Case Report

ASYMPTOMATIC RIGHT-SIDED AORTIC ARCH IN A NIGERIAN WOMAN: A CASE REPORT AND LITERATURE REVIEW.

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Abstract

Background: Right-sided aortic arch (RAA) is a rare congenital anomaly of the cardiovascular system that stems from the persistence of the right fourth branchial arch. It occurs in < 0.1% of the population. RAA with an aberrant left subclavian artery is the most frequent type. The associated aberrant left subclavian artery occurs in about 50% of RAA cases. Anomalies of the great arteries (e.g., Fallot's tetralogy) are common when RAA occurs in a normal visceral situs setting. By contrast, some cases of RAA remain asymptomatic throughout life. Echocardiography, multidetector cardiac CT, cardiac MRI, and cardiac catheterisation are the cornerstone of investigation in RAA cases.

Case presentation: This is a case report of an asymptomatic right-sided aortic arch without associated congenital anomalies detected incidentally in an adult during a pre-employment medical examination. The patient was symptomless; therefore, no treatment was given.

Conclusion: Very few cases of RAA in adults are entirely asymptomatic, as reported in this case. A review of the literature yielded scanty reports of aortic arch anomalies among Nigerians (paediatric and adults). In the previous decade (2011–2020), there were at least 32 case reports of RAA in adults. Of this number, only one was completely asymptomatic like this case report. This report might be the first documentation of an asymptomatic isolated RAA in a Nigerian.

Keywords: Right-sided aortic arch, Aortic arch variant, Cardiovascular anomalies, Computerised tomography, Case report.

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INTRODUCTION

The right-sided aortic arch is an uncommon cardiovascular anomaly that occurs in approximately 0.05% to 0.2% of the population.[1–5] Fioratti and Aglietti reported the first documented case of RAA in 1763.[6] It is one of the congenital causes of dyspnoea, but occasionally, this anatomical abnormality does not cause any symptom. When it presents with dyspnoea, it is sometimes misdiagnosed for asthma. The diagnosis of the right-sided aortic arch is often made in childhood.[7]

This is a case report of an asymptomatic right-sided aortic arch in a 32-year-old lady discovered incidentally. A review of the literature yielded scanty reports of aortic arch anomalies among Nigerians (paediatric and adults). Besides a case report of a double aortic arch,[8] most of the previous studies on congenital cardiovascular diseases in Nigeria did not document a remarkable number of isolated RAA or other aortic arch anomalies.[8–12] Even among Nigerian children with tetralogy of Fallot, RAA is apparently not a prominent feature.[13,14] RAA is seen in 10-25% of tetralogy of Fallot cases.[15,16]

CASE PRESENTATION

A 32-year-old woman presented at the radiology department for a pre-employment chest radiograph. She had no history of an acute or chronic illness. The postero-anterior chest radiograph showed a slightly hyper-inflated right lung with mediastinal shift to the left (**Figure 1**). There was no focal or diffuse lung lesion, and the pleural recesses were free (no pleural effusion). The aortic knob was not visualised at its normal position to the left of the thoracic spine, but a poorly defined right-sided aortic knob was suspected.

With a high index of suspicion for a possible right-sided aortic arch, a chest computerised tomographic (CT) scan was requested. Chest CT showed a right coursing aortic arch (**Figure 2**), a right-sided descending thoracic aorta (**Figure 3A**), and displacement of the carina to the left (**Figure 3B**). No aberrant left subclavian vessel was demonstrated.

Transthoracic echocardiography excluded any associated cardiac anomaly.

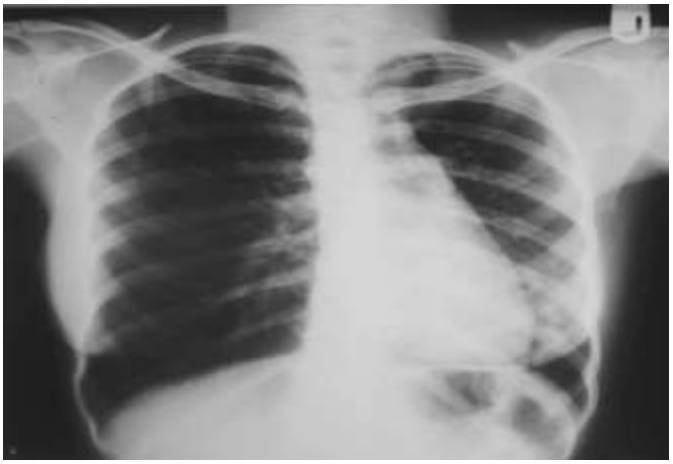

Figure 1: Chest radiograph (posteroanterior view) showing slightly hyper-inflated right lung with mediastinal shift. There is poor definition of the aortic knob on the left

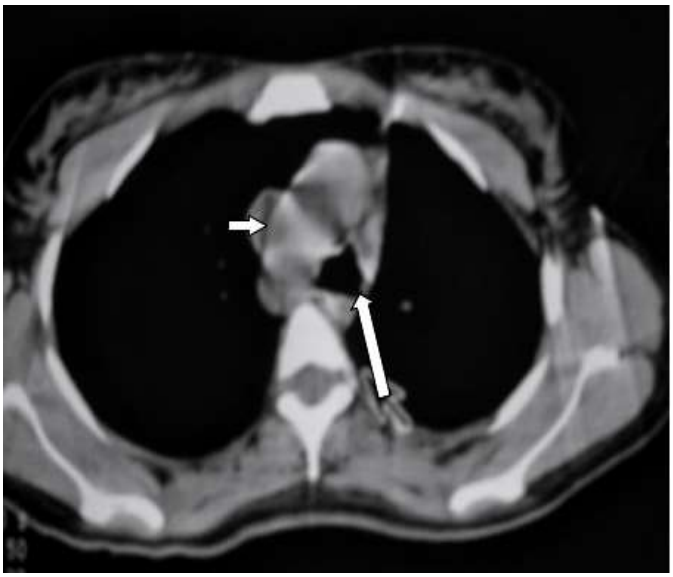

Figure 2: Axial CT scan (mediastinal window) of the chest at the level of T3 vertebra showing a right coursing aortic arch (short white arrow). The trachea is displaced to the left (long white arrow)

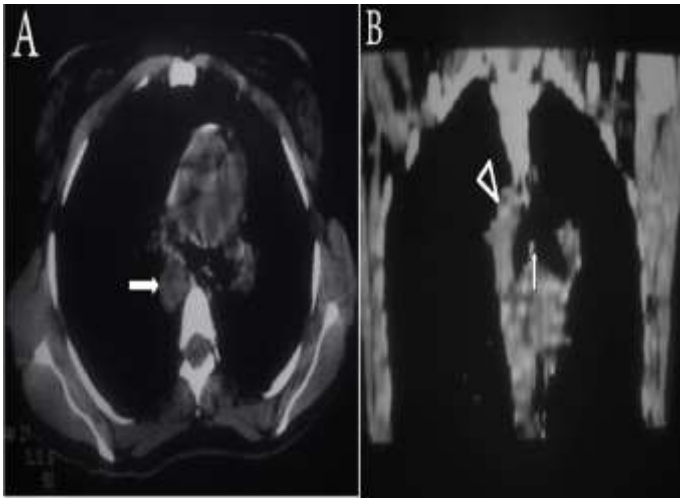

Figure 3: A: Non-contrast axial CT scan of the chest (mediastinal window) showing a right-sided descending thoracic aorta (arrow); B: Coronal Chest CT (mediastinal window) showing right-sided ascending aorta (arrowhead) with displacement of the carina to the left (arrow)

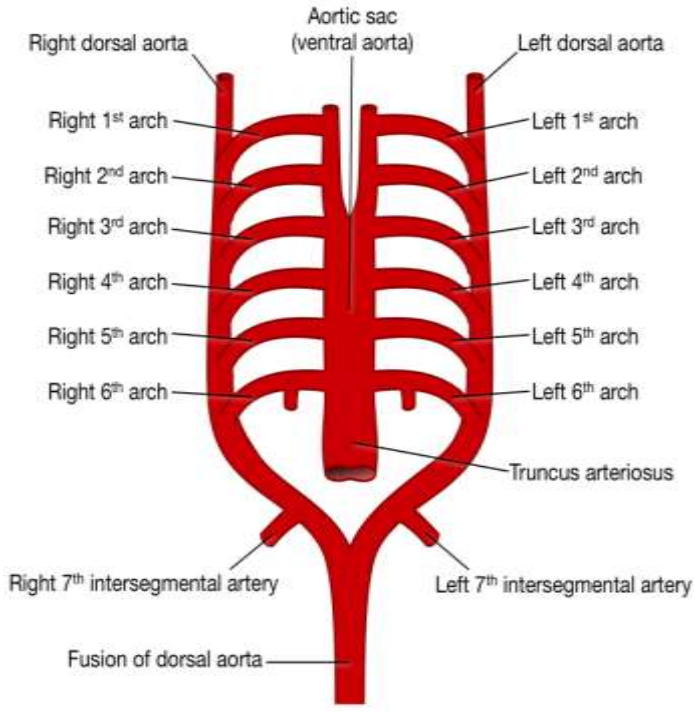

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Figure 4: Schematic illustration of the development of the aortic arch and branches from the embryonic pharyngeal arch arteries.

DISCUSSION

Six pairs of aortic arches (**Figure 4**) connect the two primitive ventral and dorsal aortae during embryonic vascular development.[17–19] Most portions of the first, second, and fifth arches regress. The third arches become the carotid arteries. A branch from the sixth arch's ventral bud unites with the pulmonary bud to form the pulmonary artery. The dorsal contribution to the sixth right arch disappears, and on the left it remains as the ductus arteriosus. The seventh intersegmental artery originates from the dorsal aorta to become the subclavian arteries. In normal development, part of the fourth right arch regresses leaving a remnant as the normal left aortic arch. If the fourth arches remain bilaterally, a double aortic arch (complete vascular ring) results.[17–19] Regression of the fourth left arch leads to the formation of a right aortic arch system, such as this case study. Different right aortic arch configurations are possible depending on the location of interruption(s) at the left fourth arch, and the branching patterns of the left subclavian artery, left carotid artery and the ductus arteriosus.[20] There are three main RAA types based on Edward's classification: type I—RAA with mirror image branching; type II—RAA with aberrant left subclavian artery; and type III—RAA with isolated subclavian artery (connected to the pulmonary artery via the ductus arteriosus).[21] Type II is the most common. Other unusual types include RAA with innominate artery isolation and right circumflex aorta.[2]

The right subclavian artery and the right common carotid artery arise directly from the right-sided aortic arch in the mirror image type. In contrast, the left subclavian and left common carotid arteries arise on the left from a common left brachiocephalic trunk, giving a mirror image of the normal vascular arrangement in the left-sided aortic arch.[2,20] Mirror image RAA is, by itself, generally asymptomatic, but it is commonly associated with congenital cyanotic heart diseases.[7] In the patient under review, there was no associated congenital heart malformation on echocardiography, and the mirror image vessels could also not be demonstrated on CT, which means it is not the type I variety. If a mirror image RAA is associated with a retro-oesophageal left ligamentum arteriosum,[22] a vascular ring is formed - this anomaly is rare but may be symptomatic.

The type II variant is often diagnosed incidentally and is usually not associated with congenital heart anomalies.[2,23] The patient under review is likely to have had the type II variant; although, an associated aberrant left subclavian artery was not demonstrated. RAA with an aberrant origin of the left subclavian artery (type II RAA) is the most common form of the right-sided thoracic aortic arch.[18,24] The (great) arteries in type II RAA originate in this order: left common carotid, right common carotid, right subclavian, and left subclavian artery. The aberrant left subclavian artery passes from the right into the left hemithorax posterior to the oesophagus and trachea and, together with the ligamentum arteriosum, forms a vascular ring.[18,24] The aberrant left subclavian artery may either arise as the fourth (last branch) of the right-sided aortic arch or from a diverticulum of Kommerell (persistent remnant segment of the sixth right arch which can develop ectasia or aneurysm).

Occasionally, adults, especially those with vascular rings, present with symptoms and signs like stridor, respiratory embarrassment, recurrent respiratory infections, or dysphagia.[24] In children, additional findings include apnoea and or a characteristic high-pitched brassy cough. Other findings include a history of asthma or recurrent pneumonia. Some children try to maintain a position in which the head is hyperextended to improve breathing and minimise the obstruction. Air trapping and pulmonary hyperinflation may also be present in one or both lungs in severe cases.[7] However, a few patients do not manifest symptoms until later in life, while others remain entirely asymptomatic like the index patient.

Radiographic findings of RAA include widening of the right superior mediastinum with an absence of the normal left aortic arch contour and a well-defined area of increased opacity in the retro-tracheal space at the level of the upper aortic arch on orthogonal plain chest radiographs.[23] Other features include slight deviation of the normal tracheal air shadow or the carina to the left as seen in this patient, narrowed or constricted air column, unilateral hyperinflation of the lung in severe cases of bronchial constriction and a right-sided descending thoracic aorta. There was slight hyperinflation of the right lung in this patient, although bronchial constriction could not be demonstrated.

Barium swallow often demonstrates anterior displacement of the oesophagus with a characteristic diagonal impression on the posterior oesophagus at the fourth thoracic vertebra level due to the obliquely coursing aberrant left subclavian artery.[1,17] Unfortunately, this patient declined barium swallow on account of further radiation exposure.

Arch aortography, CT and magnetic resonance imaging (MRI) can confirm the diagnosis, demonstrate any coexisting cardiac or vascular anomaly, and delineate the exact anatomy.[2,3,17,25,27] There was CT confirmation of this case. No aberrant subclavian vessel was seen. Echocardiography may also be useful to confirm or rule out associated congenital cardiac anomalies.

Asymptomatic patients require no form of treatment, but surgical correction/ ligation of the vascular ring to relieve the symptoms is indicated in symptomatic patients.[7] In the previous decade (2011–2020), there were at least 32 case reports of RAA detected in adulthood.[20] Of this number, only one[28] was completely asymptomatic like this case report.

CONCLUSION

Aortic arch anomalies are rare or under-reported in Nigeria. This case report might be the first documentation of this anomaly (asymptomatic isolated RAA) in a Nigerian. A high index of suspicion reduces the chances of misdiagnosis or missing the diagnosis.

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