Stanford Type B Aortic Dissection Involving a Right Aortic Arch — A Case Report and Literature Review

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Aortic dissection involving a right-sided aortic arch is a rare condition. A 50-year-old man with a history of poorly controlled hypertension was admitted with severe retrosternal pain. Computed tomographic scan showed a Stanford type B aortic dissection from the right aortic arch to the right descending aorta. As there was no indication for emergency surgical treatment, he received medical treatment to lower his systolic blood pressure to less than 120 mmHg, after which the chest pain subsided. He was discharged 2 weeks later with antihypertensive medications. There were no major complications after ten months of follow-up. We reviewed 25 similar cases reported in the literature. Although there are some differences in racial distribution, morphology, and complications, the treatment strategy for aortic dissection in a normal left aortic arch is applicable to those in a right aortic arch.

Key Words: Aortic dissection • Right aortic arch • Right descending aorta

INTRODUCTION

Right aortic arch occurs in approximately 0.1% of the general population.[1] Most cases that are diagnosed in childhood are symptomatic, with tracheoesophageal compression due to the abnormal branching pattern of the emerging vessels (i.e., aberrant left subclavian artery), a vascular ring, or other associated congenital heart diseases.[2] A symptomatic right aortic arch is only occasionally diagnosed in adulthood. To the best of our knowledge, only 24 cases of aortic dissection involving a right aortic arch have previously been reported in the literature.[3-14] Most were treated surgically because of related complications. However, given the rarity of the condition, management and prognosis are still unclear. We present here an unusual case of Stanford type B aortic dissection in a right aortic arch and right descending aorta managed medically. We also review the literature.

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CASE REPORT

A 50-year-old man with 6-year history of poorly controlled hypertension came to our emergency room in the early morning of May 19, 2002 complaining of retrosternal pain persisting for 3 hours and radiating to the back. The pain was unrelated to exertion but was associated with diaphoresis and nausea. Neither hoarseness nor swallowing difficulty was noted. On physical examination, his blood pressure was 184/96 mmHg and essentially equal in all four extremities. The heart sounds were normal and there was no audible murmur. Cardiac enzymes were within normal limits. Electrocardiography showed a normal sinus rhythm without any abnormalities. Chest X-ray (CXR) showed a normal-sized heart and a right aortic knob. There was an approximately 1.5

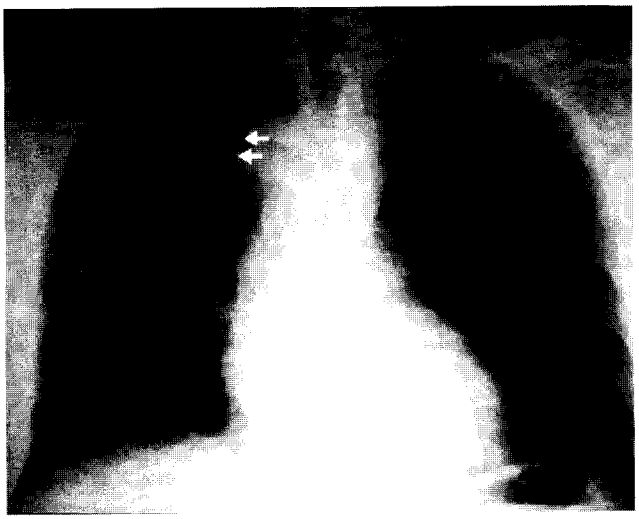

Figure 1. *CXR on admission shows a right aortic knob and a normal-sized heart. Intimal calcification of the aortic knob (white arrows) is noted.*

cm separation of the intimal calcification in the aortic knob from the outer aortic soft tissue border (Figure 1). Echocardiography showed concentric left ventricular hypertrophy with no other abnormalities. Contrast-enhanced computed tomography (CT) of the chest revealed a Stanford type B aortic dissection in the presence of a right aortic arch and descending aorta. The dissection flap was located on the lateral aspect of the arch involving Kommerel's diverticulum and the left subclavian artery and extended to the mid-descending aorta (Figure 2). On diagnosis of an acute Stanford type B aortic dissection, the patient was underwent aggressive management of his hypertension with labetolol and an angiotensin-converting enzyme inhibitor. His systolic blood pressure was maintained between 100 and 120 mmHg and his heart rate between 60 and 80 beats per minute, with rapid resolution of his chest pain. There

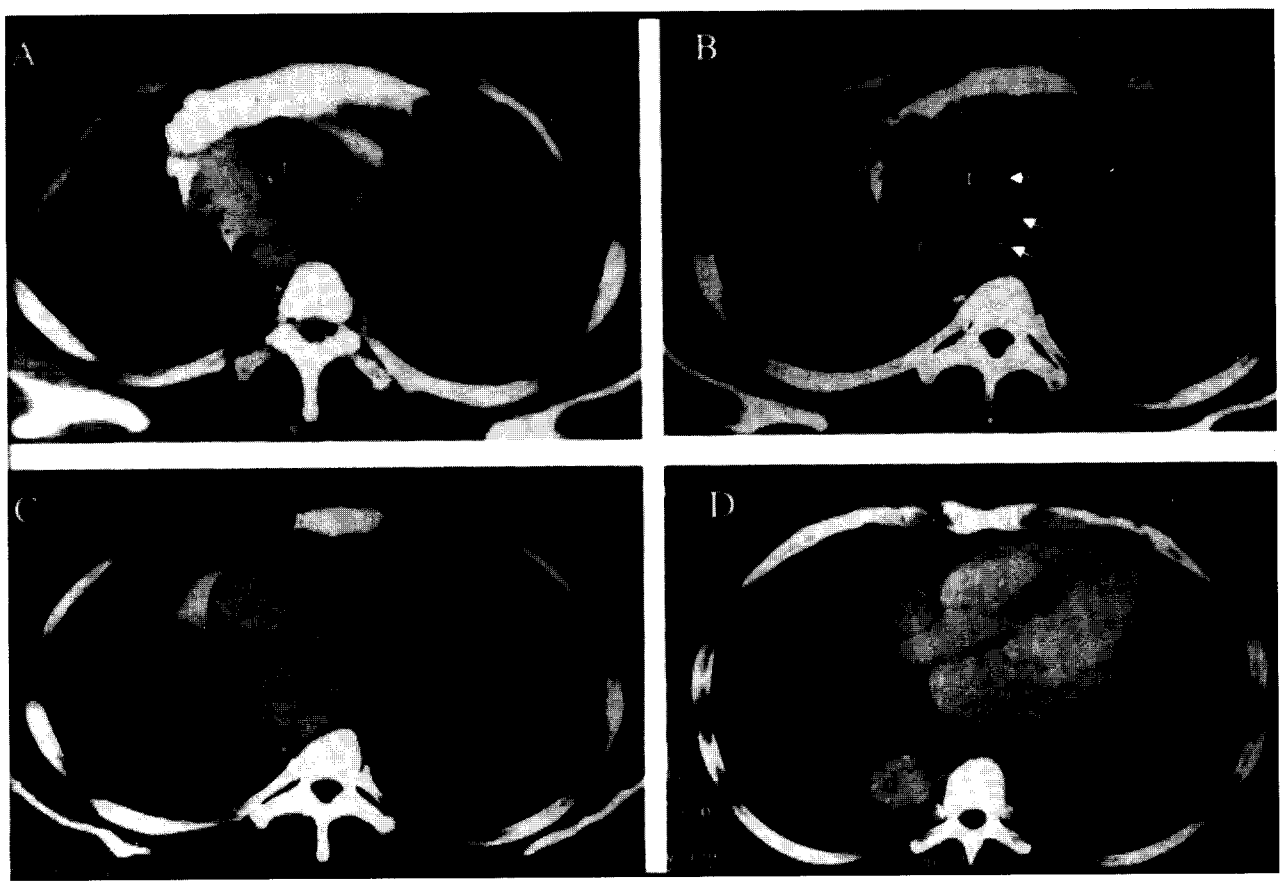

Figure 2. *CT on admission shows: (**A**) Right aortic arch with intimal calcification (long black arrow) and a dissection flap (short black arrows) at its lateral aspect. t = trachea. (**B**) Kommerell's diverticulum (K) with aberrant left subclavian artery running behind the trachea (t) and esophagus (long white arrow). The dissection flap also extends into the aberrant left subclavian artery (short white arrows). Short black arrows: dissection flap. (**C**) Aortic dissection spares the ascending aorta (a) but involves the proximal portion of descending aorta, which is separated into false (F) and true lumens (T). (**D**) The middle portion of the right descending aorta (d) is free of dissection.*

was no evidence of other end-organ damage, and he was discharged 2 weeks later. Thereafter, he complained of occasional chest pain. His blood pressure was not well controlled because of poor compliance. We informed him of the risk of recurrent aortic dissection if his blood pressure remained high. Magnetic resonance imaging (MRI) examination was also suggested, but he refused. CXR 7 months later did not show further expansion of the aortic knob or mediastinum. Because of his poor compliance, it was difficult to follow him with regularity. However, he remained stable for at least 10 months after the initial onset of symptoms.

DISCUSSION

A right aortic arch can be classified into three major anatomic types, depending on the branching pattern of the great vessels.[1,2] In the first or "mirror image" type, vessels branch from the aorta in a pattern that is a mirror image of a normal left aortic arch. From front to back, the vessels arising from the aorta are the left innominate, right common carotid, and right subclavian arteries. Congenital heart disease (CHD), especially tetralogy of Fallot, is present in as many as 75% to 98% of patients. Two of the reported cases of aortic dissection were the mirror-image type, but there were no associated congenital anomalies. The most common type shows the four great vessels arising from the arch in the following order: left common carotid, right common carotid, right subclavian, and aberrant left subclavian. The latter arises from Kommerell's diverticulum and courses posterior to the esophagus to reach the left arm. Most patients with this type are asymptomatic. However, tracheoesophageal compression may occur because of the vascular ring formed by the ascending aorta anteriorly, the transverse arch on the right, the diverticulum and aberrant left subclavian artery posteriorly, and the ligamentum arteriosum on the left. It is less frequently (5% to 10%) associated with major cardiac anomalies. The other 23 reported cases of aortic dissection belonged to this type, and two of the patients had congenital anomalies: a perimembranous ventricular septal defect and anomalous course of the left branchiocephalic vein. The third type is associated with an isolated left subclavian artery and is extremely rare. It may present with a subclavian steal syndrome.

Aortic dissection occurring in a right aortic arch is very rare. Acquired disease of the aorta does not appear to occur with increased frequency in patients with a right aortic arch.[3] The reason for the rarity of this condition might be that a significant percentage of patients have CHD that precludes survival to an age when aortic dissection is more common.[4] Of the 24 cases which have been reported since the first by Roan et. al. in 1979,[3] 18 were reported in Japan. However, the incidence of right aortic arch in Japanese wasn't different than in Western poplulations.[5] Therefore, it seems that there are racial differences in the incidence of dissection.

Of the reported cases, including ours, there have been 18 males and 7 females, with ages ranging from 39 to 75 years (mean, 56). The youngest two (39 and 45 years) had Stanford type A dissection. Seventy-two percent (18/25) had hypertension. On first presentation, most (20/25) complained of anterior chest or upper back pain. Other presentations included shock (1 case), upper abdominal pain (1), generalized malaise (1), and hoarseness, cough and dyspnea suggestive of tracheal compression (1). One asymptomatic case was discovered incidentally when a widened mediastinum was noted on routine CXR. Major complications included aortic insufficiency in 1 patient (with Stanford type A dissection), disseminated intravascular coagulopathy in 1 (who recovered after aggressive medical treatment), superior vena cava syndrome in 1 (improved after surgery), left subclavian or right iliac artery occlusion in 2 (associated with peripheral pulse deficits), aortic rupture in 5 (two of whom survived after emergency surgery), and swallowing difficulty or hoarseness in 6. The latter is the result of esophageal, tracheal, bronchus and/or right recurrent laryngeal nerve compression.[5] Acute dilatation of the ascending aorta, aortic arch, and Kommerell's diverticulum due to aortic dissection in a preexisting vascular ring may account for the increased incidence of tracheoesophageal compression. In a paper which reviewed 505 cases of aortic dissection in a normal aortic arch, only 1.2% (6/505) had hoarseness, and 1.8% (9/505) had dysphagia.[15]

Widening of the aortic silhouette and/or superior mediastinum was present in 80% (20/25) of these cases. The "calcium sign" on CXR was present in our case (Figure 1), but it is not mentioned in other published

cases. Pleural effusion was present in 4 cases. Two patients had a right-sided effusion associated with a right descending aortic dissection; one of these patients had aortic rupture. The other two both had proximal left descending and distal right descending thoracic aortic dissections. The effusion was on the left in one and bilateral in the other patient, who had aortic rupture.

Ninety-two percent (23/25) of reported cases had Stanford type B aortic dissection, a much more frequent incidence than in a left aortic arch, where only about one-third are type B.[16] Stanford type B dissection might occur more commonly in a right aortic arch for at least two reasons: (1) A right aortic arch has shorter radius and more acute curvature, and fixation at the origin of Kommerell's diverticulum by a left ductus ligament might increase aortic wall stress around the distal arch, increasing susceptibility to dissection;[6] (2) The descending aorta curves to the left at its mid-portion, which might increase local hemodynamic forces.[7] The primary intimal tear was identified in 21. It was near the junction of the aberrant left subclavian artery and Kommerell's diverticulum in 11, in the mid-portion of the right descending aorta in 4, in the ascending aorta in 2, and below the right subclavian artery in 4, including the 2 with the "mirror image" type. Two reported patients also had cystic medionecrosis.[3, 5]

Seventeen of the reported patients were operated on, of whom 2 had type A aortic dissection and 15 type B. Among the latter, the operation was performed in the acute phase (less than 2 weeks after symptom onset) in 5, and 1 patient died. The other 10 were operated on in the chronic phase, and 1 of those patients also died. The reported indications for surgery were expansion of the dissection on serial CT (6/17), tracheoesophageal compression (5/17), rupture of the dissecting aneurysm (3/17), Stanford type A aortic dissection (2/17), and compression of the true lumen of the descending aorta by the false channel (1/17). The last patient was operated on 3 years after onset of symptoms. Three others required surgery after more than 2 years of medical treatment, all because of expansion of the dissection. One patient had endovascular stent-graft placement with left subclavian artery ligation because of residual blood flow into the false lumen via the intimal tear.[10] For the other 6 previously reported patients receiving medical treatment only, the longest follow-up period was 6 months at the time the report was published. Two died within 2 months, presumably from aortic rupture precipitated by uncontrolled hypertension.[6, 9] Our patient has remained well except for occasional chest pain for 10 months.

The recommendations for medically treated patients are careful clinical observation and regular imaging studies (e.g., CXR, CT and/or MRI) as frequently as every 1 to 3 months in the first six months. The patients may then be reevaluated at 3- to 6-month intervals in the following 2 years, after which review every 6 to 12 months is appropriate. If surgical intervention is indicated, angiography is necessary to delineate the exact anatomy of the lesion (especially the great vessels) in order to minimize technical difficulties related to the anatomy.[8, 10] It is preferable to introduce the catheter through a right arm artery to avoid introduction of the guide wire into the false channel through the intimal tear.[6] Endovascular stent-graft placement is an alternative treatment for type B aortic dissection, but long-term studies are required to establish its safety and effectiveness.[8]

In conclusion, the treatment strategy for aortic dissection in a right aortic arch is similar to that for left arch dissections. It appears that right arch dissection has several epidemiologic differences, with more Japanese patients reported, Stanford type B dissection occurring more commonly, and a higher frequency of tracheoesophageal compression. Nevertheless, until more cases are available for analysis, our conclusions must remain tentative.

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右側主動脈弓併發 Stanford B 型主動脈剝離 — 病例報告及文獻整理

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右側主動脈弓併發主動脈剝離的病例相當罕見。本病例為 50 歲高血壓男性患者，因突發嚴重胸背痛而至急診室求治。經胸腹部斷層掃描檢查確定為右側主動脈弓及降主動脈合併 Stanford B 型主動脈剝離。因無緊急開刀的適應症，故依一般原則予以內科降血壓藥物積極治療。病人於二個星期後順利出院，經十個月的追蹤，並無明顯併發症產生。此篇並整理在文獻上的 25 個個案報告；雖然在人種分布、疾病類型及併發症上有些差異，但在治療原則上仍與一般主動脈剝離類似。

關鍵詞：主動脈剝離、右側主動脈弓、右側降主動脈。

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