case report – Anomalous vein – Anatomic variation – Pulmonary vein

INTRODUCTION

Partial Anomalous Pulmonary Venous Connection (PAPVC) is a congenital cardiac defect which characterizes a direct connection between the pulmonary veins and the right atrium or venous system (or its tributaries) (Sahay et al., 2012). If it affects all the pulmonary veins, it is called Total Anomalous Pulmonary Venous Connection (TAPVC) and it is classified according to Darling et al. (1957) as: supradiaphragmatic (supracardiac or type 1 and cardiac or type II) and infradiaphragmatic (or type III). In general, all the pulmonary veins with anomalous insertion are implanted in the same place in a person.

According to Spratt (2016), anomalous connections between the pulmonary and systemic venous system were first recognized by Winslow (1739), who collected 106 cases.

Partial anomalous connection of the right pulmonary veins is 10 times more frequent than that of the left pulmonary veins (Snellen et al., 1968), which is an asymmetrical occurrence caused by frequent drainage of some of the right pulmonary veins into the junctional area between the right atrium and the superior vena cava (SVC) in the presence of normal left pulmonary veins, and the complete absence of isolated left

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pulmonary venous connection to the right atrium. The prevalence of pulmonary vein anomalies is 0.2% to 0.7% (Specht and Brown, 1953; Haramati et al., 2003).

Abnormal connection of solitary pulmonary veins was always affected to the most proximal venous structure among the four possible derived from the main embryonic channels (SVC and inferior vena cava on the right side and left SVC and coronary sinus on the left side). Common pulmonary veins from one lung also drained in accordance with this proximity rule, if this may also be taken to apply to the drainage of right pulmonary veins into the right atrium. The one exception was the drainage of all right pulmonary veins into the portal venous system (Snellen et al., 1968).

PAPVC is physiologically explained in the recirculation of oxygenated blood through pulmonary vasculature to the arterial system, which is called left-to-right shunting. In patients with only this condition (with no other cardiac alterations), its effects depend on the proportion of anomalous drainage compared to total pulmonary venous recurrence, but patients are usually asymptomatic. Its consequences depend on the following factors: number and size of the variant veins involved; pulmonary segments or lobes where the alteration is originated (difference in blood distribution for each lobe) (Ward and Mullins, 1998; Talner, 1998).

These unusual connections between the pulmonary veins and many other adversely inappropriate neighboring veins are explicable only by the very early embryonic bed to form the pulmonary veins leading to the left atrium, the primitive connections with stages in which the developing foregut, trachea, and lung buds are supplied by connections through the loose mesenchyme and communicate with the primitive cardinal veins in several places.

With the development of certain channels in this primitive vascular bed to form the pulmonary veins leading to the left atrium, the primitive connections with the cardinal veins usually disappear. The unusual development of one or more of them and its retention by the adult derivative of the particular part of the cardinal system involved are probably the ones that are most responsible for the occurrence of these unusual pulmonary connections (Keith et al., 1954; Steinberg and Finby, 1956).

It is our goal to increase the awareness of the anatomical and physiological repercussions of PAPVC, because it can help medical students to diagnose it in the future.

CASE REPORT

In a routine dissection for the improvement of anatomical parts, a cadaveric anatomical piece of the neck and the chest region, which belongs to the institution's collection, was analyzed. It was possible to deduce that it was a male, by the presence of facial hair (beard and moustache). Other than that, it had no identification of age, color and origin, and it was obtained by an anonymous body donation into the 20th century – at that time, the institution did not keep identification record of the bodies. The study was started under the protocol of the ethics committee, number 12923919.8.0000.5430.

The dissection of the mediastinum began in the large vessels and the cardiac base. Subsequently, it advanced to the lower mediastinal region and pleuropulmonary cavities, emphasizing the hilum and the left pulmonary pedicle.

After the performance, a vein presenting a variable path was observed near the left lung hilum and aroused interest. Following the dissection to observe its path, it was identified as the left superior pulmonary vein (LSPV). However, the LSPV was deviating from the normal path at the left pulmonary hilum, taking on a cranial trajectory, running on the mediastinal surface of the left upper lobe, entering the mediastinum, and then draining into the left brachiocephalic vein (Fig. 1). Despite this fact, the left brachiocephalic trunk followed its normal path, joining the contralateral one and leading to the right atrium.

Evaluating the piece still, in the hilum the left lower pulmonary vein, as well as the two right pulmonary veins, had normal path leading to the left atrium. The other large vessels, bronchi and limph nodes, were dissected and observed

without finding any anatomical variations that justified the presence of the reported variation.

COMMENTS

PAPVC is a rare congenital heart condition characterized either by failure of connection or drainage of pulmonary veins (Kottayil et al., 2011). It is a condition that has an estimated incidence of 0.7% in the population (Healey, 1952). Another important fact is that it was found in 0.4-0.7% of autopsies in patients with other congenital heart diseases (Said et al., 2011; Ashrafpoor et al., 2013), and that in a recent Computed Tomography (CT) study, involving 1825 CT exams, Partial Anomalous Pulmonary Venous Connection was seen in 0.2% of adults (Haramati et al., 2003).

It is also important to note that PAPVC incidence female-to-male ratio is 2 (Senocak et al., 1993), is twice as frequent in the right lung and occurs in 10% of patients who have effects on the atrial septum (Dilman et al., 2009). It is a normally asymptomatic abnormality and it can be easily diagnosed and corrected in the first years of life due to the easy access to imaging tests. If present, the symptoms include dyspnea, fatigue, palpitations, angina and/or peripheral edema (Edwin, 2010). The symptoms depend on the number of pulmonary veins involved and the extent of fluid overload due to the left to right shunt (Brody, 1942). Spratt (2016), in the Bergman's Comprehensive Encyclopedia of Human Anatomic Variation book, cited an intraoperative finding by Savu et al. (2010) that described similar characteristics of PAPVC found in this present study. The available correction for such cases is surgical, correcting the drainage of the anomalous vein to the left atrium (Elbardissi et al., 2008; Bobylev et al., 2013). However, treatment is only indicated for symptomatic patients such as hemodynamically significant left-to-right shunt, usually presenting with right ventricle overload and recurrent lung infections due to surgical treatment of other cardiac abnormalities,

Fig. 1.- Pattern of anatomical variation found in left superior pulmonary vein drainage (drains into the left brachiocephalic vein). Abbreviations and symbols: AV: azygos vein, SVC: superior vena cava; RBV: right brachiocephalic vein, LBV: left brachiocephalic vein, LJV: left jugular vein, LSV: left subclavian vein, LSPV: left superior pulmonary vein.

assessing risks and consequences (Ward and Mullins, 1998). In the present case, as there is only one abnormality present in the left upper pulmonary vein, it is suggested that the patient was asymptomatic in life, since the characteristic had only been observed post-mortem by chance, with no correction performed.

AUTHOR'S CONTRIBUTION

D.G. Gonsalves: Project development, data collection, data analysis and manuscript writing. G.R. Ventura: Project development, data collection, data analysis and manuscript writing. R. Rissi: Project development, data analysis and manuscript editing.

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