Case Report

Unknown Partial Anomalous Pulmonary Venous Connection Causing Arterial Blood Gases During Central Venous Catheter Insertion

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Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital anomaly with a prevalence of 0.4% to 0.7% in the general population [1, 2]. Based on the location of venous drainage, anomalous pulmonary venous connections are classified into four types (supracardiac, cardiac, infracardiac, or mixed). Anomalies of pulmonary vein (PV) from the right lung are twice as common as from the left lung. Most common is a connection of the right superior PV to the right atrium or the superior vena cava and is very often (80-90%) associated with atrial septal defects of either the sinus venosus or secundum type [3-5]. The anomalous PV is reported to be right-sided in 90% of all cases and more often (2:1) in males than in females [4, 6]. No data are available to determine whether PAPVC is more common in males or females, and no associated risk factors have been identified for its development. Some patients suffer from symptoms like dyspnea, decreased exertional tolerance or peripheral edema caused by right-sided heart failure and pulmonary vascular disease [7]. Most patients with PAPVC are asymptomatic, and PAPVC is an incidental finding in patients who had undergone imaging for other indications. We report a patient with an unknown PAPVC “causing arterial blood gases” during the insertion of a central venous catheter.

Case Report

The 83-year-old patient was initially admitted to another hospital suffering from an inadequate balance with a tendency to fall, dizziness and vomiting. Because of respiratory insufficiency, the patient underwent oral intubation and respiratory therapy. Clinical symptoms and radiological findings in thoracic X-ray and cerebral CT scan revealed the diagnosis of Wallenberg syndrome and pneumonia. The
Patient was transferred to our hospital for respiratory weaning and rehabilitation two weeks later. After admission into our hospital, the patient developed fever. Therefore, the central venous catheter (CVC), initially on the right side, was changed to the left side. Function of the left jugular vein was uncomplicated. A blood gas analysis taken from the newly inserted catheter, revealed “arterial blood gases”, (Figure 1) “newly inserted central venous catheter” which were comparable to the “arterial blood gas analysis” taken from the right radial artery.

A bedside ultrasound examination confirmed the CVC (located-positioned) in the internal left jugular vein. Transesophageal echocardiography could not visualize all four pulmonary veins but showed a pulmonary flow profile in the superior caval vein. The patient underwent thoracic computed tomography (CT), verifying the catheter in the left internal jugular vein. The contrast was given through the catheter which revealed a partial anomalous venous connection of the left upper PV to the left brachiocephalic vein (Figure 2).

**Discussion**

Partial anomalous pulmonary venous connection (PAPVC) is a rare anomaly, causing a usually insignificant and asymptomatic left-to-right shunt. Right-sided PAPVCs are 10 times more often than left-sided PAPVC [7-9]. Only 3% of cases have been reported with drainage from the left lung into the brachiocephalic vein [10]. Most common is the right upper lobe vein draining into the right atrium or superior vena cava [11]. PAPVC is most commonly associated with an atrial septal defect (ASD), reported in 80-90% of cases [10]. Of these, 85% are sinus venosus type and 10-15% are secundum type [7]. An intact atrial septum, seen in our patient, is rare. Nevertheless, clinical case reports mostly report about PAPVC of the left pulmonary veins (PV) [12]. Anomalous connections in 12 patients were located in the left upper PV in 9 patients, the right upper PV in 2 patients and both upper PV in one patient [9, 12]. Only 2 of the 12 patients suffered from symptoms (dyspnea, right heart insufficiency) and underwent surgery. In 5 of the 12 patients, an abnormal position of a central venous catheter was seen. In all of them the CVC was inserted via the left jugular vein [12].

In a study of Haramati *et al.*, the prevalence of a PAPVC on CT in adults was 0.2% (4 of 1825 chest CT reports). In 23 of 29 patients (79%), the PAPVC was located on the left side [13]. Clinical significance was suggested when 50% or more of the pulmonary blood flow return anomalously, causing significant right heart volume overload, which results in dilation of the tricuspid annulus and the right ventricle leading to right heart failure and pulmonary hypertension with increased pulmonary vascular resistance [13]. Treatment is necessary for pediatric patients with anomalous blood flow return of more than 50% (Qp:Qs 1:≥1.5) [14, 15]. In adult patients, the criteria for surgical repair are less clear. Only patients who developed symptoms due to shunting or with evidence of right-sided volume overload can be considered for surgery.

In cases of impossibility to visualize all four PVs during TEE, a PAPVC should be taken into consideration [16]. Magnetic resonance tomography (MRI), if available, is an excellent noninvasive diagnostic tool to evaluate the shunt fraction and to visualize number, origin and connection of PVs [17, 18]. Computed tomography (CT), used in our patient because MRI was not available, is an equally efficient tool even though the shunt fraction cannot be quantified.

Knowledge of PAPVC is important in patients who underwent implantation of CVCs, port catheters, cardiac pacemakers, lung diseases and surgery. An impressive case report of Sakka *et al.* reports about a patient with chest trauma. Acute hypoxemia was caused by a central airway obstruction distal to the tube with bloody mucus, reaching into both main bronchi [19]. CT scan excluded pulmonary embolism and revealed an (unknown) PAPVC. The authors postulate that “likely, ventilation and consecutively perfusion of the right lung was
compromised, and this may have contributed to the acute clinical situation” [19].

Patients with unknown one-sided PAPVC may become symptomatic because of an impairment of the contralateral lung e.g. by lung disease. Surgery like a pneumectomy in patients with PAPVC could have fatal consequences because the shunt volume would dramatically increase and surgical correction of PAPVC would be necessary.

Conclusion

PAPVC is a rare congenital anomaly and should be taken into consideration in cases of “arterial blood gases” taken from a correctly inserted CVC. Most patients are asymptomatic. Adequate diagnostic tools to detect PAPVC are TEE, CT or MRI.

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