

Sudden Cardiac Death Secondary to Sarcoidosis

We describe the case of a 48-year-old woman with unremarkable medical history according to family referral. The only positive event reported was repeated episodes of syncope a week before her death.

The patient suddenly presented with nonspecific malaise and severe chest pain, and was immediately taken to hospital but died before admission; consequently, a judicial autopsy was requested to determine the cause of death.

A chest x-ray was performed, which showed a micronodular, cotton-like infiltrate in the perihilar lung parenchyma. No enlarged lymph nodes were observed. No skin lesions were detected on external examination. Internal examination revealed an enlarged heart weighing 476.93 grams, and smooth pericardium. Its cross-section showed large pearly whitish areas with up to 1 cm-thick hemorrhagic areas in the myocardium, involving both ventricles, the ventricular septum, and the papillary muscles. The lesion extended from the atrioventricular septum to 2 cm before the apex. The coronary arteries were patent. The lungs appeared congestive and edematous. No macroscopic enlarged lymph nodes were observed. The enlarged liver weighed 2.220 grams. The rest of the organs and systems showed no significant alterations.

The histological analysis of the heart revealed extensive lymphoplasmacytic infiltrate with eosinophils, histiocytes, and multinucleated giant cells with a "moth-eaten" cytoplasm, other cells with frosted-glass cytoplasm (Schaumann bodies) and others of starry aspect (asteroid bodies) which were circumscribing areas of non-caseous sarcoid granulomas involving the pericardium, septal and ventricular myocardium, papillary muscles and endocardium, dissecting the muscle fibers with areas of fibrosis. In some areas, nuclear pyknosis and homogenization of myocardial fibers were observed (Figures 1 and 2). The liver and lung parenchyma presented similar granulomas. Special staining using Ziehl Neelsen and PAS staining methods were performed, showing no specific microorganisms.

The diagnosis was sudden cardiac death secondary to systemic sarcoidosis with lethal cardiovascular involvement.

In order of increasing frequency, cardiac sarcoidosis (CS) affects: the free left ventricular wall followed by the ventricular septum, the conduction system, the right ventricle, and the pericardium. (2, 3, 6) In this case, the involvement extended to both ventricles, the septum, and the papillary muscles.

Clinically, arrhythmias are the most common manifestation; this condition should be suspected in young patients with complete atrioventricular block, ventricular arrhythmias, pathological ventricular hy-

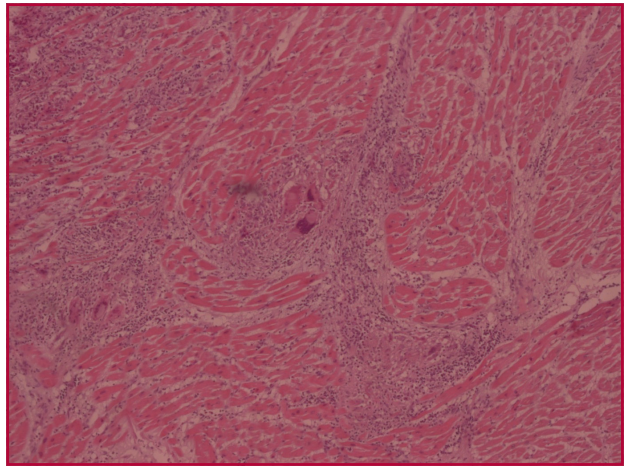


Fig. 1. Myocardial tissue histology.

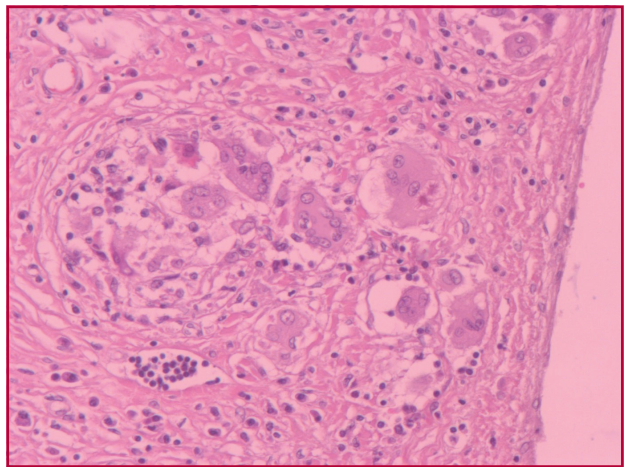


Fig. 2. Myocardial tissue histology.

pertrophy, abnormal wall motion or perfusion defect involving the anteroseptal and apical region, which improve with stress. (1, 2, 6)

In a series of autopsies analyzed by Silverman, 35% of the patients with CS were clinically silent and 4 patients presented sudden death. (3) The case presented here was clinically asymptomatic up to a week prior to death, when the patient underwent multiple syncopes that ended up in sudden death. When there is extrapulmonary involvement, and particularly cardiac involvement, an aggressive course is denoted with poor prognosis. (2)

Liver involvement is observed in 80-95% of cases, and may be asymptomatic in 2-60%, as observed in our case. (1, 4)

Steroid and immunosuppressive therapy is the treatment of choice, and in cases of heart disease, early use of intracardiac implantable devices is advised in

case of conduction disorders. For patients progressing to end-stage heart failure, heart transplantation is the therapeutic option. (2, 3, 5, 6)

We consider this case to be of interest because it is a systemic disease with a silent course and fatal outcome.

Conflicts of interest

None declared.

(See authors' conflicts of interest forms on the website/Supplementary material).

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Symptomatic Intracavitary Cardiac Metastasis 12 Years after Breast Cancer Diagnosis

We report the case of a 47-year-old female patient with history of infiltrating lobular carcinoma diagnosed in 2005, who underwent left modified radical mastectomy, received full chemotherapy and radiotherapy treatment, and is currently treated with tamoxifen.

The patient consulted her primary care physician for functional class II/III dyspnea, which worsened in dorsal recumbent position and changes in position; concomitantly, she consulted for lassitude, fatigue and general malaise. Her primary care physician requested a basic panel of complementary studies, including a transthoracic echocardiography, which showed a 2 cm x 2 cm hyperechoic image attached to the atrial septum, and was therefore referred to the cardiology department of Hospital Interzonal Especializado de Agudos y Crónicos (H.I.E. A y C) San Juan de Dios, in the city of La Plata, with presumptive diagnosis of atrial myxoma.

The patient appeared in good general condition, with vital signs within normal limits. Positive signs

at physical examination revealed a diastolic mitral inflow murmur on auscultation (2/4 intensity) and tumor plop in the mitral area in left lateral recumbent position (Pachon), with acoustic changes according to the recumbent position adopted.

A general laboratory test was requested, which showed erythrocyte sedimentation rate of 120 mm/h as only pathological finding. The electrocardiogram showed sinus rhythm at 95 bpm, with no other abnormalities.

Chest X-ray. A posteroanterior teleradiography of the chest taken with adequate technique did not show bone lesions.

Asymmetric lung fields were diagnosed, with predominance of right basal and cardiophrenic infiltrate that was inconsistent with the patient's respiratory symptoms; it was therefore interpreted as right breast images. The cardiac silhouette and the great vessels were within normal limits.

Transesophageal echocardiography revealed a 1.8 cm x 1.8 cm mobile image in contact with the atrial septum and attached to the anterior mitral valve, below the oval fossa. The rest of the examination was normal. (Figure 1)

The CT scan of the chest, abdomen and pelvis, with IV contrast showed lytic images at the level of the sternum, thoracic and lumbar spine, and sacral bone (Figure 2) and a hypodense image in the IV hepatic lobe, consistent with the secondary lesions. In addition, a hypodense tumor in the left atrium and abnormal tissue infiltrating the right breast were detected.

On the basis of the tumor found in the left atrium without morphological characteristics of cardiac myxoma, as it was not in contact with the oval fossa - a common characteristic in myxomas -, without presenting heterogeneous echo contrast, (1) and taking into account the other metastatic stages, suspected intracavitary cardiac metastasis was diagnosed.



Fig. 1. Transesophageal echocardiography at 0 degrees, showing a 1.8 cm x 1.8 cm hyperechoic image in the left atrium, in contact with the atrial septum, below the oval fossa, and closely attached to the anterior mitral leaflet.

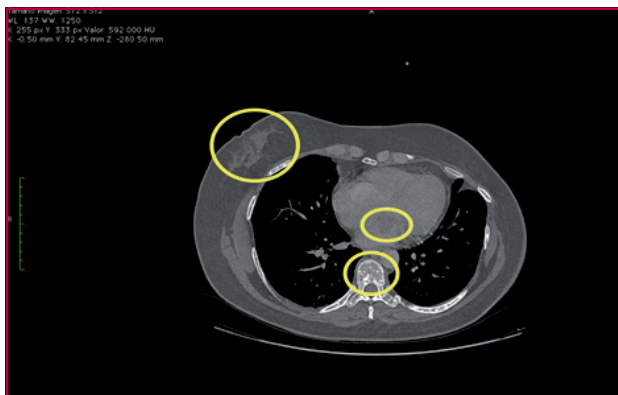


Fig. 2. Chest CT scan with mediastinal window showing rounded hypodense lesion at the left atrial level. It also shows lytic lesions at the thoracic vertebra, and image with soft tissue density, anechoic in the right breast. Left mastectomy.

A consultation request with the Oncology Department was carried out: the patient was staged and placed on a chemotherapy plan.

From the cardiac point of view, the patient was discharged from hospital under follow-up by the cardio-oncology department to monitor the cardiotoxicity produced by chemotherapy; analgesic and psychological treatments were set up.

During follow-up, 6 months after starting chemotherapy, the patient developed pancytopenia. The patient was admitted in the oncology service, where she worsened and died. The autopsy revealed infiltrating, friable tumor-like tissue, and mammary neoplastic cells were detected by microscopic examination.

Primary malignant cardiac tumors are unusual, with a frequency estimated at 0.02%; however, metastatic and secondary cardiac tumors are 20-40 times more common than primary benign or malignant tumors, as is the case in our patient. (2) Any neoplasm can cause intracavitary or pericardial -a more frequent localization-, cardiac metastasis, but melanomas, mediastinal tumors, esophageal and lung tumors, and breast tumors, as in our case, are more associated with cardiac metastases. (2)

Regarding sign-symptomatology of intracavitary tumors, only 10% of the patients have symptoms, the most common being de novo heart failure. (2)

Other less specific symptoms include fever, lassitude, general discomfort, arthralgias, and skin rash, all caused by tumor cytokine release into the bloodstream. (3)

Today, diagnostic methods par excellence are echocardiography, cardiac computed tomography and cardiac functional magnetic resonance imaging, which will provide the diagnosis of the tumor image and will guide to the type of tumor according to imaging characteristics. (4-5)

Definite diagnosis can only be made by biopsy or autopsy.

With respect to treatment, due to the spread of neo-

plasms when staging tumors, the prognosis of cardiac metastases is ominous, and the decision of an invasive or non-invasive approach should be balanced, the latter being the appropriate approach in most cases.

The case we have reported of a patient with intracavitary cardiac metastasis of a primary breast tumor is a rare finding, with a clinical and surgical management not fully researched in the current medical literature.

The therapeutic approach should be agreed upon in a multidisciplinary fashion, and the invasive approach should be evaluated, which in most cases is not appropriate.

We want to make it clear that the accuracy of the diagnosis should not be persistently searched for and all available resources should be used to avoid invading the patient's body with the simple aim of getting to the truth.

Conflicts of interest

None declared.

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Orthodeoxia and Permanent Pacemaker Implantation

Patent foramen ovale (PFO) is a common finding in the adult population, with a prevalence of around 25%, (1) evenly distributed by sex. Although PFO is, in most cases, a casual finding, it has been associ-

ated with embolic stroke, platypnea-orthodeoxia syndrome, gas embolism in divers, or migraine. (2)

Atrial septal aneurysm (ASA) has a prevalence of 1 to 4.9%, (3) and 33% is associated with PFO. (4)

The implantation of a permanent pacemaker is usually considered a minor surgical procedure, with low risk of perioperative complications. (5)

We describe the case of a patient who developed orthodeoxia (hypoxemia induced by orthostatism) during the postoperative period after permanent pacemaker implantation, in the context of PFO and ASA.

Transesophageal echocardiography (TEE) showed that the lead had displaced the atrial septum, facilitating right-to-left shunting in supine position. Agitated saline as contrast was used to confirm the shunt. Relocation of the lead was decided, resolving the condition.

Our report highlights the importance of considering the presence of PFO and ASA with intermittent right-to-left flow among the differential diagnoses of hypoxemia after pacemaker lead implantation.

This is the case of a 91-year-old male patient with history of hypertension and acute myocardial infarction, who was treated in 2010 and had chronic non-oliguric renal failure. He was admitted to the coronary care unit for high-risk syncope due to bifascicular block and prolonged PR, on a permanent pacemaker implantation plan.

Electrocardiographic complementary studies showed sinus rhythm, LAHB, CRBBB, and PR interval of 0.24 sec. Color Doppler ultrasonography revealed mild left atrial enlargement, and inferior basal and medial hypokinesia with preserved left ventricular systolic function.

Laboratory tests reported hematocrit 39%, creatinine 1.42 mg/dl, Na 135 mEq/L, K 4 mEq/L, and blood glucose 130 mg/dl.

The endocavitary device was implanted by an experienced surgeon in an operating room equipped with radioscopy. Twenty-four hours after the procedure, pulse oximetry showed a drop in oxygen saturation in sitting position, which improved with recumbent position, with BP 120/80 mmHg, HR 80 beats per minute, afebrile and lucid. A chest x-ray showed no complications associated with the procedure; in addition, a helical CT scan of the chest ruled out pulmonary thromboembolism.

Two consecutive arterial blood samples were obtained; the first one in dorsal recumbent position, whose values were pH 7.52, pCO₂ 26.7, pO₂ 63, bicarbonate 20, O₂ saturation 93.6%, and BE - 2.1; and the second sample in sitting position, whose values were pH 7.52, pCO₂ 24.7, pO₂ 42, bicarbonate 20, saturation 83.6%, BE -1.6.

Color Doppler ultrasonography revealed -after IV administration of agitated saline- the passage of bubbles through the atrial septum into the left chambers. A TEE was then performed, detecting a type III atrial septal aneurysm with atrial septum thinning at the

level of the oval fossa.

The pacemaker lead displaced the upper portion of the atrial septum toward the left atrium with a large amount of right-to-left bubble passage between the first and second heartbeats, consistent with PFO (Figure 1).

Surgical repositioning of the pacemaker lead was decided with removal of the generator, and disconnection and mobilization of the ventricular lead under radioscopy monitoring. An intraprocedure study was performed, and TEE with color Doppler confirmed right-to-left atrial shunt reduction.

Transesophageal echocardiography was repeated after the procedure, showing ASA with septal defect at the level of the oval fossa, consistent with ostium secundum type atrial septal defect. After injection of agitated saline solution, no contrast passage to the left chambers was observed (Figure 2). The patient had an asymptomatic outcome, with no variation in saturation due to body position.

The foramen ovale is a communication between the right and left atria, necessary during fetal life because it allows the passage of oxygenated blood from the placenta to the fetal systemic circulation. Immediately after birth, the decrease in pulmonary resistance and increase in left atrial pressure -as a consequence of increased venous return- produce its functional closure.

During adulthood, PFO is associated with the development of multiple conditions, including paradoxical embolism, cryptogenic stroke, and hypoxemia due to right-to-left flow.(2) It is a common finding in the adult population, with a prevalence of approximately 25%, which decreases with age, and is around 20% in adults >80. Patent foramen ovale size varies from 1 to 19 mm (mean 4.9 mm); in the first decade, mean diameter is 3.4 mm and increases to 5.8 mm in adults >90 years. (1)

Atrial septal aneurysm is defined as a mobile pro-

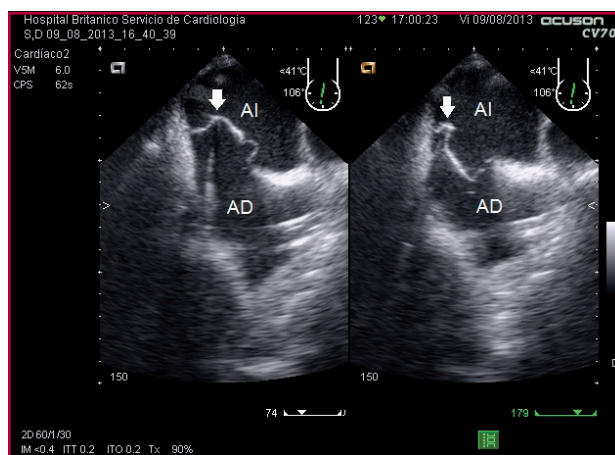


Fig. 1. Displacement of the upper portion of the atrial septum into the left atrium, caused by the pacemaker lead (arrow). RA: Right atrium. LA: Left atrium

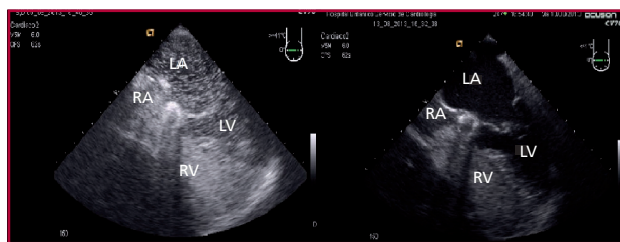


Fig. 2. Contrast agitated saline solution pre- and post-repositioning of pacemaker lead.

RA: Right atrium. LA: Left atrium. RV: Right ventricle. LV: Left ventricle.

trusion of the septum primum tissue into the atrium measuring at least 10 to 15 mm or a septal excursion of at least 15 mm occurring at some point during the cardiorespiratory cycle. The definitions of ASA vary widely in the literature. Certain conditions such as pulmonary stenosis, pulmonary hypertension, chronic obstructive or restrictive pulmonary disease, acute or chronic recurrent pulmonary embolism, and right ventricular infarction, worsen hypoxemia in the presence of PFO.

In the absence of pulmonary hypertension, right-to-left flow may be caused by mechanisms such as anomalies of the systemic venous return to the left atrium through the superior vena cava, or blood flow from the inferior vena cava to the left atrium through an atrial septal defect. In the latter case, the anatomy of the inferior vena cava, the Eustachian valve or the septal defect allow for a marked preferential right-to-left flow in the absence of pressure gradient. (2) It is crucial to determine the presence of right-to-left flow due to the risk of hypoxemia and paradoxical embolism.

The treatment of choice for PFO is not well defined, and further information is needed to elucidate which is the best therapeutic option. (2, 6)

We assume that the orthodeoxia in our patient was the result of atrial septum displacement caused by the pacemaker lead, which increased the pre-existing defect and triggered preferential right-to-left flow during supine position. Since he had no symptomatic component related to positional changes, his orthodeoxia cannot be strictly framed within the platypnea-orthodeoxia syndrome. As orthodeoxia remitted with the repositioning of the lead, it was not necessary to consider other therapeutic approaches for PFO.

This case reveals the importance of clinical and biological control, with oxygen saturation measurement in different positions in patients with PFO who require permanent pacemaker implantation.

Conflicts of interest

None declared.

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Subcutaneous Implantable Cardioverter Defibrillator in Patients with Cardiomyopathy

Subcutaneous implantable cardioverter defibrillator (S-ICD) is recommended as an option to the implantable cardioverter defibrillator (ICD) in patients who do not require stimulation, antitachycardia stimulation therapies, or resynchronization, as well as in young patients who will need a device for a long time or who have problems due to infection or endovascular access.

The patient characteristics detailed below turned him into the recipient of the first S-ICD in Argentina, authorized as a "compassionate" treatment.

This was the case of a 29-year-old male patient, with a history of cleft lip and palate that underwent surgical treatment at 8 months of age, with no family history of sudden death, who performed non-competitive cycling (up to 20 km/day), and swimming three times a week.

The patient consulted with a 4-month history of short duration palpitations, initially on exertion and later also at rest, and occasional functional class II dyspnea.

The electrocardiogram (Figure 1) showed sinus rhythm, right and left atrial overload, right bundle branch block, epsilon wave and negative T-waves in V1 to V6. The echocardiogram revealed severe dilated right chambers and hypokinesis of the right ventricular free wall. Anomalous venous drainage was ruled out by this method.

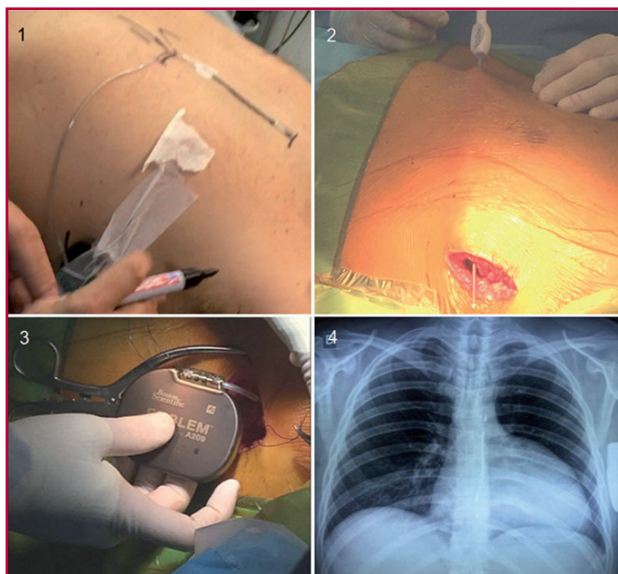


Fig. 2. Subcutaneous implantable cardioverter defibrillator implantation. 1. Cutaneous marks; 2: Subcutaneous tunneling; 3: Generator implantation in mid-axillary line; 4: Rx control.

reported in 2002 with the publication of Bardy and Capato's experience with a prototype, and was first commercialized in 2009.

In order to study this new technology, two pioneering registries have been performed: the EFFORTLESS study in June 2009, with 472 patients, and the IDE registry, with 197 patients, from January 2010 to May 2011, which provided the basis for the implantation and follow-up of patients with this therapy, leading to the FDA approval for the use of S-ICD in 2012. The 2015 European guidelines (6) include its use with a class IIa recommendation, level of evidence C, as an alternative to the traditional ICD, in patients who do not require stimulation, antitachycardia stimulation therapies, or resynchronization. It is also suggested, with IIb recommendation, for young patients who will need a device for a long time or who have problems due to infection or endovascular access. It is important to point out that, in addition to the populations that are not eligible for this device, some patients have inadequate ECG signals for this therapy. In order to identify these patients, an automatic screening in the programmer, instead of manual screening, is currently used, which is performed in dorsal recumbent position, sitting position, and eventually, during exercise. Exclusion predictors include obesity, hypertrophic cardiomyopathy and inadequate R/T wave amplitude ratio. Our patient underwent automatic screening and an adequate recording was obtained in two of the three leads, two of which are established between the carcass and the two electrodes, on both sides of the coil, and a third one between them.

Today, four studies are being developed to answer questions about the S-ICD: the PRAETORIAN study (transvenous ICD vs. S-ICD) in patients with primary

and secondary prevention, which has already finished inclusion and will be published next year; the UNTOUCHED study, with a registry that evaluates the performance and effectiveness in patients with left ventricular dysfunction; the S-ICD System Post Approval Study and the MADIT S-ICD, a registry that compares S-ICD with medical therapy in patients with ischemic heart disease and diabetes with moderate LV systolic functional impairment.

Even though Argentina was in 1985 a pioneer in ICD implantation in Latin America, the national situation in this regard delayed the start of the S-ICD implants. The patient described has been the first implanted case in Argentina. His particular severity motivated the management of implantation authorization through established channels for compassionate therapies. We hope that our experience will be the precedent that will encourage the use of this technology for patients who require it.

Conflicts of interest

Dr. Claudio Muratore discloses that he works for Medtronic. (See authors' conflicts of interest forms on the web/Supplementary material).

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