CASE REPORT

ACTA MÉDICA PORTUGUESA 1994; 7: 57-60

RECURRENT INTRACARDIAC MASS IN A PREGNANT WOMAN WITH ANTIPHOSPHOLIPID SYNDROME

VÍTOR MATOS, BEATRIZ PINHEIRO, PEDRO MAIA, HELENA LEITE, ÁLVARO COELHO, ANTÓNIO FERNANDES

Medicine and Cardiology Departments. Coimbra Hospital Centre. Coimbra

SUMMARY

Clinical manifestations of the antiphospholipid syndrome result from thromboembolic phenomena that occur in all vascular territories. Cardiac manifestations frequently associated with this syndrome include valvular and myocardial lesions. We present a case report of primary antiphospholipid syndrome in a pregnant young woman with a right atrial mass detected by echocardiography. Its morphological characteristics presented problems in establishing differential diagnosis within atrial thrombus and atrial myxoma. This case was complicated by pulmonary embolism and recurrence of the mass within four months. Cardiac surgery was performed and two masses excised. Histopathological studies showed them to be thrombotic in nature.

INTRODUCTION

The Antiphospholipid Syndrome, described by Harris and cols. in 1987 [1], is defined by the association of arterial and/or venous thrombosis, spontaneous recurrent abortion, thrombocytopenia and presence of antiphospholipid antibodies (anticardiolipine antibodies, lupus anticoagulant and false positive VDRL) in the serum of patients with Systemic Lupus Erythematosus or other diseases of the connective tissue (Secondary Antiphospholipid Syndrome) or in patients without criteria for the diagnosis of any other disease (Primary Antiphospholipid Syndrome).

We present a case report of a pregnant woman with Antiphospholipid Syndrome, recurrent intracardiac mass and pulmonary embolism.

CASE REPORT

A 21 year old pregnant woman was admitted to hospital because of an intracardiac mass visualized on echocardiography. She had a history of two episodes of severe lower limb thrombophlebitis when she was 15 and 18 years old. A routine laboratory examination disclosed thrombocytopenia (95 x 10^3 platelets/L), prolonged partial thromboplastin time (53 sec) that led to her referral to the outpatient clinic of Medicine Department (Centro Hospitalar de Coimbra). Further laboratory tests, in March 1989, revealed positive type I Lupus Inhibitor, negative Antinuclear Antibody, negative VDRL, negative Rheumatoid Factor, normal C3 and CH100 and a low C4 complement fractions. The hypothesis of Antiphospholipid Syndrome was raised and the patient started treatment with acetylsalicylic acid 100 mg daily. She remained assymptomatic and in August 1991 delivered a pre-term (31 weeks) low weight baby (1,020 kg), that died during the neonatal period with Respiratory Distress Syndrome. In August 1992 she became pregnant again, taking acetylsalicylic acid 100 mg and prednisolone 30 mg daily. On routine clinical examination she presented persistent tachycardia (110/min), however assymptomatic. The chest x-ray was normal except for slight increase of cardiothoracic ratio. An echocardiogram was performed, revealing the presence in the right atrium of an ecodense, pedunculated, mobile mass (30 x 10 mm), occupying one third of the atrial chamber, prolapsing into the right ventricle during diastole *(Fig 1)*.

DIFFERENTIAL DIAGNOSIS- cardiac tumor, intracardiac thrombus, vegetation

The patient was admitted in August 1992 to the Medicine Department.

On physical examination she was conscious, and appeared well with no dyspnea or cianosis. Temperature was 36.5 °C. Blood pressure was 100/70 mmHg. Lungs were clear and a grade 2 mid-systolic murmur was heard at the lower left sternal border, unchanged by body

Received for publication: 6 May, 1993

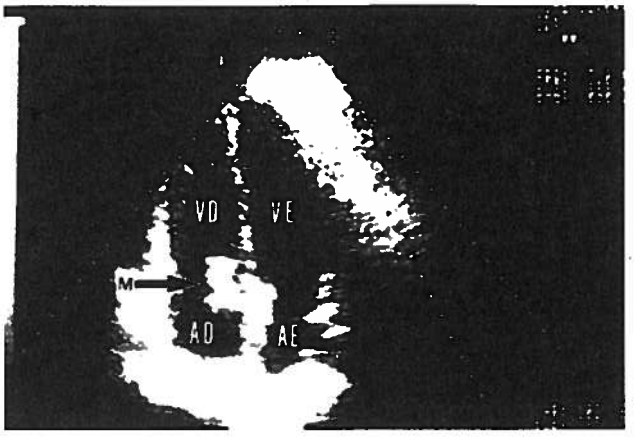

Fig. 1B – mode echocardiogram (4 chamber view): right atrial mass (M) adherent to the atrial septum. AD (right atrium), VD (right ventricle), VE (left ventricle).

position. Abdomen examination was normal. Extremities were normal(Homans sign negative) except for livedo reticularis in the lower limbs. Neurologic examination was negative.

Hematologic laboratory values were :hemoglobin 13.5 g/dL, RBC 4.1 x 10^6/mL, WBC 10.7 x 10^3/mL, platelets 141 x 10^3/L, prothrombin time 16 sec, partial thromboplastin time 34 sec. Liver and renal function tests were normal. The immunological tests ANA, anti ds-DNA, anti-Ro, anti LA, anti SM, VDRL and anticardiolipin antibody were negative and Type I lupus inhibitor was positive. Complement fractions C3, C4 and CH100 were normal. Urinalysis was normal. The electrocardiogram showed sinus tachycardia (110/min).

Besides acetylsalicylic acid 100 mg daily and prednisolon 10 mg daily, anticoagulant therapy was started with continuous intravenous heparin.

This case was discussed in collaboration with the Cardiothoracic Surgery Department (Coimbra University Hospital) and it was decided to maintain the treatment already in course. Cardiac and fetal ultrasound control should be made and cardiac surgery and cesarian should be performed when fetal viability was attained.

EVOLUTION

In September 1992, with adequate anticoagulation levels, the patient suffered an episode of sudden dyspnea, with severe right sided pleuritic chest pain, dry cough, central cyanosis and fever (38.5°C). Blood pressure was 80/40 mmHg and pulse was 130/min. Breath sounds were abolished at the rigth hemithorax base. Arterial blood gases showed PaO2 of 51 mmHg, PaCO2 24.8 mmHg with pH 7.489. Serum aspartate aminotransferase was 63U, alkaline aminotransferase 118U and lactate dehydrogenase 440U. The partial thromboplastin time was 48 sec. The electrocardiogram showed sinus tachycardia (130/min), frontal right axis deviation and negative T waves at inferior and right precordial leads. The chest X-ray revealed cardiomegaly with dilatation of pulmonary artery and enlargement of the lower right border of cardiac silhuette *(Fig 2)*. The cardiac echo-Doppler showed the disapearance of the right atrial mass, dilatation of right cardiac chambers and pulmonary hypertension (estimated pulmonary artery systolic pressure 64 mmHg)*(Fig 3)*.

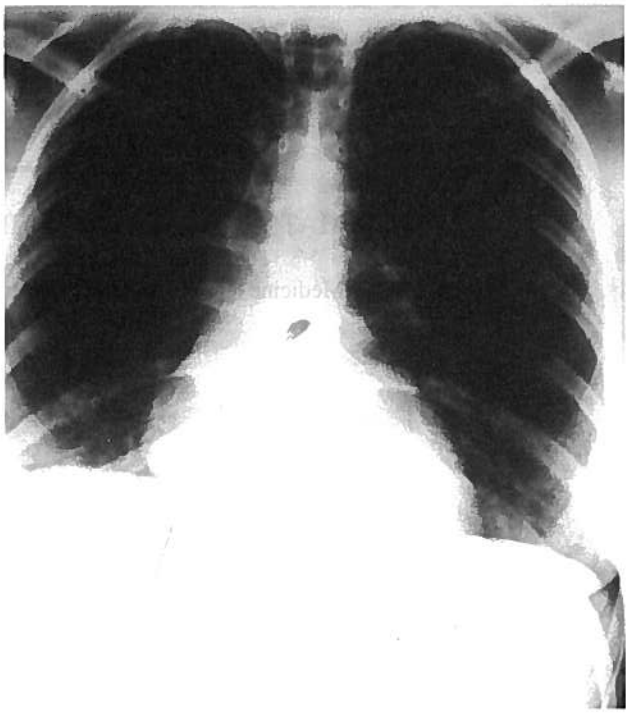

Fig. 2 – Chest X-ray: cardiomegaly, dilatation of pulmonary artery and the right lower border of cardiac silhuette.

CLINICAL DIAGNOSIS: pulmonary embolism due to the detachment of a right atrial mass.

The patient was transferred to the Coronary Care Unit of the Cardiology Department and started oxygen therapy through Venturi mask (FiO2 35%), maintaining intravenous heparin, acetylsalicylic acid, prednisolone and ceftazidime. Clinical status was slightly improved but the patient maintained dispnea for small efforts, dry cough, central cyanosis and hypoxemia (PaO2 70 mmHg) despite oxygen therapy. Obstetric evaluation showed good fetal status. At 30 weeks of pregnancy she started painful uterine contractions and elective cesarian section was performed without complications (female newborn weighing 1,200 Kg, Apgar score 9). The patient stayed in the Intensive Care Unit during the immediate post-operative period, medicated with intravenous heparin and prednisolone. Mechanical ventilation was stopped at the end of the first post-operative day and the patient was transferred to the ward. Ten days after surgery she had right sided pleuritic chest pain, cough, mucopurulent sputum, fever (39°C)and tachypnea. Blood pressure was 120/80 mmHg and a grade 2 mid-systolic murmur was heard at the lower left sternal border. Breath sounds were abolished at the lower two thirds of right hemithorax (Fig 4). The chest X-ray showed a triangle shaped opacity with external base in the middle of the

right lung field. Arterial blood gases (without oxygen) were: PaO2 60 mmHg, PaCO2 30 mmHg with pH 7.470)

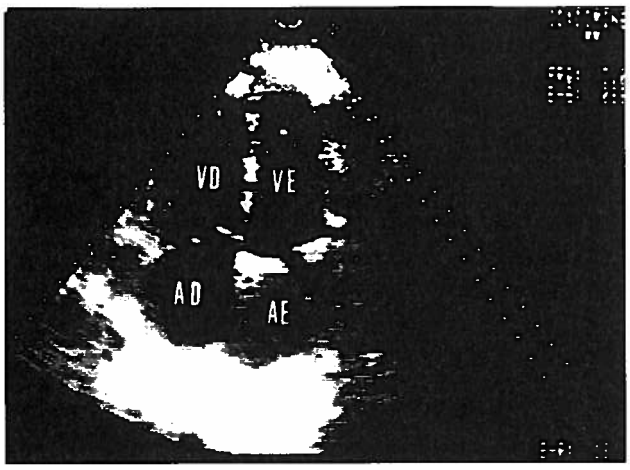

Fig. 3 – B-mode echocardiogram (4 chamber view): disappearance of the right atrial mass and dilatation of right cardiac chambers (see legend in fig 1).

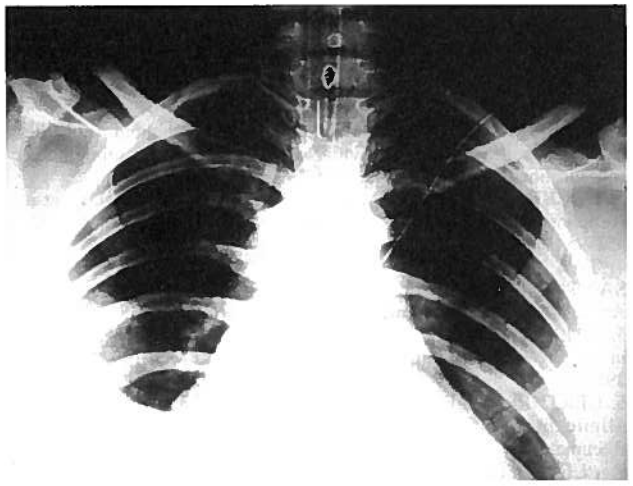

Fig. 4 – Chest X-ray: triangle shaped opacity in the middle of the right lung field

DIFFERENTIAL DIAGNOSIS: Recurrent pulmonary embolism, secondary infection of a prior pulmonary infarction.

The patient was readmitted to the Intensive Care Unit for mechanical ventilation and returned, two days later, to Medicine Department without chest pain, maintaining dry cough and reduction of breath sounds at the right hemithorax despite the clearing of the opacity on the chest X-ray and the improvement of blood gases.

She began oral anticoagulation with warfarin. Functional respiratory tests showed hypoxemia at rest (PO2 75 mmHg), a 30% decrease of total lung capacity and reduction of the alveolo-capilar diffusion of carbon monoxyde. Echocardiogram was normal.

She was discharged on November 1992 complaining of dry cough and dyspnea for medium efforts, taking warfarin 5 mg, and prednisolone 30 mg daily. The echocardiogram performed two months later showed the reappearance of two masses in the right atrium, one of them (15x10 mm) attached to the atrial free wall and the other (10x10 mm)to the atrial septum.

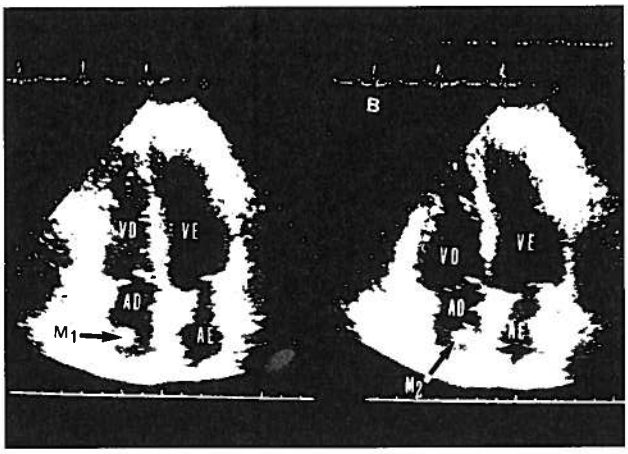

Fig. 5 - B-mode echocardiogram:(4 chambers): two right intra-atrial masses (M1 and M2)

Readmitted for anticoagulation with continuous intravenous heparin plus acetyilsalicilic acid and prednisolone. The case was once again presented to the Cardiothoracic Surgery Department of the Coimbra University Hospital and the patient was accepted for surgery.

Cardiac surgery was performed in February 1993 and two masses were excised from the right atrium near the openings of coronary sinus and inferior vena cava. The pathological examination showed them to be thrombi.

The post-operative period was uneventful and the patient was discharged taking warfarin 5 mg daily. Until now, the follow-up echocardiographic examinations showed no recurrence of intracardiac masses and the patient remains assymptomatic.

DISCUSSION

The presence of anthiphospholipid antibodies in the blood (lupus anticoagulant, anticardiolipin antibody false positive VDRL) interacts with the hemostatic system, altering the balance between the coagulation and the fibrinolytic pathways contributing to the formation of intravascular thrombi [2].

The clinical interest of antiphospholipid syndrome, apart from its linkage to adverse pregnancy events (spontaneous recurring abortions, stillbirths, intrauterine fetal growth retardation, pre-term births)[3], increased in recent years due to its protean clinical manifestations. The formation of thrombi in the multiple body systems: neurological (stroke, transient ischemic attacks, dementia, transverse mielopathy, migraine, chorea)[4], cardiac (myocardial infarction, heart failure, valvular lesions, early occlusion of coronary by-pass)[5], respiratory (pulmonary embolism and hypertension), renal (arterial and vein thrombosis), endocrine (adrenal insufficiency) and osteo-articular (aseptic bone necrosis) explain these multiple clinical consequences.

The recognition of the presence of an intracardiac thrombus related to the Antiphospholipid Syndrome [6,7] is of great interest due to the serious complications that may arise from its detachment and embolization (as exemplified by this case report) and to the fact that they can mimic the clinical aspects of cardiac tumors, (namely myxoma) [8]. The constitutional symptoms (fever, myalgia, arthralgia, fatigue, weight loss), elevated erythtrocyte sedimentation rate, anaemia, leucocytosis, thrombocytopenia and hypocomplementaemia are frequent in both clinical conditions [8].

The pulmonary or systemic embolic phenomena related to cardiac tumors may be secondary to the fragmentation of the tumor itself or to the deposition and detachment of thrombi on its surface.

The presence of intravascular thrombi in Primary Antiphospholipid Syndrome has been frequently reported in recent medical literature [9], however, intracardiac mural thrombi in this clinical situation has rarely been described [5,6]. In this case report, the presence of an intracardiac mass implied the differential diagnosis between a primary cardiac tumor, an intracardiac thrombus or endocarditis vegetation. Non invasive laboratory tests did not provide reliable clues to a definite pre-operative diagnosis. The echocardiographic morphological characteristics of the mass, (mobile, peduncled, with smooth surfaces, attached to the atrial septum) may lead to the conclusion that it was a cardiac myxoma, which is the most frequent cardiac tumor. Cardiac myxomas are more prevalent in younger women and located in the atrium in most cases. However, the presence of an underlying disease that predisposes to the formation of intravascular thrombi and the early recurrence of two masses should argue in favor of the hypothesis of a thrombus. This was confirmed by the pathological examination of the two masses excised. Computerized Tomography scan and Magnetic Resonance Imaging could provide high resolution images of intracardiac masses [10] due to their ability to distinguish different adjacent structures, but couldn't be used because the patient was pregnant.

The adverse pregnancy events are very frequent in women with Antiphospholipid Syndrome and the accomplishment of a full term pregnancy with the birth of a normal newborn is very rare.

Management of patients with Antiphospholipid Syndrome has not been standardized yet, but there is a general consensus that the patients who do not have thrombocytopenia, thromboembolic phenomena or pregnancy should not be submitted to any treatment or should only take low doses of acetylsalicylic acid. In the presence of any one of the conditions above, various treatment regimens have been used (corticosteroids, dipyridamole, heparin, warfarin, cytotoxic drugs, plasmapheresis, high dose human immunoglobulin) according to the severity and individuality of the case.

In this case, due to the contraindication for the use of oral anticoagulants in a pregnant woman, we used continuos intravenous heparin during pregnancy and warfarin after, in association with acetylsalicylic acid and prednisolone.

At present, the management of Antiphospholipid Syndrome is based on the experience of isolated cases and standardized treatment should be achieved when multicentre randomized trials are available.

ACKNOWLEDGMENTS

The authors thank Manuel Antunes (Cardiac Surgeon) and Matos Beja (Pathologist) for their valuable collaboration.

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