

Large mediastinal teratoma causing recurrent pericarditis (RCD code: VI)

Dawid Kudliński¹, Pawel Rubis^{1*}, Lucyna Rudnicka-Sosin², Agata Leśniak-Sobelga¹, Magdalena Kostkiewicz¹, Piotr Podolec¹

¹ Department of Cardiac and Vascular Diseases, Institute of Cardiology, Jagiellonian University Medical College, Centre for Rare Cardiovascular Diseases, John Paul II Hospital, Krakow, Poland; ² Department of Pathology, John Paul II Hospital, Krakow, Poland

Abstract

The mediastinal tumors are rare but potentially fatal diseases. Teratomas are derived from multipotent germ-line cells. In majority of cases mediastinal tumours are incidentally detected on chest X ray. We present 19-year-old patient with recurrent pericarditis recurrent pericarditis. After comprehensive diagnostic process it was figured out that the reason for recurrent pericarditis was large mediastinal teratoma. Once the correct diagnosis has been made, the patient could have been completely cured. JRC D 2014; 1 (6): 29–32

Key words: recurrent pericarditis, mediastinum, tumour, teratoma

Background

The mediastinal tumors are rare but potentially fatal diseases. Teratomas are derived from multipotent germ-line cells and for this reason may contain in their structure different tissues, such as muscles, epithelial tissues, as well as cartilage, and even teeth or hair. Benign teratomas usually grow slowly and are asymptomatic for long periods of time or present only minimal symptoms, such as non-specific chest pain, dyspnoea or tachycardia when the vagus nerve is oppressed by the tumor. In majority of cases mediastinal tumors are incidentally detected on chest X rays exam.

Case presentation

We present 19-year-old patient, who was admitted urgently due to recurrent pericarditis (the third episode in the last nine months). In the previous months, the patient was hospitalized twice due to syncope episodes, non-specific chest pain, fast palpitations, sustained subfebril state. Moreover, while being in coal mine during the apprenticeship, he lost consciousness. In district hospital, he was diagnosed with idiopathic pericarditis as moderate pericardial effusion was revealed on standard echocardiography. Thus he was prescribed standard treatment for pericarditis with antibiotics, colchicine, and non-steroidal anti-inflammatory

ry drugs with good effect. This time patient was admitted with similar but more intensified symptoms of severe chest pain and breathlessness. The chest pain was exacerbated with supine position. His vital signs were stable with slight tachycardia of 120/min, arterial blood pressure of 130/80 mm Hg, saturation of 98% on room air, and accelerated breathing rate to 22 per minute, and temperature of 37°C. The physical examination was not very remarkable and revealed a normostenic body composition, normal heart sounds without murmurs including pericardial rub, vesicular murmur over the lungs and Killip 1 class, no peripheral edemas, the abdomen was not painful on palpation and liver and spleen were of normal size. The standard 12-lead electrocardiography revealed sinus tachycardia with heart rate between 100 to 130 bpm, normal cardiac axis and intriguingly no other pathological findings. The blood test revealed a significantly elevated level of inflammatory markers such as C-reactive protein (CRP) up to 182 mg/L (norm: <3.0 mg/L), as well as high levels of white blood cells ($14.62 \times 10^3/l$ with normal level up to $10 \times 10^3/l$), high level of neutrophil granulocytes ($11.66 \times 10^3/l$ with norm up to $7.80 \times 10^3/l$), high level of monocytes ($1.32 \times 10^3/l$ with norm up to $1.00 \times 10^3/l$). The levels of other morphotic and biochemical parameters such as haemoglobin, amount of erythrocytes, liver and kidney function tests were normal. The viral serology for human immunodeficiency virus (HIV), cytomegalovirus (CMV), hepatitis C virus (HCV) were all negative. Initially, suspicion was

Conflict of interest: none declared.

* Corresponding author: Department of Cardiac and Vascular Diseases, Institute of Cardiology, Jagiellonian University Medical College, Centre for Rare Cardiovascular Diseases, John Paul II Hospital, Pradnicka str. 80, 31-202, Krakow, Poland; tel. 0048 12 614 22 87, fax 0048 12 423 43 76; e-mail: pawelrub@poczta.onet.pl

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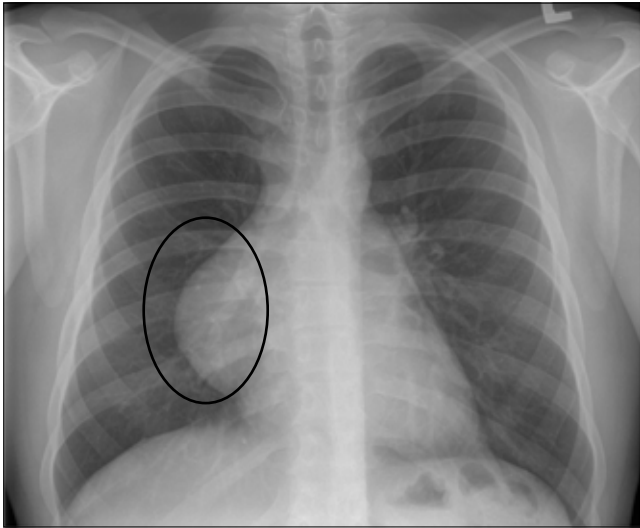


Figure 1. Chest X-ray in the anterior-posterior view. Tumor mass originating from the mediastinum causing rightward “bulge” of cardiac silhouette (circle)



Figure 4. Resected mass of the tumor. Irregular shape indicating complex inner morphology

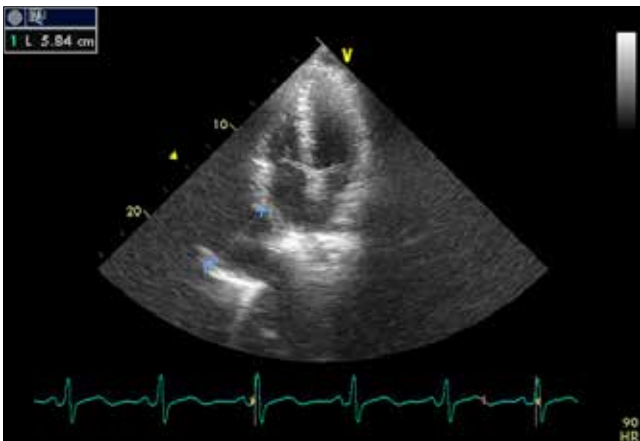


Figure 2. Transthoracic echocardiogram, apical four chamber view. Hypoechoic structure over right atrium (blue line)

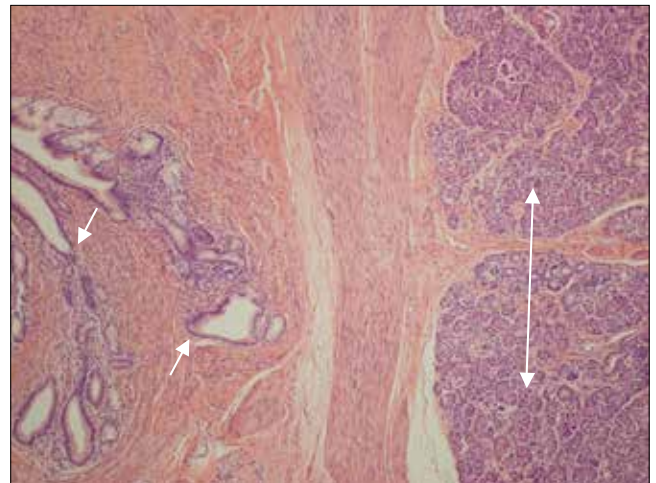


Figure 5. Light microscopy of the excised tumor. Two types of tissues – pyloric mucosa (white arrows), and pancreas tissue (white double arrow)

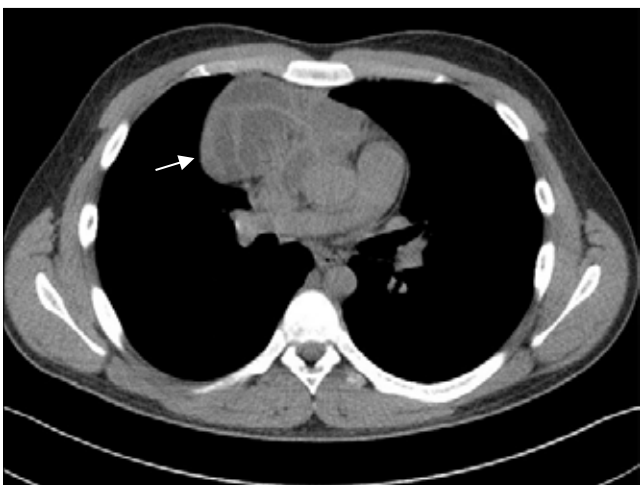


Figure 3. Computed tomography of the chest. Multi-cystic tumor with evident contact with large portion of the pericardium (white arrow)

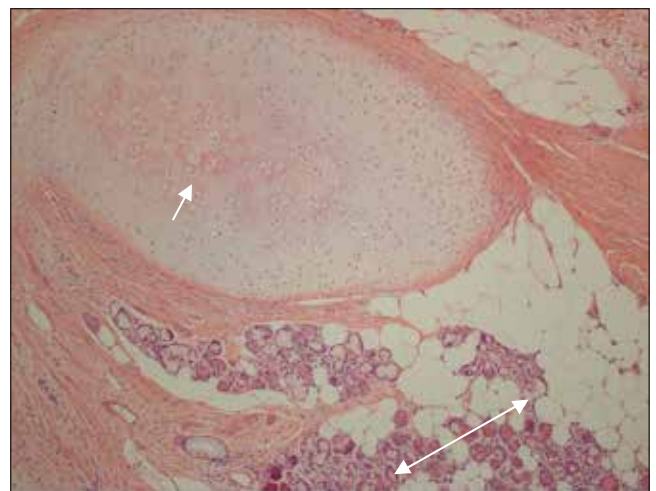


Figure 6. Hyaline cartilage (white arrow), intestinal glands (white double arrow)

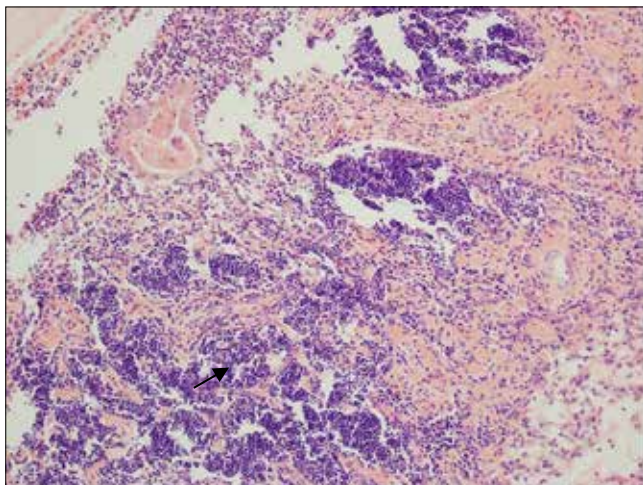


Figure 7. Thymical tissue (arrow)

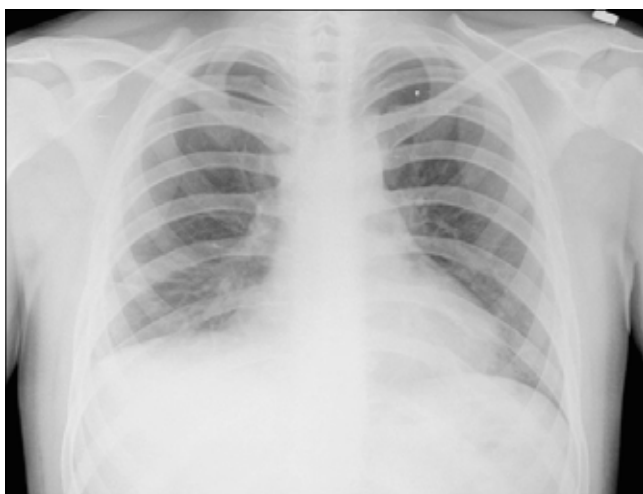


Figure 8. Post-operation control chest X-ray in the anterior-posterior projection. Free mediastinum without tumour mass

made for an atypical bacterial infection and an empirical antibiotic therapy was introduced. The standard chest X-ray was performed and showed an improper silhouette of the heart, causing 'bulge' of the right atrium (Figure 1). Transthoracic echocardiography revealed normal size of cardiac chambers, preserved left ventricular contractility, no heart valves disease, thickened and hyperechogenic pericardium with minimal pericardial effusion to 5 mm. Moreover, in the apical four chamber projection between right atrium and the right lung, was seen an hypoechogenic structure resembling a cyst with at least two chambers (Figure 2). At this point it became clear that there is an additional structure in the mediastinum and more advanced examinations and tests were initiated. In order to accurately visualize the structure and surroundings chest computed tomography (CT) was performed. The study clearly confirmed the presence of a large, approximately 16 cm in diameter cyst, with easily defined chambers and fluid-filled compartments (Figure 3). The chest CT confirmed previous suspicion of mediastinal tumor 'compressing' or at least hav-

ing close contact with the right atrium. Although the tumor was identified, its origin was unclear. As the patient was a young adult, the suspicion of germ cell tumor was made and ultrasonography of abdomen and pelvis, including testis were performed but did not provide any new data. At the same time blood tests for human chorionic gonadotropin (hCG) and alpha-1-fetoprotein (AFP) were performed but similarly the results were within normal limits. Additionally, the patient was investigated for echinococcosis and the molecular testing for *Echinococcus* spp. was performed but results were also negative. In order to optimize patient's treatment a multidisciplinary team meeting was organized and it was commonly agreed that the patient should be promptly referred for the thoracic operation of cyst/tumor removal. Fortunately, the operation was uneventful and resection of the tumor was complete without any major complications. The resected structure was a large tumor of at least 15 cm in diameter (Figure 4) but more importantly the histopathological examination revealed typical tissue composition for benign teratoma. Inside the tumor several types of tissues were found, including pyloric mucosa, pancreas tissue (Figure 5), hyaline cartilage, intestinal glands and Paneth cells (Figure 6), as well as thymical tissue (Figure 7). Furthermore, the post-operative period was also short and without any sequels he was discharged home a week later without any prolonged medications (Figure 8). The patient has been under close follow-up for the last months and he is in good health and without any episodes of recurrent pericarditis.

Discussion

Epidemiology

Among all chest tumors teratomas account for 8–13% [1]. The world-wide annual incidence is one in 4000 live births [2]. Teratomas are primarily located in the anterior mediastinum and affect young people [3].

Mediastinal germ cell tumors

The incidence of specific tumors differs between studies and series [2]. Neoplasms in the anterior mediastinum are thymic lesions (most frequently thymoma), lymphoma, pheochromocytoma, germ cell tumors, thyroid tissue, and parathyroid lesions [1]. Masses in this area are more likely to be malignant than those in other compartments. The most common type of mediastinal tumor is the neurogenic tumor (21%), followed by thymoma (19%), lymphomas (13%), and germ cell tumors (10%) [4, 5, 6, 7, 8]. Mediastinal germ cell tumors are a heterogeneous group of benign and malignant neoplasms that originate from primitive germ cells. This group include neoplasms such as seminoma, teratoma, non-seminoma malignant germ cell tumor. Teratomas are most common mediastinal germ cell tumors [1, 2, 4, 9]. Their histologic structure is complex and consist of three primitive germ cell layers. More than two thirds of teratomas are mature that means that they do not contain foetal tissue whereas the remaining one third is built to some extent from embryonic tissue. Importantly, mature teratomas are benign. However, their special location in the mediastinum with close proximity to vital organs can result in serious problems. Teratomas are known to secrete digestive

enzymes that irritate and damage the structure of bronchi, pericardium, pleura, or lung and can lead to rupture of those organs. We believe that this is the particular pathomechanism of recurrent pericarditis in our patient.

Clinical manifestations

Less than half of patients with mediastinal tumors are symptomatic on presentation. The absence of symptoms correlates closely with benign neoplasms. The frequency of symptoms vary with the precise location of the tumor in the mediastinum. The anterior-superior position is associated with the most frequent occurrence of symptoms because more than two-thirds of patients are symptomatic, whereas middle mediastinal or posterior mediastinal masses cause symptoms in approximately only half of patients. The most common symptoms are chest pain, cough, and dyspnoea. Chest pain usually suggest invasive spread of malignant neoplasm. Other less common symptoms are hemoptysis, dysphagia, hoarseness, Horner's syndrome, vena caval obstruction, arrhythmias, fever, weakness, and weight loss. Besides symptoms that are caused by compression or invasion, other signs and symptoms, such as endocrinologic abnormalities, cardiovascular lesions (as in our patient), and associated systemic syndromes, e.g. thymoma and myasthenia gravis can occur [7]. It is of paramount importance to search for gonadal malignancy when malignant mediastinal tumor is diagnosed. As was done in our case, except for ultrasound examinations, the measurements of α -fetoprotein (AFP) and β -human chorionic gonadotropin (β -hCG) are examinations of choice [3].

Diagnostic process and surgical treatment

In chest X-rays those structure tend to be rounded or lobulated with well-defined boundaries that protrude to one side of the mediastinum and can reach large sizes. At least one forth have calcifications and some have gross radio-dense areas that can be explained as fragments of bones or teeth [1, 2, 3]. Most commonly teratomas are accidentally detected on routine chest X-ray. There are usually no evident abnormalities in the standard transthoracic echocardiography. The pericardial effusion or thickened pericardium can occasionally be observed but as most teratomas are benign tumors, they are well separated from the surrounding tissue. If a tumor is particularly large, it can compress the heart, and as a result atrial or ventricular silhouette of the heart can be distorted (as was observed in our patient). Chest CT is the imaging technique of choice in the evaluation of mediastinal abnormalities. On CT teratomas are usually multi-locular cystic tumors. Mediastinal mature teratoma manifests on CT as a heterogeneous anterior mediastinal mass containing soft-tissue, fluid, fat or calcium attenuation. Fluid-containing cystic areas, fat and calcification occur frequently [2, 3]. Cystic lesions without fat or calcium are presented in 15% of tumors. On microscopic examinations of excised tumor mesodermal, ectodermal, and endodermal elements are seen in varying proportion.

The surgical approach to these neoplasms has altered significantly over last decades. In the beginning of thoracic surgery, the major causes of unsuccessful operation and death were tumor mass effect or complication of general anaesthesia. With the advent of modern surgical technique and more importantly novel anaesthetic and in-

tensive care approach, the mortality rate has significantly dropped. Generally, mature mediastinal teratomas (the majority of cases) are benign and encapsulated and can be resected completely with good results [10]. The long-term prognosis, after successful tumor resection, is excellent [2, 10].

In summary of this interesting and educational case, it can be said that before any diagnosis of idiopathic pericarditis or in the broadened meaning any kind of disease is made, all other potential causes should be actively sought with all available diagnostic tests. In our patient the reason for recurrent pericarditis was large mediastinal teratoma, which 'compressed' or 'irritated' pericardium causing recurrent inflammatory state. Once the correct diagnosis has been made, the patient could have been offered the complete cure of this disease by resecting the tumor that could have had potentially even more devastating and life-threatening consequences.

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