Linear Naevus Sebaceous Syndrome with Aortopathy and Cystic Kidney Disease A Rare Presentation

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INTRODUCTION

Linear naevus sebaceous syndrome (LNSS) is characterized by linear sebaceous naevus which usually presents at birth. Its association with various neurological, ophthalmological and cardiovascular anomalies is better described. Aortopathy and cystic kidney disease, however, is not reported so far in the literature.

HISTORY

We reported an 8-year old girl who had multiple well-defined yellowish plaques over body at birth. Ophthalmological examination showed right chorioretinal coloboma and left eyelid lipodermoid. She had coarctation of aorta (COA) required surgical intervention on Day 7. She had epilepsy with MRI Brain showing right frontotemporal pachygyria. With combination of these clinical features, she was diagnosed to have LNSS.

She had long-standing hypertension, which was not totally explained by her history of COA. Further investigations with ultrasound and MR angiogram revealed severe aortopathy with bilateral renal artery stenosis (RAS) and multiple right renal cysts. The cysts measured up to 1.5 cm in size. There was no intra-cystic solid component, with preserved renal cortical thickness. MAG3 scan showed impaired right renal perfusion (Differential function Left-Right: 68%:32%). There was severe narrowing of whole aorta involving bilateral renal arteries, making balloon angioplasty for RAS difficult. However, her renal function test remained normal. Her blood pressure was controlled with amlodipine and atenolol. There was no family history of cystic kidney disease.

MRI showing multiple right renal cysts

MRA showing aortopathy with bilateral RAS

CONCLUSION

Linear sebaceous naevus syndrome is associated with cystic kidney disease and renal artery stenosis. Early ultrasonography is recommended to look for associated renal anomalies. Hypertension in this group of patient can be related to RAS.