Angioembolisation for Bilateral Angiomyolipoma in TSC patient

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ABSTRACT

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Introduction: Bilateral Angiomyolipoma is a rare entity, usually associated with tuberous sclerosis. The condition is seen more in females and the tumours show progressive increase in size over a period of time bringing with it considerable risk of complications like haemorrhage and morbidity in the form of pain, haematuria, palpable mass, shock and hypotension.

Objective: To report a case of multiple large bilateral angiomyolipoma in a case of Tuberous sclerosis treated successfully with transarterial embolization and to review the literature concerning treatment of AML.

Case report: A 23 year old female patient, on treatment for epilepsy and a known case of tuberous sclerosis with family history too presented with abdominal pain and shock in a peripheral hospital. She was resuscitated and evaluated. CT Scan Abdomen revealed bilateral multifocal AMLs largest over 4 cm and with evidence of pseudo aneurysm with intralesional bleeding. After a preliminary angiogram confirming the intralesional bleed transarterial embolization was done with good results. A mild post embolization syndrome was managed conservatively. Imaging was repeated after 3 months.

Conclusion: Transarterial embolization is a safe option in the management of selected cases of bilateral multifocal AMLs seen in hereditary syndromes like tuberous sclerosis.

Keywords: Tuberous sclerosis, Angiomyolipoma, Intralesional bleeding, Transarterial embolization

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INTRODUCTION

Angiomyolipoma is a tumour composed of varying proportions of blood vessels, smooth muscle cells and adipose tissue. Pathologically it is considered a choriostoma, a disordered arrangement of mature tissue appearing at a site where that tissue does not normally reside. The frequency of angiomyolipoma of kidney varies from 0.7 to 2% depending on whether cases were incidental detection or associated with tuberous sclerosis. Patients are predominantly female. The average age at diagnosis is 41 years. Angiomyolipoma have been observed in patients with several hereditary disorders including Von Recklinghausen disease, Von Hippel Lindau syndrome and autosomal dominant polycystic kidney disease. The association is particularly strong with tuberous sclerosis (mental retardation, epilepsy, cutaneous hamartoma, depigmented spots and subungal fibromas of finger. Angiomyolipoma occurs in 80% of individuals with tuberous sclerosis and in most cases it is bilateral.[1-3]

Tuberous sclerosis complex (TSC) a rare autosomal dominant disorder with variable penetrance, affects approximately 1 in 10,000 people.[4] TSC is caused by mutations of TSC1 or TSC2 genes. Products of these genes, hamartin and tuberin, create a complex that inhibits mTOR, a key protein engaged in regulation of the cell cycle. Mutation of TSC genes lead to activation of mTOR resulting in uncontrolled proliferation, differenciation and migration of cells. TSC is a multisystem disease characterised by the presence of benign tumours in many tissues, mostly localised in the skin, brain, kidney and lungs.[4,5,6,7] Renal angiomyolipomas (AMLs) are benign but progressive tumours consisting of smooth muscle, fat and vascular elements, that are commonly associated with TSC. The size of the tumour varies and they may be classified based on size as small (<4 cm), medium (4-8 cm) and large (>8 cm). Generally AMLs below 4 cm are asymptomatic. In symptomatic patients the common symptoms are pain retroperitoneal haemorrhage, haematuria, hypovolemic shock, hypertension, palpable mass, anaemia, acute pyelonephritis and fever. AMLs over 8 cm are symptomatic, cause significant morbidity, require treatment, prior to development of symptoms and potential complications, the large ones are more common in females and

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their rapid growth in pregnancy suggests a hormone influence.

The management options include observation, embolization and surgical resection. Recommendations for treatment are based on the size of the lesions and the symptoms observed.

CASE REPORT

A 23 year old female patient, a known case of tuberous sclerosis on treatment for epilepsy since childhood developed left loin pain and presented with shock in a peripheral hospital. She was anaemic on examination. She had classic features of Bournevilles disease like adenoma sebaceum (Fig 1), convulsive disorder and mental deficiency. She had shagreen patches in the low back (Fig 2). CT scan abdomen revealed multiple bilateral AML the largest over 4 cm with evidence of focal pseudo aneurysm formation within the left renal artery producing intralesional haemorrhage (Fig 3). She was resuscitated and considering all evidence the decision was taken to try intra arterial embolization.

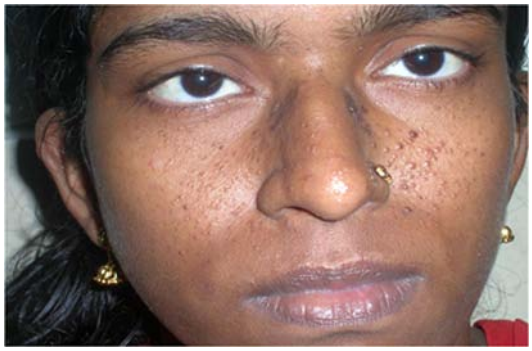

Figure 1. **Showing Adenoma Sebaceum**

A preliminary angiogram done through the right common femoral artery with 7F sheath using 5F pig tail and 935 terumo exchange wire showed evidence of intralesional bleeding which was embolised (refer

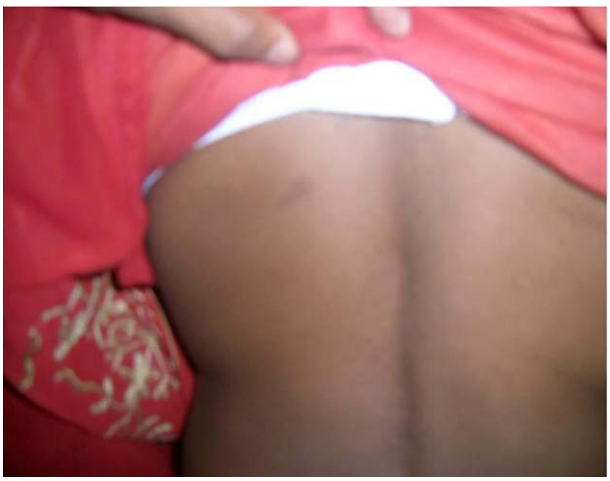

Figure 2. **Showing Shagreen patch**

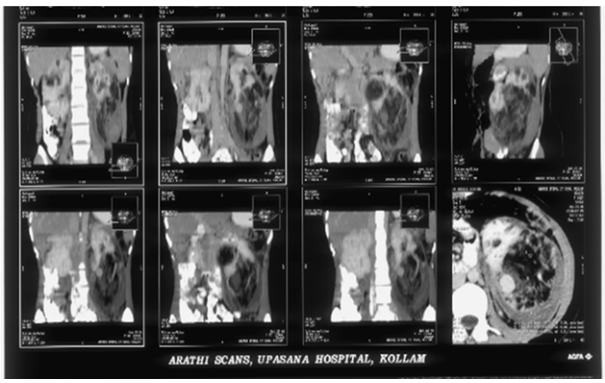

Figure 3. **Showing CT scan Abdomen with bilateral multiple AML and intralesional haemorrhage.**

figures 4, 5 and 6). She had mild fever second post procedure day which improved with antibiotics. Post embolization syndrome consisting of nausea, vomiting, pain and fever was managed conservatively. Followup imaging was done after 3 months.

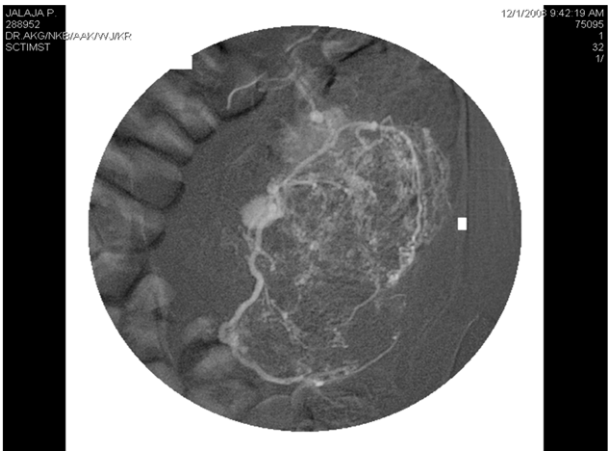

Figure 4. **Showing selective arteriogram**

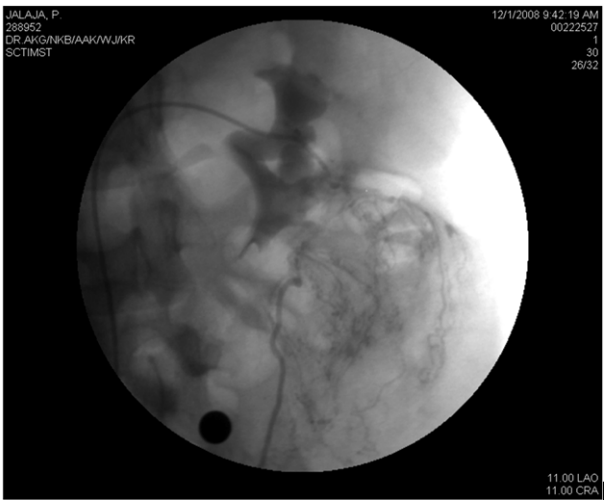

Figure 5. **Showing more details**

DISCUSSION

Tuberous sclerosis is frequently complicated by AML, which is known to occur more frequently as multiple

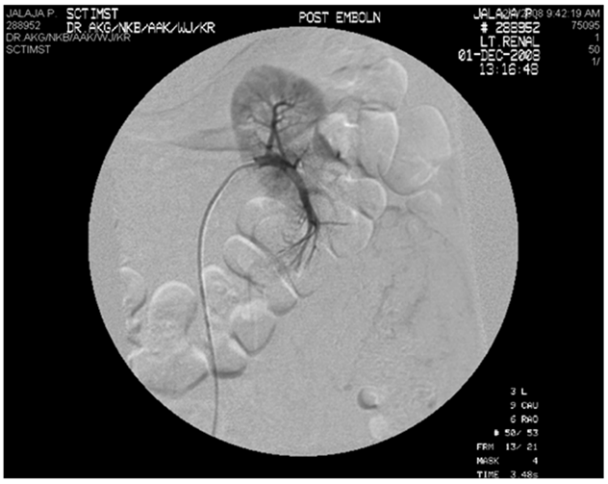

Figure 6. **Showing reduced bleeding post procedure**

lesions and to grow to larger sizes as compared to idiopathic AML. It has also been documented that AML associated with tuberous sclerosis shows a high likelihood of rupture during the clinical course, with resultant retroperitoneal haemorrhage and hematuria leading to haemorrhagic shock.[4] The management options in a case of AML are based on the lesion size and the presence of complications. Here the presence of multiple bilateral large lesions in a tuberous sclerosis patient with intralesional bleeding mandated a procedure with minimal parenchymal damage that was repeatable. Hence intra arterial embolization was the best option. Selective embolization provides embolization of the tumour vasculature and reduction of tumour size and preservation of normal renal parenchyma to the best possible extent. Long term results have been excellent for this procedure.[8,9,10,11,12] David Ewalt et al reported in the J Urology Nov 2005 about 16 patients with 27 tumours who had 18 procedures with average tumour size 4-21 cms.[11] Jacob Ramon etal in European Urology 2009 reports on 48 kidneys treated in 41 patients with ethanol & PVA mixture, the average tumour size being 10.8 cms with 4 – 8 years followup.[12] Danforth et al from Cleveland Clinic reports on two patients with TSC who had giant asymptomatic AMLs and followed for over 20 years on conservative treatment.[13] Kothary et al reported in the Journal of Vascular Intervention Radiology January 2005 reported on 30 renal AMLs treated over 10 years.[10] The recommendation is for lifelong surveillance for recurrence after AML embolization in patients with TS.

CONCLUSION

Cases of multiple bilateral AMLs like in this case require a procedure that is minimally invasive, safe and repeatable for multiple attempts. Trans arterial embolization is an excellent option in this situation. Followup is needed to study their clinical course.

END NOTE

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Conflict of Interest: The authors declare that they have no competing interests.

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