

Imaging of a Rare Case of Gluteal Myxoid Liposarcoma

F. Akhatar^{1*}, MA. Nouri¹, Y. Bouktib¹, A. Elhajjami¹, B. Boutakioute¹, M. Ouali Idrissi¹, N. Cherif Idrissi El Ganouni¹

¹Department of Radiology, Arrazi Hospital, Mohamed VI University Hospital, Faculty of Medicine and Pharmacy, Cadi Ayyad University, Marrakech, Morocco

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*Corresponding author: F. Akhatar

Department of Radiology, Arrazi Hospital, Mohamed VI University Hospital, Faculty of Medicine and Pharmacy, Cadi Ayyad University, Marrakech, Morocco

Abstract

Case Report

Myxoid liposarcoma is a distinct subtype of soft tissue sarcoma, characterized by a myxoid stroma, delicate capillary networks, and scattered lipoblasts. It has demonstrated a notable pattern of spread to soft tissue and bony structures. MRI examination is a highly reliable method in the diagnosis of these tumors. We report the case of a 61-year-old patient with a history of intra-abdominal liposarcoma of the right iliac fossa, surgically treated three years prior, who presented with a slowly enlarging, painless right gluteal mass evolving over one year, in whom magnetic resonance imaging played a pivotal role in the detection and characterization of the lesion, highlighting some imaging features of myxoid liposarcoma. Through this rare case, we review the imaging findings of myxoid liposarcoma and emphasize the value of MRI in the evaluation of soft tissue sarcomas.

Keywords: Myxoid, Liposarcoma, Mri, Gluteal, Imaging.

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INTRODUCTION

Recurrent thrombosis of mechanical heart valves during pregnancy presents a significant clinical challenge due to the high risk of maternal and foetal complications. Pregnancy induces a hypercoagulable state, increasing the risk of thromboembolic events in patients with mechanical heart valves (Brenner, 2004; van Hagen *et al.*, 2015). Anticoagulation is crucial, but managing it during pregnancy is complex because of potential risks to the foetus (Steinberg *et al.*, 2017).

Prosthetic heart valve thrombosis (PVT) is uncommon, occurring in 0.1% to 5.7% of patients annually. However, during pregnancy, the procoagulant changes elevate the PVT risk to as high as 10%. Risk factors for valve thrombosis during pregnancy are the same as in nonpregnant women, which are a prosthetic valve in the mitral position and subtherapeutic anticoagulation (Casais & Rolandi, 2013).

This case report details a pregnant patient with a mechanical heart valve who experienced recurrent thrombosis and had a history of recurrent miscarriages. During our investigations, we suspected antiphospholipid syndrome (APS) as an underlying cause. We also review the current literature on managing

similar cases to provide insights into optimal treatment strategies.

CASE REPORT

A 34-year-old woman, gravida 4 para 0, at 33 weeks of gestation, presented to the emergency department with progressive exertional dyspnoea. She had a history of mechanical mitral valve replacement nine years earlier due to rheumatic heart disease, along with three prior miscarriages and an episode of acute limb ischemia two years earlier related to inadequate anticoagulation, which was managed with surgical embolectomy. One week prior to admission, her cardiologist switched her anticoagulation therapy from Acenocoumarol (a vitamin K antagonist) to weight-adjusted low molecular weight heparin (Enoxaparin, twice daily) in anticipation of delivery.

On examination, she was tachypnoeic at 30 breaths per minute and in atrial fibrillation with a heart rate of 140 bpm. Blood pressure was stable, and pulmonary auscultation revealed bilateral crackles. She was admitted to the maternal intensive care unit, placed in a semi-sitting position, and started on oxygen therapy. Transthoracic echocardiography revealed a high trans-mitral gradient (maximum 27.89 mmHg, mean 16.44 mmHg), increased peak velocity (2.64 m/s), good left

ventricular function, and a dilated left atrium (Figure 1). A transoesophageal echocardiogram confirmed the diagnosis of mechanical mitral valve thrombosis.



Figure 1: Transthoracic Echocardiography on Admission

The echocardiographic image displays a continuous transmitral Doppler flow, indicating an elevated trans-mitral gradient with a peak value of 27.89 mmHg and a mean value of 16.44 mmHg.

A multidisciplinary discussion involving cardiology, cardiac surgery, anesthesiology, and obstetrics led to the decision for urgent surgical valve replacement. Unfractionated heparin (UFH) was initiated, targeting an activated partial thromboplastin time (aPTT) 2–3 times the upper limit of normal. Informed consent was obtained from the patient, with clear explanation of maternal and fetal risks.

Preoperative fetal heart monitoring showed positive cardiac activity.

Under general anesthesia, the patient underwent replacement of the thrombosed valve using cardiopulmonary bypass (CPB) (Figure 2). Anesthetic management was adapted for pregnancy, including bispectral index (BIS) monitoring to minimize anesthetic exposure, maintenance of normothermia, and mean arterial pressure >75 mmHg. CPB flow was maintained between 3 and 3.5 L/min. CPB time was two hours. Postoperative transthoracic echocardiography showed a mean mitral gradient of 7 mmHg with preserved ventricular function. The patient was extubated and resumed on UFH 12 hours later.

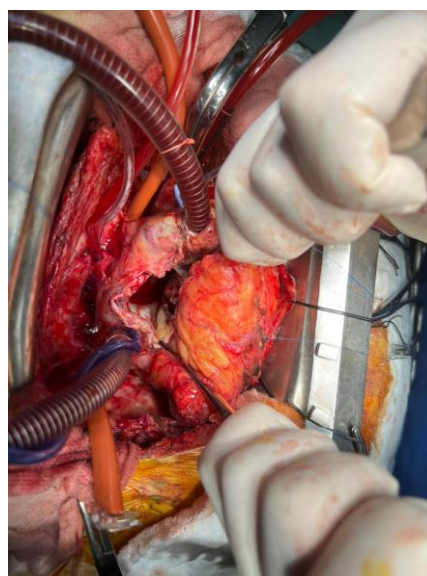


Figure 2: Intraoperative Image During Valve Replacement Surgery

The image captures the first surgical procedure involving the replacement of a thrombosed valve with a new mechanical prosthesis, performed under cardiopulmonary bypass.

On postoperative day 1, fetal ultrasound revealed absence of cardiac activity. A decision was made to perform an emergency cesarean section under general anesthesia after a 4-hour UFH suspension to permit timely re-initiation postoperatively. The surgery was uneventful.

Four days after surgery, the patient developed respiratory deterioration with tachypnoea and desaturation. Lung ultrasound revealed signs of pulmonary congestion. Echocardiography showed a new trans-mitral gradient of 20.87 mmHg, and transoesophageal imaging confirmed recurrent thrombosis of the newly implanted mechanical valve (Figure 3). Thrombolysis with continuous infusion of alteplase was administered (1 mg/h for 25 hours), but no clinical or echocardiographic improvement was observed.

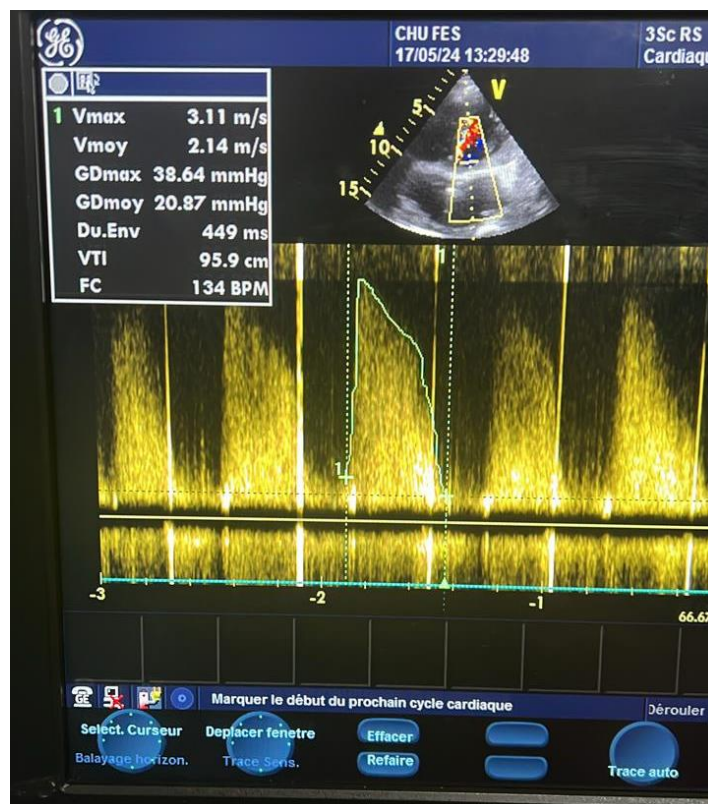


Figure 3: Transthoracic Echocardiography on Postoperative Day 4

The echocardiographic image reveals continuous transmitral Doppler flow with an elevated trans-mitral gradient, showing a peak value of 38.64 mmHg and a mean value of 20.87 mmHg.

A second surgery was performed to replace the mechanical valve with a bioprosthesis. The CPB time was three hours, and postoperative echocardiography showed a mean gradient of 7 mmHg without paravalvular leak. The patient was extubated, resumed on UFH at 12 hours, and transitioned to vitamin K antagonists within 24 hours.

An etiological assessment revealed positive lupus anticoagulant antibodies, indicating antiphospholipid syndrome (APS). The patient was transferred to the internal medicine department for further evaluation and long-term management.

DISCUSSION

This case highlights the complex management of recurrent mechanical heart valve thrombosis during pregnancy, a rare but life-threatening condition. A 34-year-old woman at 33 weeks of gestation was diagnosed with mitral valve thrombosis shortly after her anticoagulation therapy was switched from Acenocoumarol to Enoxaparin. Despite surgical replacement of the valve and initiation of unfractionated heparin, the patient experienced a second episode of valve thrombosis. Fibrinolytic therapy failed, necessitating a second surgical replacement with a bioprosthesis. Further investigations revealed positive lupus anticoagulant antibodies, suggesting antiphospholipid syndrome (APS) as a contributing factor.

Pregnancy significantly heightens the risk of thromboembolic events in patients with mechanical heart valves due to the hypercoagulable state it induces (van Hagen *et al.*, 2015). Hormonal changes, increased blood volume, and modified hemodynamics collectively contribute to a higher propensity for clot formation (Brenner, 2004). Mechanical valves themselves are inherently thrombogenic, and pregnancy exacerbates this risk, making vigilant anticoagulation management paramount. However, anticoagulation during pregnancy presents a complex challenge because of the potential teratogenic effects of certain anticoagulants and the increased bleeding risk for both the mother and foetus (Steinberg *et al.*, 2017).

The anticoagulation therapy in pregnant patients is addressed in the Guidelines of the European Society of Cardiology (ESC) 2021 and Guidelines of the American Heart Association/American College of Cardiology (AHA/ACC) 2020 (Otto *et al.*, 2021; Vahanian *et al.*, 2022). The guidelines recommend that women should be switched from vitamin K antagonists (VKA) to low molecular weight heparin (LMWH) at least one week before their planned delivery to minimize bleeding risk. However, maintaining therapeutic levels of LMWH can be challenging, and inadequate anticoagulation increases the risk of valve thrombosis (Xu *et al.*, 2016). In our case, the patient's history of recurrent miscarriages and acute limb ischemia underscores the critical importance of rigorous monitoring and precise adjustment of anticoagulation therapy during pregnancy. Targeting anti-Xa activity levels between 0.8 and 1.2 U/mL (measured 4 to 6 hours post-dose) is essential for effectively managing thrombotic risks and ensuring optimal maternal and fetal outcomes (Otto *et al.*, 2021).

The management of valve thrombosis during pregnancy necessitates a multidisciplinary approach involving cardiologists, cardiac surgeons, anesthesiologists and obstetricians. The primary options for managing thrombosis include surgical intervention or fibrinolysis. Surgical valve replacement, although associated with significant maternal and foetal risks, can be life-saving and may be preferred in cases of severe valve obstruction, as seen in this patient (Otto *et al.*, 2021; Vahanian *et al.*, 2022).

The use of CPB during surgery is particularly risky due to potential hemodynamic instability and adverse effects on the foetus. However, the risk can be minimized with continuous monitoring of the mother, foetus, and uterine tone. Sustained uterine contractions during CPB, particularly during the cooling and rewarming phases, are a leading cause of foetal death. To prevent early labour, tocolytic therapy can be discussed, and post-operative hormonal administration may be considered (Yuan, 2014). Minimizing CPB time is crucial. Hypothermia during CPB can adversely affect placental perfusion through acid-base imbalances,

alterations in the coagulation pathway, and sustained uterine contractions. Maintaining normothermia or mild hypothermia ($T > 35^{\circ}\text{C}$) has been shown to improve foetal outcomes. Ensuring adequate pump flow ($> 2.5 \text{ L/min/m}^2$), mean arterial pressure ($> 70 \text{ mmHg}$), and haematocrit levels $> 28\%$ during CPB is recommended for optimal utero-placental perfusion. Despite these precautions, the reported foeto-neonatal mortality remains high, around 20% (Jafferani *et al.*, 2011; Yuan, 2014). In our case, unfortunately, the foetus did not survive.

Fibrinolysis with agents such as alteplase offers a non-surgical alternative, potentially avoiding the immediate risks associated with surgery (Özkan *et al.*, 2015). However, the efficacy of fibrinolysis can be variable, and the risk of bleeding complications remains a significant concern (Karthikeyan *et al.*, 2013). In this case, despite initial fibrinolytic therapy, the persistent high trans-mitral gradient necessitated surgical intervention, ultimately leading to the replacement of the mechanical valve with a bioprosthetic valve. This highlights the complexities and limitations of fibrinolysis in managing valve thrombosis during pregnancy.

Recurrent thrombosis of a mechanical heart valve during pregnancy, while rare, poses a significant clinical challenge. The incidence of PVT in the general population ranges from 0.1% to 5.7% annually, but pregnancy elevates this risk to approximately 10% due to the procoagulant changes inherent in pregnancy (Casais & Rolandi, 2013). The literature on managing recurrent thrombosis specifically during pregnancy is limited, with few case reports addressing this issue (Jimenez *et al.*, 1988; Kalcik *et al.*, 2014; Lisowska *et al.*, 2004; Sathananthan *et al.*, 2019; Vrkcova *et al.*, 2016). Treatment typically involves fibrinolysis or surgical intervention, combined with optimized anticoagulation strategies to prevent future events.

The etiological factors contributing to recurrent thrombosis can include mechanical valve dysfunction, subtherapeutic anticoagulation, and underlying hypercoagulable states such as APS and systemic lupus erythematosus (SLE) (Gencbay *et al.*, 1998). APS, characterized by recurrent thrombotic events and pregnancy morbidity, was suspected in this patient due to her history of recurrent miscarriages and acute limb ischemia (Ruiz-Irastorza *et al.*, 2010). While initial tests for anticardiolipin antibodies were negative, further investigation revealed positive lupus anticoagulant antibodies, suggesting an underlying autoimmune pathology contributing to the thrombotic events.

CONCLUSION

Managing recurrent thrombosis of a mechanical heart valve during pregnancy presents significant clinical challenges, requiring a delicate balance between

anticoagulation, fibrinolysis, and surgical intervention. This case illustrates that even with appropriate anticoagulation, patients may suffer severe complications, especially when underlying prothrombotic conditions such as antiphospholipid syndrome are present. The recurrence of thrombosis and need for multiple surgeries highlight the limitations of current protocols and the importance of early recognition and individualized treatment. A thorough etiological workup is essential to guide management decisions, particularly in high-risk patients. Multidisciplinary coordination among cardiologists, cardiac surgeons, anesthesiologists, obstetricians, and internists is critical to optimize outcomes for both mother and fetus. While both surgical and medical approaches remain valid, each carries significant maternal and fetal risks, reinforcing the need for personalized, risk-adapted strategies. Future research should focus on refining anticoagulation protocols and exploring targeted therapies for patients with underlying autoimmune or hypercoagulable disorders.

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Conflict of interest

The authors declare that they have no conflict of interest.

Ethics, Informed Consent, and Data Confidentiality Statement

We confirm that oral and written informed consent was obtained from the patient and her relatives for the publication of this clinical case. All clinical data were handled in compliance with confidentiality standards, ensuring that the patient's privacy and personal information remain protected.

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