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Case Report

ATRIAL SEPTAL DEFECT IN A 50 YEARS OLD MALE PRESENTING AS OBLIQUE SEPTAL CANAL THAT APPEARED TO BE REGULATED BY FIVE LUMINAL BANDS

Ainory Peter Gesase *[1], Gabriel J. Mchonde [2], Alfred Meremo [3], Mange Manyama [4].

[*1] Professor of Anatomy, Department of Biomedical Sciences, University of Dodoma, Tanzania.
[2] Lecturer, Department of Biomedical Sciences, University of Dodoma. Tanzania.
[3] Lecturer, Department of Internal Medicine, University of Dodoma. Tanzania.
[4] Senior Lecturer, Department of Anatomy and Cell Biology, Catholic University of Health and Allied Sciences, Tanzania.

ABSTRACT

To date seven different types of atrial septal defects (ASD) have been described and they include septum primum, septum secundum, superior vena cava, inferior vena cava, coronary sinus and patent foramen ovale types of ASD. One feature in common among these ASD's is that they all present with a hole that may allow communication between the left and right atria. The current observation reports what appears to be a new type of ASD that is characterized by the presence of an oblique septal canal that opens into the right and left atria. The right atrial opening was D-shaped and measured about 0.9 cm wide and the left atrial opening was crescent-shaped and measuring about 0.5 cm wide. In addition to this the left atrial opening was associated with five luminal bands; the last three bands lies on the roof of the oblique septal canal. The action of pulling the first luminal band resulted into closure of the left atrial opening an indication that the bands prevented blood coming from the lungs from entering into the right atrium. The presence of oblique septal canal and luminal bands has not been reported in relation to the ASD's. Continued documentation of such anomalies remains clinically important particularly in African settings where unexplained illnesses are easily attributed to endemic diseases.

KEYWORDS: Atrial septal defects, Oblique septal canal, Luminal bands, Heart.

Address for Correspondence: Prof. Dr. Ainory Peter Gesase, Professor of Anatomy, Department of Biomedical Sciences, University of Dodoma, College of Health Sciences, P.O. Box No. 11029, Dodoma, Tanzania. **E-Mail:** agesase@yahoo.com

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BACKGROUND

The human heart has four chambers; right and left atria and the right and left ventricles. Under normal circumstances the two atria are separated by the interatrial septum, whereas interventricular septum separates the ventricles [1]. These heart chambers communicate both during fetal and adult life. In adult life it occurs between the right atrium and right ventricle and between the left atrium and the left ventricle, while in fetal life it occurs between right and left atria through foramen ovale, and between the right atrium and right ventricles, and between left atrium and the left ventricle [2, 3].

Congenital heart diseases (CHD) are well documented and in most cases are associated with symptoms and signs. However, others may pass unnoticed until adulthood and may manifest in acute situations but others may remain silent until the end of life of an individual or observed during medical examination [4]. CHD occurs in approximately 8-10 per 1000 live births;

and include septal and vulvular defects, patent ductus arteriosus, tetralogy of Fallot, and disposition of great vessels [5, 6, 7].

Septal defects have been described and include ventricular and atria septal defects and are caused by spontaneous malformation of the interventricular and interatrial septum respectively during development [2, 3].

Atrial septal defects (ASD) are a form of a CHD characterized by the presence of a hole that enables blood to flow mainly from the left to the right atria. They include; ostium primum, ostium secundum patent foramen ovale, superior vena cava, inferior vena cava and coronary sinus ASD types [8, *9, 10, 11, 3*]. ASD is more commonly recognized congenital cardiac anomalies presenting in adult account for 22% among adults with CHD but rarely diagnosed [12]; more than 70% are detected by the 5th decade of life [13,14,6].

The present case reports the observation of the atrial septal defect that was found in the heart of a 50 years old male person who died of severe malaria attack.

CASE REPORT

The external observations made on the heart and other organs including major vessels of the heart of a 50 years old male cadaver, were found to be normal. The heart appeared to be normal with respect to the size, shape and its position in the left chest and there was no evidence suggesting enlargement of its chambers.

Fig. 1: External Appearance of the Heart.

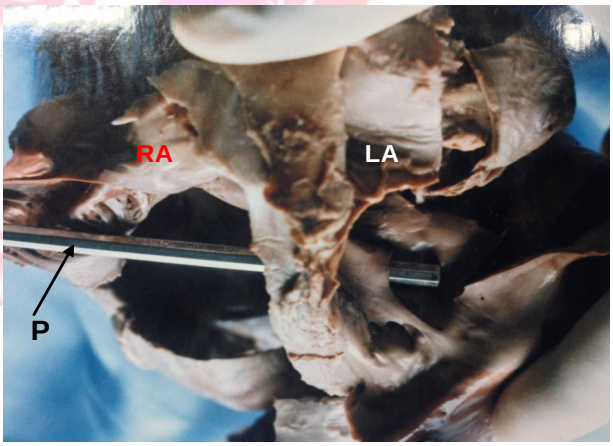
Photograph of the heart that contained the atrial septal defect from a 50 years-old male who died of malaria showing the right (R) and left (L) ventricles.

It measured 15cm long from the base to the apex and about 10cm wide at the atrioventricular junction [**Figure 1**].

Anatomy of the right atrial opening

After opening the heart during dissection, close observations revealed the presence of a large opening at the middle of the interatrial septum that allowed a probe to pass from the right to the left atria [**Figure 2**]. Presence of interatrial communication necessitated examination of both the right and left atrial. The atrial septal opening on the right atrial side showed a D-shape configuration and appeared larger than that on the left side; it measured about 1.3cm wide [**Figure 3**]. The margins of a D-shaped opening were smooth and did not contain redundant tissue materials or valve-like structure. The right atrium showed a normal looking openings of the coronary sinus, superior and inferior vena cavae.

Fig. 2: Internal Appearance of the Atria.



Photograph of the heart that contained the atrial septal defect from a 50 years-old male who died of malaria showing the internal appearance of the *right (**RA**)* and *left (**LA**) atria*. The *probe* ***(P)*** was inserted through the oblique septal canal that allowed communication between the two atria.

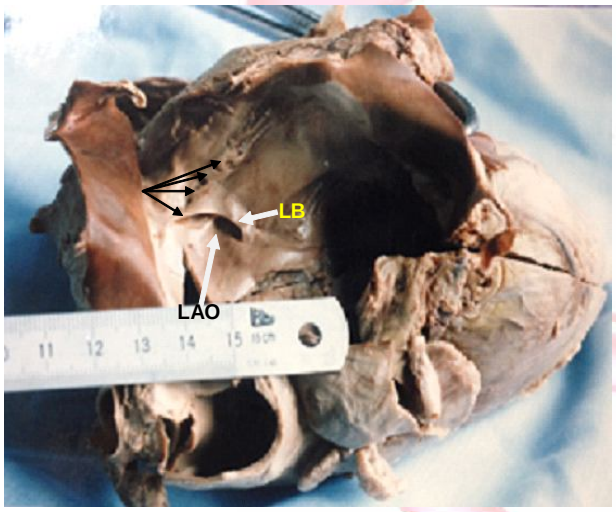
Fig. 3: Internal Appearance of the Right Atrium.

Photograph of the heart that contained the atrial septal defect from a 50 years-old male who died of malaria showing the *D-shaped right atrial opening (RAO).*

Anatomy of the left atrial opening

The interatrial septum showed a crescent-shaped profile that measured 2cm long from the vertebral surface of the heart and close examination revealed that it was associated with the atrial opening and the luminal bands. The left atrial opening was located at the inferior portion of the crescent-shaped margin and did not occupy the entire length of the crescent-shaped profile [**Figure 4**]. The opening measured about 0.9cm long and 0.5cm wide, at its widest diameter. The left atrial opening communicated with a larger right atrial opening and the right atrial opening was located superior to the left atrial opening [**Figure 2**]. The two left and right atrial openings were connected by a oblique canal that appeared to run in the atria septal wall.

Fig. 4: Internal Appearance of the Left Atrium.



Photograph of the heart that contained the atrial septal defect from a 50 years-old male who died of malaria showing the crescent-shaped profile of the left atrial opening (**LAO**) and the 5-luminal bands (**Black Arrows**). The first luminal band (**LB**) lies within the crescent-shaped profile and remaining 4-bands (**Arrows**) are arranged along the oblique septal canal.

Close examination of the crescent-shaped margin revealed also the presence of five bands that were formed by the luminal thickening [**Figure 5a and 5b**]. The first band formed the margin of the crescent-shaped profile and the fifth band was located 5cm away from the first band. The third, fourth and the fifth bands appeared to lie along the wall of the oblique septal canal [**Figure 5b**]. Each band showed two areas of attachments that can be compared with the pints of insertion and origin seen in skeletal muscles. Pulling of the first luminal band resulted into closure of the left atrial opening and pulling of the second, third, fourth and fifth bands resulted into obliteration of the interatrial canal.

Fig. 5: Internal Appearance of the Left Atrium.

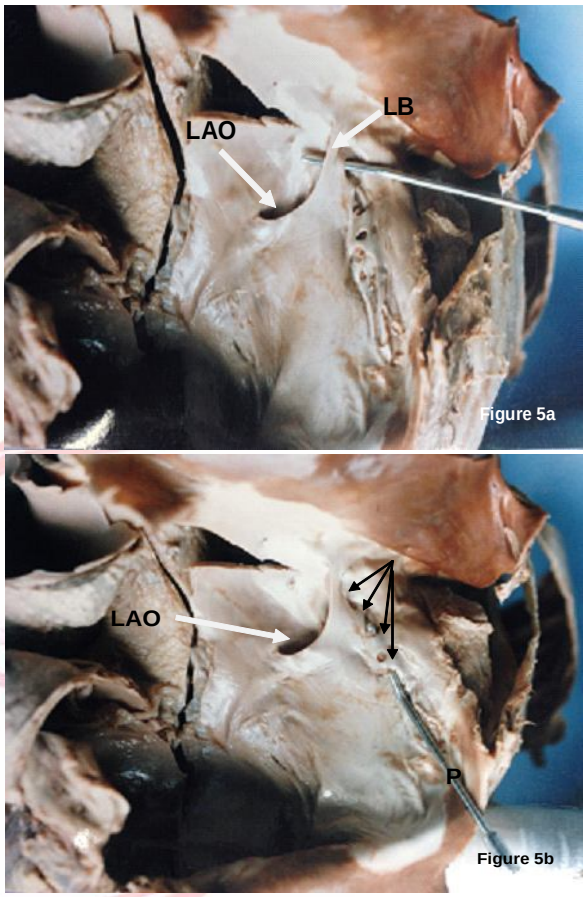


Figure 5a and 5b: The photographs of the heart that contained the atrial septal defect from a 50 years-old male who died of malaria. **a**. shows the first luminal band (**LB**) and the left artial opening (**LAO**). **b**. Shows the left atrial opening (**LAO**) and the remaining 4-bands (**Black Arrows**) and the blunt probe (**P**) display the unattached free areas and the attached parts of the bands.

The color of blood clots that were found in the atria lumina was different; blood clots observed in the left atrium appeared paler than those from the right atrium which were darker. The walls of the atria and ventricles appeared normal and there was no indication for abnormal dilatation of the heart chambers or hypertrophy of the myocardium.

DISCUSSION

The current observation has documented a case of a 50 years old male who presented with an atrial septal defect, characterized by what appeared to be the oblique septal canal that opened into the right and left atria via the right and left atrial openings. Observations on the defective site of the left atrium revealed the presence of five luminal bands that were associa-

ated with margins of the left atrial opening and the oblique septal canal. Presence of the oblique septal canal and luminal bands is a novel observation that has not been documented before.

The anatomy of atrial septal defects has been extensively studied and one observation in common is that they all contain a hole that may allow communication between the left and right atria [9]. The current observation showed the presence of the right and left atrial openings that were connected with an oblique septal canal. The left atrial opening was associated with five luminal bands. Review of literature indicates that there are seven types of atrial septal defects (ASD) that have been described [9]. They include ostium primum ASD; ostium secundum ASD; patent foramen ovale; superior vena cava sinus venosus ASD; inferior vena cava sinus venosus ASD and the coronary sinus ASD [15,*16,17,11*]. Mixed defects occurs when two or more of the atrial septal zones are involved, and account for only 7% of all atrial septal defects [18]. The current ASD appears to be located in the middle of the interatrial septum an indication that it can be considered to be the ostium secundum ASD type [11]. But the presence of an oblique canal and the luminal bands makes it to be a unique type of ASD. Ostium secundum ASD type has been showed to be associated with strands of tissues and mitral valve prolapsed [19, 20]; presence of luminal bands has not been described. It is suggested that the current ASD observed in the heart of a 50 year old male cadaver is characterized by the presence of an oblique canal and the luminal bands is a novel type of ASD. Why we are describing this for the first time is not known, but it is important to note that many descriptions of the ASD's relied on radiological technique and it is possible that such anatomical features can be missed. The cause of death for this individual was determined to be malaria, an indication that despite having a large defect in the interatrial septum he was asymptomatic. Studies have showed that 70% of individual with ostium secundum ASDs remain asymptomatic during early adulthood [21]. The reason for this has not been determined; it is possible that the presence of structures similar to the luminal bands observed in the current report may prevent blood coming from the lungs to enter the right atrium. The current study shows the importance of combining different methods in assessing the congenital heart anomalies.

The current observation reported also the presence of five luminal bands; all of them located in the left atrium side and associated with the left atrial opening and the oblique septal canal. The bands appeared smooth and covered with the endocardium, similar to the ones covering the lumen of the atrium. Past studies have reported the presence of strands in relation to the ostium secundum ASD, creating the fenestrated appearance [15], and redundant septum has been reported to occur in patent foramen ovale malformation [22]. At the moment the exact significance of the luminal bands during the living heart is not clearly known. But it was revealed that pulling of the larger band resulted into closure of the left atrial opening, an indication that the five bands could work together to prevent blood from entering the right atrium. The suggestion shows that much as this heart developed a canal between the two atria, it also formed the five luminal bands that appeared to regulate the flow of blood. Symptoms and signs related to the ASD develop because blood coming from the lungs enters the right atrium causing overload to the right chambers of the heart [23,24].

CONCLUSION

Congenital heart defects involving the atrial septum are implicated in several serious clinical syndromes, including stroke, myocardial infarction, systemic embolism and migraine. It has been shown that 40% of all patients suffering from ischaemic strokes (80% of all stroke victims) remain without clearly identifiable cause [25]. The current observation reports the existence of another oblique septal canal type of ASD in human heart. Continued documentation of such anomalies remains clinically important particularly in African settings where unexplained illnesses are easily attributed to endemic diseases.

List of Abbreviations

ASD: Atrial septal defects
CHD: Congenital heart defects
PFO: Persistent foramen ovale

Conflicts of Interests: None

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