THE TUMORAL ETIOLOGY OF THE SUPERIOR CAVA VEIN SYNDROME PRESENTING AS EXSUDATIVE PERICARDITIS IN A YOUNG PATIENT

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Abstract

We present the case of a 36-year-old patient admitted with dyspnea, subjective sensation of edema of the cervical region, and precordial pain with sudden onset. Pathological antecedents revealed a respiratory intercourse three weeks ago. Laboratory findings indicated an inflammatory syndrome accompanied by hypoproteinemia. We performed an echocardiographic examination which revealed important pericardial effusion and swinging heart aspect with diastolic collapse of the right atrium and ventricle. According to the existing guidelines pericardiocentesis was immediately required. The hemorrhagic pericardial effusion had an intense positive Rivalta reaction with frequent macrophages and malignant atypical cells. The next step was a full-body computer tomography scan, which revealed an expansive mediastinal mass, invading the upper cava vein and pericardium, with mediastinal adenopathy. A biopsy was performed and the pathology examination concluded there was a classical Hodgkin's lymphoma nodular sclerosis type. Finally the patient was directed to oncology department.

Keywords: pericardial effusion, cardiac tamponade, Hodgkin's lymphoma, pericardiocentesis, swinging heart.

Rezumat

Relatăm cazul unei paciente de 36 de ani care prezintă următoarele acuze: dispnee, senzație subiectivă de edem al regiunii cervicale și durere precordială cu debut brusc. Antecedentele patologice au relevat o intercurență respiratorie în urmă cu trei săptămâni. Explorările paraclinice au indicat un sindrom inflamator însoțit de hipoproteinemie. Am efectuat o examinare ecocardiografică care a decelat o importantă colecție pericardică și aspectul de swinging heart însoțit de colaps diastolic al atriului și ventriculului drept. Conform ghidurilor din literatura de specialitate, pericardiocenteza a fost indicată de urgență. Lichidul pericardic drenat a prezentat un aspect hemoragic cu o reacție Rivalta intens pozitivă și au fost găsite celule macrofage frecvente și celule atipice maligne.

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Următorul pas a impus realizarea examenului computer tomografic toracic care relevă o masă mediastinală expansivă, invadând vena cavă superioară și pericardul, decelând și adenopatii mediastinale. A fost efectuată o biopsie pericardică, iar examenul anatomopatologic a concluzionat prezența unui limfom Hodgkin clasic de tip scleroză nodulară. În cele din urmă, pacienta a fost îndrumată spre un department oncologic avizat pentru inițierea tratamentului de specialitate.

Cuvinte cheie: pericardită, tamponadă cardiacă, limfom Hodgkin, pericardiocenteză, swinging heart

Introduction

Pericardial diseases are relatively common in our clinical practice but the real incidence of them is difficult to appreciate because there are many cases which remain undiagnosed. Men are usually more affected than women. It is mandatory to determinate whether this pathological condition is idiopathic or part of some systemic diseases⁽¹⁾. The main types of pericardial diseases involve infectious or non-infectious causes. Neoplastic pericardial effusion is a concerning problem and it can develop from direct extension or metastatic spread of the underlying malignancy via lymphatics into the pericardium⁽⁵⁾. In autopsy series, the prevalence of pericardial involvement varies from 4% in general autopsies to 15%–30% in autopsies of cancer patients⁽⁴⁾.

Pericardial involvement in malignancy is actually a common finding especially in non-Hodgkin's lymphoma, breast cancer or pulmonary neoplasia, whilst the primary pericarditis is more rarely revealed.

Case presentation

A 35 years old non-smoker female was admitted to our department presenting severe dyspnea, subjective sensation of edema in the cervical area, precordial pain accentuated by deep breathing and palpitations with sudden onset, approximatively 3 days ago. Medical history revealed a respiratory intercourse three weeks ago, condition which required antibiotics. At presentation, physical examination outlines the following features: a body temperature of 37.9°C, the

<i>Material</i>	<i>Analyses</i>	<i>Result</i>	<i>Values</i>
Blood	Haemocultures - bacteria	Negative	
	Haemocultures - fungus	Negative	
	RFC Coxiella Burneti	Negative	
	RFC Mycoplasma pneumoniae	Positive	1/640
	RFC Leptospira icterohemoragie	Negative	
	RFC Leptospira pomona	Negative	
Pericardial effusion	Macroscopic	Hemorrhagic	
	Culture	Sterile	
	Cytology	Nonspecific inflammation, malignant cells	
	Biochemistry	LDH	816 mg/dl
		Glucose	90 mg/dl
		Proteins	4.1
Urine	Culture	Sterile	
	Biochemistry	Normal	

Table 1. The laboratory findings including blood culture, pericardial liquid examination and urine analysis

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respiratory rate 35/min, low intensity heart sounds; blood pressure was 100/60 mmHg, and heart rate 75 beats per minute.

Electrocardiogram reflected the presence of sinus rhythm with a ventricular rate of 75/min, diffuse negative T wave (Figure 1).

We performed a set of global laboratory findings in order to identify more elements for the final diagnosis. There was an inflammatory syndrome certified by increased CRP, erythrocyte sedimentation rate, with normal leucocytes, neutrophil, lymphocytes, TSH, fT4, fT3 and mild hypoproteinemia. We wanted to exclude other sites of infection so we indicated a urine analysis which was normal. Also protein electrophoresis was the next step due to decreased amount of proteins in the blood; the only elements which were mildly raised were the alpha 2 globulins. The blood cultures were negative and those for atypical bacteria demonstrated the presence of *Mycoplasma Pneumoniae* (Table 1).

Radiographic exam identified the ascension of the right diaphragm with 6 cm, without focal lung injury and enlarged cardiac silhouette (Figure 2). In some cases the classic enlarged cardiac silhouette may not appear in acute tamponade due to the fact that the process requires at least 200 mL of pericardial fluid⁽³⁾.

Transthoracic echocardiography revealed reduced ejection fraction (<35%) measured

by Simpson's rule, pericardial effusion 19 mm laterally of right ventricle appearing in systole, with swinging heart, collapse of the lateral wall of the right atrium, diastolic collapse of the right ventricle, wave E decrease by more than 25% in inspiration and increase in expiration, which is consistent with cardiac tamponade (Figure 3). Pericardiocentesis was quickly demanded and the macroscopic appearance was that of hemorrhagic fluid with atypical malignant cells⁽¹⁾. After the procedure the BP got normal values (130/80 mmHg) with improving the dyspnea and the initial accuses.

Another element needed was the abdominal echography showing no modification. We requested a more reliable examination such as thoracic CT, an important element for this case management which showed pericardial effusion, an expansive mass in the anterior mediastina which was compressing the superior cava vein and aortic adenopathy, reflecting the suspicion of lymphoma (Figure 4). The abdominal CT didn't reveal notable elements.

Thoracic surgeons were the ones who succeeded to obtain some biopsies from the pericardial layer through a mini-thoracotomy. Considering the invasion of the mediastinal formation in the pericardium and upper cava vein, without the possibility of pericardiotomy or fenestration, the best solution was to perform a ultrasound guided pericardial

Figure 2. The chest X ray showing an enlargement of the cardiac silhouette

Figure 3. ECG showing sinus rhythm, 75 beats/min, diffuse negative T wave in inferior and anter-septal limbs

Figure 3. TTE - Apical 4 chambers view - the pericardial effusion is illustrated as well as the diastolic collapse of the right ventricle

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Figure 4 . Axial thoracic CT examination revealing a mediastinal mass invading the upper cava vein (arrows)

drainage using a catheter of 7 Ch⁽⁴⁾. The results of these maneuvers have materialized in the considerable improvement of dyspnea and the inflammatory markers showed a declining trend.

The pathological exam gave the final verdict: classical Hodgkin's lymphoma- nodular sclerosis type, describing tumor cells with nuclear atypia and intratumor necrosis. Reed Sternberg cells positive to CD15 and CD30 markers were also discovered. The patient was directed to oncology department in order to find the best therapeutic option for this disease.

Discussions

Neoplastic etiology of the pericardial effusion is a common finding in some malignancies but there are a few cases of HL discovered due to the cardiac tamponade manifestation. Only 5% of the patients with HL have associated pericarditis⁽⁶⁾.

Moreover non-Hodgkin's lymphoma is rarely responsible for the pericardial involvement. The incidence of pericarditis was evaluated in a systematic review gathering 1600 patients with leukemia where the follow up

included periodically echocardiography examination. The analysis of this retrospective study reveals that 325 patients (20%) had pericarditis, most of the reported cases showed mild pericardial effusion, only 3% required drainage of the pericardial fluid⁽⁷⁾.

Pericardial diseases are difficult to detect in hemodynamically stable patients with normal physical examination, the only element that could raise the suspicion of a pericardial effusion is the unexplained tachycardia⁽⁶⁾. In the presented case the medical history describing a recent infection could have given the fake impression that the substrate was an infectious pericarditis. Usually the neoplastic pericarditis is asymptomatic.

The cardiac tamponade represents a medical emergency and requires immediate medical interventions that can improve the patient condition. This is the result of the pericardial liquid pressure that overcomes the cardiac filling pressure causing an improper filling of the ventricle with drastically drop in the cardiac output. Pericardiocentesis is the immediate solution.

The mechanisms leading to pericarditis in malignancies are various. There can be a metastasis of the tumoral cells via the lymphatic or hematogenous circulation⁽⁸⁾. Affecting the circuit of the pericardial fluid by diminishing the amount of liquid which is reabsorbed in the venous and lymphatic vessels may represent another hypothesis. Finally the mediastinal large adenopathy can impair the lymphatic drainage of the pericardial fluid without invasion of the pericardium layer⁽⁹⁾.

The major problem concerning this pathology is the high recurrence rate. A pleuropericardial window or intrapericardial infusion of the antineoplastic agent and pericardiectomy

may represent important elements for the treatment of pericardial neoplasia. Usually in lymphomas systemic chemotherapy is suitable due to the fact that the most important mechanism of metastasis is the hematogenous pathway. Other solid tumors like lung cancer require both local and systemic chemotherapy⁽¹⁰⁾.

The particularity of this case relies in:

- the initial presentation of the underlying HL as an exudative pericarditis complicated with cardiac tamponade in a young patient without medical history.
- the fact that the initial suspicion based on a recent respiratory infection may have mislead the diagnostic algorithm and the treatment if there wasn't the clinical suspicion of another underlying cause of the massive hemorrhagic pericardial effusion.
- the normal values for the BP and the immediate relief of symptoms were obtained after the drainage of the large pericardial effusion.

Conclusions

The best therapeutic choice for treating pericardial diseases requires the proper identification of the major cause responsible for the disease. Even if the given information leads to a final conclusion we should always consider the patient's symptoms. Occasionally, the initial clinical evaluation reveals a distinct finding that fits into a more circumscribed problem⁽⁴⁾. Cardiac tamponade presence and the hemorrhagic aspect of pericardial effusion are the major findings that abolished the initial presumption of infectious pericarditis⁽²⁾. All the possible locations that may constitute the starting

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point for an infectious pericarditis must be ruled out. There are some cases of large pericardial effusion without an obvious cause following routine evaluation and they suggest the presence of an malignant process⁽³⁾. Malignant causes of pericardial disease represent a rare condition in young people, but this fact shouldn't stop us from eliminating the possibility when we are trying to find out the causes of the diseases. We should consult the guidelines and corroborate the data with our clinical judgement in order to find the successful approach for our patients.

The authors declare that there are not conflicts of interest.

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