

Cardiac tamponade: A rare manifestation of Castleman disease

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A 13-year-old male patient was transferred to the Department of Pediatric Cardiology in emergency mode due to exacerbation in breath shortness and deterioration in exercise tolerance observed for 2 months. He had been treated earlier with macrolide due to pneumonia and bronchitis. Laboratory tests showed elevated levels of C-reactive protein (206 mg/l), white blood cells ($14 \times 10^3/\text{ul}$), urea (95 mg/dl), creatinine (2.02 mg/dl), and hypoalbuminemia (3.4 g/dl) with proteinuria. Levels of N-terminal pro B-type natriuretic peptide and troponins were within the normal range. Moreover, we spotted severe anemia, and two units of red blood cells from whole blood were transferred, without any complications. The examination showed an audible pericardial friction murmur, persistent fever without reaction to antipyretics, and pale, sweaty skin.

On the electrocardiogram, we observed regular sinus rhythm (107/min), nomogram, and ST-segment elevation in leads II, III, aVF, V5–V6 (Figure 1A).

Echocardiography (ECHO) revealed fluid in the pericardial sac. The contractility of the left ventricle was within normal parameters, but the right ventricle flexed under the influence of accumulated fluid, which was an indication for urgent pericardiocentesis (Figure 1B). Pericardial drainage was performed, and 750 ml of serosanguinous high-protein fluid was drained. Control ECHO showed no fluid in the pericardial sac (Figure 1C). An ultrasound examination revealed other fluid areas in Morisson's sinus and both pleural cavities. Moreover, there were numerous small lymph nodes in the liver area, which made us extend diagnostics. Computed tomography showed an image suggestive of lympho-

proliferative disease. Numerous bundles of lymph nodes were found along the trachea, axillary cavities, and in the right lung (Figure 1D). Cooperation with the Department of Hematology enabled us to perform differential diagnosis. Cytometric examination of the bone marrow showed a low percentage of lymphocytes, signs of B cell line regeneration, and suppressed erythropoiesis. Those findings with previous tests were suggestive of idiopathic multicentric Castleman disease [1], which was confirmed by histopathological analysis of a lymph node sample collected in mediastinal biopsy. There was neither evidence of accumulation of dendritic cells (CD 123+) nor the presence of HHV-8 virus, so the final diagnosis was the hyaline vascular variant with atypical morphology [2].

There is no clear treatment plan for the generalized form of Castleman disease. First-line treatment for disseminated disease most often includes corticosteroids [1, 3]. Therefore, we implemented steroid therapy with prednisone (30 mg twice a day), which resulted in significant improvement in the patient's clinical condition. However, most patients require doses of steroids that are too high to be tolerated in the long term, and relapse is virtually inevitable during dose reduction [3, 4]. A follow-up computed tomography scan was performed and showed regression of changes. No signs of fluid in the pericardiac sac were observed on further ECHO.

Cardiac tamponade in the pediatric population is a life-threatening condition and requires special attention. It is necessary to expand diagnostics to search for potential causes, including lymphoproliferative diseases [5]. Interdisciplinary cooperation resulted in applying a comprehensive approach,

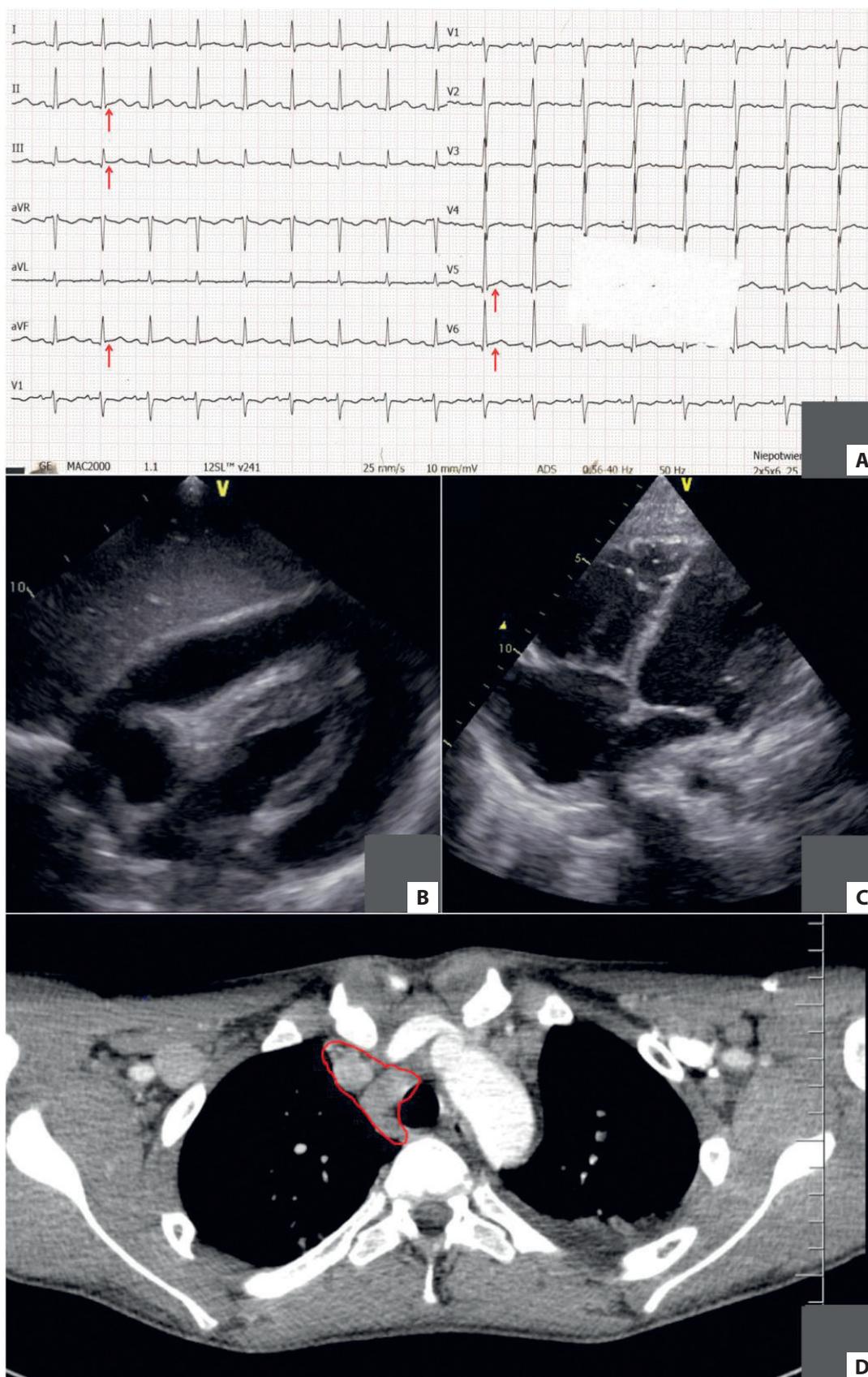


Figure 1. **A.** 12-lead electrocardiogram with no characteristic signs of cardiac tamponade. ST-segment depression in II, III aVF, V5, V6 (red arrows) mimicking myocardial ischemia. **B.** Echocardiography — subcostal long axis view showing a large circumferential pericardial effusion measuring more than two centimeters. Preserved left ventricular ejection fraction (68%). **C.** Echocardiography — apical four-chamber view, 2 days after pericardiocentesis, no presence of pericardial effusion. **D.** Computed tomography of the chest after pericardiocentesis — bilateral pleural effusion, higher amount of fluid in the left pleural cavity. Enlarged cardiac silhouette with air bubbles after pericardiocentesis. Numerous node packets in the image of the right lung

which allowed us to diagnose Castleman disease despite the unusual clinical manifestation, which resulted in the stabilization of the patient's clinical condition.

Article information

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