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Research Article

COMPLICATIONS OF PARKES WEBER SYNDROME

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ABSTRACT

The article presents a clinical case of congenital arteriovenous dysplasia – Parkes Weber syndrome of the lower extremities. The features of its clinical picture and the difficulties of diagnosis are described. It is emphasized that Parkes Weber syndrome is a rare congenital disease of the vascular system, in some cases with the absence of its typical clinical manifestations and combined with other pathologies of the veins.

KEYWORDS

Parkes Weber syndrome, venous angiodysplasia.

INTRODUCTION

Parkes Weber syndrome is a congenital pathological development of arteriovenous shunts. With the normal development of the vascular system, the arteries and veins are connected to each other through the arteriole-capillary network, with an abnormal one, a direct message appears between the arteries and veins, as a result of which varicose veins develop. Most often it occurs on the upper and lower extremities [1,3]. During the intrauterine development of the foetus, the circulatory system is initially not differentiated, due to which arterial blood is discharged into the venous system. During the growth of the body, the functions of the vessels change, and the arteries begin to connect with the veins through a

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multitude of capillaries. At a certain stage, these connections are interrupted, but some vessels remain connected to each other. This is how the shunts described above appear between veins and arteries. Arteriovenous fistulas can be located in any of the organs, and often the symptoms of their functioning can be masked as another disease. More often than other parts of the body and organs, Parkes Weber syndrome affects the limbs, both upper and lower. There are several types of pathological anastomoses connecting arteries and veins. The direct connection through a small isthmus of a large artery and vein is carried out by the so-called fistula. If two large vessels communicate through a cavity, then this type of pathological anastomosis is called an aneurysm. And the third type of communication of the artery and vein is a conglomerate of numerous arterioles and venules. If the anastomosis carries out communication between the artery and the vein for a long time, then their functioning and structure are disrupted. In the arteries, the muscle layer is thinning, and collagen grows, and the walls of the veins thicken due to the hypertrophied muscle layer. Dysfunction of cardiovascular activity occurs as a result of a large and prolonged discharge of blood from the arteries into the veins. Initially, overload leads to hypertrophy of the myocardium and subsequently to the expansion of chambers and the development of chronic heart failure [2,4].

THE MAIN FINDINGS AND RESULTS

As an example, the medical history N 4003 of patient S. born in 1977, from the Jizzakh region, was presented. Patient S. entered the 1st cardiology department of the Multidisciplinary Clinic of the Tashkent Medical Academy with complaints of shortness of breath, heaviness in the abdomen, and weakness. In 2005, he noticed the pronounced expansion of the veins of the lower extremities and the appearance of shortness of breath and contacted the Republican Specialized Surgery Center where the Parkes Weber syndrome was diagnosed. In 2007, surgery was performed for varicocele, phlebectomy on the left and on the right lower limbs in 2013 and 2015, respectively. Dilated cardiomyopathy was diagnosed at him in 2013. Due to episodes of a rare pulse and dizziness, he contacted the Republican Specialized Cardiology Center in February, 2017. ECG Holter monitor revealed bradysystolic atrial fibrillation, polytopic premature ventricular contractions of grade IV by Lown grading, and the patient was referred to the RSSC, where a pacemaker (electrocardiostimulator (ECS)) was implanted. Due to the deterioration of his condition and the increase in shortness of breath, he was admitted to the cardiology department. As a child, he grew up a sickly child, often suffered from colds. Married, no children. He denies bad habits. Heredity is not burdened. Worked as a driver. The condition is severe, asthenic, height 187 cm, undernourished. Pronounced acrocyanosis, the skin is dry, and jaundiced. The sclera of the eyes are icteric. There are varicose pulsating veins on the anterior wall of the abdomen, scrotum, and lower extremities. Some veins bleed, and therefore he had to seek surgical care before. On the lower extremities, varicose veins are enlarged in the form of conglomerates, the skin above them is warm.

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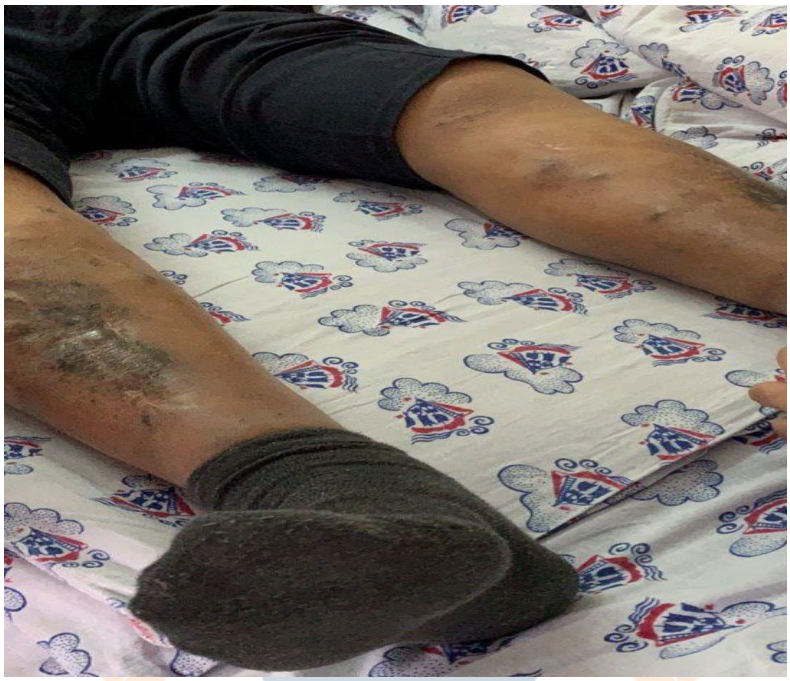

There is a harsh breath in the lungs. Implanted ECS is on the left under the collarbone. The heart is enlarged in all sizes. The heart sounds are muted, the pronounced accent of the II tone over the pulmonary artery. Heart rate is 70 per minute, the rhythm is correct. Blood pressure 120/80 mmHg. cyanotic tongue, overlaid. The abdomen is enlarged in size. The liver protrudes to the umbilical line, there is fluid in the abdominal cavity.

Laboratory and instrumental examinations.

Total blood count: Hb-125 g/l; Erythrocytes- 4,03x1012/l; Ht-35.9; Leukocytes - 2,9x109/l; Platelets -288 x109/ l; ESR -2 mm/hour. Urinalysis: relative density -1021; protein -0; epithelium -0-1-2; leukocytes -1-2. Blood lipid spectrum: Total cholesterol -116 mg/dl; TG-8 mg/dl; HDL-40 mg/dl; VLDL-11 mg/dl; LDL-64 mg/dl; Fasting blood glucose -4.3 mmol/l. ALAT-12 U/L. ASAT -10 U/L. Total bilurubin is 47.6 mmol/l. Creatinine -78 mmol/l. Urea 5.7 mg/dl. Acute phase indicators: RF 5 U/ml; CRP - 4.33mg/dl. Electrolytes: K+-4.31mmol/l. Na+ - 136.1mmol/l. Cl- -109.3mmol/l. Ca2+-1.23 mmol/l. Coagulogram: PT-18.8.sec; PTI-64.2%; PTR-1.39; INR-1.44; APTT-38.9sec; fibrinogen-2.4g/l. Markers of infectious agents: HBsAg-negative. Anti-HCV-negative. WR-negative.

ECG. The rhythm imposed by an electrocardiostimulator with a heart rate of 70 beats per min.

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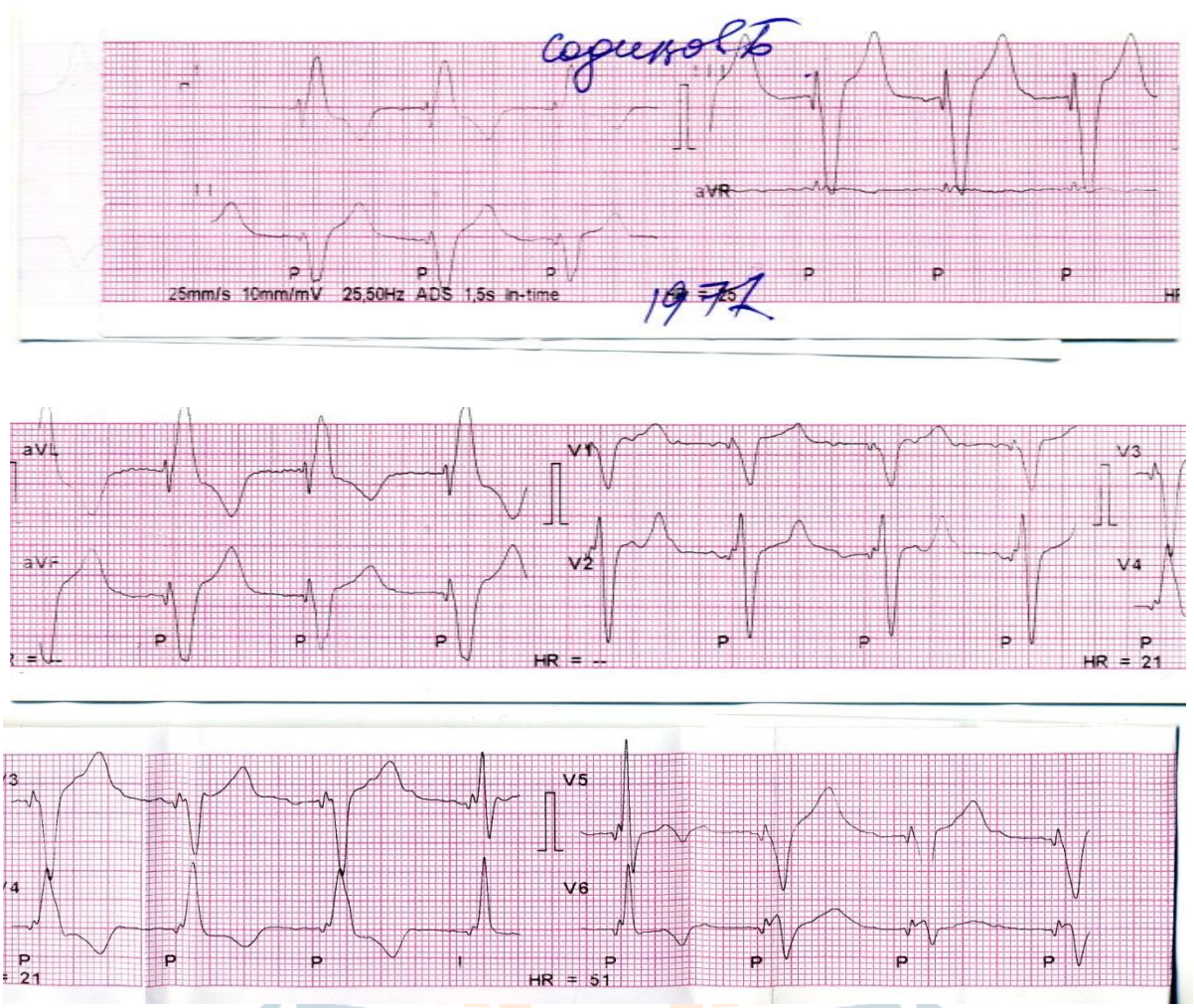

EchoCG: Ao-27mm; LA-47mm; LV EDS-67mm. ESS-51mm. IVS-10.6 mm. LVPW-11mm. RV-46mm. RA-57*69mm. LV mass-329.03gr. EDV-231.4ml. ESV-123.8ml. EF-46.5%. E-1.05m/s. PVaort. -1.06m/s. The walls of the ascending aorta and the flaps of the aortic and mitral valves are sealed. The pulse movement of the aorta is preserved. Systolic opening of the semilunar valves is 18mm. Doppler EchoCG: Mitral regurgitation grade 2-3, Pressure gradient - 84 mmHg. Tricuspid regurgitation grade 2; Pressure gradient - 34 mmHg. Systolic pressure of the pulmonary artery - 54 mmHg.

MSCT: on a series of multi-slice tomographic sections with 1mm resolution, images of abdominal organs were obtained with the use of oral contrast. The liver is enlarged - the right lobe is -170 mm, the left one is -67 mm, the caudate lobe is 69.2x50.6 mm. Its contours are uneven, and clear. Parenchyma structure with a density of 57x59 units. Intra- and extrahepatic bile ducts are not dilated. The inferior vena cava is expanded with a diameter of 40.0 mm. The gallbladder is pear-shaped, its walls are thickened to 4.0 mm, and the contents are compacted with sediment. The portal vein is expanded to 16 mm. The spleen is enlarged

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(109x52 mm), its contours are smooth, clear, the texture is not changed, the density is 44x46 units. Pancreas - the head is not clearly differentiated, the body is 22.5 mm, the tail is 16.7 mm. Density 49x55 units. The adrenal glands are of the usual shape and size. The position, shape and size of the kidneys have not been changed. The calyx-pelvic system of both kidneys is not deformed or expanded, concretions are not detected. The pelvis of the left kidney is up to 14 mm wide. Paranephric space - without features. Abdominal aorta and other large vessels of the abdominal cavity with the presence of calcified atherosclerotic plaques. The lymph nodes of the abdominal cavity and retroperitoneal space are not enlarged. Inferior vena cava, mesenteric veins are dilated. At the level of examination of the lower-basal segments of the chest, pronounced dilatation of all chambers of the heart, mainly the right parts, with the presence of calcification and compaction of the pericardium, more along the right contour of the heart, is determined. Conclusion: hepatosplenomegaly (manifestations of cirrhotic changes and congestion in the liver), free fluid in the abdominal cavity. Constrictive pericarditis. Cardiomegaly.

Chest radiography: right-sided hydrothorax. Cardiomegaly.

Diagnosis: Parkes Weber syndrome

Complications: Secondary cardiomyopathy. Condition after ECS implantation. CHF III stage, pulmonary hypertension II grade, congestive cirrhosis of the liver, splenomegaly, ascites, right-sided hydrothorax, constrictive pericarditis. Bleeding from varicose veins of the lower extremities.

CONCLUSION

Thus, the congenital pathology of the development of arteriovenous anastomoses led to bleeding from varicose veins of the lower extremities, constrictive pericarditis, the development of cardiomegaly, cardiac insufficiency and complex rhythm disturbances, and therefore an ECS was implanted in the patient.

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