

POSTER PRESENTATION

POEMS -syndrome in a patient with hypertrophic cardiomyopathy.

Andrew Lysenko, Gennady Salagaev, Pavel Lednev, Yurii Belov, Giurikhan Magomedova, Nargiz Mamedova.

B.V. Petrovsky National Research Center of Surgery, Moscow, The Russian Federation

INTRODUCTION

POEMS-syndrome is a rare disabling paraproteinemic disease syndrome based on modern diagnostic criteria. POEMS-syndrome distinguishes itself from other paraproteinemic and inflammatory nephropathies by its damage to lots of organs, which caused by increased levels of proinflammatory and angiogenic cytokines. Most often it affects people aged 40 to 50 years, mostly men. In total there are about 200 thousand patients in the USA.

AIM

This is our first experience of surgical treatment of a patient with HCMP with POEMS-syndrome, who underwent septal myoectomy.

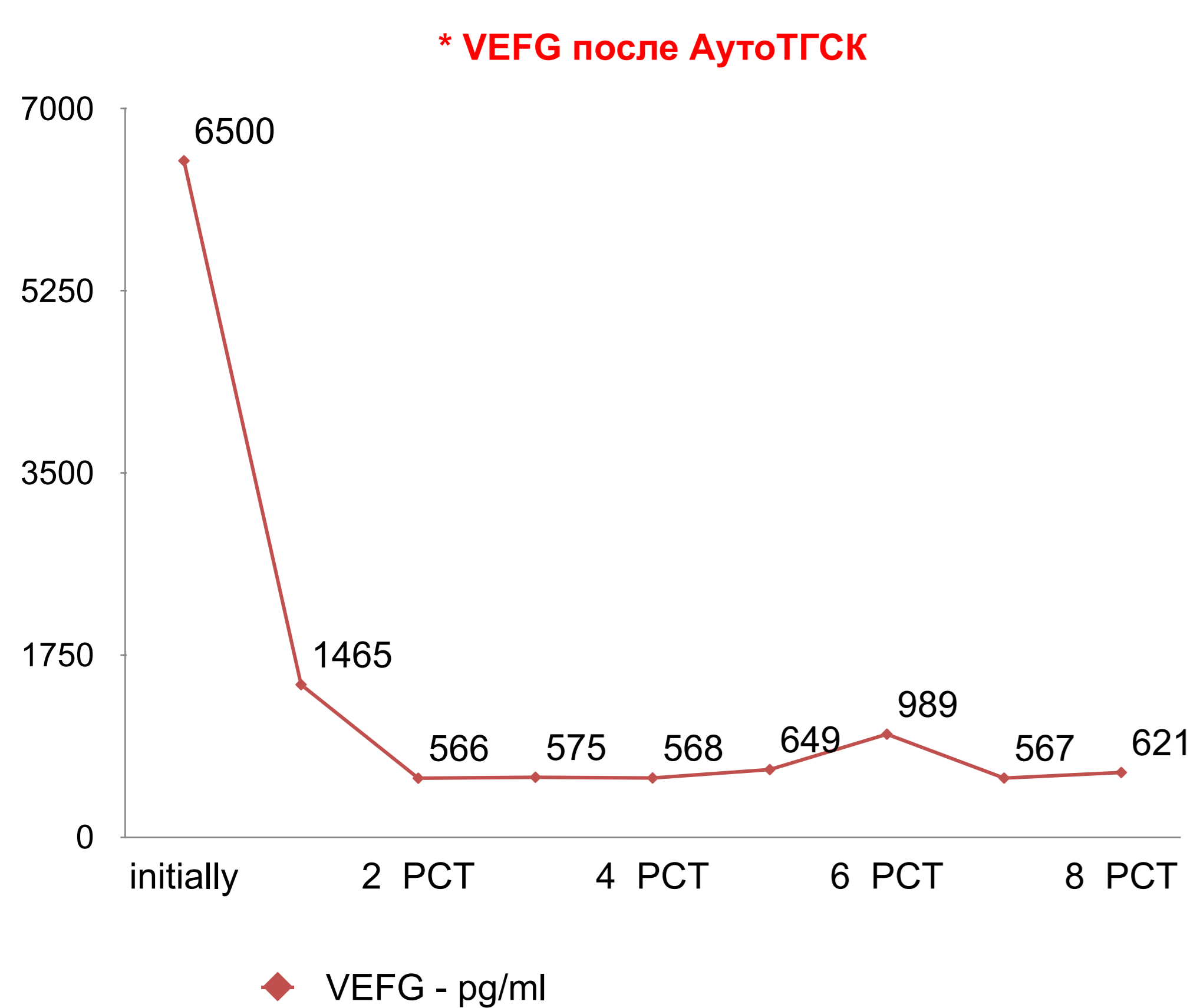


Fig. 1. Dynamics of VEGF throughout therapeutic treatment.

METHODS

Patient G., 40 years old, was admitted with complaints of shortness of breath with minimal physical exertion, weakness, sweating, intermittent palpitations. From history: he note the above complaints during the last year . A hypertensive crisis with BP 200/100 mm Hg first appeared In March 2022. POEMS-syndrome was diagnosed in May 2017. In the period from 2017 to 2018, the patient underwent 8 courses of chemotherapy, and in 2018 autologous hematopoietic stem cell transplantation was performed.(fig.1 and 2) ECG and CAG without features. ECHO: an asymmetric for m of latent obstructive HCM .Moderate anterosystolic movement of the anterior mitral valve flaps (SAM). LVOT: Vmax 5.6 m /s ,PG max /min 127.6 / 62.8 mmHg. mitral insufficiency of the 2nd degree, systolic pressure LA up to 40 mmHg. NTproBNP– 10.8 pg/ml. TEE:EDV - 101 ml, ESV- 38 ml, LVEF 62.38%, IVS1.9 cm. The patient was reffered septal myoectomy surgery.

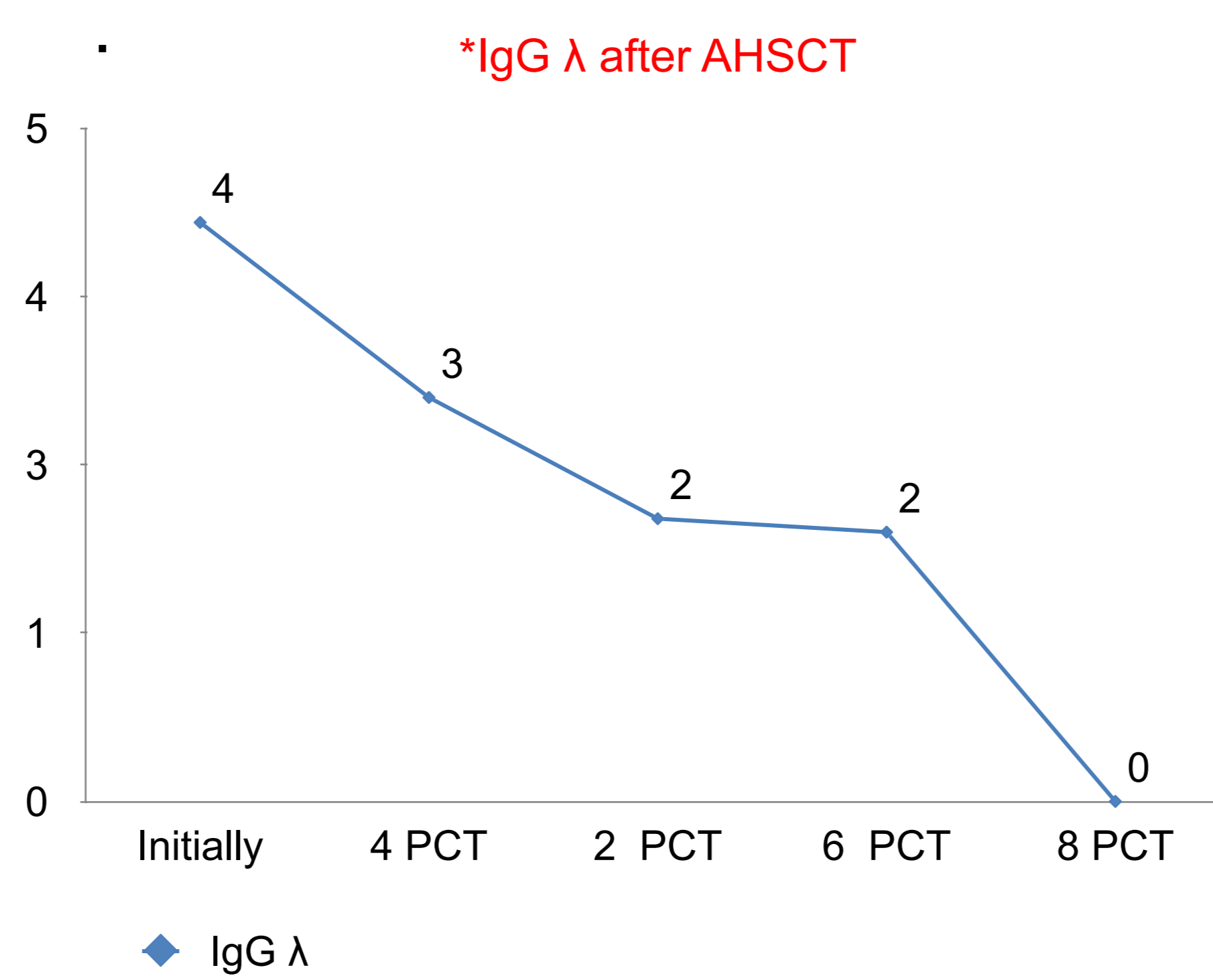


Fig. 1. Dynamics IgG lambda

RESULTS

Considering severe hypertrophy with LVOT obstruction and clinical signs of circulatory disorder (CD), the patient underwent septal myectomy under cardiopulmonary bypass and blood cardioplegia. According to the data of intraoperative TEE after restoration of effective cardiac activity:EDV 90 ml, ESV 34 ml, LVEF 62.22%, V max 1.7 m/s PG max/min 11.6/6.5 mm Hg. Histological results: An uneven thickening of the endocardium due to sclerosis is determined in a fragment of the interventricular septum. There are areas with hypertrophied cardiomyocytes with large hyperchromic nuclei.(fig.3) In a satisfactory condition, the patient was discharged on the 4th day of the postoperative period under the supervision of a cardiologist at the place of residence.

CONCLUSIONS

Cardiac manifestations in POEMS-syndrome are heterogeneous: cardiomyopathy, pericarditis, pericardial effusion and congestive heart failure, which in one third of patients led to death. Most endomyocardial biopsy samples in POEMS-syndrome are not stained with VEGF or other specific proteins, demonstrate hypertrophy and fibrosis- paraneoplastic process.

BIBLIOGRAPHY

- Levene J, Murray N, Desai S, et al. Pericardial Tamponade and Other Cardiac Complications in POEMS Syndrome. JACC Case Rep. 2021;3(2):286-290. <https://doi.org/10.1016/j.jaccas.2020.12.027>
- Abdelahad M, Pearson R, Mauri B, et al. Heart Failure With Cardiogenic Shock as a Manifestation of Untreated POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal Protein, Skin Changes) Syndrome. Cureus. 2021;13(9):e18046.
- Tanus T, Miller HJ. POEMS syndrome presenting with cardiomegaly Land cardiomyopathy. J Intern Med. 1992;231(4):

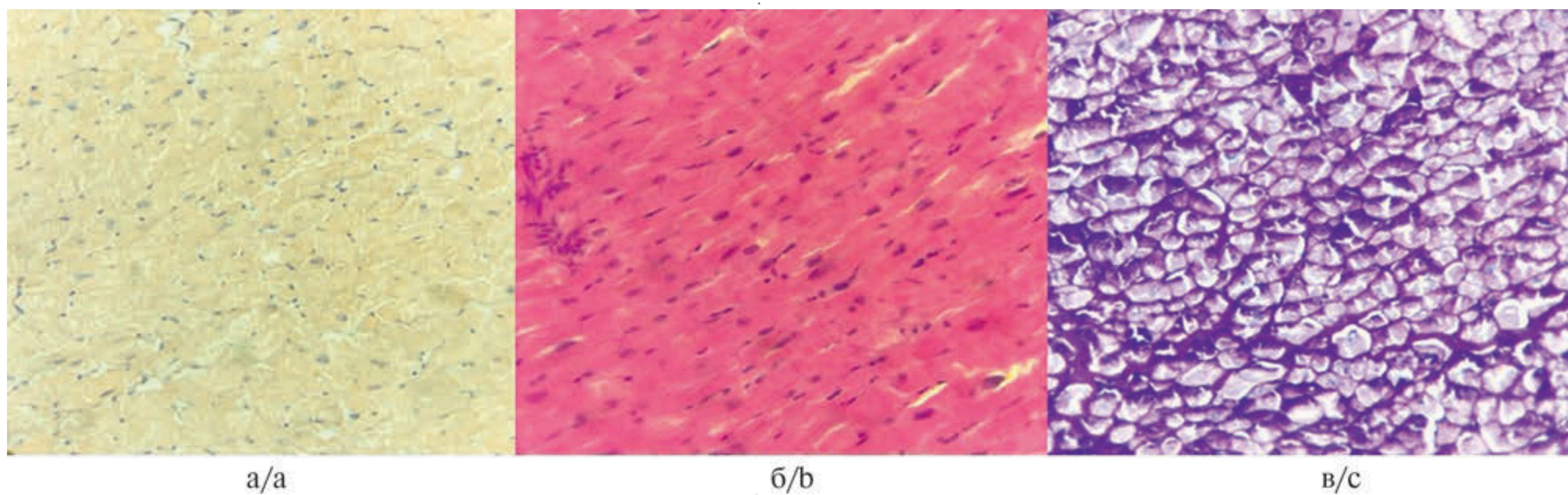


Fig. 3. Interventricular septum. a — Van Gieson's staining, perivascular foci of sclerosis; b — staining for amyloid, negative reaction; c — positive PAS reaction within the normal range for glycogen granules in cytoplasm of cardiomyocytes.