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# LENTICULO STRIATE VASCULOPATHY IN ASSOCIATION WITH HAEPATORENAL MALFORMATION (ANGIOMIOLYPOMA) A STIGMATA OF TUBEROUS SCLEROSIS: A RARE ENTITY

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**ABSTRACT**
Lenticulostriatal vasculopathy ( LSV) has been proposed as a marker of diffuse insult to the fetal and neonatal brain. LSV have a higher incidence of asphyxia, respiratory disease, congenital heart disease, fetal TORCH infection, chromosomal aberrations, and congenital malformations. Here we present a rare LSV association, a congenital malformation of kidney "Angiomyolipoma" with fatty change of liver. Again Angiomyolipoma a soft tissue tumor with stigmata of tuberous sclerosis involving kidney, liver and other organ. So more study require to know any real association.

## INTRODUCTION

The vasculature supplying the thalamus and basal ganglia is normally indistinct from brain parenchyma on the head ultrasound exam of a newborn. However, linear and branching echodensities have been described in the thalami and basal ganglia of a small percentage of newborn undergoing HUS examination. This sonographic finding is known as lenticulostriate vasculopathy . It has been proposed that these bright areas represent a vasculitis of the lenticulostriate branches of the middle cerebral arteries, which occurs in association with a range of perinatal cerebral insults, including infections.

Angiomyolipoma (AML) is a rare well-known soft tissue tumor involving the kidneys, liver and other organs which is classified as typical (triphasic), with three components: mature fatty tissue, blood vessels and smooth muscle; and atypical (monophasic or epithelioid) [1,2]. The epithelioid angiomyolipoma (AMLE), is a atypical histological variant, with aggressive behavior; associated in more than half of the cases to Tuberous Sclerosis , with mutations in the p53 gene and a high rate of distant metastases [3,4].

In our report, we present a case of congenital malformation of kidney "angiomyolipoma" with fatty change of liver in association with lenticulostriate vasculopathy which is an unusual association.

## Case Report

A 24 days old male baby was admitted with history of cough and not passed stool for 7 days. .He is a term 37 wks gestation baby, delivered to a primi mother by NVD. He cried immediately after birth, with apgar score 4 , 6 at 1, 5 min ,respectively . There was no history of any maternal illness complicating the pregnancy. He was on breast feed at 2 hrs of life. There was no history of delayed passage of stool. Physical examination revealed weight 2.5 kg, length 48 cm with no external congenital abnormalities. At the time of admission, his heart rate was 140/min, respiratory rate 76/min, Spo2-80% and blood pressure 60/40 mm hg. Sepsis screen was positive .Cardiovascular examination revealed no abnormality. Respiratory examination revealed increased rate with both intercostal and subcostal retraction and decreased breath sound in lt

infrascapular area. Gastorintestinal system was normal .Laboratory investigation showed random blood sugar 57mg/dl, hemoglobin-17.3 gm%, and white blood cell count 9,000with normal differential and platelets. Serum urea nitrogen was 15 mg%, creatinine 0.9 mg%. Serum electrolytes were Na- 138mg/dl and K- 4.7 mg/dl .Cerebrospinal fluid analysis was normal. Urinalysis showed a specific gravity of 1.015, pH 7, and trace protein and RBC cells with urine output >2 ml/kg/hr.

Abdominal and pelvic ultrasound revealed a focal hyperechoic area noted in rt lobe of liver of size 1.3 ×0.6cm may be focal fatty change (fig 2) and a small well defined rounded hyperechoic lesion of size 0.6 ×0.5cm noted in rt kidney (fig 1) likely to be "angiomyolipoma".

**Figure 1. Rounded hyperechoic lesion of size 0.6+0.5cm noted in rt kidney**

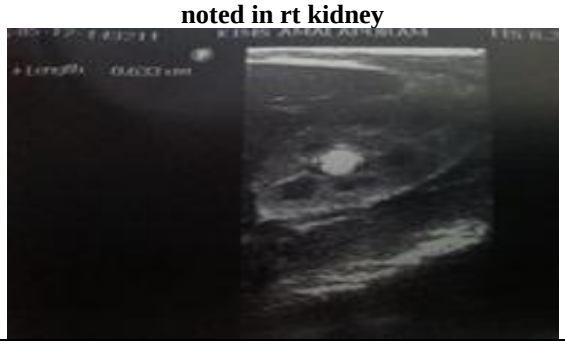


**Figure 2. Rt lobe of liver with focal fatty change of size 1.3+0.6cm.**

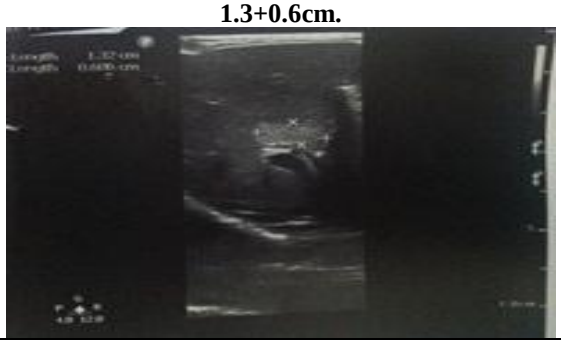
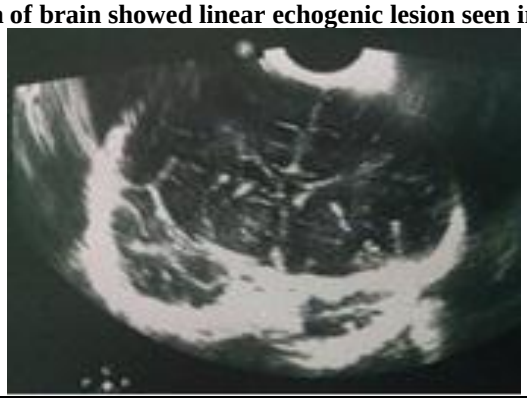


**Figure 3. Ultrasonogram of brain showed linear echogenic lesion seen in the b/l lentiform nuclie**



## DISCUSSION AND CONCLUSION

LSV has been proposed as a marker of diffuse insult to the fetal and neonatal brain. Various underlying perinatal and neonatal conditions have been reported to be associated with LSV, mainly fetal TORCH (toxoplasma, other viruses, rubella, cytomegalovirus, herpes virus) infections, chromosomal aberrations, congenital malformations, congenital heart disease, and asphyxia [5]. LSV was first described by Teele et al [6] the reported incidence of LSV has varied between 1.8% and 5.8%.

In one study, Sonographic follow up of patients with LSV up to 15 months of age showed progression of LSV in 14.7–85% of cases, no change in 15–86.5% [7], or resolution of lesions in 36.4–50% of cases [8]. However, the extent of LSV progression was determined sonographically Follow up studies reported a neurodevelopemental delay in 18.9–55% of patients with LSV [9]. Furthermore, in premature infants of birth weight < 1250 g, those with LSV had lower scores for mental development, motor quality, and emotional regulation [8]. Prematurity by itself, with or without respiratory distress syndrome, does not appear to adversely affect the short term developmental outcome of patients with LSV [7].

In the absence of severe underlying conditions such as fetal TORCH infections, chromosomopathy, major malformations, or hypoxic-ischaemic states [10], survival without adverse clinical outcome is the rule in patients with LSV.

AML is present in 50 to 80% of cases of TSC, and their partnership is closer with epithelioid variant of renal AML [11]. AMLE affects both sexes equally. Patients are symptomatic, complaining of flank pain, palpable mass, and less than 15% will present renal failure due to compression and replacement of renal parenchyma.. The imaging studies simulate clear cell carcinoma scanty fatty tissue [12,13]. Macroscopically, tumors are medium to large size, yellowish-orange with large areas of hemorrhage and necrosis. There may be extrarenal tumor extension or involvement of the vena cava or renal vein. Microscopically, is an infiltrating tumor, very cellular, consisting mainly of polygonal cells with vacuolate cytoplasm, eosinophilic granular or clear, with plenty of glycogen, and other multinucleated similar to the ganglion cells. Immunohistochemistry is important to characterize this tumor.

The presence of immunoreactivity positive for HMB45, HMB50, CD117, CD63, and the negativity to epithelial markers and cytokeratins confirm the diagnosis [14]. The management of renal AML in asymptomatic tumors, evaluation with abdominal ultrasound and/or CT-scan every six or twelve months, depending on the size of the tumor, greater or less than 4 cm., respectively, is necessary. In symptomatic and/or bilateral tumors, artery embolization, selective kidney or conservative surgery (nephron sparing) are the treatments of choice. Radical nephrectomy is reserved for those cases with hemodynamic instability due to massive bleeding, large tumors, or coexistence with carcinoma in the same kidney [15].

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