L'échocardiographie faite devant l'apparition de trouble du rythme à type d'extrasystoles auriculaires et à la recherche d'une atteinte cardiaque de l'amylose ou de la sarcoïdose, a conclu à l'infiltration avec une cardiomyopathie hypertrophique et un aspect granité du cœur. Le diagnostic de l'association de sarcoïdose avec une amylose, un syndrome de Sjögren et de récidive d'angiomyxome agressif a été retenu. L'indication d'une éventuelle corticothérapie n'a pas été retenue, mais devant la nécessité de la reprise chirurgicale de l'angiomyxome pelvien, le patient a été traité en per opératoire par des boli de solumédrol à visée anti-inflammatoire. Le patient a été opéré en Décembre 2006. L'histologie de la pièce opératoire à confirmé la récidive de l'angiomyxome. L'évolution immédiate était compliquée d'une embolie pulmonaire ainsi qu'un syndrome occlusif ayant bien évolué sous traitement médical. L'évolution ultérieure est bonne avec un recul actuel de 10 mois.

Conclusion

Les arguments chronologiques : amylose précédant la sarcoïdose et la nature localisée de l'amylose ainsi que le type non AA plaident en faveur du caractère fortuit de cette association. Nous pensons que la révélation de cette association par l'angiomyxome agressif pelvien n'est qu'une coïncidence, hypothèse qui ne peut être infirmée ou confirmée que par l'étude de plus grandes cohortes.

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Calcific constrictive pericarditis

Pericardial inflammation secondary to mycobacterium tuberculosis infection is a rare condition but still observed in developing countries (1).

Constrictive pericarditis develops in three stages, an initial stage involving acute pericarditis with pericardial effusion and fibrin deposition, a subacute stage with organization and reabsorption of the effusion and chronic stage with fibrosis and thickening of the pericardium and calcium deposition (2).

This disease is associated with scarring and loss of elasticity of the pericardium, causing impaired cardiac filling.

Computed tomography is the gold standard imaging tool for cardiac calcification, it allows a nice anatomic delineation of the pericardium and its calcifications which may be very useful in determining the optimal surgical approach for pericardial resection. We submit this case not only for its striking appearance on computed tomography of the pericardium but also because tuberculosis remains a relatively common leading cause of pericarditis in Tunisia.

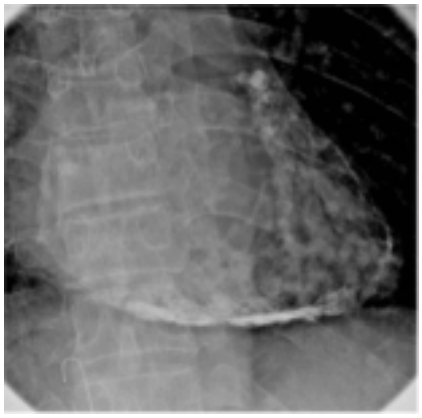
Observation

A 55-year-old male was admitted to our department for palpitation and dyspnea on exertion He had had pulmonary tuberculosis 17 years earlier. Physical examination revealed distension of the jugular vein, hepatomegaly, bilateral leg edema , Diminished and irregular heart sounds.

Electrocardiogramme showed atrial fibrillation and no specific repolarization changes.

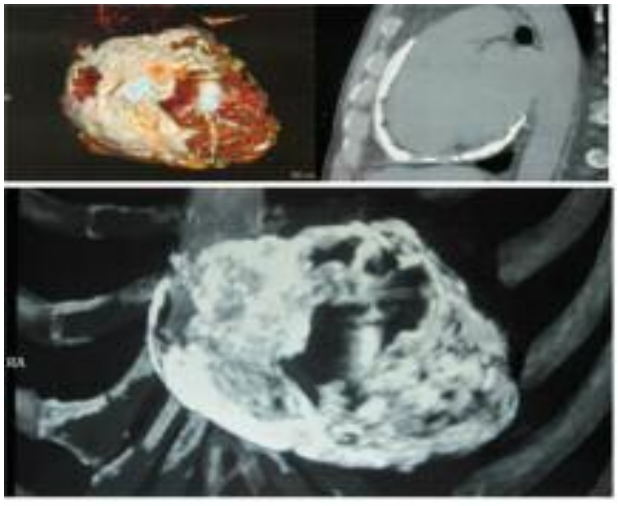
Chest radiography and fluoroscopy revealed heavy calcification of the pericardium (fig 1).

Figure 1 : The combination of imaging and hemodynamic findings established the diagnosis of constrictive pericarditis.



Cardiac Computed Tomography evidenced severe and extensive pericardial calcification giving " eggshell" appearance (fig 2).

Figure 2 : Severe dense calcification of the pericardium.
With "eggshell" appearance in multislice computed tomography



Catheterization and echocardiography confirmed the diagnosis of constrictive pericarditis which presumed to be due to old Tuberculosis infection .The patient underwent pericariectomy with good results.

Conclusion

Constrictive pericarditis should be suspected in patients with clinical features of right-sided heart failure.

Computed tomography (CT) is a useful method for accurate measurement of pericardial thickness and calcium distribution before pericardiectomy.

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Case report of a patient presenting both schizophrenia and Charcot-Marie-Tooth disease

Charcot-Marie-Tooth disease (CMT) is the most frequent inherited neuromuscular disorder (prevalence 17–40/100000) [1]. CMT comprises a group of disorders that affect peripheral nerves. CMT can be categorized into subtypes according to gene mutations [2]. Diagnosis is based on clinical findings and electromyography (EMG) and nervous conduction velocity characteristics.

CMT can rarely be associated with psychotic manifestations.

Psychosis is mainly caused by schizophrenia but can also be secondary to medical conditions or drug abuse.

Schizophrenia is the most frequent and debilitating psychotic disorder. It is the result of a complex group of genetic, psychological, and environmental factors.

Late adolescence and early adulthood are peak periods for the onset of schizophrenia. Its symptoms comprise the so-called positive symptoms or psychotic domain of schizophrenia. Negative symptoms of blunted affect, anhedonia, apathy, and alogia are a second symptom domain in schizophrenia.

Comorbidity between CMT and schizophrenia appears to be exceptional. This case report raises questions about the possible links between both conditions.

Case report

C. D. was diagnosed with CMT at the age of 16. He used to suffer from gait disturbances, while being an excellent student with no symptoms suggestive of a psychotic disorder.

Clinical features included weakness of the foot and lower leg muscles, foot deformities: high arches and hammertoes. The clinical examination noticed absent achilles' tendon reflex and a superficial sensory disturbance. In addition to this peripheral neurogenic syndrome, a quadripyramidal syndrome was also noticed consisting of tendon hyperreflexia in four limbs (except the achilles' tendon reflexe which was absent) and a positive bilateral Babinski sign.

An EMG was performed and showed a motor and sensory axonal neuropathy. Family tree suggested an autosomic dominant transmission.

The cerebro-medullar Magnetic Resonance Imaging was normal.

Thyroid function was normal.

HbS, HIV and syphilis were negative.

The patient was diagnosed with CMT associated with a pyramidal syndrome.

C.D. was diagnosed with schizophrenia in 2009, at the age of 27. The prodromal period lasted about one year consisting of deterioration of social functioning and social withdrawal.

The patient's clinical psychiatric features were: auditory hallucinations, persecutory delusion, behaviour disturbances consisting of self-neglect, obscene language and exhibitionism.

All other causes of psychosis were discussed and ruled out. The patient didn't take any medication that could account for his neurological nor psychiatric symptoms.

He was put on antipsychotics. Now he is on amisulpride 400mg qd and is well stabilized.

Family history was negative for schizophrenia.

Thus, C.D. had both schizophrenia and a form of CMT associated with a pyramidal syndrome. This involvement of the CNS in CMT does raise questions about the pathophysiology of both conditions and the possible links between them [3-5].

Conclusion

Comorbidity between schizophrenia and CMT is rare. It is even rarer and worth reporting when CMT with CNS involvement is concerned. It raises questions and suggests new theories about the pathophysiology of both diseases and the possible link between them [6]. No definite answer has been found so far. Additional research is needed to improve our knowledge of both diseases.

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