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A Rare Case of Aortic Arch Anomaly with Subclavian Steal Syndrome

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This paper discusses a rare case of aortic arch anomaly with subclavian steal syndrome. The images of our diagnostic findings are presented and compared with findings in healthy individuals.

Key words: *aortic arch anomaly, subclavian steal syndrome, functional diagnostics, blood pressure.*

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Introduction

Aortic arch anomaly is a rare form of cardiovascular system (CVS) congenital disorders [1–3]. Subclavian steal syndrome (SSS) is an occlusion or stenosis of the subclavian artery (SA) causing retrograde flow in the vertebral artery (VA) and vertebrobasilar insufficiency (VBI) [4, 5]. In this paper we present a clinical case of SSS due to aortic arch anomaly that was diagnosed with non-invasive procedures.

Case report

A 53-year female with a history of diffuse systemic scleroderma with sclerodactyly, hyperpigmentation, telangiectasia, tightening of the mouth area, Raynaud's phenomenon, achalasia, and fibrosing alveolitis. She had stage 0–1 dyspnea and a history of pneumonitis.

The patient's complaints at admission and during hospital stay were typical for systemic scleroderma.

The patient has had systemic scleroderma for over 20 years. She's been previously treated with cytostatics, systemic glucocorticoids, antifibrates, vascular agents, calcium-containing agents, and peripheral vasodilators. The patient wasn't adherent to the recommended treatment altering the dose or timing of a medication. For over 9 years there were signs of bilateral interstitial lung disease.

The patient has been overweight for many years. For the last 5 years she has been diagnosed with Cushing syndrome. The patient underwent menopause at age 42. She was diagnosed with osteoporosis due to menopause and glucocorticoid treatment 3 years ago. Her blood pressure (BP) was usually within normal limits with rare elevations up to 130/90 mmHg. The patient denied having a history of stroke, coronary artery disease (CAD), or diabetes.

Physical exam findings matched the clinical diagnosis. Excessive accumulation of adipose tissue in the abdominal region was noted. Body mass index (BMI) was 31.2 k^2. Body temperature, oxygen saturation, respiration rate, and heart rate were within normal limits during the hospital stay. BP was always checked at the left arm and normal. Evaluation and clinical management were performed according to current guidelines and standard algorithms for systemic scleroderma management. Laboratory and instrumental findings also confirmed the clinical diagnosis. Electrocardiography and echocardiography (Echo-CG) [6] revealed no abnormalities. 6-minute walking test was 430 m.

The presence of obesity, BP elevation episodes, dyslipidemia, long-term use of glucocorticoids with Cushing syndrome necessitated carotid artery duplex scan for cardiovascular disease (CVD) risk stratification. Carotid artery duplex showed normal blood flow in the common carotid (CCA), external carotid (ECA), and internal carotid arteries (ICA). Blood flow linear velocity (BFLV) was also normal without significant asymmetry (Table 1). Intima-media ratios (IMR) of the right and left CCA were normal (0.9 mm on the right and 0.99 mm on the left). Thickness and echo density were even, layers well-differentiated. However, we found that left and right CCA arised from the common trunk with 7.4 mm diameter and 8.0 length from aortic arch (Figures 1, 2). Left subclavian artery (LSCA) was 7.6 mm in diameter and had normal location (started from the aortic arch) and angle with aortic arch; BFLV was 96 cec. Right subclavian artery (RSCA) was 5.0 mm in diameter, but we couldn't determine its origin. It probable arised from the posterior wall of the descending aorta. RSCA turned out to be collateral with reduced systolic BFLV (46 cec) and no phases (Figures 3b). It normally should have high amplitude and three phases (Figure 3a). Right extracranial vertebral artery had retrograde flow; left — antegrade flow (Figure 4).

Table 1. **Extracranial carotid artery duplex scan parameters**
(GE Vivid S70 Ultrasound Machine with GE 11L–D Linear Probe, 1–6 MHz)*

Vessel	Right BFLV				Left BFLV			
	PS, CEC	DIAST, CEC	RI	D, MM	PS, CEC	DIAST, CEC	RI	D, MM
CCA	78	19	0.75	6.8	99	20	0.80	6.4
ICA	64	24	0.61	4.4	68	21	0.68	4.5
ECA	—**	—	—	3.5	—	—	—	4.5
VA	73	11	0.85	4.0	54	15	0.72	4.9

Note. *— reference values were determined according to critical parameters specific for this pathology [7]. ** — no clinical significance. BFLV — blood flow linear velocity, CCA — common carotid artery, d, mm — vessel diameter; Diast, cec — diastolic velocity; ECA — external carotid artery, ICA — internal carotid artery, Ps, cec — peak systolic velocity; RI — resistive index; VA — vertebral artery.

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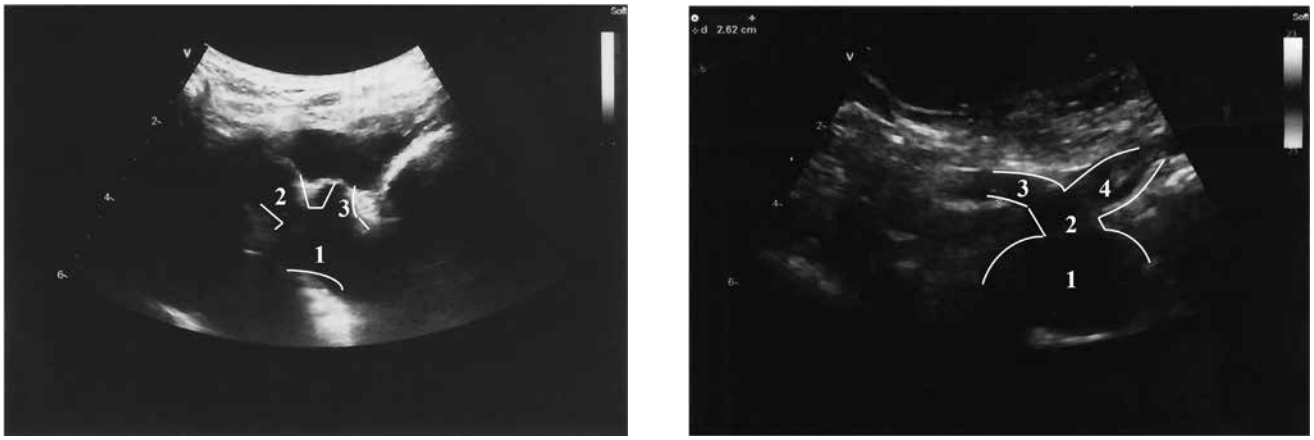
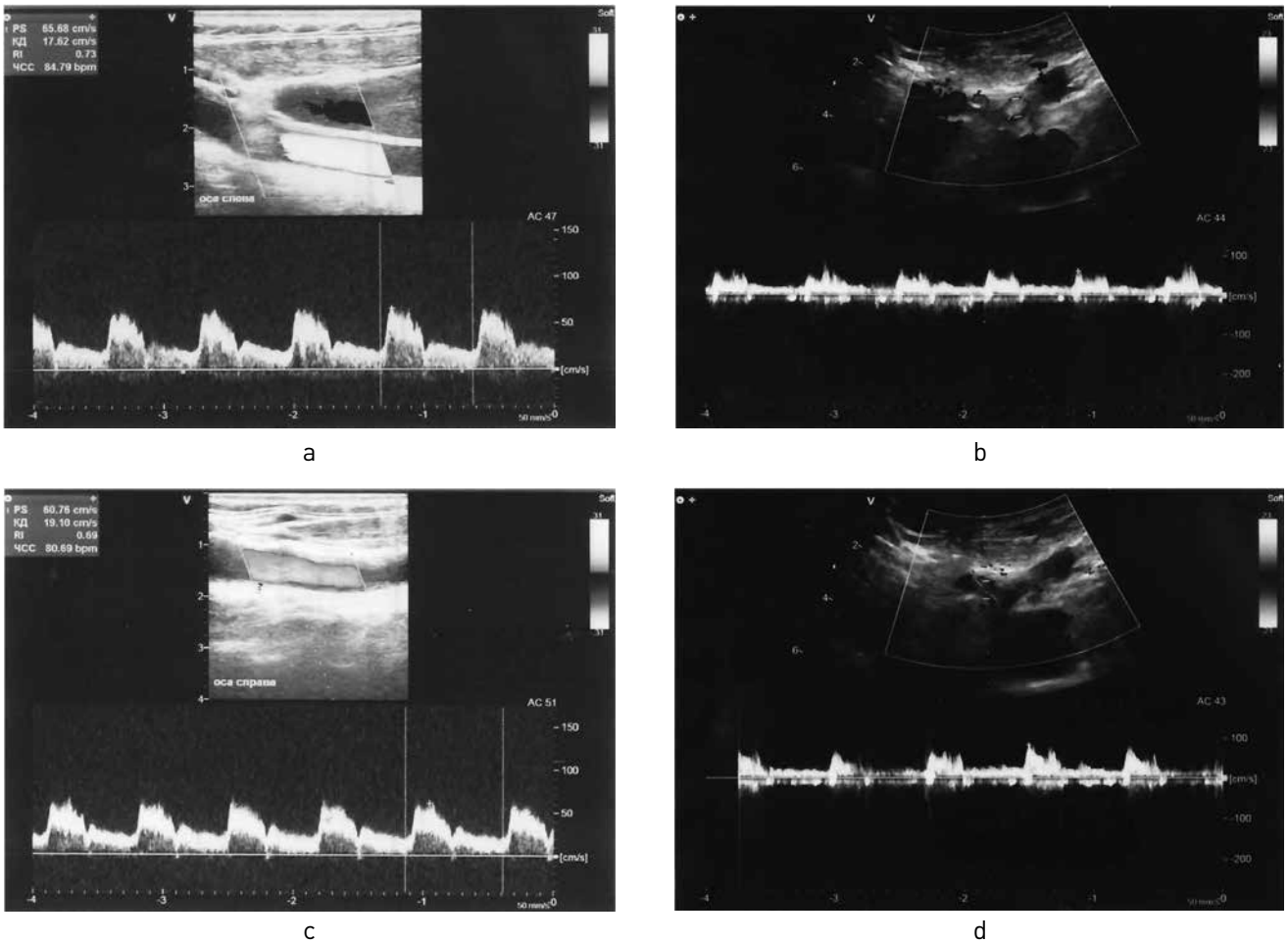


a b

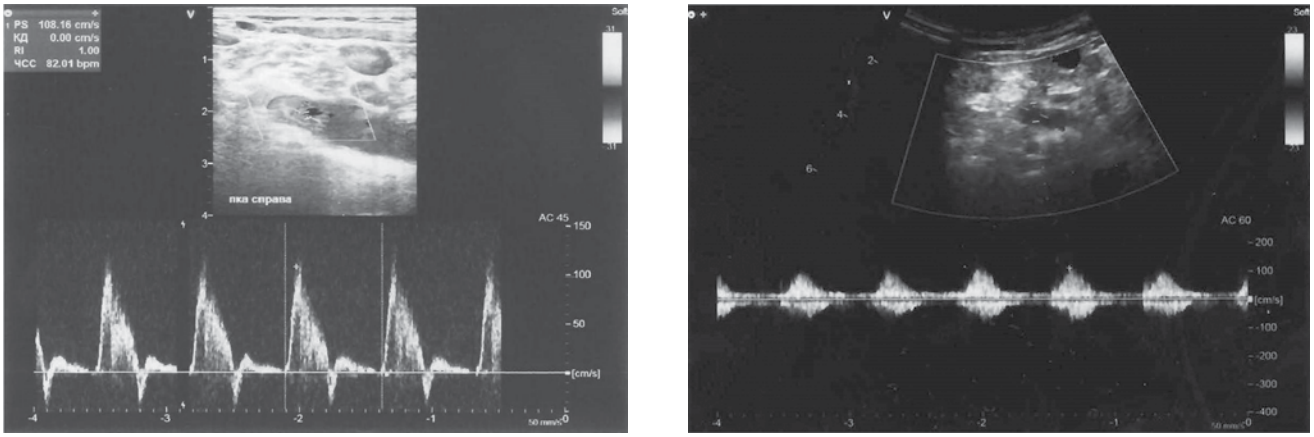
Figure 1. Carotid artery duplex scans showing aortic arch in: a — a healthy individual (1-aortic arch, 2 — brachiocephalic trunk, 3 — left CCA); b –aortic arch anomaly (1 — aortic arch, 2 — common trunk of the right and left CCA, 3 — right CCA, 4 — left CCA) (further from here — scans are compared with images from a healthy female patient of the same age).



a b

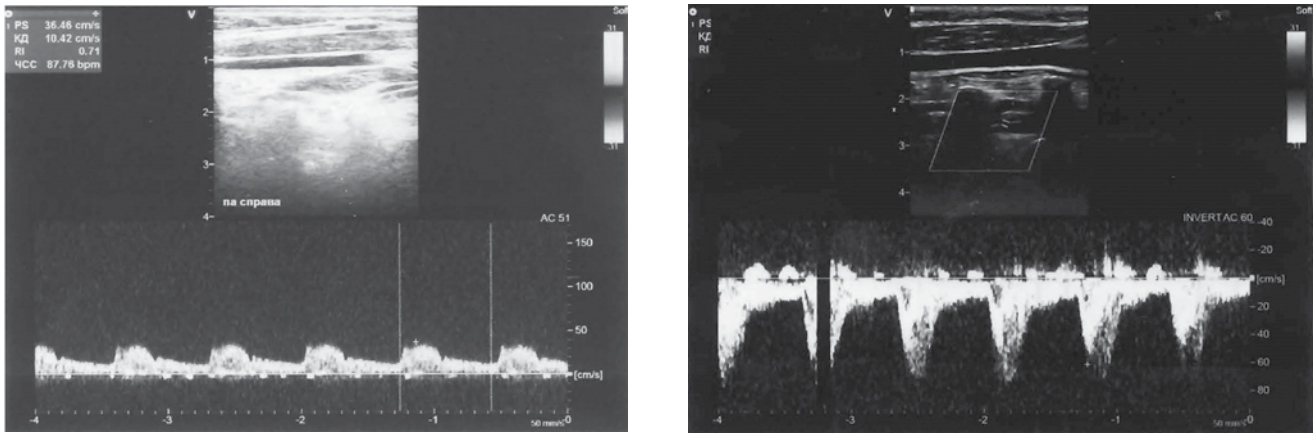
c d

Figure 2. Blood flow in the common carotid arteries (CCA): a — left CCA in a healthy individual; b — left CCA in our patient; c — right CCA in a healthy individual; d — right CCA in our patient



a b

Figure 3. Blood flow in the right subclavian artery: a — in a healthy individual; b — in subclavian steal syndrome



a b

Figure 4. Blood flow in the right vertebral artery in extracranial carotid artery duplex scan: a — in a normal individual; b — in subclavian steal syndrome (retrograde flow)

Therefore, we found an aortic arch branch anomaly: both CCA originated form a single trunk (truncus arteriosus) with aberrant (most-probably, left-sided) RSCA from the posterior wall of the descending aorta with a complete right subclavian steal syndrome.

In order to visualize vertebral intracranial blood flow, we performed transcranial doppler (TCD) ultrasound. The results are presented in Table 2. SSS was confirmed with retrograde blood flow in the right vertebral artery (Figure 5), antegrade blood flow in the left vertebral artery, increased BFLV in both vertebral arteries.

Suprasternal view echocardiogram was performed to determine the anatomy of branches of aortic arch (Figure 6). Right and left CCA originate from a single trunk. LSCA arises to the left from the common trunk. Blood flow was normal in all arteries. Brachiocephalic trunk wasn't found in the typical lo-

Table 2. **TCD parameters (GE Vivid S70 Ultrasound Machine with M5Sc-D probe)**

Parameter	Vertebral artery			Main artery	
	NORMAL*	RESULT		NORMAL*	RESULT
	LEFT, RIGHT	LEFT	RIGHT		
VPs, cec	56.3±7.8	81	97	59.5±17.0	98
VPd, cec	27.0±5.3	22	19	29.2±8.4	40
RI	0.52±0.16	0.82	0.87	0.49±0.12	0.54
Flow direction	ANTEGRADE	ANTEGRADE	RETROGRADE	—	—

Note. * — age-related reference range [8]; RI — resistive index, TCD — transcranial doppler, VPd, cec — diastolic velocity, VPs, cec — peak systolic velocity.

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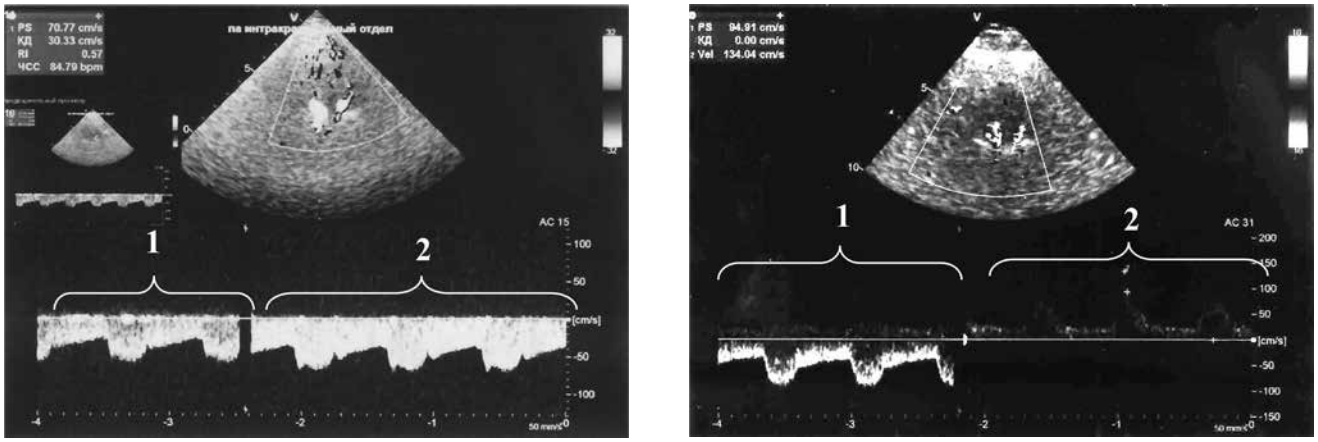
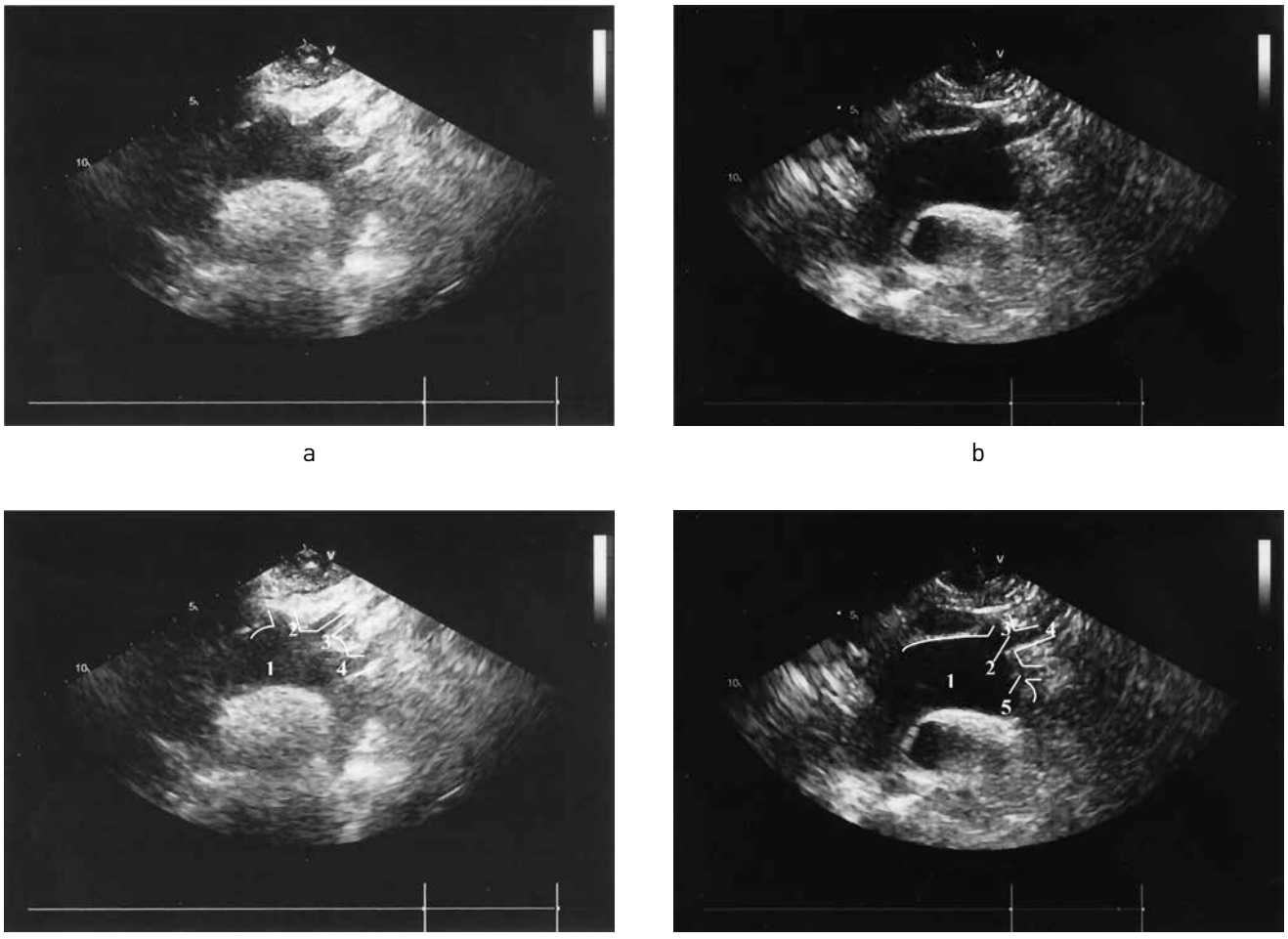


a b

Figure 5. Blood flow in the vertebral artery in transcranial doppler ultrasound on the extracranial level on the left (1) and right (2): a — in a healthy individual; b — in subclavian steal syndrome



a b

c g

Figure 6. Suprasternal view echocardiograms showing aortic arch branches: a, c — in a healthy individual (1-aortic arch, 2 — brachiocephalic trunk, 3 — left CCA, 4 — LSCA); b, g — in our patient (1 — aortic arch, 2 — common trunk of the right and left CCA, 3 — right CCA, 4 — left CCA, 5 — LSCA)

cation. RSCA wasn't visualized due to right internal jugular vein.

Discussion

This case report describes the abilities of functional diagnostics. Although the anomaly described was congenital, it was only first discovered at the age of 53. Detailed review of the patient's medical history didn't reveal any complaints specific for SSS such as any signs of vertebrobasilar insufficiency. We then checked BP on both arms simultaneously: BP on the right arm was lower (90/50 mmHg) then on the left arm (130/80 mmHg) after taking 5 mg/day of amlodipine. We couldn't determine whether BP was previously checked only on one hand and if so, which one, or on both hands, and how were the measurements evaluated. If BP difference was determined at the first place, the sequence of diagnostic evaluations would've been different. Differential diagnosis also included atherosclerosis and Takayasu disease.

As for the further management, the patient will require cardiovascular surgery consultation, regular BP monitoring and review of the medications that may affect BP on the left arm.

Conclusion

Vertebrobasilar system anatomy variations are well-described and are usually diagnosed using the invasive procedures [3, 4]. This case report demonstrates that congenital aortic arch anomalies can be diagnosed with non-invasive techniques.

Conflict of interest: none declared.

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