Unusual association of diseases/symptoms

A patient with bilateral facial palsy associated with hypertension and chickenpox: learning points

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Summary

Bilateral facial nerve paralysis is an uncommon presentation and even more so in children. There are reports of different causes of bilateral facial nerve palsy. It is well-established that hypertension and chickenpox causes unilateral facial paralysis and the importance of checking the blood pressure in children with facial nerve paralysis cannot be stressed enough. The authors report a boy with bilateral facial nerve paralysis in association with hypertension and having recently recovered from chickenpox. The authors review aspects of bilateral facial nerve paralysis as well as hypertension and chickenpox causing facial nerve paralysis.

BACKGROUND

We found this a good opportunity to educate ourselves about bilateral facial palsy in children and share our learning experience. We performed a literature search and found no reports of hypertension causing bilateral lower motor neuron facial palsy. To the best of our knowledge this is the first case of bilateral facial palsy associated with hypertension to be reported in children or adults.

CASE PRESENTATION

We report an 8-year-old boy with bilateral lower motor neuron palsy who initially presented to our department with a 4-day history of headaches described as bilateral heaviness in the frontal area and was responding to paracetamol. He had recently recovered from chickenpox a few days prior to this presentation. The headache was not associated with fever, dizziness, vertigo or abnormal behaviour. There was no history of clumsiness or falling and his gait and speech were appropriate. Examination showed a blood pressure (BP) of 116/80 mm Hg and 123/67 mm Hg and no focal neurological signs. There were a few scabbed chickenpox lesions. Systemic examination was otherwise unremarkable. Following a period of observation, his headache subsided with some paracetamol and he was allowed home with advice on when to return.

He returned to the ward 3 days later still having headaches, which became worse and required more pain relief. Parents reported that his lower lip went numb followed by numbness of his upper lip and he could not eat as he could not put his lips together and found it difficult to even suck through a straw. His parents reported that he was sleeping with his eyes half open. His BP was between 127/90 and 138/89 and his examination showed a bilateral lower motor neuron paralysis of his facial nerve. All other cranial nerves were intact, his motor system examination was normal, he had a normal gait and speech and co-ordination was normal. He had a few scars of chickenpox but no fresh vesicles

and no lesions in his ears. Systemic examination was otherwise unremarkable.

He lives with both parents and a 5-year-old sister who are all healthy. He suffered with recurrent urinary tract infections and bilateral vesicoureteral reflux. Four years earlier a mercaptoacetyltriglycine reported the left kidney contributing 72% and the right kidney 28% of the renal function with no evidence of obstruction; however, there was reflux into the right ureter during micturition. A technetium dimercaptosuccinic acid scan (DMSA) showed similar uptake and loss of cortex from the upper and lower poles of the right kidney. He was no longer on prophylaxis as he had no recent urinary tract infections and is under annual review.

An ophthalmology opinion was obtained, which was satisfactory.

The regional paediatric neurology and nephrology departments advised to start him on losartan and if his BP became higher to give him a stat dose of nifedipine. He was also started on hypomellose eye drops. Overnight his BP measurements were between 122/80 mm Hg and 147/89 mm Hg.

Following transfer to the regional paediatric tertiary centre, a diagnosis of bilateral lower motor neuron facial palsy associated with raised BP with no other focal neurological signs was confirmed.

INVESTIGATIONS

Full blood count, urea and electrolytes, bone profile, liver function tests and glucose were all within normal limits. CT brain was normal. MRI of the brain was normal.

TREATMENT

In view of the recent history of chickenpox, he was started on valaciclovir for 5 days as a precaution. Steroids were not given. The nephrology team advised to continue with his antihypertensive medication.

OUTCOME AND FOLLOW-UP

Four weeks later parents reported that his facial palsy resolved within less than a week of starting treatment and his BP was around 110/75 mm Hg within a week. On examination his BP was 113/60 mm Hg and his facial palsy made a complete clinical recovery. He was advised to continue with the antihypertensive treatment as he is hypertensive secondary to his underlying renal scarring. His BP was well controlled since then and he is under regular follow-up.

DISCUSSION

There are a plethora of publications describing the relation between hypertension as well as chickenpox and facial nerve palsy and others describing the causes of bilateral facial nerve palsy. However, we found no case reports in the literature describing hypertension causing bilateral facial nerve palsy.

The facial nerve is predominantly a motor nerve with some parasympathetic fibres of sensory-autonomic functions. Therefore, it is not only responsible for facial expression but also has an influence on lacrimal and salivary glands, auditory reflex and taste over the anterior two thirds of the tongue. Lesions at the facial nerve nucleus and nerve distal to it result in a lower motor neuron palsy (forehead and lower facial muscles). Some motor neurons supplying the forehead cross to the contralateral side at the level of the brainstem, which means forehead enervation comes from both hemispheres. As a result, supranuclear lesions will spare the forehead on the affected side.

Definition and incidence of facial nerve palsy

Bell's palsy accounts for 40–70% of cases of facial nerve palsy.^{1–3} It is an idiopathic, acute lower motor neuron palsy of the facial nerve. Sir Charles Bell was the first to describe it, as well as the anatomy and functions of the facial nerve.⁴ The incidence is between 2.7–4.2 per 100 000 children under the age of 10 years and 10.1–15.3 per 100 000 people aged 10–20 years with a prevalence rate of 10–30 per 100 000.^{5 6}

Natural history of facial nerve palsy

Unilateral idiopathic facial nerve palsy occurs over a few hours to a day with the complaint that the face is numb despite no actual sensory loss. The paralysis progresses over 17 days until complete. Depending on the location of the facial nerve lesion, lacrimation, taste, salivation and hearing may be affected.^{7–10} In children with Bell's palsy prognosis is usually excellent with almost 90% complete clinical recovery compared to 80–85% in adults.¹⁰ Recovery usually begins within 2–4 weeks reaching its maximum within 6–12 months.^{9 11} Lower rates of recovery are seen with patients who present with complete paralysis and are more likely to benefit from steroid treatment.^{10 12}

History and examination

A detailed history and physical examination are essential to rule out recognisable and treatable causes. The onset, rapidity of progression, history of viral illness, trauma and associated systemic symptoms should be documented. Physical examination should document extent of facial nerve involvement¹³ as well as other cranial nerves and neurological signs. BP,¹⁴ otoscopic examination, including inspection

of the auditory canal, palpation of the parotid gland, inspection of the mastoid area, fundoscopy and joint examination, as well as skin inspection for tick bites in endemic areas are all essential.^{8 10} Faster recovery in the upper face, leaving predominantly lower facial weakness, could be confused with a supranuclear lesion.¹⁵ No further investigations is recommended unless guided by findings of history and examination.^{7 8}

Causes of BP and differential diagnosis of facial nerve palsy

Idiopathic facial nerve palsy is thought to be caused by inflammation of the facial nerve leading to oedema and entrapment in the facial canal.¹⁶ In recent years infection with herpes simplex virus type 1 has been suggested as a cause after researchers found the viral genomic sequence in samples from the endoneural fluid, facial muscle and oral sample of patients with Bell's palsy using PCR DNA testing.^{17 18} Facial nerve palsy in children could be congenital or postnatally acquired. The acquired palsy is most commonly due to Bell's palsy (42%), trauma (21%), infection (13%) and neoplasms (2%).³ Response to inflammation has also been documented in Guillain-Barre syndrome and vasculitis.¹⁰ Lyme disease is the commonest cause in endemic areas.¹⁹ Other causes include hypertension, benign intracranial hypertension, leukaemia, neuromuscular disorders and cerebral disorders.¹⁰

Bilateral facial palsy in children

Bilateral simultaneous facial nerve palsy is defined as a facial paralysis involving both sides of the face within 4 weeks.²⁰ Bilateral simultaneous facial nerve palsy is uncommon and reported at only 0.3–2.0% of facial nerve palsy cases by some²¹ whereas others report it to be less than 1% of cases.²² The annual incidence is approximately 1 per 5 million.²³ It is most often a sign of systemic disease and seldom idiopathic or Bell's type.^{24 25} It often indicates a serious underlying medical condition and may be a medical emergency warranting urgent investigation and medical intervention. Further investigation should be guided by history and examination.²⁶ On the other hand, Kean²² approximates that 20% of bilateral simultaneous facial nerve palsy is due to Bell's palsy and should not be ruled out as a cause according to a report by May and Hardin²⁵ in 1976. Lyme disease is currently the most commonly reported cause of bilateral facial paralysis in children and in endemic areas should be considered the cause until proven otherwise; hence, the importance of eliciting a travel history.^{19 26} Other common causes in children are idiopathic Bell's palsy, trauma, otologic, Guillain-Barre syndrome, Miller Fisher syndrome, multiple idiopathic cranial neuropathy and brain stem encephalitis.^{22 26} Other causes include posterior fossa tumours, leukaemia, neuromuscular disorders and neurosarcooidosis.¹⁰ Chickenpox has been reported to cause bilateral sequential facial palsy in a 5-year-old girl.²⁷ Rare infectious aetiologies include poliomyelitis, tuberculous meningitis and cryptococcal meningitis.^{10 22} Epstein-Barr virus, cytomegalovirus, HIV and mycoplasma pneumonia have also been reported.^{26 28–30} Idiopathic bilateral facial paralysis is a diagnosis of exclusion and can be seen in up to 23% of paediatric bilateral facial paralysis.^{22 26} Electrophysiological testing is of limited prognostic value and poorly tolerated by children and can be omitted from the

evaluation.²⁶ As with unilateral disease, investigations should be limited to findings on history and examination.²⁶ Bilateral Bell's palsy appears to have the same favourable prognosis as unilateral paralysis with one side recovering faster than the other.³¹ There are no high quality trials on the benefit of steroids in paediatric patients even with unilateral palsy and treatment should be directed at the cause of the bilateral disease. However, in view of documented benefit in adult patients, short duration of treatment and good safety profile, it may be advisable to prevent rare long-term deficit. Eye care and protection is recommended in all patients as with unilateral disease.

Prognosis of Bell's palsy and bilateral simultaneous facial nerve palsy in children and adults

In children with Bell's palsy prognosis is usually excellent with almost 90% complete clinical recovery compared to 80–85% in adults.¹⁰ Recovery usually begins within 2–4 weeks reaching its maximum within 6–12 months.^{9 11} Lower rates of recovery are seen with patients who present with complete paralysis and are more likely to benefit from steroid treatment.^{10 12} Bilateral Bell's palsy appears to have the same favourable prognosis as unilateral paralysis with one side recovering faster than the other.³¹

Hypertension and facial nerve palsy and other reported neurological complications

A neurological sign in the presence of hypertension in a child should be regarded as a complication.³² Signs include but are not limited to headaches, convulsions, altered level of consciousness, hemiplegia or cranial nerve palsies. Failure to recognise hypertension as a cause of facial nerve palsy could cause recurrence or serious complications especially if steroids were prescribed.^{33 34} Lloyd *et al* reviewed 35 consecutive patients with severe hypertension over 10 years and revealed 7 (20%) patients who had one or more episodes of hypertention.¹⁴ Facial palsy was the presenting symptom in three of those patients with the onset of the paralysis coinciding with the rise in BP and improved when it was controlled. The paralysis was always unilateral. In the same period, there were 87 postnatal cases of lower motor neuron facial paralysis—that is, 8% caused by hypertension. Five of the seven patients died within a year of the diagnosis. In another series of 47 postnatal cases over 7 years, only 2 (5.4%) were associated with hypertension.¹⁵ Both reported haemorrhage into the facial canal as a possible mechanism based on two necropsy reports of hypertensive patients with facial palsy (Moxon 1869 and Puech 1930).¹⁴ Other suggested mechanisms include pressure of a vessel against the nerve itself or raised intracranial pressure.¹⁵ Two (3%) out of seventy hypertensive children reported by Rance *et al*³⁵ over a period of 14 years had lower motor neuron facial palsy. Trompeter *et al*³² reviewed 45 hypertensive children who suffered neurological complications over a 10-year period and of those only 2 (4%) had facial palsy. Facial nerve paralysis due to hypertension can be intermittent and independent of BP control.³⁶ The earlier reports had a high mortality reflecting the poor control of hypertension in those children but the modern antihypertensive agents changed that to a favourable outcome.³⁶ There are contrasting publications on the usefulness of steroids in idiopathic Bell's palsy; they are contraindicated in

patients with hypertension and so we did not consider them in our patient.

Chickenpox and facial nerve palsy and bilateral simultaneous facial nerve palsy

Ogita *et al*³⁷ noted that varicella zoster virus associated facial palsy was found in 36% of a series of Japanese children. In a study by Gomez *et al* of complications of varicella that led to hospital admission in children,³⁸ central nervous complications occurred in 19.27% of which only one patient suffered a bilateral facial palsy.

Van der Flier *et al*²⁷ reported a 5-year-old girl who developed a bilateral sequential facial palsy during chickenpox who exhibited a right facial palsy followed by the appearance of the characteristic chickenpox exanthem and subsequently she suffered a left facial palsy. Ganjoo *et al*³⁹ has also reported bilateral facial palsy following chickenpox. Pre-eruptive haematogenous and neurogenous spread of varicella zoster virus are the two different pathophysiological mechanisms involved and they demonstrate the variable timing of presentation of facial palsy in relation to the exanthem.

Kim *et al*²⁰ reported that the prognosis of bilateral simultaneous Bell's palsy was better than that of bilateral simultaneous facial nerve palsy due to varicella zoster, which was in keeping with previous reports that unilateral Bell's palsy has a better prognosis than unilateral Ramsay Hunt syndrome.

Learning points

- ▶ To our knowledge this is the first case report of bilateral facial nerve palsy associated with hypertension.
- ▶ Facial nerve palsy, unilateral or bilateral, simultaneous or sequential may occur in association with chickenpox whether before, during or after the exanthem.
- ▶ Hypertensive neurological complications can occur even with mild to moderate elevations in BP. Hence, serial documentation of the BP is essential as errors of measurements could lead to a false reassurance that BP is normal. This is more likely in smaller units where BP measurement is not routine. All children with facial nerve palsy must have their BP measured. This is a known fact that cannot be stressed enough.
- ▶ Bilateral simultaneous facial nerve palsy is most often a sign of systemic disease and seldom idiopathic or Bell's type. It often indicates a serious underlying medical condition and may be a medical emergency.
- ▶ Bilateral Bell's palsy appears to have the same favourable prognosis as unilateral paralysis with one side recovering faster than the other.

Competing interests None.

Patient consent Obtained.

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