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CASE REPORT

Rupture of Renal Angiomyolipoma Post Fibrinolysis

A rare complication of fibrinolytic therapy

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ABSTRACT: Angiomyolipoma is a common benign solid tumour that accounts for up to 3% of all renal tumours; most of the cases are sporadic. However, it can be part of other diseases. Angiomyolipomas are usually found incidentally through unrelated clinically indicated images but also, they can be diagnosed after complications have occurred. We report the case of retroperitoneal haemorrhage following the rupture of renal angiomyolipoma post-fibrinolysis, we are highlighting such a rare condition, the management options and the follow-up plan. The management of angiomyolipomas ranges from conservative treatment to surgical intervention depending on the patient's condition and the tumour's radiological features. Following-up on patients with angiomyolipomas depends on the symptoms and the tumour size. Till date, there is no reported cases of renal angiomyolipoma rupture post-fibrinolysis therapy as a treatment for myocardial infarction in non-percutaneous intervention capable facility.

Keywords: Angiomyolipoma; Myocardial Infarction; Fibrinolysis, Retroperitoneal; Hemorrhage; Artery Embolization; Percutaneous Intervention.

Angiomyolipoma is a benign tumour that accounts for 0.3% to 3% of all renal tumours.[1] However, 80% of the angiomyolipomas are sporadic, and the rest are associated with tuberculosis complex or lymphangioleiomyomatosis (LAM).[2] Most of the angiomyolipomas occur among females and rarely develop before puberty.[3]

Angiomyolipomas are solid tumours composed of dysmorphic blood vessels, adipose tissues and smooth muscles, and which are strongly expressing B progesterone, oestrogen, and androgen receptors which suggest a hormonal influence for their growth.[3,4] The diagnosis of angiomyolipomas is incidental in more than 80% of the cases through the cross-sectional imaging, as the finding of macroscopic fat within a lesion is the hallmark of all modalities.[2,5] However, the rest of the cases are diagnosed after a complication has happened such as spontaneous retroperitoneal haemorrhage, where the patients will present with the following signs and symptoms; flank pain, palpable mass, haematuria, anaemia, renal failure or urinary tract infection.[2] Following-up patients with angiomyolipomas depends on multiple factors, such as the tumour size and if the patient is symptomatic; as for the tumours which are more than 4 cm in size and they are asymptomatic, then a six-month radiological follow-up is recommended, but if they are symptomatic then surgical or endovascular interventions are recommended.[5] In this paper, we are reporting a case of a retroperitoneal haemorrhage caused by ruptured renal angiomyolipoma post-fibrinolysis, which was treated transcutaneously with arterial embolisation.

Case Report

A 66-year-old male who is an active smoker and with a past medical history of type two diabetes mellitus and hypertension, presented to the regional hospital with a typical chest pain, so an electrocardiogram (ECG) was done which showed an anterior wall myocardial infarction (MI), as the previously mentioned regional hospital is a non-percutaneous intervention (PCI) capable facility, therefore, he was admitted and managed with fibrinolysis 10 IU at 0 and at 30 minutes, which resulted in more than 50% resolution of the initial ST-segment elevation, nevertheless; the patient continued to have intermittent chest pain, therefore, a rescue PCI was arranged and he was transferred to our cardiac centre which is PCI capable facility.

On arrival at the cardiac centre, he was conscious, well-oriented and haemodynamically stable, however, he was constantly complaining of chest pain and right flank pain. Clinically he looked dehydrated evident by the bedside transthoracic echocardiography (TTE) which showed under filled and kissing ventricles. Shortly after the initial assessment and resuscitation, the patient developed a vasovagal arrest, for which he received a brief cycle of cardiopulmonary resuscitation (CPR) and then initiated noradrenaline.

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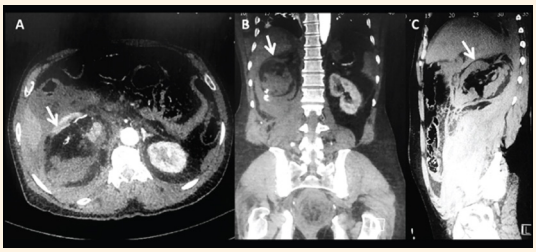

Figure 1: Abdominal computed tomography scan with contrast in (**A**) axial, (**B**) coronal and (**C**) sagittal views showing actively bleeding right renal angiomyolipoma and haematoma.

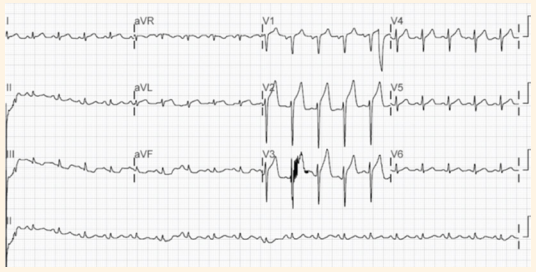

Figure 2: 12-lead electrocardiogram, showing ST-segment elevation in lead V1, V2 and V3.

Repeated ECG showed a new ST-segment elevation in the anterior leads, so the catheterisation lab was activated for emergency PCI, and he was shifted to the lab. Before starting, the team received a call from the haematology lab that the patient's haemoglobin was 5.3 g/dL (previously 13.7 g/dL), with severe metabolic acidosis of pH 7.14 and bicarbonate of 7 and high lactate of 5.6 mmol/L. Given the fact that the patient was complaining of right flank pain and with the evidence of acute blood loss, acute intra-abdominal bleeding was suspected; therefore, the patient was electively intubated and kept on inotropes, and a massive blood transfusion protocol was activated. PCI was abandoned and an urgent CT abdomen showed actively bleeding large right renal angiomyolipoma along with large perinephric and retroperitoneal hematoma and 12-lead ECG showed anterior wall STEMI [Figures 1 and 2].

Based on CT findings, the patient was shifted immediately to the interventional radiology suite, for angioembolisation of the bleeding angiomyolipoma. Access was made through right common femoral artery and a 5 Fr vascular sheath was inserted over a wire. Catheterisation of right renal artery was made with 5Fr C2 catheter (Merit Medical). Renal angiogram confirmed presence of actively bleeding small segmental arterial branch in the mid pole. The bleeding branch was selectively catheterised with a Progreat™ microcatheter (Terumo Corporation, Tokyo, Japan) size 2.8 Fr inserted coaxially through the main

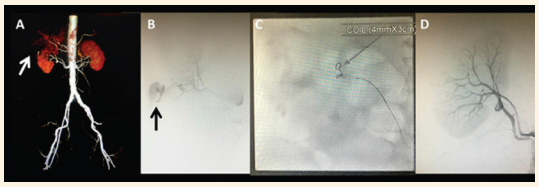

Figure 3: A: Renal angiogram (bleeding from the right renal angiomyolipoma). **B:** Renal arteriography showing extravasation in the right kidney (by an interventional radiologist). **C:** Coil and foam application by an interventional radiologist. **D:** Post-procedure (no extravasation).

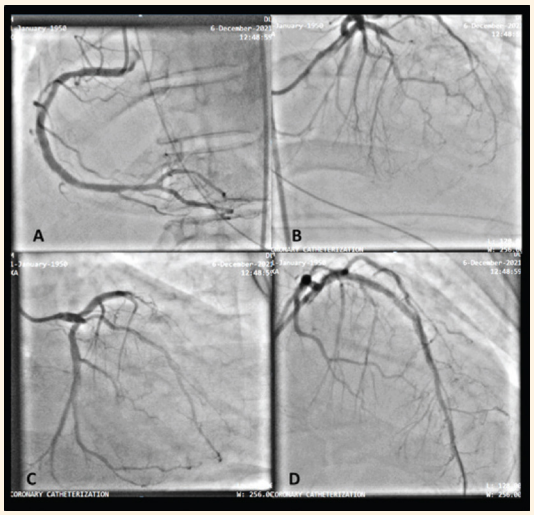

Figure 4: A: Dominant right coronary artery with non-obstructive diffuse atheroma. **B:** Occluded left anterior descending artery (LAD) at the proximal segment. **C:** Showing minor disease in the circumflex artery. **D:** percutaneous intervention of the LAD.

C2 catheter and coil embolisation was performed with a small size 4 mm. Final control post-embolisation angiogram showed no active bleeding [Figure 3].

Post-arterial embolisation, the patient was shifted to the coronary care unit (CCU) for supportive care. Repeated ECG showed partial ST-segment elevation resolution and therefore PCI was further delayed. His stay in CCU was complicated by acute kidney injury (AKI), so continuous veno-venous haemofiltration dialysis (CVVHD) was started, and other supportive measures were continued.

TTE showed low normal systolic function with an estimated left ventricular ejection fraction (LVEF) of 40–45%, regional wall motion abnormalities (RWMA) could not be assessed due to tachycardia at that time. Repeated renal CT angiographies showed no active extravasation. After spending a few days in CCU, the patient's condition improved gradually and eventually weaned down from inotropes and dialysis and then extubated.

As the patient stabilised, he was stepped down from CCU to the intermediate care unit. While on the floor, the patient unfortunately had ventricular fibrillation (VF), so CPR was started and the patient received one DC shock of 200 J. Once return of spontaneous circulation (ROSC) was achieved, the patient was intubated and shifted back to the CCU.

The following day, the patient was taken to the catheterisation lab. His coronary angiogram (CAG) showed minor disease in the right coronary artery, the circumflex artery and the left anterior descending artery (LAD) was occluded at the mid-segment. It was decided to proceed to PCI the LAD. This was a challenging and complex procedure as the LAD was heavily calcified artery and multiple wires were used till it was successful to cross the lesion. Lesion preparation was done with high-pressure non-compliant balloons and cutting balloons. Then, proximal to mid LAD was stented with two drug-eluting stents in an overlap fashion with good results [Figure 4]. The patient was kept on aspirin for lifelong and plavix for 1 year.

Post-PCI, the patient was kept in the CCU for a few days. He was then extubated and underwent aggressive rehabilitation with regular physiotherapy and a dietary programme. Before discharge, he was referred to the electrophysiology (EP) team for assessment of secondary prevention implantable cardiac defibrillator (ICD). EP team suggested that there was a secondary cause for the episode of VF, which was ischaemia, and given that the patient underwent PCI, ICD was not warranted. Subsequently, the patient was discharged home on aspirin for lifelong and clopidogrel for 1 year. He was in a stable condition accompanied by his family.

Two weeks later, we saw the patient in the cardiology clinic. He continued to do well with a stable Hb level and angina-free.

Consent for the publication of this case report was obtained from the patient.

Discussion

This case was challenging as the patient had recent acute MI post fibrinolysis with active intra-abdominal bleeding. Although fibrinolysis was successful initially, the patient had re-ST-segment elevation after a significant haemoglobin drop, which was most likely supply-demand ischaemia. This was evident when the ST-segment elevation subsided after resuscitation and blood transfusion.

The other challenging decision was the timing of starting the dual antiplatelets (DAP) for the management of acute coronary syndrome (ACS). So given the high risk of bleeding from the renal angiomyolipoma site compared to the benefit of DAP in the management of ACS, we decided to hold all the DAP and anticoagulation medications, and we kept the patient on mechanical deep vein thrombosis (DVT) prophylaxis instead of antithrombosis until the patient was clinically stable with no evidence of extravasation in the repeated abdominal CT scan. Then we started on aspirin initially and subsequently proceeded to clopidogrel with continuous monitoring for any signs of bleeding or haemodynamic instability.

Another challenge that we faced was the timing of the PCI, as it became more urgent when the patient developed ventricular tachycardia (VT) along with a new STEMI, although there was no evidence of re-bleeding that can explain it. So, we only proceeded with PCI after the patient was haemodynamically stable, and PCI to the LAD was done successfully.

The most important complication of angiomyolipoma is rupture and retroperitoneal haemorrhage, and it is not only influenced by the tumour size, but also by other risk factors such as aneurysm formation, pregnancy, coagulopathy, trauma, hormonal level and the comorbidity with tuberous sclerosis complex (TSC) and lymphangioleiomyomatosis (LAM).[6] In the presented case, the cause of the intra-abdominal bleeding was the coagulopathy induced by the fibrinolysis as a management of the ACS.

Embolisation is considered the first-line therapy for bleeding angiomyolipoma and is also used as a primary prevention measurement, the advantages of this procedure include: low complication rate, preserved renal function, less trauma and satisfactory short-term outcome; but it is not devoid of limitations, as embolisation has a higher rate of recurrence of the haemorrhage despite the shrinkage of the tumour compared to the surgical option. Therefore, the nephron sparing surgery (NSS) is now considered the optimal operation in clinical practice, as it is associated with acceptable complications, preservation of renal function, and low recurrence rate. Conservative management can be applied in cases where the tumour size is less than 4 cm, or in larger but stable cases, or in cases of limited bleeding that responds to the fluid resuscitation.[6] In our case, the tumour size was large (i.e. >4 cm), so there was a controversy between embolisation by the interventional radiologist and NSS, but considering the patient's critical condition and multiple comorbidities, the embolisation was the optimal choice compared to the NSS.

A yearly follow-up for 3 years with a CT scan is advised after embolisation. If the size of the tumour has reduced significantly after 3 years and there is no evidence of regrowth, then there is no need for

further follow-up. In case a minor reduction is only observed, then a continuation of follow-up is advised, however, if a significant regrowth is demonstrated, further imaging may be required.[7] Therefore, we are planning to follow-up this patient yearly for 3 years with abdominal CT scan, to monitor the tumour size and to decide if any intervention is needed accordingly.

ICD implantation in this case as a secondary prevention was inapplicable, as the cause of his cardiac arrest was reversible, and he underwent PCI successfully.[8]

Conclusion

Renal angiomyolipoma is a benign tumour that is mostly found sporadically through unrelated clinically indicated imaging, or after complications have occurred like retroperitoneal haemorrhage, in which the risk increases after treatment with systemic fibrinolysis for any indications. Hence, close monitoring for any signs of bleeding including changes in mental status, or vital signs raises the suspension of acute bleeding, which warrant immediate action depending on the case. Management of renal angiomyolipoma varies from conservative treatment, embolisation of the bleeder, to surgical extraction, and the choice depends on the patient's condition and the tumour's characteristics, and the recommended follow-up is by imaging modalities yearly for 3 years to decide if any intervention is needed.

AUTHORS' CONTRIBUTION

MH and AJ managed the case. SK collected the data and drafted the manuscript. MH and AJ revised and edited the manuscript. All authors approved the final version of the manuscript.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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