Cardiac Extension of Primary Mediastinal Seminoma Compressing the Right Ventricular Outflow Tract

Lurildo R. Saraiva, Djair Brindeiro Fº, Thiago B. Saraiva, Mauro B. Arruda, Vital Lira

Recife, PE - Brazil

We report the case of a 33-year-old male with primary seminoma of the anterior mediastinum with initial clinical manifestations suggestive of heart disease.

Tumors of the anterior mediastinum are currently represented by seminoma or primary germinoma 25 to 30% of the time, with a marked preponderance in the male sex[1].

These neoplasms are invasive and poorly differentiated, characteristics accounting for their excellent regressive response to chemotherapy or radiotherapy. These neoplasms are almost always related to the thymus, hence the name thymic seminoma has also been proposed for their designation [1].

Involvement of the pericardium by tumoral tissue is not rare [2], but myocardial and endocardial invasion up to the point of obstructing the right ventricular outflow tract and compressing the pulmonary artery, mimicking a stenotic lesion, is an uncommon event, rarely reported in the literature. Describing the occurrence of such a tumor is the motive of this report.

Case Report

The patient is a 33-year-old male laborer from Jaboatão dos Guararapes, Pernambuco State, who sought the cardiology outpatient clinic complaining of fatigue on exertion, tachycardic palpitations, and precordialgia for 50 days. The patient reported that the precordial pain increased in intensity with inspiration, and was alleviated with ordinary analgesics. He also reported asthenia, anorexia, and recent weight loss of approximately 2 kg in the last 15 days. He denied a rheumatic antecedent, alcohol abuse, smoking, venereal diseases, and diabetes mellitus.

On physical examination, the patient weighed 62.0kg and was 1.73m tall (body mass index = 20.7kg/m^2). He was in regular condition, eupneic, anicteric, and with no peripheral cyanosis. His pulses were symmetrical and regular, his blood pressure was 140/90mmHg, and his heart rate was 88bpm. No jugular stasis occurred at 45°.

On precordial examination, clear systolic impulsions were observed on the pulmonary area, and no thrill existed. The cardiac rhythm was regular; the first cardiac sound had a normal intensity in the mitral area, and the second cardiac sound was split, variable, with a muffled pulmonary component. A moderate (++/4) rude ejection systolic murmur was auscultated in the pulmonary, aortic, and mesocardial areas. A forth cardiac sound could be heard in the mitral area.

The lungs were clear and no hepatosplenomegaly existed.

Laboratory tests were as follows: hemoglobin, 14.3g/dL; hematocrit, 43%; leukocytes, 5,700/mm^3; normal levels of urea and creatinine; antistreptolysin O, 200 U Todd.

Chest X-ray in the posteroanterior view depicted a suggestive enlargement of the right ventricle, bulging of the middle arch (+/2), decreased pulmonary circulation, and mild enlargement of the mediastinum.

The electrocardiogram showed sinus rhythm and was compatible with right ventricular hypertrophy and clockwise rotation of the heart, and an elevation of the ST segment with superior concavity in the inferior and lateral leads (QRS axis of +100°) was observed.

Doppler echocardiography (fig. 1) revealed enlargement of the right chambers, hypertrophy of the interventricular septum, a small pericardial effusion, and a tumor invading the right ventricular outflow tract, compressing the pulmonary artery trunk at the origin of its branches. Systolic pressure in the right ventricle was estimated in 95mmHg.

On thoracotomy, a large nonresectable tumor was observed involving the pericardial sac and the entire heart. A histopathological examination of a section of the tumor (fig. 2) revealed a probably primary neoplasia of the thymus comprising sheets of polygonal cells and septa in a delicate fibrous stroma with lymphocytic infiltrate. The neoplastic cells had large nuclei with fine chromatin and clear slightly acidophilic cytoplasm.

Universidade Federal de Pernambuco and Serviço de Ecocardiografia do Recife (SECOR)
Mailing address: Lurildo R. Saraiva - Hospital das Clínicas da Universidade Federal de Pernambuco - Av. Moraes Rego, SN - 50670-420 - Recife, PE - Brazil
English version by Stela Maris C. Gandour

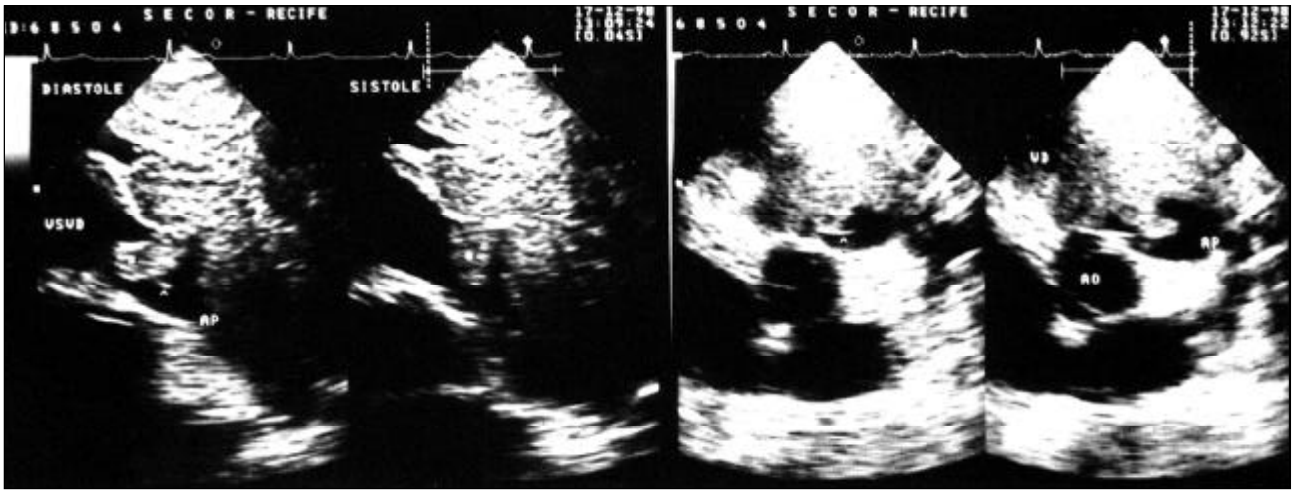

Fig. 1 – Two-dimensional echocardiogram, paraesternal longitudinal view, at the level of the pulmonary valve: a large tumor mass is seen in the superior part of the anterior mediastinum invading the right ventricular outflow tract (RVOT, asterisks), reaching the pulmonary valve (arrows), and compressing the pulmonary artery (PA) at the origin of its 2 branches.

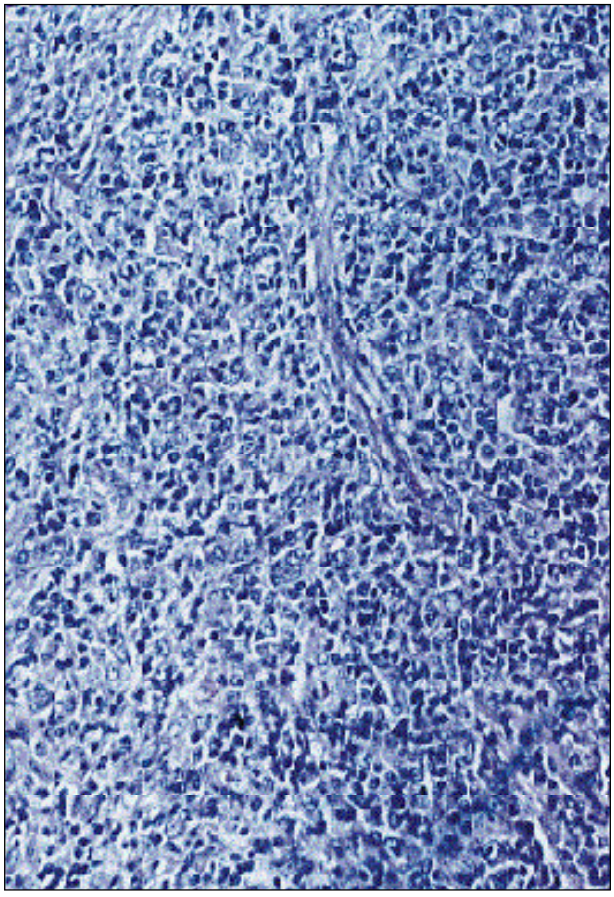

Fig. 2 – Polygonal cells in an alveolar pattern with pleomorphic nuclei and fibrous septa with lymphocytic infiltrate. (HE, approx. 250 x).

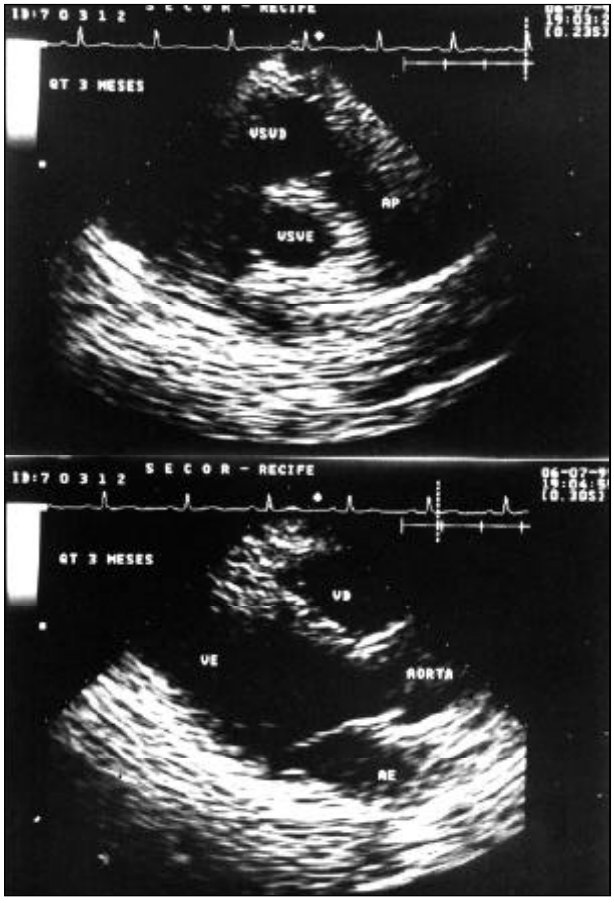

Fig. 3 - Normal echocardiographic study, 90 days after chemotherapy.

The patient was referred to the oncology unit, where he underwent periodical cycles of chemotherapy consisting of ectoposide, bleomycin, and cisplatin. The outcome was successful with a normal echocardiogram 3 months after chemotherapy (fig. 3). One year and 3 months after his admission to the hospital, the patient had no complaints and his cardiac assessment was normal.

Discussion

The curious finding in this case is that the attention of the physician was first drawn to a probable diagnosis of heart disease in a young male, who, in reality, had a malignant neoplasia of the mediastinum. For this initial false impression, findings resulting from the physical examination

of the heart contributed, as did the electrocardiogram and the nature of the alterations in the cardiac silhouette on roentgen examination, which were very suggestive of narrowing of the pulmonary valve. However, precordialgia with pleural features and recent onset does not constitute a usual symptom of such a disease [3], except for the retrosternal pain in severe congenital stenotic forms [4]. This pain along with the mild enlargement of the mediastinum oriented the clinical investigation in a diverse direction.

The echocardiographic alterations (fig. 1) are very significant. In addition to providing a rapid diagnosis of neoplasia, its localization in the right ventricular outflow tract and pulmonary artery trunk allows understanding of the semiologic findings, such as the systolic impulsions, the rude systolic murmur, the decreased intensity of the second heart sound in the pulmonary area, and the precordialgia. These findings resulted from the right ventricular obstruction and compression of the pulmonary artery in one side, and from pericardial involvement in the other.

According to Roberts [5], extension of the neoplasia to the heart is most frequently seen in leukemias and breast and lung carcinomas; in the latter, pericardial constriction may occur. According to Cox [6], the primary malignant germinal tumors, formerly included under the broad term "teratomas", clinically manifest as chest pain with a pleural component and cough. Hemoptysis, pneumonia, and dyspnea on exertion are rarer findings in these tumors, which are preferentially located in the anterior mediastinum, and more rarely in the middle mediastinum [1].

Even though this is not the case of a cardiac metastasis – the term "extension" here is more appropriate [5] – in the 14 truly metastatic cases of diverse neoplasias (pancreatic and colon adenocarcinomas, and melanoma, among others) reported by Labid et al [7], the finding of systolic murmur with characteristics similar to those of our patient's cardiac murmur in the presence of dyspnea on exertion is the major clinical feature. Another unique observation in their patients was the usual electrocardiographic finding compatible with complete or incomplete right bundle-branch block and right ventricular hypertrophy, as seen in our patient, who also had morphological changes in the ST segment, recalling pericardial involvement, which might have contributed to the precordialgia.

Of unknown histogenesis, the scarce extragonadal germinomas, according to Kitami et al [1], are currently managed with initial surgical biopsy and subsequent chemotherapy. Hachiya et al [2] recognized the rare possibility of spontaneous regression in a 22-year-old Japanese male, in whom the severe necrosis related to tumor aspiration could explain regression due to restriction in blood flow.

Even though the neoplasia is radiosensitive, one third of the patients may relapse or even show metastases, hence the use of chemotherapy, which is in fact currently more appropriate [1].

On the basis of what has been reported, we think that exclusion of such a neoplasia is necessary whenever clinical findings compatible with obstruction of the right ventricular outflow tract are present in a young male complaining of asthenia, dyspnea, and thoracic pain with pleural characteristics, accompanied by radiological mediastinal enlargement.

References

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