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Cardiac tamponade: Rare manifestation of Castleman's disease

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

In the electrocardiography was observed regular sinus rhythm (107/min), normogram, ST segment elevation in leads II, III, aVF, V5–V6 were observed (Figure 1A).

Echocardiography (ECHO) revealed fluid in the pericardial sac. The contractility of the left ventricle was within normal results, 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). The ultrasound examination revealed another fluid areas in Morisson's sinus and in both pleural cavities.

Moreover, there were numerous small lymph nodes in the liver area, which gain our attention to extend diagnostics. Computed tomography showed image suggestive for lymphoproliferative disease. Numerous bundles of lymph nodes were found along the trachea, axillary cavities and in the right lung (Figure 1D). Cooperation with Department of Hematology enabled to perform differential diagnosis. Cytometric examination of bone marrow showed low percentage of lymphocytes, signs of B cell line regeneration and suppressed erythropoiesis. Those findings with previous tests were suggestive for idiopathic Multicentric Castleman's Disease [1]. Further confirmed with histopathological analysis of the lymph node collected in mediastinal biopsy. There was neither evidence of accumulation of dendritic cells (CD 123+) nor presence of HHV-8 virus, so the final diagnosis was hyaline vascular variant with atypical morphology [2].

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

Cardiac tamponade in pediatric population is a life-threatening condition and requires special attention. It is necessary to expand diagnostics in order to search for potential causes, including lymphoproliferative diseases [5]. Interdisciplinary cooperation resulted in comprehensive approach, which allowed for the diagnosis of Castleman's disease, despite the unusual clinical manifestation, and caused the stabilization of the patient's clinical condition.

Article information

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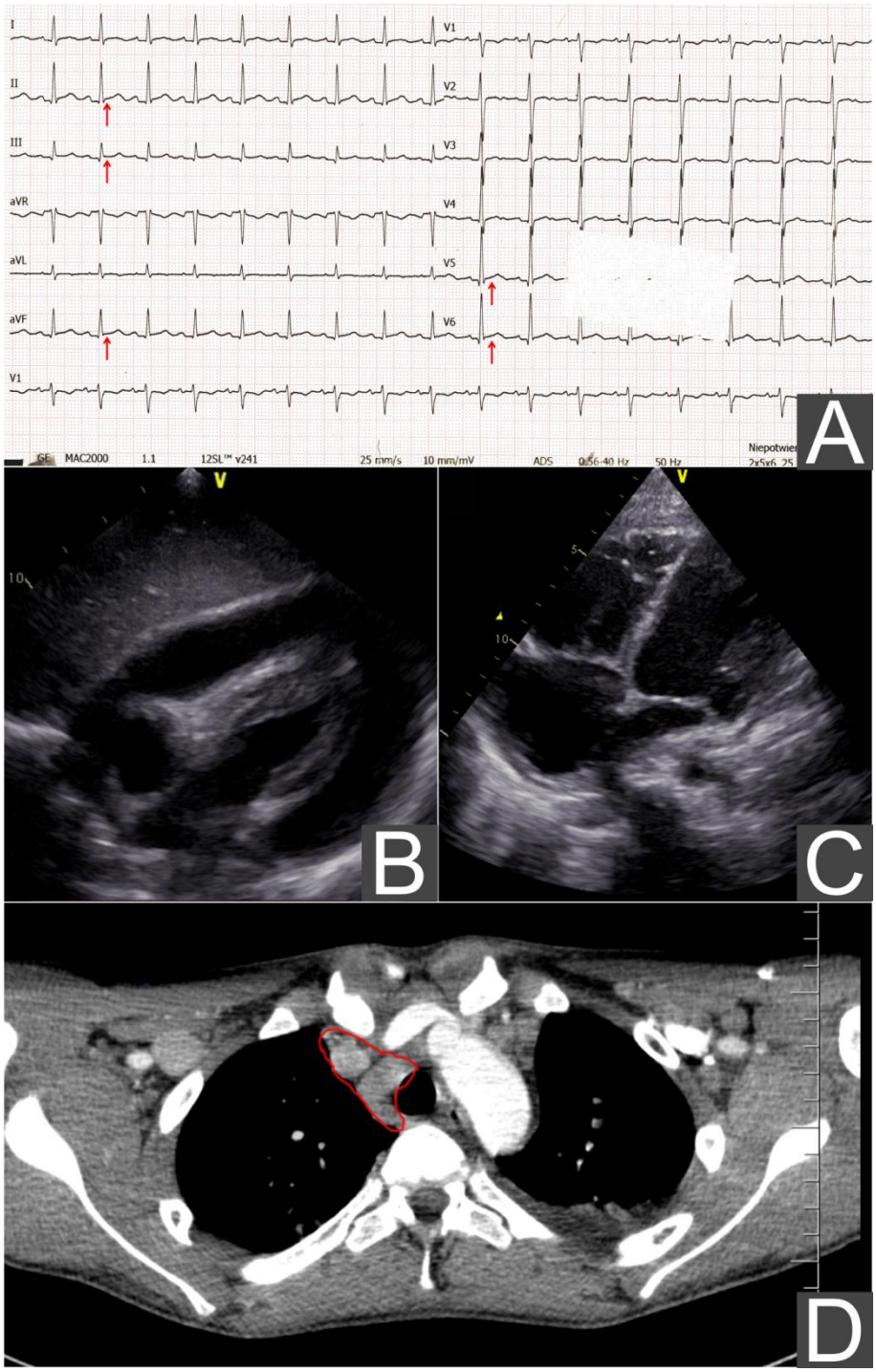

Figure 1. A. 12-lead electrocardiogram with no characteristic signs of cardiac tamponade. ST segment depression in II, III aVF, V5, V6 (red arrows) mimicking myocardial ischemia. **B.** Echocardiography — Subcostal long axis view: showing a large circumferential pericardial effusion measures more than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