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Aortic arch syndrome — Takayasu's arteritis in Nigeria

O. A. OGUNBIYI* AND A. O. FALASE†
*Departments of *Radiology and †Medicine, University College Hospital, Ibadan, Nigeria*

Summary

Four cases of aortic arch syndrome (Takayasu's arteritis), all in Nigerian females, are presented. Our clinical and roentgenological findings are described and the disease briefly discussed.

Résumé

Quatre cas du syndrome de la crosse de l'aorte (Takayasu's Arteritis), tous les quatre chez des femmes nigériannes, sont exposées. Nos observations cliniques et radiologiques sont décrites et la maladie brièvement discutée.

Introduction

Takayasu's arteritis is a rare disease of the aorta and its main branches. It has a patchy distribution causing coarctations, occlusion and aneurysmal dilatation with stenosis as its main characteristics [1,3]. First described in 1908 by a Japanese Ophthalmologist, Takayasu [4], it became known as a pulseless disease in 1952 after a description of its brachiocephalic manifestations [5]. Its many other names include atypical coarctation, aortic arch syndrome and primary or non-specific arteritis [6–8].

Its distribution is world-wide and has no territorial or racial boundaries [3]. It has been described most frequently in adolescent and young adult females, but it also occurs in the middle-age of this sex. It has occasionally been reported in the male [5,9].

Ladapo [10] in a recent roentgenological communication described a possible case of Takayasu's arteritis in a Nigerian but, surprisingly, without radiological representation. In this communication we report four such cases, encountered over a period of 5 years, as the first series ever clearly illustrated in Nigeria.

Correspondence: Professor A. O. Falase, Department of Medicine, University College Hospital, Ibadan, Nigeria.

Case Reports

Case 1

BO is a 16-year-old girl admitted with left heart failure. She also gave a history of dizzy spells and occasional fits. She was hypertensive with a blood pressure of 180/120 mmHg (taken over the right popliteal artery) and had scars of healed tuberculous cervical adenitis.

The radial, brachial and the axillary arteries as well as the left carotid artery were not palpable, the right carotid artery being palpable with a thrill and a bruit heard over it. There was clinical cardiomegaly with a biventricular heave and a raised jugular venous pressure. The electrocardiogram (ECG) showed sinus tachycardia, left axis deviation and left ventricular hypertrophy. The Heaf test was grade II positive.

The packed cell volume (PCV) was 34%, white blood cell count (WBC) 7900 per cmm with normal differentials, and erythrocyte sedimentation rate (ESR) 112 mm/h (Westergreen). The urinalysis, serum electrolytes and urea, and liver function tests were within normal limits. The Venereal Disease Research Laboratory test (VDRL) and Lupus erythematosus (LE) cell test were negative. Histological examination of biopsies of the deltoid muscle and temporal artery were also normal.

The chest X-ray showed biventricular cardiac enlargement with pulmonary congestion and basal pleural effusion. Intravenous urogram showed a normal right kidney but a small left kidney with delayed excretion and increased

concentration of contrast medium. The ureters and bladder were normal.

Total aortography revealed a normal aorta with occluded subclavians, vertebrals, left common carotid artery, and a narrowed tapered innominate artery (Fig. 1). The left renal artery was also completely occluded (Fig. 2).

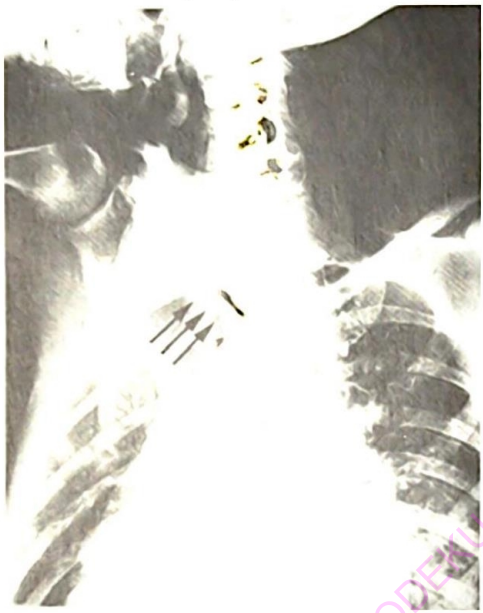

Fig. 1. Arch aortogram on Case 1. Note the characteristic flame-shaped tapered narrowing of the innominate artery (↑) and occlusion of the left common carotid, subclavian and vertebral arteries.

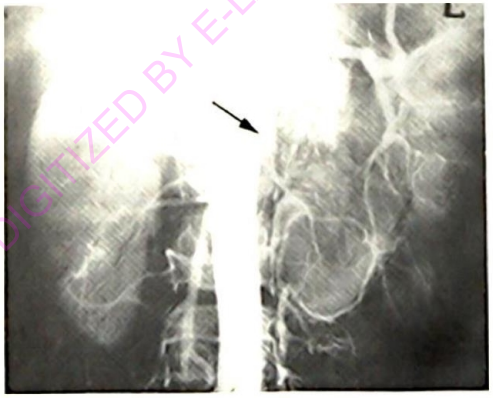

Fig. 2. Abdominal aortogram on Case 1. Note the complete occlusion of the left renal artery (↑).

Case 2

EE is a 45-year-old female admitted for subtotal thyroidectomy. She had a painless goitre and wanted it removed for cosmetic reasons. She was otherwise asymptomatic.

On examination, she was normotensive from blood pressure taken over the right popliteal artery. Cardiac examination was normal, but she had absent radial, brachial and axillary arterial pulsations on both sides. ECG showed left ventricular hypertrophy while her Heaf test was grade I positive. Her PCV was 36%, WBC 12,450 per cmm and ESR 98 mm/h (Westergreen). The urinalysis, serum electrolytes and urea, and liver function test were normal. The VDRL test was negative and no LE cells were detected in her blood, while biopsies of the deltoid muscle and temporal artery were normal on histological examination.

Her chest X-ray showed slight left ventricular cardiac enlargement and bilateral notching of the inferior aspect of the 4th–7th ribs. An intravenous urogram was normal. Total aortogram showed normal aorta, innominate and carotid arteries but occlusion of the subclavian and left vertebral artery with extensive scapular collaterals, and tortuous and dilated intercostal arteries which gave rise to the notched ribs (Fig. 3). All the branches of the abdominal aorta were normal.

Case 3

SS is a 50-year-old woman admitted for resection of a rectosigmoid carcinoma. She was

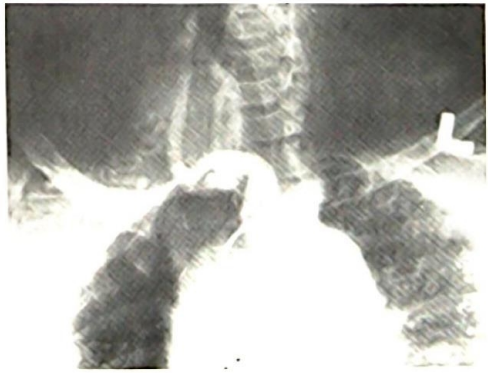

Fig. 3. Arch aortogram on Case 2. Note occluded subclavians and left vertebral artery, extensive scapular collaterals and tortuous intercostal arteries.

normotensive from blood pressure taken over the right popliteal artery, she had no clinical cardiomegaly and her optic fundi were normal. The radial and brachial arterial pulsations were absent. ECG showed right axis deviation and her Heaf test was grade II positive. Her PCV was 32%, WBC 7000 per cmm with normal differentials, and ESR 130 mm/h (Westergreen). The urinalysis, serum electrolytes and urea, and liver function tests were normal. The VDRL test was negative and no LE cells were detected in her blood. Biopsies of the deltoid muscle and temporal arteries were normal on histological examination. There was a slight left ventricular cardiac enlargement on her chest X-ray which also showed notching of the 3rd, 4th and 5th ribs inferiorly on both sides (Fig. 4). Her intravenous urogram was normal, a barium enema confirmed the presence of a recto-sigmoid carcinoma while a total aortogram revealed normal aorta, innominate and carotid arteries but occlusion of the subclavian and vertebral arteries with tortuous intercostal arteries. The branches of the abdominal aorta were all normal.

Case 4

AO is a 40-year-old female admitted in coma with a right-sided hemiplegia. She was normotensive, and had neither clinical cardiomegaly nor abnormal heart sounds.

Both radial, brachial, axillary and left carotid arterial pulsations were absent. Fundoscopy revealed temporal pallor of the discs and tortuous retinal veins, but no new blood vessel formation was seen.

Her ECG was normal while the Heaf test was grade II positive. PCV was 30%, WBC 6400 per cmm with normal differentials and ESR 95 mm/h (Westergreen). Urinalysis, serum electrolytes and urea, and the liver function tests were normal while the VDRL was negative, and no LE cells were detected in her blood. Biopsies of the deltoid muscle and temporal artery were normal histologically. Her chest X-ray showed a slight left ventricular cardiac enlargement.

An intravenous urogram was normal while a total aortogram showed a normal aorta and right carotid artery but occluded left carotid and both vertebral arteries, and characteristic flame-shaped narrowing of the innominate artery (Fig. 5).

Discussion

All our patients were female Nigerians. One was an adolescent girl of 16 and the remaining three were middle-aged ladies.

The exact aetiology of Takayasu's arteritis is unknown. Malnutrition [11] and auto-immune reactions [7] have been implicated and an association with tuberculosis considered [7,11]. It is possible that tuberculosis could have caused the arteritis of Case 1, although her tuberculosis was no more active when she presented in hospital. Of the four patients described, the first had associated tuberculosis, the second a goitre, the third a rectosigmoid carcinoma while no associated lesion was found in the fourth. This suggests that Takayasu's disease is a reaction, possibly auto-immune, to a number of varied factors [7].

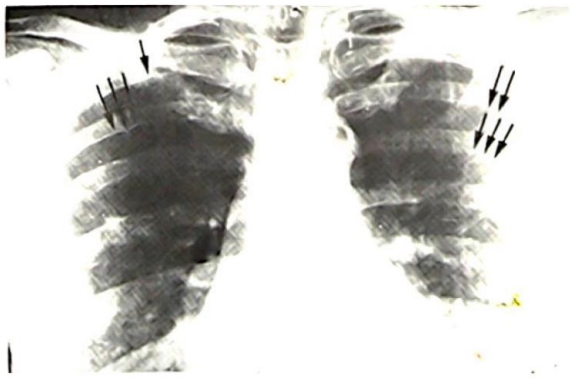

Fig. 4. Chest X-ray on Case 3, showing inferior notching of 3rd, 4th and 5th ribs bilaterally.

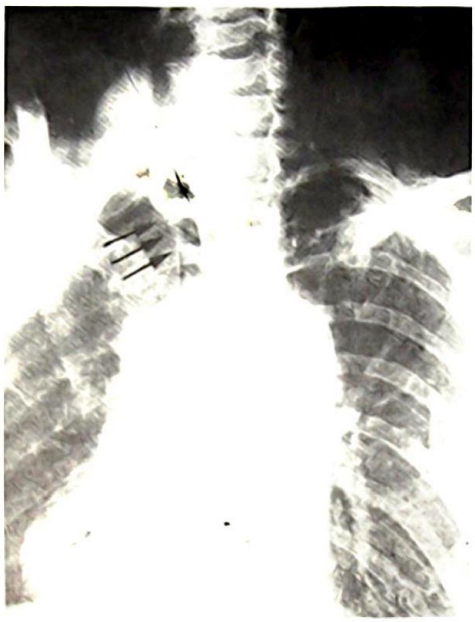

Fig. 5. Arch aortogram on Case 4. Note the occlusion of the left carotid, subclavian and vertebral arteries and the tapered narrowing of the innominate artery (↑).

Other possible causes of aortitis, i.e. atherosclerosis, syphilis, tuberculosis, chronic dissecting aneurysm, collagen disorders, idiopathic aortitis in the African, etc. were considered and excluded. In particular, differentiation of Takayasu's arteritis from idiopathic aortitis in the African was made. Aortic arch syndrome is a rare clinical presentation in idiopathic aortitis in the African where the majority of cases have involvement of abdominal aorta only or abdominal and thoracic aorta with maximal changes in abdominal portion — the characteristic roentgen findings being thickening of the aortic wall, dilatation and irregularity of diffuse or localized stenosis of the abdominal or thoracic aorta [6,8].

Roentgenological examinations were most useful in arriving at the diagnosis in all cases. The plain radiographs of the chest may be normal or there may be secondary cardiac changes. Rib-notching due to dilated intercostal arteries secondary to subclavian obstruction has been described.

Total aortography is of immense value in the diagnosis of Takayasu's arteritis [2,12] as it also shows the extent and severity of the disease. All four of our cases had stenotic and occlusive lesions with peaking, tapering or flame-shaped appearances — in the brachiocephalic arteries — characteristic of Takayasu's disease [3,12]. Three cases had normal abdominal aortographic findings but Case 1, the adolescent girl, had complete occlusion of the left renal artery. Of particular interest is the fact that she was the only hypertensive patient in our series. Stenosis or occlusion of one or both renal arteries occurs in 25–75% of cases [7,13]. Unlike the other major branches of the aorta, the renal artery is an end-artery and a collateral supply rarely develops sufficiently to prevent renal ischaemia and renovascular hypertension.

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