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Case Report

Incidental finding of double aortic arch and tract from noncoronary sinus to left atrium: A case report ☆

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ABSTRACT

The double aortic arch with vascular ring is a rare but documented aortic arch variant, traditionally presenting with difficulty swallowing or breathing due to extrinsic compression. Tracts from the noncoronary sinus to left atrium are very rare, with limited case reports to compare against. We report an incidental finding of double aortic arch in an elderly woman who underwent a cerebral angiogram for symptoms of a right-sided stroke, with a further anomaly identified on subsequent CT gated aortogram of a possible tract between the non-coronary sinus and left atrium. It is worth noting that the aortic arch abnormality was missed on previous plain radiographs, which can happen even among experienced radiologists. This case illustrates the need for a thorough, systematic approach to interpreting chest radiographs to avoid missing mediastinal lesions, such as aortic abnormalities.

Introduction

The double aortic arch is a rare congenital vascular anomaly where a complete vascular ring encircles the trachea and esophagus. The compression of these structures can lead to various degrees of swallowing and breathing difficulties. Case reports of elderly patients are extremely rare. We report an incidental finding of double aortic arch in an elderly woman who underwent a cerebral angiogram for symptoms of a right-sided stroke, with a further anomaly identified on subsequent CT gated aortogram of a possible tract between the non-coronary sinus and left atrium.

Case report

An 89-year-old woman was admitted to our hospital following an 18-hour history of left-sided arm and leg weakness.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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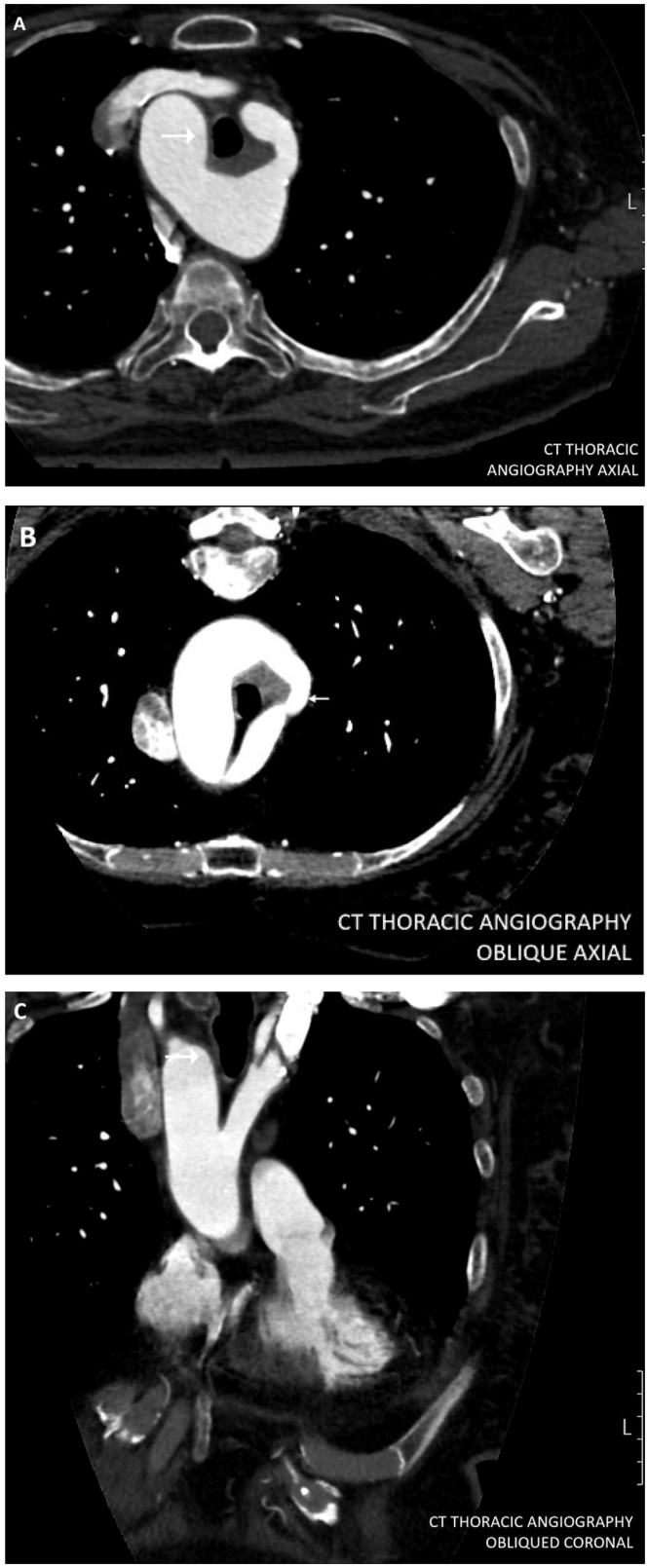

Fig. 1 – CT thoracic angiography images demonstrating the presence of a double aortic arch encircling the trachea, indicated by arrows. (A) Axial section of the aortic arch on CT thoracic angiography. (B) Obliqued axial of double aortic arch. (C) Obliqued coronal of ascending aorta. RA, right aortic arch; LA, left aortic arch.

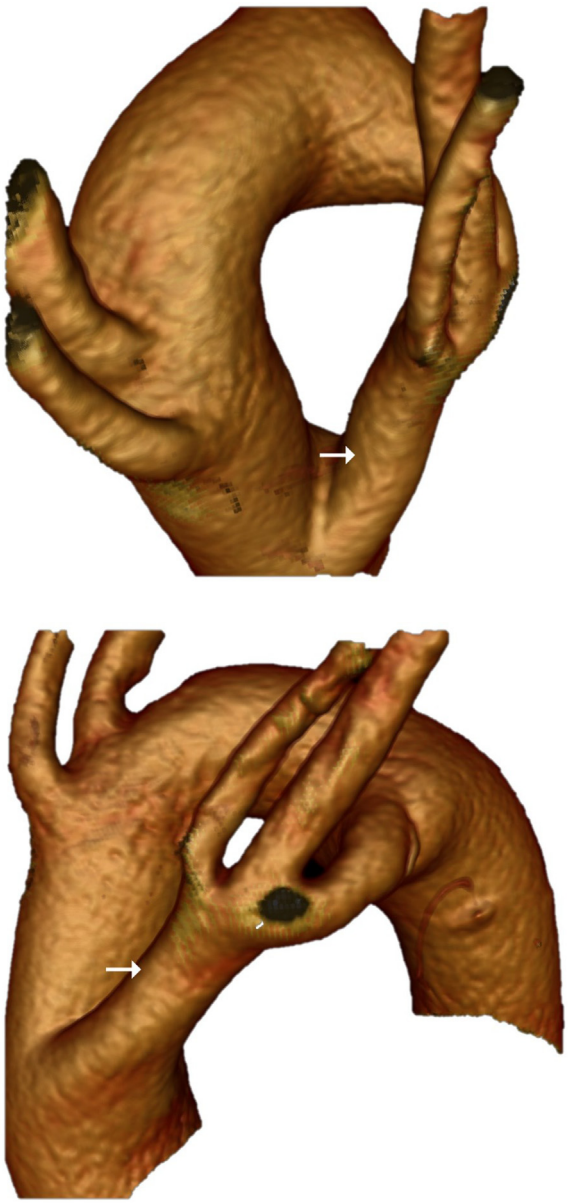

Fig. 2 – Three-dimensional computed tomography (3D-CT) reconstruction of the arch of the aorta. The double aortic arch is visualized here (see arrows).

Her past medical history included type-2 diabetes mellitus, ischemic heart disease and Barrett's esophagus. She has a strong family history of cardiovascular disease. Physical examination demonstrated left-sided dense hemiparesis with reduced reflexes but intact sensation. Electrocardiogram (ECG) and laboratory results were unremarkable. Initial clinical impression was an acute right-sided ischemic stroke with left-sided hemiparesis. Computed tomography (CT) of her head showed age-related involutional changes and small vessel disease, however, was unable to confirm acute infarction. Subsequent Magnetic Resonance Imaging (MRI) of her head showed multifocal small areas of acute ischemic within the right cerebral hemisphere, and Carotid Doppler ultrasound showed a complete right internal carotid artery (ICA) occlusion. A CT intracranial angiogram was performed from aortic arch to cerebral vertex which confirmed the right internal carotid artery occlusion, but also identified as an incidental finding an unusual anatomical configuration to the aortic arch. As this was incompletely visualized, a CT thoracic aortogram was performed gated in systolic phase (Fig. 1). The ascending aorta was seen to bifurcate at the level of the main pulmonary arteries, with a particularly prominent "right sided" aortic arch passing abnormally to the right of the trachea and esophagus seen in combination with a second less dominant arch passing to the left of the trachea. The right common carotid and subclavian arteries originated separately from the right sided arch (without formation of a brachiocephalic artery), with separate origins of the left common carotid and subclavian arteries from the left sided arch. The 2 arches subsequently joined together distal to the great vessel origins forming a vascular ring, exhibiting mild compression to the trachea and esophagus. A 3D volumetric reconstruction of the CT imaging delineates the anatomy more clearly (Fig. 2). Reviewing the rest of the visualized anatomy for congenital abnormalities of the heart, an additional incidental finding of a possible small tract between the non-coronary sinus and the left atrium was also identified, although this finding has to be regarded as equivocal due to the presence of motion artifact (Fig. 3).

The patient was promptly managed with aspirin and clopidogrel, alongside physiotherapy and rehabilitation which she showed improvement in her physical function. Important risk factors that predisposed this patient to a stroke included her old age, hypertension, type-2 diabetes mellitus, personal and family history of cardiovascular disease. She is on a 3-year surveillance program for her Barrett's esophagus which she initially presented with dyspepsia. She has not reported of any breathing or swallowing difficulties.

A chest X-ray was not performed during this admission, however on retrospective review of the patient's previous chest X-rays dating back as far as 2012, an abnormal appearance to the aortic knuckle consistent with a right-sided aortic arch was noted not to have been previously reported upon (Fig. 4).

Discussion

A double aortic arch is a rare but documented congenital vascular anomaly. It is the most frequent form of vascular ring malformation, accounting for 1% of congenital heart diseases and 42% of all vascular rings [1,2]. The formation of a vascular ring around the trachea and esophagus can give rise to a variety of symptoms according to the degree of tracheal and/or esophageal compression present. The condition can come to light in early childhood with dyspnea, stridor, dysphagia, feeding difficulties, and recurrent chest infections [3,4], or later in life with chronic swallowing difficulties. As this case has highlights however, patients can however also be asymptomatic and remain undiagnosed when the compression is less pronounced [5,6,7].

Current models of embryonic development suggest that 6 pairs of arches emerge from the truncus arteriosus and regress

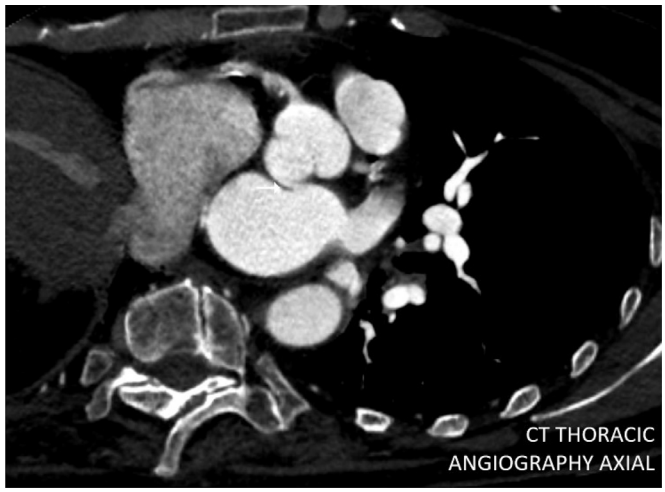

Fig. 3 – CT thoracic angiogram demonstrating the short axis view at the level of the coronary sinuses of Valsalva. Although equivocal due to motion artifact, a possible tract from the noncoronary sinus to the left atrium (see arrow).

sequentially between weeks 2 and 7 of fetal development, the remaining structures forming the aorta and great vessels [8]. In normality at week 5 of gestation the right side of the fourth pair of arches regresses leaving behind a left-sided vessel that forms the normal left-sided aortic arch. Deviation from this process forms the basis of understanding for aortic arch variants and anomalies: a failure to regress the right side of fourth pair of arches giving rise to a bilateral aortic arch configuration. Conversely, a solitary right-sided aortic arch may remain if the left-sided arch regresses while the right-sided arch persists [8,9].

The 3 subtypes of the double aortic arch can be classified depending on the relative size of the 2 arches and their position in relation to the trachea: right-dominant aortic arch (75% of cases), left-dominant aortic arch (20% of cases), and balanced aortic arches (5% of cases) [10].

On plain chest radiographs, the right-sided aortic arch can present itself with absence of the normal left sided aortic knuckle, tracheal bowing to the left at the level of the right aortic arch, soft tissue indentation on the right side of the distal trachea, and a right-sided descending aorta [11,12]. The right aortic arch is often seen as high riding and projecting as a mass in the right paratracheal region [13]. In the case presented here, the patient's chest radiograph demonstrates the aortic knuckle is abnormally projected straddling both sides of the trachea silhouette, rather than as in normality the knuckle residing fully to the left of the trachea. It is pertinent that this key finding although clearly present was not identified on any of the patient's multiple previous historic chest radiographs, despite being reported by several different radiologists. It is therefore evident that a failure to recognize this feature is a common oversight even among experienced radiologists.

It is well established in the literature that the incidence of radiologists overlooking key findings may be as 30% [14,15], with substantial interobserver variability in the identification of chest radiograph findings. This highlights the challenges in the accurate detection of abnormalities on plain film imaging even among experienced observers, with interpretation and reporting of chest radiographs particularly prone to missing mediastinal lesions. As mediastinal lesions can be subtle and overlooked, a thorough and systematic approach to following and interpreting mediastinal contours with a good knowledge of normal mediastinal anatomical features is required.

The use of formalized checklists has been suggested as an effective approach to reducing errors in clinical settings [16]. Checklists, in the form of structured reports with standardized templates, have been suggested as a means of potentially improving reporting consistency [17], with one study attempting to compile a checklist based upon commonly missed chest radiograph findings [18].

Once an aortic arch abnormality is identified, careful review of the cardiac anatomy is also required as it is recognized that this can be associated with other congenital heart defects such as a ventricular septal defect, tetralogy of Fallot, truncus arteriosus, and transposition of the great arteries [3]. Aortic arch anomalies can also be associated with chromosomal abnormalities such as DiGeorge syndrome (22q11 deletion) [19].

Three outpouchings formed at the root of the ascending aorta immediately above aortic valve are known as the aortic sinuses (colloquially known as a sinuses of Valsalva); it is thought the anatomy of these is such that the adjacent aortic valve leaflets do not occlude the ostia of the coronary arteries during systole [20]. Whilst the left and right aortic sinuses give rise to the left mainstem and right coronary arteries respectively [21], in normality the third outpouching deemed the "non-coronary sinus" does not gives rise to any coronary vessels. Anatomical variations to the non-coronary sinus are thought rare [22]. At present, only a small number of case reports appear in the literature detailing incidental findings of tracts from the non-coronary sinus into the left atrium [23–26]. It is noted that reports of these fistulas were found in both adults and infants as an incidental finding since the patients were asymptomatic. In our case, whilst the tract appears more convincing on multiplanar reformatting (MPR), the presence of motion artifact makes the finding equivocal. Assuming the visualized fistula from the non-coronary sinus to left atrium is

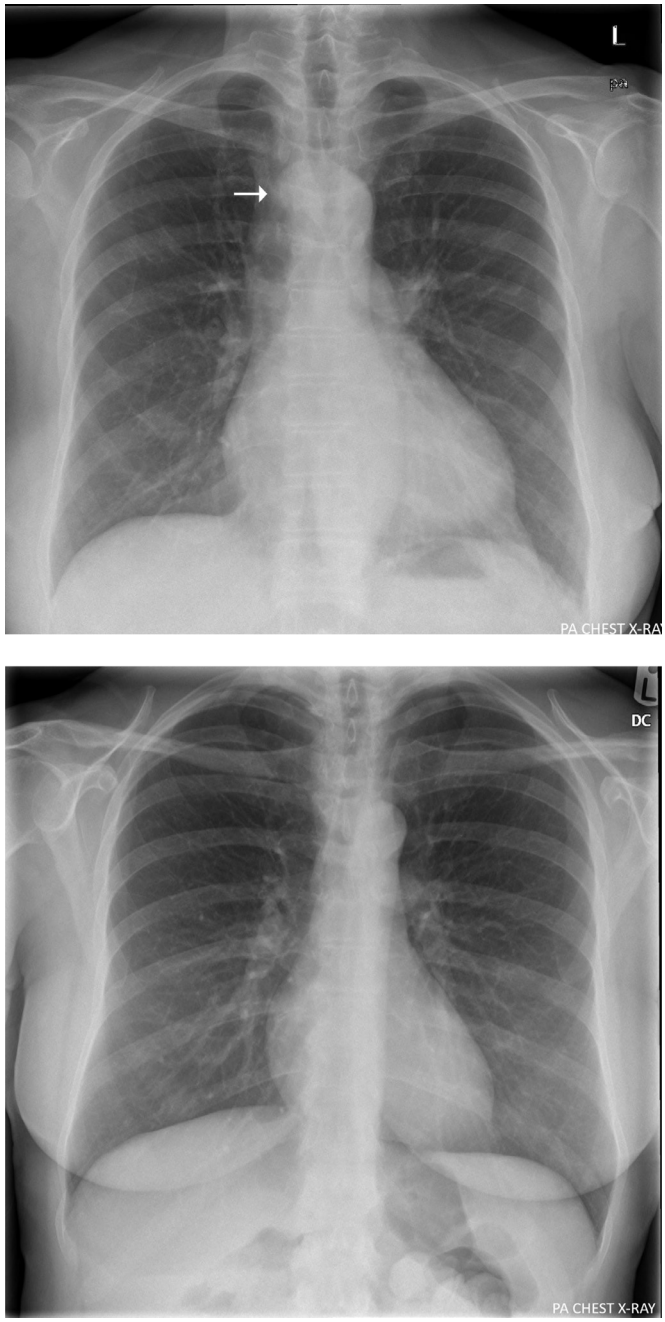

Fig. 4 – Comparison between this patient's chest radiograph (PA view) (left) and a normal chest radiograph for comparison (right). Note the rounded profile of a normal aortic knuckle should always pass to the left of the trachea silhouette. In this case the aortic knuckle traverses the tracheal silhouette (see arrow).

indeed real, the small caliber of the tract is unlikely to exhibit a clinically significant degree of shunting.

Patient consent

Written informed consent was obtained from the patient for publication of this case report, including accompanying images.

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