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Right aortic arch: CT. diagnosis

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Summary

Computer Tomography (CT) can be helpful in the detection and diagnosis of aortic arch malformations, particularly the retro-oesophageal right side aortic arch with aberrant left subclavian artery. Three adult Saudi patients with this anomaly where recently encountered and are herein presented. All the three patients had CT scans with clear demonstration of the right aortic arch. CT can confirm the presence of this aortic arch anomaly suspected from plain chest radiographs or can detect the vascular anomaly when the radiographs suggest the presence of a mediastinal mass. The condition of congenital anomalous right aortic arch is also briefly discussed.

Resume

La Tomographie Computee (TC) peut assister a la detection et au diagnostic des malformations des ares aortiques, surtout l'arc aortic retro-oesophageal de la droite avec l'artere subclavienne aberante. Trois patients adultes d'origine de l'Arabie Saodite manifestant cette anomalie ont ete examines et presentes ici. Tous les trois ont subi le balaye TC avec une claire demonstration de l'arc aortic de droite. Les balayages TC peut confirmer la presence de cette anomalie de l'arc aortic qui est vaguement soupconne dans les simples radiographie de la poitrine. Ces balayages peuvent detecter l'anomalie vasculaire quand la radiographie suggere la presence d'une masse mediastinale. La condition de l'anomalie congenitale de l'arc aortic de droite est aussi brievement discutee.

Introduction

Anomalous development of the embryonic paired aortic arches can result in a number of malformations of the aortic arch and great vessels. These include, amongst others, right aortic arch with (i) mirror-image branching, (ii) aberrant origin of the left subclavian artery or (iii) isolated left subclavian artery. In some patients, a mediastinal vascular anomaly may be mistaken for a mediastinal mass on plain radiographs and a CT scan performed; in others an anomaly will be detected incidentally on scans performed for another reason. Furthermore, when an aortic arch malformation is suspected clinically or radiographically, CT can provide a non-invasive method for its evaluation.

Some authors[1-4] have demonstrated the unique capabilities of CT to display anatomy and pathology in the lungs, pleura, mediastinum and the great vessels in the chest. The first reported CT diagnosis of a retro-oesophageal right aortic arch was by Taber *et al.* [5] in 1979. A search of the literature yielded seven other cases reported in three separate articles in the 1980s[6,7,8]. We are therefore reporting three cases of this relatively uncommon anomaly recently seen in our centres, all of which were confirmed, in a non-invasive manner, by CT.

Case reports

Case 1: A 60-year old Saudi male presented with a two-month history of vague chest pain and breathlessness on exertions. Physical examination and systemic review revealed no abnormality but a rather fit old man. The chest radiographs however showed upper mediastinal widening with left anterior deviation of the trachea by a large rounded retro-tracheal soft tissue mediastinal mass. A right aortic arch anomaly was considered the most likely diagnosis but the differential diagnosis of thoracic aortic aneurysm and mediastinal tumour was also entertained. CT of the chest and mediastinum with

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contrast enhancement (Fig. 1) demonstrated the absence of the usual left sided position of the aortic arch and a right aortic arch presenting as an unusual mass to the right side of the trachea, crossing the midline behind the trachea and oesophagus, displacing them forward away from the spine. In the region of the absent left aortic arch, an opacified dilated origin of the aberrant left subclavian artery was noted. There was no evidence of a thoracic aortic aneurysm or a mediastinal tumour. The descending aorta was identified lying to the right of the spine.

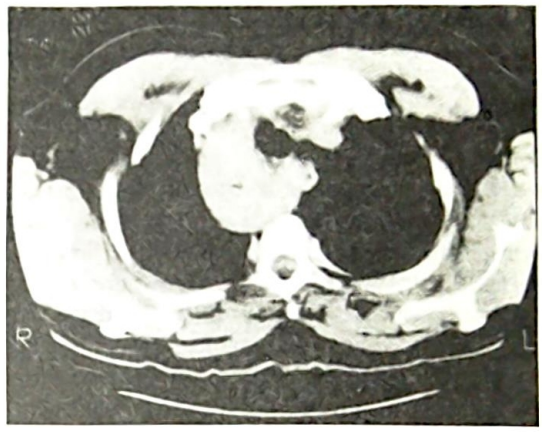

Fig 1: CT Scan on Case 1 showing right-sided aortic arch (AO) displacing both oesophagus and trachea (T) forward, away from the spine. (S) = dilated orgin of aberrant left subclavian artery.

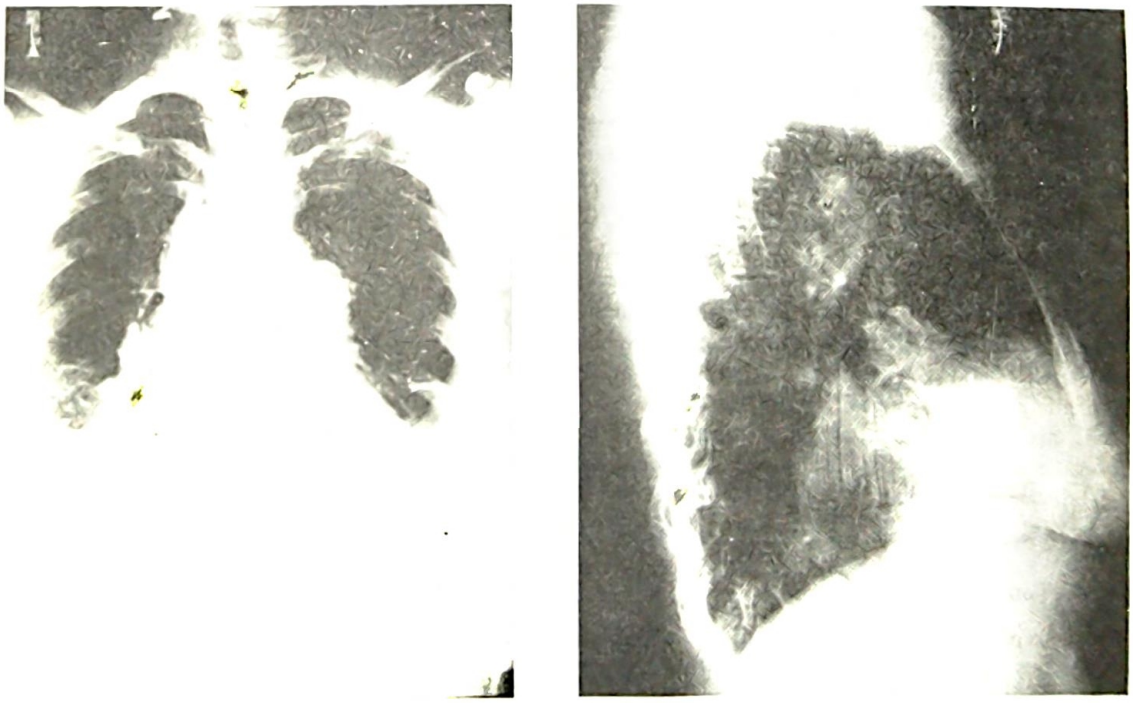

Fig 2 (a & b): Chest Radiograph on Case 2 (PA & Lateral) showing widened mediastinum, right aortic arch (A), bulky left hilum and a rounded retro-tracheal soft tissue mass (M). Note cardiomegaly.

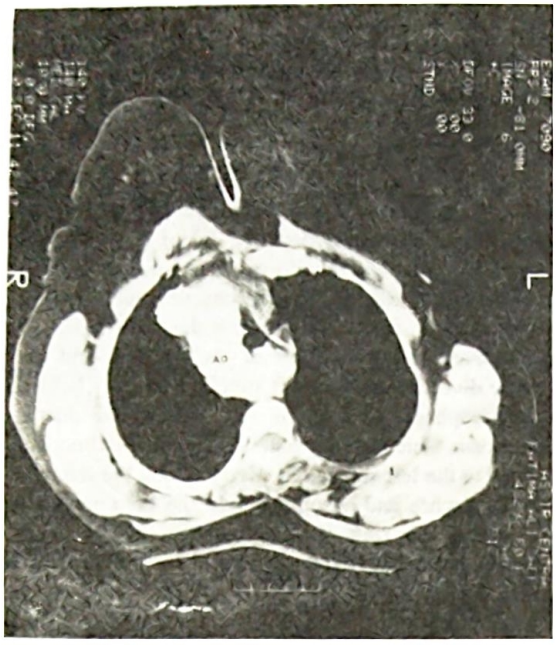

Fig 2c: CT Scan on Case 2 showing only Right-sided aortic (AO); no hilar mass or aneurysm.

Case II: A 65-year old Saudi female was referred from a district hospital to Asir Central Hospital, Abha because of a widened mediastinum and a suspected left hilar mass lesion. The admission chest radiographs (Fig. 2a+b) showed slight cardiomegaly, a widened mediastinum due largely to an unfolded aorta. The left hilum was indeed bulky suggestive of the presence of a soft tissue mass but was probably all vascular. A large rounded retro-tracheal mediastinal soft tissue mass displacing the trachea forward and laterally to the left was also seen. A right aortic arch anomaly was the entertained diagnosis but there was concern over the possible co-existence of either a mediastinal tumour of thoracic aortic aneurysm. CT scan with contrast enhancement (Fig. 2c) revealed a right aortic arch, postero-lateral to the trachea and oesophagus and an opacified structure to the left of the trachea considered the aberrant left subclavian artery. Other mediastinal mass lesions including a thoracic aortic aneurysm were confidently excluded. The descending aorta was seen lying to the right of the spine.

Case III. A 75-year old Saudi male, a known hypertensive for years, was admitted with a stroke. The previous chest radiographs had been interpreted as merely showing mediastinal widening due to marked aortic unfolding. However, the admission chest radiograph raised suspicion of a right aortic arch anomaly in addition to the aortic unfolding. The arch showed atheromatous changes with fairly extensive calcifications.

CT scans with contrast enhancement confirmed the clearly demonstrated calcified right aortic arch, postero-lateral to the trachea and oesophagus. The descending aorta was also identified lying to the right of the spine.

Discussion

All the three adult patients in this series had fairly characteristics plain chest radiographic appearances of retro-oesophageal right aortic arch and widened mediastinal shadows largely due to aortic unfolding but co-existence of a mediastinal tumour mass could not be excluded in each case. CT, in a non-invasive manner, was used to evaluate the suspected mediastinal shadows, confirming the right-sided aortic arch and confidently excluding other mediastinal lesions including thoracic aortic aneurysm.

The diagnosis of retro-oesophageal right aortic arch was made by identifying the lack of the usual left-sided arch and clearly demonstrating the right arch and the dilated origin of the aberrant left subclavian artery behind the oesophagus and trachea with both structures being displaced forward away from the spine. The shadow of the descending aorta on serial CT scans was seen along the right side of the spine then swinging over to the left side before reaching the diaphragm to pass through the normal aortic hiatus. It would seem that invasive angiographic diagnosis is no longer indicated for this usually asymptomatic condition.

In 1963, Felson and Palayew[9] reviewed the embryology and discussed the two types of right aortic arch commonly observed — namely the anterior or right aortic arch with mirror-image branching of the major arteries and the posterior or right arch with an aberrant left subclavian artery. Stewart and co-workers[10] in 1964 added a rarely encountered third type — the right aortic arch with isolation of the left subclavian artery. It is so rare that it is usually ignored as a possibility when interpreting the radiographs in patients with right aortic arch although one should be aware of this anomaly. It will no longer be referred to in this discussion.

At an early stage in the development of the heart and great vessels, the ascending and descending portions of the aorta are connected by two aortic arches, one on either side of the trachea and oesophagus. As the embryo matures, the right arch regresses almost completely while the left one remains and forms the adult aortic arch. The proximal segment of the embryonic right arch normally persists and establishes continuity with the root of the right subclavian artery. If, contrary to the normal, the left arch regresses the right persists the adult aortic arch will lie on the right side of the mediastinum. The anatomic configuration of a right aortic arch is determined largely by the manner in which the left arch regresses[9,10].

The anterior or right aortic arch with mirror-image branching results from interruption of the distal left arch adjacent to the descending aorta. The developmental process is essentially a mirror image of the normal and the order in which the vessels originate from this type of arch is the exact reverse of a normal left arch. There is therefore a right arch with three branches namely a left innominate artery, a right common carotid artery and a right subclavian artery in that order. The proximal portion of the left arch forms the left innominate artery and the left common carotid and left subclavian arteries become its two branches. In this type the right arch is anterior and to the right of the trachea and oesophagus. As a result there is no retro-oesophageal component and the oesophagus and trachea are not compressed from behind. The descending aorta is almost always entirely right-sided. The cardiac silhouette is rarely normal since about 98% of cases usually have associated cyanotic congenital heart disease especially tetralogy of Fallot and truncus arteriosus[9,10,11].

The posterior or right arch with an aberrant left subclavian artery is the most common type of right aortic arch, and usually exists as an isolated anomaly. The cardiac silhouette is usually normal since it is seldom associated with congenital heart disease[10,11]. It results from interruption of the left arch in its proximal portion near the ascending aorta between the left common carotid and left subclavian arteries. In this type the four vessels arise from the right arch in the following order: a left common carotid artery, a right common carotid artery, a right subclavian artery and an aberrant left subclavian artery. The proximal portion of the embryologic left arch forms the left common carotid artery which arises as the first branch of the aorta. The distal portion of the left arch may persist as an enlargement — the so called diverticulum of Kommerell[12]. The left subclavian artery arises from this aortic diverticulum at the junction of the right arch and descending aorta[19].

The descending aorta then usually descends to the right of the spinal column before entering the normal aortic hiatus. An important feature of this posterior type of aortic arch is the formation of a vascular ring encircling the trachea and the oesophagus. The ring is formed by the aortic arch on the right side, the pulmonary artery in front of the trachea, and the aortic diverticulum and left subclavian artery behind the oesophagus. The circle is closed by the ductus arteriosus which usually extends from the pulmonary artery to the left subclavian artery, along the left side of the trachea and oesophagus. In the great majority of cases, the ring is loose and does not cause significant compression evidenced by the fact that most arches of this type are first discovered as an incidental finding. All the three cases in this series have posterior type of right aortic arch, were asymptomatic and discovered as an incidental finding late in adult life.

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