ABSTRACT

Introduction: Morphological variations involving co-existence of left atrial appendage (LAA) and patent ductus arteriosus (PDA) are extremely rare congenital anomaly. Case Report: This manuscript presents a case of a massive chicken-wing left atrial appendage associated with a patent ductus arteriosus observed during a routine dissection of a 41-year-old embalmed male cadaver. The massive LAA overlaid the left cardiac artery and associated branches and great cardiac vein. Conclusion: Cardiac and vascular dysgenesis may arise due to activation or inactivation of embryonic transcription factors by any atypical factor. The resulting anomalies may present with clinical symptoms or could be asymptomatic. Knowledge on the co-existence and developmental variations of these two anatomical structures (LAA and PDA) is of importance in clinical presentation.

Keywords: Left atrial appendage, Persistence ductus arteriosus, Variation

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INTRODUCTION

Left atrial appendage (LAA) is a flap, finger-like extension of the left atrium (LA). Earlier, it was considered as a vestigial and insignificant structure of cardiac anatomy. It was thought to have a storage function. However, recent advances have shown that LAA is actively contracting and plays a crucial role in cardiac hemodynamic [1, 2, 3] and also an endocrine organ [4]. It has been described as one of the most lethal structures in human body [5, 6] due to its association with the development of atrial fibrillation [7–10] with risk of cardio embolic complications.

Variations on the size, shape, and relation of LAA with other cardiac structures have been reported previously [5, 7, 11, 12]. However, this case reports describes a giant LAA associated with a persistent ductus arteriosus observed in a course of examination of specimens in a laboratory of gross anatomy.

With the increasing pathological association, clinical and surgical importance, and the evolution of transesophageal echocardiography and transcatheter-LAA occlusion, the information on existing morphological variations of LAA anatomy is essential for cardiologist when planning for intervention in patients with difficulties in anticoagulation treatment.
CASE REPORT

A massive LAA with atypical shape and size was observed during a routine thoracic dissection classes for undergraduate medical students, of a 41-year-old preserved adult male body with no previous history of surgical or cardiac problem.

Anatomic measurements of the LAA

The shape, length, width and number of lobes of the LAA (Figure 1) were recorded similar to previous studies on the LAA [12]. The shape and number of lobes were examined by external observation and confirmed after LAA was opened. A single flattened tubular body with a blind-ending tail sac was noted protruding from the LA and overlying the upper portion of the LV. Length was measured as the distance perpendicular from the junction of LA and LAA to the apex of the LAA tail. The average external width was also recorded.

The shape was observed to have a single lobe with an elliptical finger-like tail extension process that bend in its proximal folding backwards towards the LA and LV. The LAA base had a width of 2.6cm at the junction with the tail process. The tail had an average width of 1.8cm (taken from four different position). LAA had a total length of 9.4 cm from the base to the tail apex (Figure 1).

Relationship of the LAA and pulmonary trunk

The pulmonary artery/trunk was located superior to the LAA. The two had a contact at the junction of the base and tail portion of the LAA. At this point the LAA had an indentation (depression) which marked the end of the base and the beginning of the tail portion (Figure 2). On the superior aspect of this point on the pulmonary trunk, the patent ductus arteriosus arise to connect the descending aorta and the pulmonary trunk (Figure 2).

Relation with the coronary arteries and veins

The LAA was observed overlying the main stem of the left coronary artery, circumflex artery and proximal part of the anterior interventricular artery. It also overlies the branches arising from the circumflex artery (left marginal and anterior ventricular arteries) (Figure 1) and all the associated veins including the great cardiac vein.

Patent ductus arteriosus

Patent ductus arteriosus (PDA) was also observed to arise from the pulmonary trunk 1cm before the origin of left pulmonary artery and connects to the proximal descending aorta 1.2 cm after the origin of the left subclavian artery (Figure 2). It passes from the anterosuperior aspect of the pulmonary artery to the posteriorinferior aspect of the aorta. It measured 1cm in length and 4 mm in diameter.

DISCUSSION

A detailed knowledge of the existing variations on the anatomy of the LAA is essential during planning, management or intervention of pathologies associated with LAA. The present article reported a case of an adult male with a giant LAA which was accompanied with a PDA. Based on the existing literature, this is the first time these two anomalies are reported together.

Variation on size, shape, width and volume of LAA has been documented. However, few reports have documented the existence of massive LAA [3, 13, 14]. In these cases, LAA measured 1.6 cm to 5.1 cm in total length, which is shorter than the current observation. In contrast, neither of these have reported the existence of both giant/massive LAA and PDA concurrently.

The massive LAA have been previously linked with the thrombus formation particularly in patients with non-
valvular atrial fibrillation mostly due to its larger volume, size and depth [8, 9, 15, 16] and this could be the reason why it was regarded as “most lethal human attachment” [6]. However, some researchers have disputed this, by reporting that cardioembolic events are related more with the shape rather than the size of the LAA [17, 18]. In their retrospective studies, cauliflower shaped LAA was trumped as the most associated with cardioembolic events. This could be the possible explanation on the current observation whereby the subject had chicken-wing LAA (wing-and-hook) shape and have never been reported to have a cardiac event. This further supported by Parsonet et al., study which suggested that, “a massive LAA is a lesion that individuals can be comfortable with no cardiac symptoms for a period of years” [14].

Spatial anatomical relationships for the LAA have been also of a concern particularly with structures overlaid by it [5, 11, 19, 20]. In the current observation the LAA overlaid the main stem left coronary artery (LCA), anterointerventricular artery, left circumflex coronary artery and great cardiac vein. This may result in compression of these structures when the LAA is filled with blood or during LV contraction (systole). Furthermore, there’s a potential risk of compression or damage during LAA exclusion procedures.

PDA in adulthood may present with clinical symptoms or maybe silent [21, 22] depending on the location, size, and magnitude of the shunt [23, 24] but also associated cardiac defects. However, in the present observation the PDA was associated with the massive LAA but unfortunately, the defects were asymptomatic. Thus the physicians planning for surgical or percutaneous transcatheter occlusive devices of PDA or LAA should be aware of the existence of the two defects concurrently, so that to achieve their intended goal without damaging the other major structures.

The LAA exist as a remnant of the original embryonic left atrium and has a different embryological origin with the left atrium proper which arise from the outgrowth of the primordial pulmonary veins. The LAA derived from the mesodermal tissue during the third week of embryonal development of the heart [25–27] which begins with fusion of paired cardiac mesodermal tubes. It originates from the superolateral (left side) wall of the primary left atrium and then matures with the formation of the trabeculae, secondary to the cellular protrusion into the lumen and surrounding vasculature [9, 17].

Mutations on regulatory genes required for early embryonic cardiac development (particularly the LAA) may results into morphological variation of the LAA manifested in the adult. The transcriptional repressors Tbx2 and Tbx3 are known to suppress gene expression of specific cardiac chambers [28] development allowing other cardiac structures to develop. Failure on activation of these transcriptional repressors could lead to over expression of certain genes that may result into cardiac dysgenesis as observed in current report.

Several studies have associated HIRA (histone cell cycle regulator) gene present in the cardiogenic mesoderm with the cardiac dysgenesis [29–32]. The uncharacteristic under or over expression of the HIRA gene in the cardiogenic mesoderm may have effects in cardiac genes which could result into morphological malformation of the heart as observed in the present report.

Endogenous cardiac progenitor cells have been previously shown to play key role in cardiac regeneration and cardiac repair [33]. Another study conducted in murine adult hearts have found that LAA possess large number of cardiac progenitor cells (CPC) of different populations which could be differentially grown from deeper layers of the LAA [34]. These findings indicate that any atypical factor which could activate embryonic transcription factors (such as Nkx-, Gata-, Hand-, T-box and Mef2 family) with effects on CPC could results in morphological difference in the adult LAA and aortic and pulmonary conduits as observed in this case.

CONCLUSION

In conclusion, continuous documentation of the existing anatomical variation involving the LAA is of important for medical literature in order to highlight unknown clinical presentation and the anatomical awareness. Furthermore, knowledge on concurrent existence of LAA and PDA seems to be of importance...
particularly when planning the appropriate management or intervention of either of the two conditions.

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Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES


