Bilateral Facial Nerve Palsy in Kawasaki Disease

Dear Editor,

A previously well, developmentally normal 6-year-old Indian boy was admitted for evaluation of fever. He was first seen at our outpatient service at the onset of fever. At that time, upper respiratory tract symptoms of rhinorrhoea and nasal congestion together with bilateral non-purulent conjunctivitis suggested a viral aetiology. He was initially treated symptomatically but empiric antibiotics were started on day 4 of illness when his fever persisted and a leukocytosis of 19.7K/μL was noted on complete blood count. Haemoglobin (Hb) was 10.3g/dL and platelet counts were 554K/μL. It was thought that he might have an early acute bacterial sinusitis. A chest radiograph performed was also normal. He was admitted to hospital on day 7 of illness when his fever did not resolve despite the above measures.

Our patient's fever was unremitting and peaked around 39ºC, with transient response to routine antipyretics. On admission, leukocyte count was 24.3 K/μL, platelet count was 705 K/μL and Hb 9.4g/dL. Westergren sedimentation rate was 101 mm/h. We considered the possibility of incomplete Kawasaki Disease (KD) in view of the persistent conjunctivitis, raised inflammatory markers, anaemia and rising platelet levels. At no time did our patient demonstrate any rash, acral or mucosal change, or any significant lymphadenitis.

An echocardiogram on day 8 of illness showed aneurysms of the left main coronary artery (LCA), right main coronary artery (RCA), left anterior descending coronary artery (LAD) and left circumflex coronary artery(LCx) thus confirming the diagnosis of incomplete KD. We started our patient immediately on high-dose aspirin (100 mg/kg/day) and administered intravenous immunoglobulin (IVIg) at a dose of 2 g/kg according to published guidelines. Following IVIg, fever continued but at a lower grade than before, with a maximum temperature of 38ºC. On day 10 of illness, right-sided facial asymmetry was noted. Our patient was unable to shut his right eye and experienced drooping of the right side of his mouth. An examination revealed features of a pure lower motor neuron palsy of the right facial nerve (FNP). There was no decreased salivation, hyperacusis, ear pain or loss of taste. He was otherwise asymptomatic and a detailed neurological assessment did not reveal other cranial nerve defects. The auditory canal and tympanic membrane were normal.

When our patient's temperature started to spike towards 39ºC on day 11 of illness, we administered a second dose of IVIg. This time, there was a complete resolution of fever.



Fig. 1. Patient attempting to smile and close both eyes.



Fig. 2. The same patient demonstrating complete resolution of facial nerve palsy after 3 months.

On day 12 of illness, our patient was noted to be unable to close both eyes and had bilateral drooping of the sides of his mouth. A left FNP was consequently diagnosed (Figure 1 shows our patient attempting to smile and close his eyes).

A two-dimensional echocardiography performed on day 13 of illness showed the development of a giant LAD aneurysm (8 mm) as well as aneurysms of the LCA (5.6 mm), LCx (4mm) and RCA (4.8 mm and 4.1 mm). Anticoagulation with warfarin as well as an anti-platelet regime of aspirin was commenced. Prednisolone at 1 mg/kg twice a day for 3 days and once a day for 7 days was given

in an attempt to ameliorate the vasculitic inflammation of the facial nerve.

The left FNP took 23 days while the right FNP took 82 days to resolve. Recovery was complete with no evidence of aberrant reinervation (Fig. 2). Cardiac wise, he was wholly asymptomatic with normal effort tolerance and there were no symptoms of myocardial ischaemia on exertion. He is currently maintained on warfarin and aspirin under standard precautions. To the best of our knowledge, this is the first reported case of incomplete KD causing bilateral FNP.

Discussion

The multi-system vasculitis known as KD was first described in 1967 and its exact aetiology is still a mystery.[1] Although KD's predilection for the coronary vasculature has been the preoccupation of most clinicians and researchers, its vasculitic pathology means it can conceivably affect any organ system. The neurological complications of KD are rare but have been fairly well described, ranging from aseptic meningitis to cerebrovascular accidents and facial nerve palsy.[2]

The first case of facial nerve palsy was described in 1974 and in the 30 years since, only slightly more than 30 cases have been described.[3] The mechanism for FNP has been speculated to be due to ischaemia of the facial nerve distal to the facial canal and outside the stylomastoid foramen. This is consistent with our physical findings of upper and lower facial weakness without any sensory deficit. Ischaemia is likely the result of vasculitic inflammation of attendant arteries. All previous cases of FNP described have been unilateral in distribution.

In the vast majority of previously described KD patients with FNP, their disease had been 'complete' in nature, meaning it wholly satisfied available case definitions (AHA and Japanese KD foundation) of KD. Children with 'incomplete' KD are those who do not fulfill the classic criteria for KD but who have coronary artery involvement.[4]

Our patient, although unfortunate, is thus unique in two aspects. His KD was 'incomplete' and his FNP was bilateral. Our patient is also much older than previously reported patients, the vast majority of them being younger than 2 years of age.

We venture that the unique and unusual features in our patient are but indicators of a florid multi-system inflammatory activation. This case clearly demonstrates that 'incomplete' KD does not in any way mean 'mild' KD. On the other hand, 'incomplete' KD more likely means severe KD as these cases already demonstrate coronary artery involvement whereas the risk of coronary artery involvement in 'complete' KD is 25% without IVIg.

It has been previously suggested that FNP is an indicator of higher coronary risk in KD as a greater proportion of KD patients with FNP have coronary artery complications compared to KD patients without FNP. We think however that FNP itself is probably not a risk factor per se for coronary artery complications, but rather FNP is simply a part of the spectrum of immune activation consequences of KD.[5] Simply put, if KD was severe enough to cause vasculitic ischaemic effects on the facial nerve, it is also more likely to be severe enough to result in coronary artery effects as well.

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