Takayasu's Arteritis: Angiographic and Duplex Finding

Tarek Rashad Saleh, MD, Asma Al-Dabbagh, MD, FRCR
Hassan Al-Prince, MS, Hassan Ali Al-Zahrani, FRCS(G)

To our knowledge, this is the first report of Takayasu's arteritis from Saudi Arabia. Seven patients out of ten were Saudis. Seven were females and three were males. Chest radiograph is invaluable in the evaluation of this disease since it allows visualization of the contour of the aorta with calcification of aortic wall. Carotid duplex can initially suggest the diagnosis based on characteristic sonographic features of the disease, however, panarteriography helps in evaluation of the extent of the disease and the state of collateralization.

Introduction

This peculiar arteritis was first noted in 1908 by the Japanese ophthalmologist Takayasu[1], subsequently this disease entity has been described by a variety of terms that reflect some of its many features, i.e. aortic arch syndrome, pulseless disease, reversed coarctation, occlusive thromboaortopathy, young female arteritis as well as Takayasu's arteritis[2]. This disease occurs world wide, the majority of the reports of this condition have come from Japan, Korea, China, Southeast Asia, Mexico and India with occasional reports from North American and European Caucasians[3,4]. To our knowledge, not a single report came out from Saudi Arabia.

Materials and Methods

Ten pateints with Takayasu;s arteritis were seen at King Abdulaziz University Hospital, or referred for angiography from King Fahd General Hospital, Jeddah. Saudi Arabia between the period from October 1991 and October 1993. All patients were evaluated by duplex scan and angiography. Seven were Saudis and three were Non-Saudis (2 Palestinians and 1 Sudanese). Their age ranged from 21 to 42 with an average of 32 years. There were seven females and three males. The diagnosis was made on the basis of the proposed criteria for the clinical diagnosis of Takaysu's arteriography[5]. These included clinical symptoms and signs, laboratory findings and the results of angiography. The diagnosis was also based on the characteristic duplex findings[6]. In six patients, the diagnosis was proven by surgical arterial biopsy. Patients who had only single vessel involvement without histologic confirmation were excluded from the study. Two patient went for arterial bypass operations.

All patients were examined by DSA thoracic and abdominal aortography through intraarterial route. For thoracic aortography, a pigtail catheter tip was placed at the aortic root and 30 ml of omniopaque 240 (Iohexol) was injected at a rate of 15/sec. For thoraco-abdominal aortography, the catheter tip was placed in the thoraco-abdominal aorta at the level of 11th thoracic vertebra and 20 ml Omniopaque 300 was injected at a rate of 10/sec. Other DSA runs were taken when necessary. Also, selective catheterization of either common carotid or subclavian were done according to the clinico-radiological circumstances. Pulmonary scintigraphy was done only in one patient who suffered three attacks of pulmonary embolism.

Results

Clinically, eight patients presented by diminished or absent pulses; 4 of them showed audible bruit, six patients had recurrent headache (mostly either due to brain ischemia or hypertension); two of them had

Address for correspondence: Dr. Tarek Rashad Saleh, Radiology Department, King Abdulaziz University Hospital, P.O.Box 6615, Jeddah 21452, Saudi Arabia.

Table 1: Distribution of Arterial Lesion in 10 Patients with Takayasu's Aortitis

Nos	Age	Sex	Nationality	Thoracic Aorta	R.CCA	L.CCA	R.SC	L.SC	Abdominal Aorta	Pulmonary
1	29	F	S	-	-	+	*	+	-	-
2	28	F	S	-	-	+	-	-	-	-
3	36	F	S	-	-	-	-	*	-	-
4	35	F	Palestinian	-	-	+	+	-	-	-
5	33	M	S	-	+	+	-	+	-	-
6	21	F	S	-	+	-	+	-	-	-
7	42	F	S	+	-	-	*	+	-	-
8	37	M	Sudanese	-	+	*	-	*	-	-
9	27	F	S	-	-	-	-	-	+	-
10	32	M	Palestinian	-	-	-	-	+	-	+

Legend

F	: Female	+	: Occluded vessel
M	: Male	-	: Patent Lumen
R.CCA	: Right common carotid artery	*	: Narrowed vessel
L.CCA	: Left common carotid artery	S	: Saudi
R.SC	: Right subclavian artery		
L.SC	: Left subclavian artery		

syncopal attacks. Four patients presented by ischemic manifestation of the brain (three TIA and one extensive-stroke). Two had abdominal angina and one showed aortic valve regurgitation. Only one patient suffered pulmonary manifestation who presented by recurrent attacks of shortness of breath which was secondary to pulmonary embolism. None of our patients presented in the acute systemic initial phase. Laboratory findings showed that seven patients had elevated erythrocyte sedimentation rate at presentation. On chest radiography, two patients showed linear, irregular calcification of the aortic arch and descending thoracic aorta.

On carotid Duplex, all the affected common carotid arteries had diffuse, homogenous, circumferential vessel wall thickening, causing a long segment of smoothly narrowed patent lumen or thrombosed lumen. Internal, external and carotid bifurcation were spared in all cases. The flow in the narrowed segment had high velocities and increased resistance with turbulence, while distal to the narrowed segment, the flow was dampened.

By angiography, according to Nasu classification[7], eight patients had Type I with involvement of the branches of the aortic arch only, one patient has Type II with involvement of the thoracic aorta and its branches, and one patient had Type III with involvement of the abdominal aorta and its branches, however, no patient showed Type IV with extensive involvement of both thoracic and abdominal aorta.

According to Yamato, et. al.[8], we did not include involvement of pulmonary artery in the classification of Takayasu's aortitis. Only one patient with Type I aortitis had involvement of the pulmonary artery which was proved by scintigraphy.

Table I shows distribution of vessel involvement.

Discussion

In our study, we reported 10 cases of Takayasu's arteritis; seven of them were females and three were males with a mean age of 32 years. This agreed with Yamato et. al[8] who stated that Takayasu's arteritis is much common in young females. Ishikawa[5] pointed out that patient's age should be ≤40 years at the onset of characteristic signs and symptoms. in our study, all patients were below 40 years except one (42 years). If we reviewed her initial symptoms, we found that she was symptomatic 9 years earlier and was misdiagnosed. This confirms Hall et. al.[4] statement that the classical signs and symptoms of Takayasu's arteritis had frequently been noted for several years prior to diagnosis with delay between first symptom and time of diagnosis ranging from 1 month to 16 years.

The chest scintigraphy is invaluable in the evaluation of this disease since it allows visualization of the contour of the aorta, calcification in the aortic wall, rib notching and pulmonary arterial and cardiac changes[8,9,10].

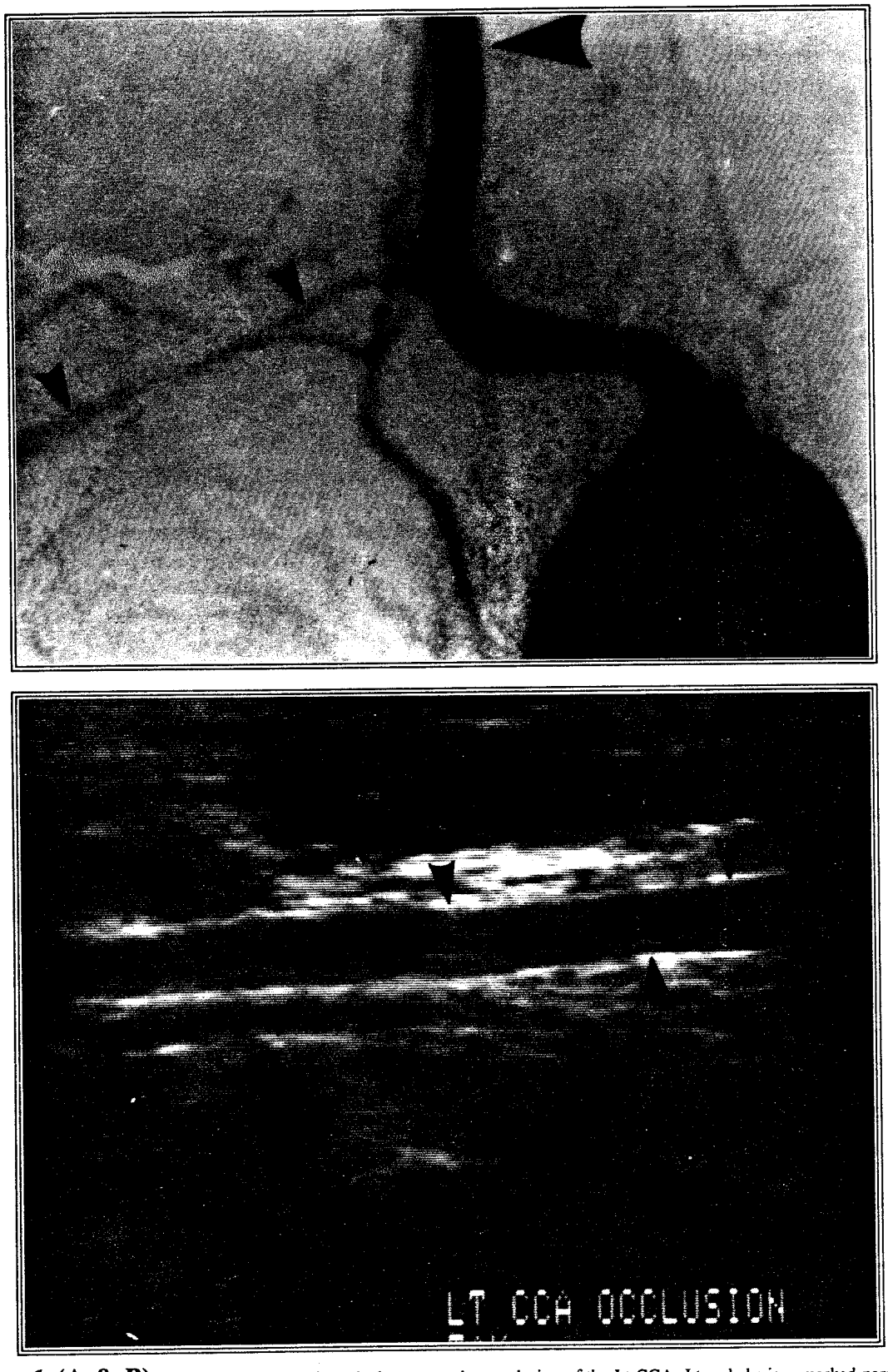

Figure 1 (A & B): [A] DSA for the aortic arch shows complete occlusion of the Lt.CCA, Lt. subclavian, marked narrowing of Rt. subclavian and occluded Rt. vertebral. (Small arrow heads point to Rt. subclavian and large one points to Rt.CCA). This 29 year old female was proved to have Takayasu's arteritis. [B] Long B-scan for Lt.CCA shows concentric wall thickening with narrowed thrombosed lumen.

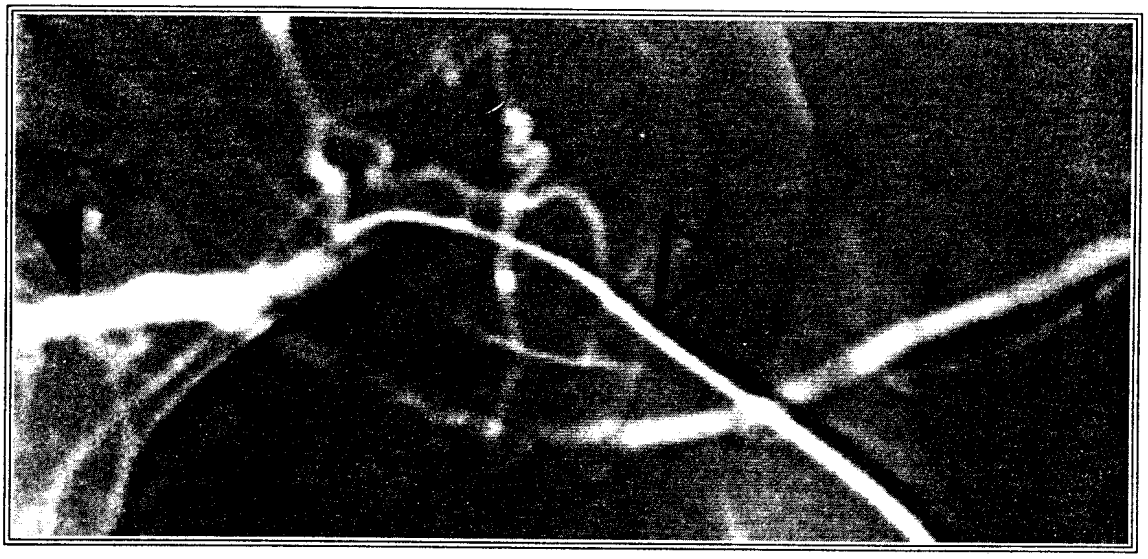

Figure 2: Selective catheterization of right subclavian (large arrow head) visualizes retrograde filling of the venous graft (small arrow heads) that carried blood flow to the left axillary artery.

Figure 3: 32 year old male who was proved to have Takayasu's arteritis. Post operative DSA for the aortic arch which was done to ensure patency of the venous graft (black arrow heads) that shunted blood from right subclavian to left axillary artery. It also demonstrates complete occlusion of right CC (open arrow), left CCA and left subclavian (long arrow). The small arrow points to the patent right vertebral.

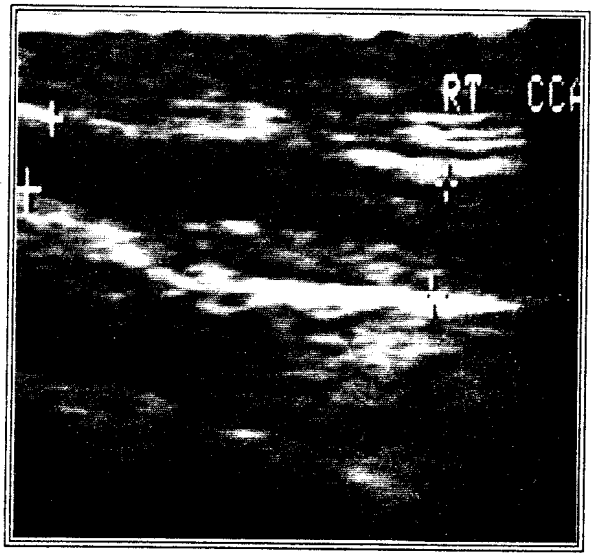

Figure 4: Long B-scan for the right common carotid artery shows marked concentric wall thickening with thrombosed narrow lumen.

Although, pulselessness of the upper extremities or hypertension are the usual clues that suggest the diagnosis, the chest radiography may be the first examination to suggest this disease[9,11]. Irregular contour and calcification in the aorta, very unusual in premenopausal women[10], should alert the radiologist to the diagnosis, although they are not early manifestations of this disease. In our study, two patients (20%) showed linear irregular calcification of the aortic arch and descending thoracic aorta. This agreed with the previous report of Hachiya[12] who reported that the incidence of calcification ranges from 10% to 25% and contour irregularity ranges from 10% to 73%.

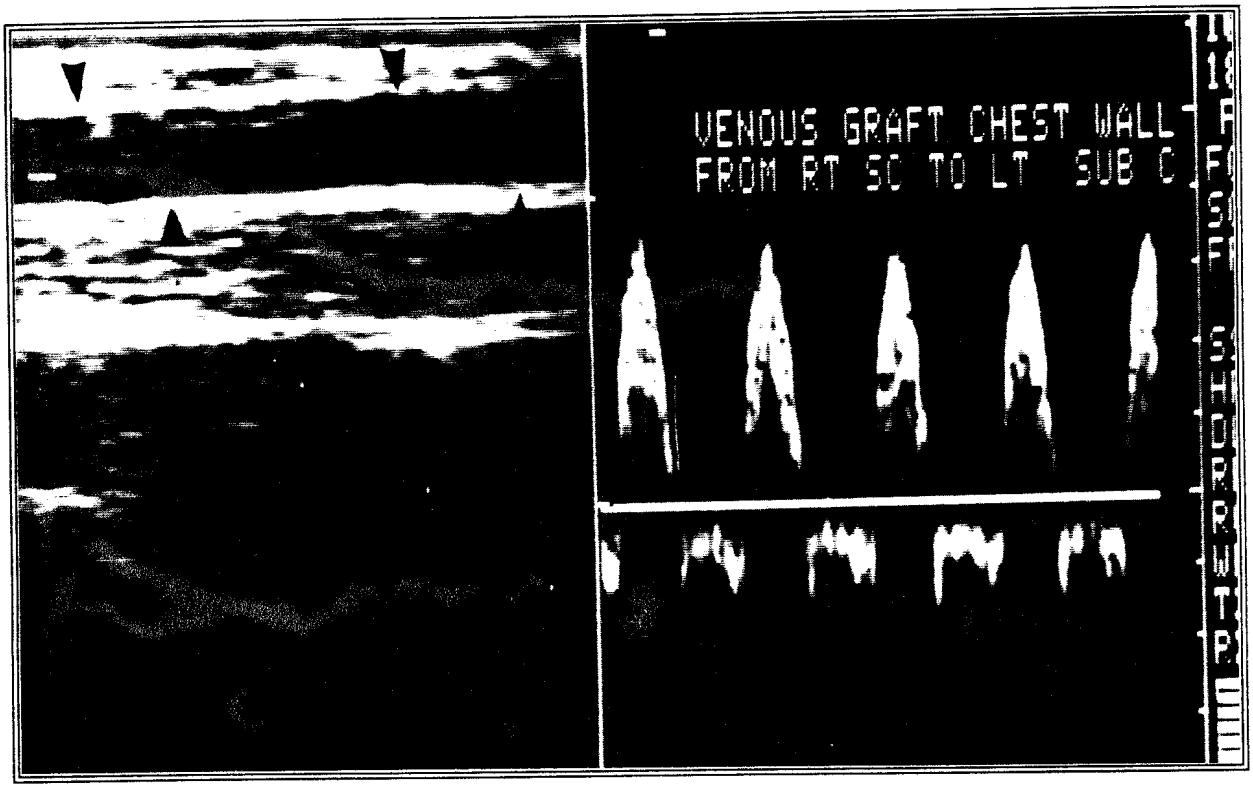

Figure 5: Duplex for the venous graft, that runs transversely and subcutaneously on the chest, demonstrates its patency. The flow during diastole is negative as there is no elastic recoil in the graft wall.

Figure 6: DSA for the aortic arch shows occluded brachycephalic trunk. The left vertebral arises directly from the aortic arch.

Figure 7: Long B-scan for the right CCA demonstrates marked intimal proliferation that leads to concentric wall thickening and tight thrombosed lumen.

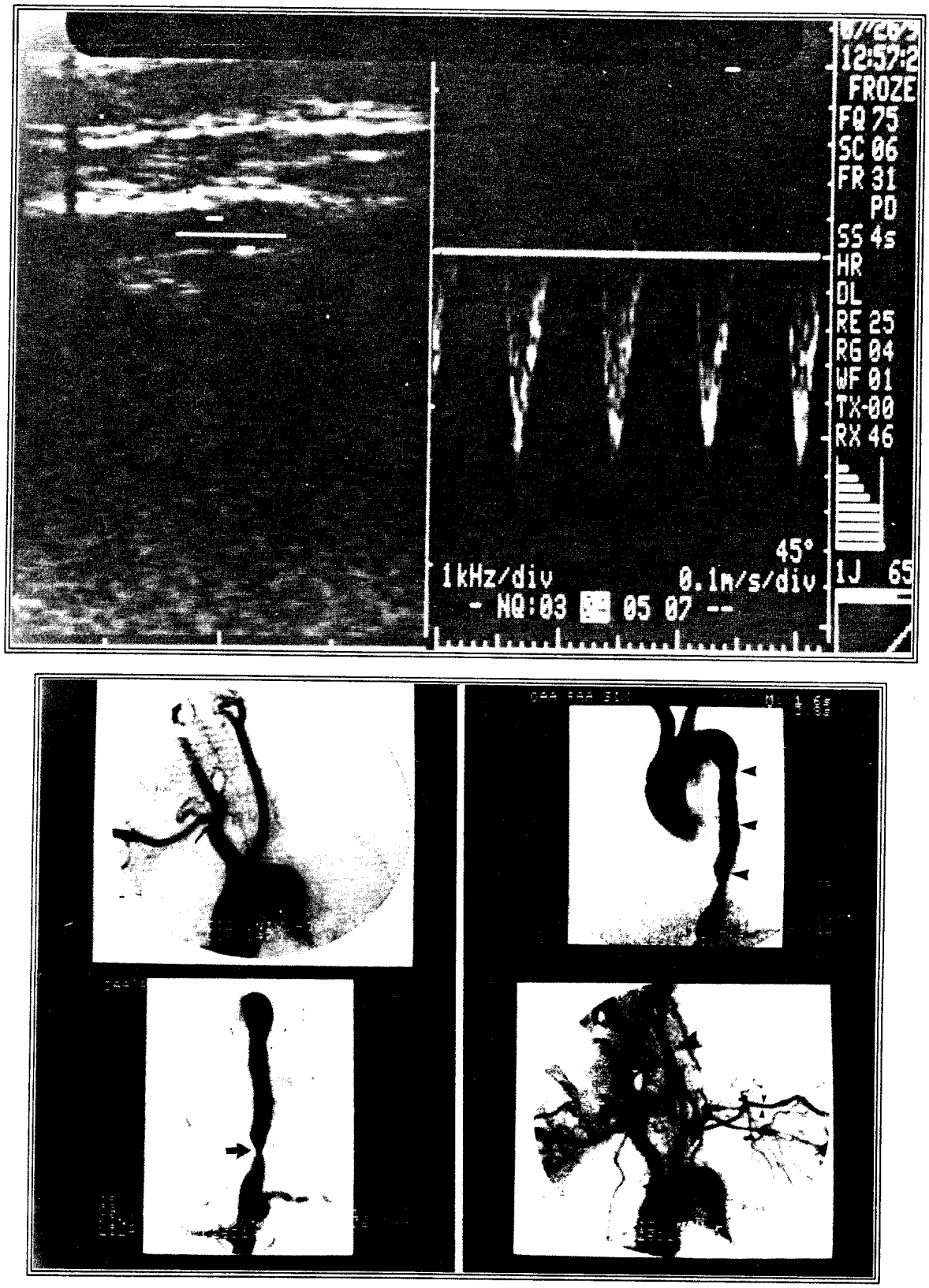

Figure 8 (A & B): [A] Demonstrate reversed flow in left vertebral (subclavian steal). [B] In the first frame, complete occlusion of left subclavian, severe stenosis of RSC origin (small arrow). The second and third frames show the narrowed descending thoracic aorta. The last frame shows retrograde filling of left vertebral (arrow head) and left subclavian. That picture confirmed the presence of subclavian steal.

On carotid Duplex, the common carotid artery was affected, while internal and carotid bifurcation were spared on all cases. The affected vessel showed diffuse, homogenous, circumferential wall thickening with long segment or narrow or occluded lumen. These diagnostic sonographic features were previously described by Bond et. al.[6]. They correlate well with the described pathological and anatomical features of Takaysu's arteritis. Actually, these specific sonographic features help to differentiate this disease from other disease entities specially atherosclerosis where it usually affect bifurcation, fibromuscular dysplasia and idiopathic carotid dissection involving the internal carotid, while temporal arteritis affects the external carotid artery. The common carotid artery affection was found in 40% of the patients (8 out of 20 vessels). Five on left and three on right. In the previous reports, the CCA involvement ranges from 45% to 70%. The most pertinent to this discussion is the fact that CCA is the easiest artery to be examined by duplex, as it is relatively large, superficial with clear surface anatomy. Park et. al.[14] reported that the left common carotid was frequently more affected than the right one.

The findings in our angiographic studies are similar to those in Nasu's autopsy series[7] where the aortic arch vessels are affected in all types except Type III which is much less common. This type was only recorded in one patient in our series (10%) while aortic arch vessel affection was found in the remaining nine patients. Since this disease can involve the entire aorta and its branches, total aortography is mandatory for diagnosis, evaluation of the extent of the disease and the state of collateralization specially when surgical grafts or bypass are considered.

References

1. Takayasu M. A Case with peculiar changes of central retinal vessels(In Japanese). Acta Societatis Ophthalmologicae Japonicae, 1908;12:554-555.
2. Baker RA, Rumbaugh CL, Kido DK. Pathology of cerebral vessels. In:Abrams HL. Abrams angiography, vascular and interventional radiology. 3rd ed,:Little, Brown and Company, Boston 1983: 278-280.
3. Lande A, Bard R, Rossi P, Passariello R, Castrucci A. Takayasu's arteritis: A worldwide entity. NY State J Med 1976;76:1477.
4. Hall S, Ban W, Lie JT, Stanson AW, Kazmier FJ, Hunder GG. Takayasu's arteritis: A study of 32 North American patients. Medicine 1985;64:89-99.
5. Ishikawa K. Diagnostic approach and proposed criteria for clinical diagnosis of Takayasu's arteriopathy. J Am Coll Cardiol 1988;12:964.
6. Bond JR, Charboneau JW, Stanson AW. Takayasu's arteritis: Carotid duplex sonographic appearance, including color doppler imaging. J Ultrasound Med 1990;6:625-629.
7. Nasu T. Aortitis syndrome: Pathological aspect (In Japanese). Gendai Iryo 1976;8:1143-1150.
8. Yamato M, Lecky JW, Hiramatsu K, Kohda E. Takayasu's arteritis: Radiographic and angiographic finding in 59 patients. Radiology 1986; 161;329-334.
9. Kosuka T, Nosaki T, Sato K, Ihara K. Roentgenologic diagnosis of atypical coarctation of the aorta. Acta Radiologica 1966; 4:497-507.
10. Linuma J. Chest roentgen symptoms in the pulseless disease (In Japanese). Nippon Igaku Hoshasen Gakkai Zasshi 1961;20:2548-2569.
11. Gillanders LA, Strachan RW. The role of radiology in the diagnosis of Takayasu's arteriopathy (pulseless disease). Clin Radiol 1965; 16:119-129.
12. Hachiya J. Current concept of Takayasu's arteritis. Semin Roentgenol 1970;5(3):245-259.
13. Srur MF, Sos TA, Saddekni S, Cohn DJ, Rozenblit G, Wetter EB. Intimal fibromuscular dysplasia and Takayasu's arteritis: delayed response to percutaneous transluminal renal angioplasty. Radiology 1985; 157:657-660.
14. Park JH, Han MC, Kim SH, Oh BH, Park YB, Seo JD. Takayasu's arteritis: Angiographic findings and results of angioplasty. AJR 1989; 153:1069-1074.