A case of Dyke-Davidoff-Masson syndrome with ectopic kidney

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Introduction

Unilateral cortical atrophy in the intrauterine period with compensatory hypertrophy of the contralateral brain, skull and sinuses was described by Dyke-Davidoff and Masson in 1933[1]. There are many case reports in the literature but ectopic kidney as in our case is a very rare association.

Case report

An 8 year old girl was admitted with left sided recurrent focal seizures originating in the left upper limb followed by secondary generalization. She was on multiple antiepileptic medications since 6 months of age. Motor milestones were delayed on the left side consistent with a left sided hemiparesis with early hand preference of the right hand. Neurological examination revealed right-sided spastic hemiparesis with brisk tendon reflexes and an extensor plantar response. At present, with occupational and physiotherapy, she can use the left side with grade II limitation of activity[2]. Magnetic resonance imaging (MRI) at 5 years of age showed right sided diffuse cortical atrophy, porencephalic cyst with dilatation of lateral ventricle, and compensatory hypertrophy of the left hemi-cortex with normal cerebellar structure[1] (Figure 1).

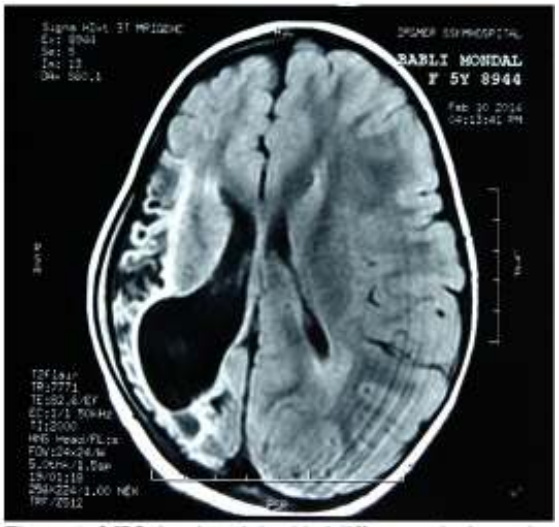

Figure 1: *MRI showing right sided diffuse cortical atrophy, porencephalic cyst with dilatation of lateral ventricle and compensatory hypertrophy of left hemicortex*

X-ray paranasal sinuses (Figure 2) at this admission showed right sided dilatation of all sinuses with enlarged marrow spaces of all calvarial flat bones[3]. Routine ultrasonography of abdomen showed that the left kidney was absent below the spleen (Figure 3), ectopically situated in the left iliac fossa (Figure 4), and smaller in size, measuring 7.72cm×3.95cm, whereas the right kidney was 9.41cm×4.70cm. Renal function and electrolytes were normal.

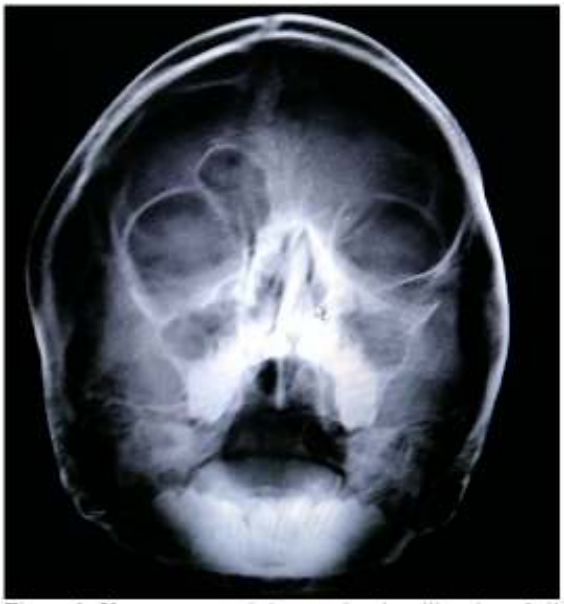

Figure 2: *X-ray paranasal sinuses showing dilatation of all sinuses with enlarged marrow spaces of calvarial flat bones*

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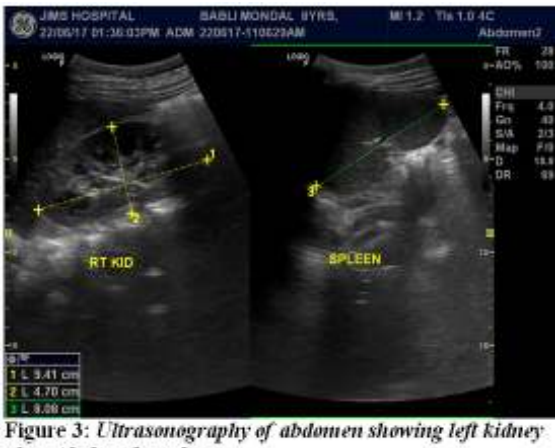

Figure 3: *Ultrasonography of abdomen showing left kidney absent below the spleen*

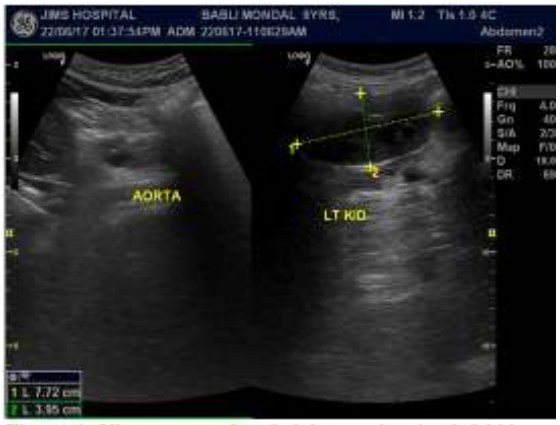

Figure 4: *Ultrasonography of abdomen showing left kidney ectopically situated in left iliac fossa*

The child had a normal delivery at term with delayed cry needing neonatal intensive care admission. There was no history of any fever of long duration or central nervous system (CNS) infection after the neonatal period.

Discussion

This spectrum of signs and symptoms arising out of unilateral cortical atrophy in the intrauterine period, with compensatory hypertrophy of the contralateral brain and ipsilateral hypertrophy of skull and sinuses, is described by Dyke-Davidoff and Masson in 1933 by observing the skull x-ray and pneumato-encephalographic changes in 9 persons[1]. In 1939, Alpers and colleagues described 2 types of cerebral atrophy[4]. In the primary type, the entire cerebral hemisphere is characteristically hypoplastic and in the secondary type, it is due to a vascular insult, cranial trauma or progressive inflammatory neurodegenerative disease. In the primary type, the primary pathology of unilateral cerebral atrophy is lack of development rather than atrophy. The earlier the vascular lesion in early gestation, the more will be the cerebral atrophy.

In this case, refractory seizures were controlled with multiple anticonvulsants. Along with drugs, occupational-physical therapy and speech therapy play a significant role in the long-term management of the child. But no existing literature explains the association with ectopic kidney and its prognosis. Hence this association is very rare.

References

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