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Case Report

LMN Facial Palsy - Initial Presentation of Posterior Reversible Encephalopathy Syndrome (PRES) and Obstructive Hydrocephalus in a Young Child Due to Focal Segmental Glomerulosclerosis (FSGN)

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Keywords

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- Posterior Reversible Encephalopathy Syndrome (PRES)

Abstract

Lower Motor Nerve (LMN) facial palsy is a rare presentation of systemic hypertension in children. A 8.5 year old boy, initially had isolated right LMN facial palsy, progressed over 2 weeks to hypertensive encephalopathy, obstructive hydrocephalous, status epilepticus, renal injury with proteinuria treated symptomatic in PICU diagnosed as Focal Segmental Glomerulosclerosis (FSGS). Subsequently Child recovered neurologically without deficit. Blood pressure measurement is required in every child with isolated facial palsy. Obstructive hydrocephalus is a rare association with PRES.

INTRODUCTION

Isolated Lower Motor Nerve (LMN) facial palsy is mainly idiopathic, while may be a very rare initial manifestation of systemic hypertension [1-3]. Acute, acquired isolated LMN facial palsy is most often labelled as Bell's palsy by office paediatricians and treated with short course of steroids without extensive investigations. Routine blood pressure measurements may not be done in children in office practice unless high index of suspicion. Our case, who initially presented as isolated LMN facial palsy later progressed to near fatal Posterior Reversible Encephalopathy Syndrome (PRES) with status epilepticus needing prolonged intensive care management, re-emphasise importance of blood pressure measurement for early diagnosis and timely intervention.

CASE REPORT

Eight year and six months old boy who had right LMN facial palsy followed by hypertensive emergency with PRES, obstructive Hydrocephalous, status epilepticus, renal injury with nephrotic range proteinuria; 2 weeks after initial presentation. He had presented 15 days prior, as sudden onset inability to close right eye with disappearance of naso-labial fold and with jaw deviation to left indicating right Lower Motor Neuron (LMN) facial palsy without history suggestive of acute infection. On examination his higher functions, rest cranial nerves and motor system with gait was normal. Child has normal birth and development history with normal growth. This was diagnosed as Bell's palsy in a paediatrician's clinic where blood pressure was not recorded and oral prednisolone 2 mg/ kg /day was prescribed without any further investigations.

Two weeks later, he presented to emergency department with status epilepticus, de-cerebrate posturing, respiratory failure needing intubation and mechanical ventilation. He was managed with airway clearing, arresting seizure with IV Midazolam, IV Phosphenytoin, along with rapid sequence intubation was followed and child was shifted to PICU for ventilation. On stabilization child had persistent hypertension 180/130 mm of Hg (> 99 centile for age and sex without significant difference between extremities) with normal pupils and fundus examination. Na-nitroprusside was started with arterial blood pressure monitoring along with subsequent substitution of antihypertensive agents (amlodipine, metoprolol). His complete haemogram, BSL, coagulation profile, liver functions, electrolytes (Na, K, Cl, Mg, Ca, P), compliments (C3,C4), ESR, lipid profile and electrocardiogram, cardiac 2D echo were normal. Child had high urine protein to creatinine ratio (11mg/ mg nephrotic range), high blood urea (140 mg %) & creatinine (3.09 mg %), low serum albumin (2.6 gm %) (Figure 1).

Ultrasound showed bilateral bulky kidneys, increased parenchymal echogenicity with altered cortico-medullary

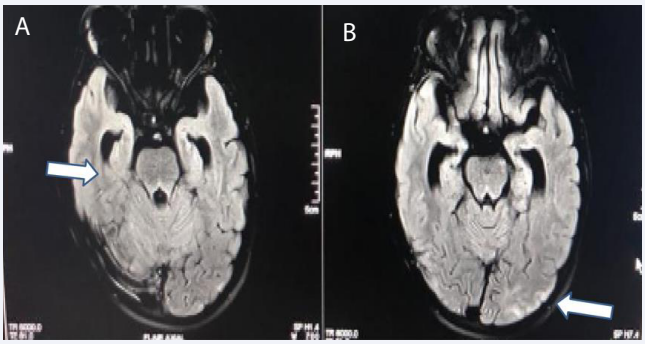

Figure 1 MRI Flair images showing. A) Dilated lateral ventricles and B) Hyper intensities in parieto-occipital cortex.

differentiation suggestive of medical renal disease with normal renal artery Doppler. MRI brain, showed Posterior Reversible Encephalopathy Syndrome (PRES) changes, arrested obstructive hydrocephalus with peri-ventricular ooze. Child required haemodialysis (8-10days) due to progressive renal impairment. His sensorium remained impaired for 3 weeks and needed tracheotomy for maintenance of airway. During ward stay he become cognitively normal by 6 weeks with maintained BP around 50th centile on oral antihypertensive medications and started ambulation by 8 week with complete recovery of facial palsy. Child was discharged on 4 oral hypertensive medications (amlodepin, metoprolol, clonidine and prazocin). Renal biopsy was done on follow up showed Focal Segmental Glomerulosclerosis (FSGS) with interstitial fibrosis requiring immunosuppression.

DISCUSSION

Our patient (without significant co-morbidity) presented with LMN facial palsy started on oral steroids who later developed PRES due to underlined new onset acquired renal parenchymal disease secondary to FSGS. This is youngest FSGS complicating into hypertensive crises with PRES who initial presented with LMN facial palsy reported in literature till date. Hypertension presenting as isolated LMN facial palsy has been reported earlier in fewer case series [1-3]. Obstructive hydrocephalous with changes of PRES on MRI which could be result of congestion of posterior fossa structures as postulated earlier [4,5]. Patient was managed symptomatically and supportively with final diagnosis of right LMN facial palsy with hypertensive encephalopathy secondary to FSGS. Our patient recovered completely without neurological deficits.

LMN facial palsy is common neurological condition seen at clinical practise with incidence up to10 to 20/1,00,000 populations per year. While prevalence of systemic hypertension is reported around 1-2% [1]. Usually it presents as Headache which is the most common neurological complication in children with severe arterial hypertension, followed in decreasing order by altered level of consciousness and vomiting, convulsions, focal central nervous system deficit, and peripheral facial nerve palsy. First case was described in 1869 [3], systemic literature review revealed around 70 reported cases worldwide [4-11].

Jorg et al, in a systemic review of the facial palsy in hypertension including 5 largest case series also states that it is an uncommon presentation with median age of 9.5 years, incidence and median time to recovery 2 months in 26 patients. The median time to elapse between the first facial symptoms and diagnosis of systemic hypertension was 45 days. Commonest cause of hypertension was ascribed to congenital anomaly of kidney and urinary tract (7/26) while others been renal artery stenosis (3/26), GBS (3), coarctation of aorta (3) and primary hypertension (4/26) rarely due to neoplasm like phenochormocytoma (1/1) unknown (3/26) mid aortic syndrome (1/26) . Neuro-radiology showed infracts (3/26), bleed (1/26) and root enhancement (2/26) and PRES (1/26). Its pathogenesis remains unclear with possible proposed explanations like Swelling of the facial nerve in its bony canal associated with vessel engorgement, haemorrhage into the facial canal, an ischemic stroke affecting the post nuclear fibres of the nerve and bleeding in the facial nerve nucleus [4,5].

CONCLUSION

Blood pressure is not routinely measured in busy paediatric outpatient clinic unless in obvious high risk group like renal disorders, obesity etc. LMN facial palsy, being a rare sole presenting clinical feature of hypertension, can be misdiagnosed as idiopathic Bell's palsy during initial evaluation without careful BP measurement during busy office practise. In order to prevent life threatening hypertensive crises we strongly recommend BP measurement for children presenting with isolated LMN facial palsy.

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