Neuroendocrine carcinoma revealed by a tamponade: a case report

Carcinome neuroendocrine révélé par une tamponnade: à propos d'un cas

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SUMMARY

A 48-year-old man presented with prolonged retrosternal chest pain, dyspnea and asthenia. Physical examination showed tachycardia and polypnea. The patient's ECG showed diffuse microvoltage. Chest x-ray film showed cardiomegaly, para-hilar and para-cardiac opacities. An echocardiogram showed a abundant heterogeneous circumferential pericardial effusion. A puncture drainage was made urgently and a cytopathological study of fluid showed tumor cytology. CT scan revealed an anterior mediastinal mass primarily suggesting infiltrating thymoma. A scanno-guided biopsy showed neuro endocrine thymoma.

KEYWORDS

Pulmonary artery aneurysm, surgery

RÉSUMÉ

Un homme de 48 ans s'est présenté avec des douleurs thoraciques. L'examen physique a montré une tachycardie et une polypnée. L'ECG montrait un microvoltage. La radiographie thoracique montrait une cardiomégalie et des opacités. Une ETT a montré un épanchement péricardique circonférentiel hétérogène abondant. Un drainage par ponction a été réalisé en urgence et une étude cytopathologique du liquide a montré une cytologie tumorale. Le scanner a révélé une masse médiastinale antérieure évoquant principalement un thymome infiltrant. Une biopsie scanno-guidée a montré une tumeur neuro endocrine.

MOTS-CLÉS

Tamponnade

Carcinome neuroendocrine ; Thymome

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INTRODUCTION

Thymoma is the most frequent primary tumor in the anterosuperior mediastinum (1). Thymomas are usually asymptomatic. However, one-third of the patients present with symptoms owing to tumor compression or invasion of adjacent structures. Among the complications of thymomas, cardiac tamponade is unusual (2).

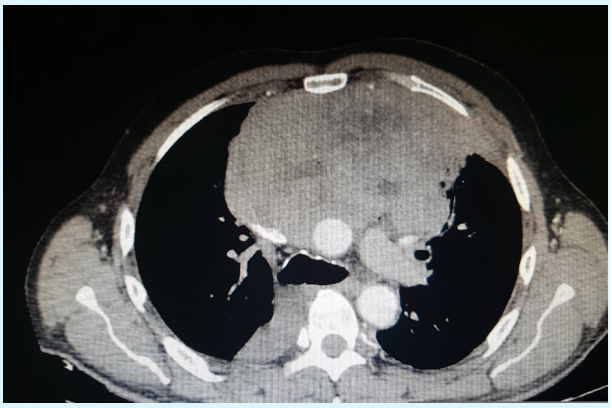
We report here a rare case of a patient with thymoma revealed by a tamponade as the first manifestation.

OBSERVATION

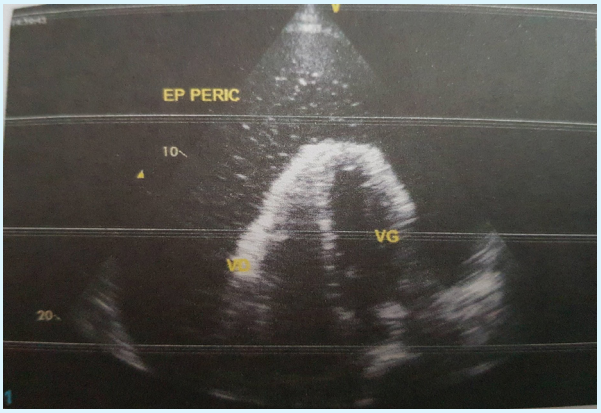
A 48-year-old man was admitted to our hospital, with complains of prolonged retrosternal tingling-type chest pain without irradiation, aggravated on deep inspiration and relieved by the lean forward position associated with dyspnea, dysphagia first to liquids then to solids, weight loss and asthenia since 2 months aggravating gradually.

Physical examination showed tachycardia (heart rate was 100 bpm), polypnea, muffled heart sounds, and turgid jugular veins.

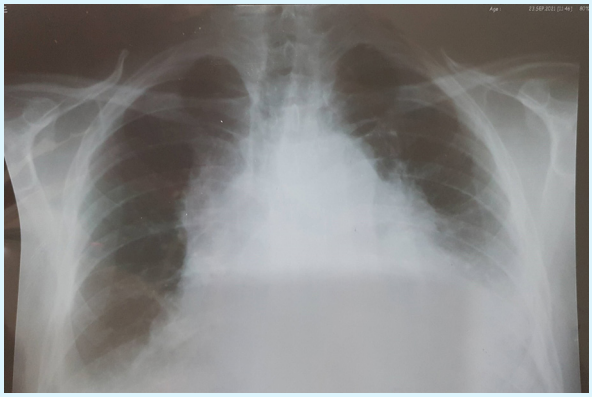
The patient's electrocardiogram showed diffuse microvoltage. Chest x-ray film showed cardiomegaly, para-hilar and para-cardiac opacities (Figure 1).

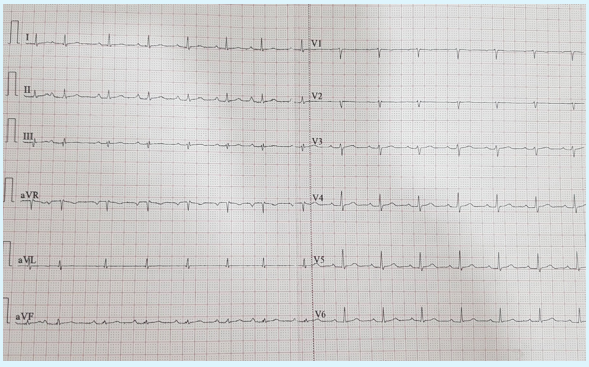


Laboratory data revealed C-reactive protein 7,23 mg/dl, white blood cells count are normal. An emergency echocardiogram showed a very abundant heterogeneous circumferential pericardial effusion (Figure 2). A puncture drainage was made urgently bringing back 750ml, the fluid was bloody and contained 45 g/l of protein. Fluid Cytopathological study showed tumoral cells.



A thoraco-abdomino-pelvic computerized tomography scan showed an anterior mediastinal mass primarily suggesting infiltrating thymoma with pericardial and pleural effusions of carcinomatous origin (Figure 3).





One week after the puncture, a control echocardiogram showed recurrence of large circumferential effusion, variation in mitral flow 23%, dilated and compliant IVC, paradoxical septum. He was presented to the thoracic surgery staff for surgical drainage and biopsy but this was not accepted since the mass was locally invasive and even the pleuro-pericardial window was impossible. A second echo guided puncture drainage was performed and a computerized tomography guided biopsy was performed revealing a neuroendocrine thymoma. Patient was offered a radio-chemotherapy protocol (cisplatin + doxorubicin with 40 Gy of thoracic radiotherapy) and died few weeks later.

DISCUSSION

It is crucial to diagnose thymic neuroendocrine carcinomas early to offer the best therapeutic options. The clinical presentation is varied and atypical. There are no specific clinical signs of thymic carcinoids, and they are present in about third of cases (3).

Most thymomas are seen in adults aged 45 to 50 years and are discovered on routine chest X-ray, especially with the lateral view (4,5).

Symptoms are various: chest pain or dull ache are the most frequent symptoms and are the result of local extension of the tumor, patients can also present with cough, fatigue, weight loss, or superior vena cava syndrome (4,5). The paraneoplastic syndrome of thymic carcinoma has been seen only rarely. Pericardial effusion or cardiac tamponade as an initial presentation of this cancer is extremely rare and appears more in the elderly (6). Particularly in our case, the severity and frequent recurrence of the pericardial effusions as well as the delayed diagnosis led to repeated hospitalizations and a delay in the therapy, which had a negative impact on the patient's prognosis.

Even in the presence of usual symptoms, cardiac tamponade diagnosis has consistently been delayed in reported cases. In the case series by Campbell et al, pericardial biopsy provided a diagnosis in 93% of the patients, whereas only in 15 (26%) patients (11 of whom had malignant effusions) diagnosis was made on pericardial fluid cytology (7,8). In our case, pericardial fluid investigation did not identify the cause of the cardiac tamponade at the time of presentation and the computerized tomography -guided biopsy confirmed the diagnosis.

In an advanced stage of tumor development, surgery would be incomplete or even impossible as is our case. So , definitive chemoradiotherapy is the standard of care in the absence of metastases (9). In our case, we used a combination of cisplatin and doxorubicin with 40 Gy of thoracic radiotherapy.

The most effective order of the multimodal treatment ultimately depends on the response to chemotherapy, degree of invasion, and likelihood of obtaining total resection, just like in locally advanced non-small cell lung cancer (9).

CONCLUSION

Neuroendocrine tumors of the thymus are aggressive and have a poor prognosis. As a result of unsatisfactory results with adjuvant therapy, surgical resection of the tumor and its extension remains the treatment of choice and the management of patients requires multidisciplinary approach.

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