Case report – Congenital

Facial palsy as a presenting feature of coarctation of aorta

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Abstract

We report the unusual presentation of coarctation of the aorta with facial nerve palsy in an infant and a child. The facial nerve palsy and hypertension resolved spontaneously after relief of the aortic coarctation. Our two cases are the first reports of unidentified coarctation of the aorta presenting as facial nerve palsy, with the infant being the youngest to be reported.

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1. Introduction

Acute lower motor neurone facial nerve palsy is a recognized complication of hypertension in adults. It has been less frequently reported in children, where the hypertension is usually secondary to an underlying pathology [3]. We present two contrasting cases of children with coarctation of the aorta and hypertension who presented with acute lower motor neurone facial nerve palsy. The hypertension and facial nerve palsy fully resolved after relief of the aortic coarctation.

2. Case reports

2.1. Case 1

An 11-week-old male infant presented with left-sided facial weakness and weight loss. His weight had dropped from 3.75 kg (75th centile) at birth to 5.4 kg (25th centile) on admission. There was no other significant history or family history of either cardiac or neurological problems.

On examination the radial pulse was 120 beats/min, and femoral pulses were not palpable. Blood pressure was 144/54 mmHg in the right arm, 133/94 mmHg in the left arm, 79/55 mmHg in the right leg and 86/54 mmHg in the left leg.

His heart sounds were normal and he had an ejection systolic murmur loudest at the right sternal edge, which radiated to the neck. Tone was increased in all four limbs with hyperreflexia. A left-sided lower motor neurone facial palsy was present. Chest X-ray revealed a large cardiac shadow. Transthoracic echocardiogram confirmed the presence of juxta-ductal coarctation of the aorta with mild left ventricular hypertrophy and dilatation (Fig. 1). Magnetic resonance imaging of the brain showed no evidence of intracerebral pathology. The facial nerve palsy was felt to be an isolated lesion.

Blood pressure was controlled with a labetalol infusion. A surgical correction was performed by resection and extended end-to-end anastomosis through a left posterolateral thoracotomy. On completion no residual gradient was detected. The facial palsy resolved over the next few days. Post-operative echocardiogram showed good bi-ventricular function and follow-up at 9 months of age was clinically normal.

2.2. Case 2

A 13-year-old Caucasian boy presented with a 12 h history of left-sided facial weakness and lid lag. He had complained of a mild frontal headache for 2 days prior to admission but was otherwise fit and well. Apart from mild asthma, he had no significant past medical history.

On examination, he had obvious left-sided lower motor neurone facial nerve palsy. No other neurological abnormality was detected. On cardiovascular examination, the...
pulse rate was normal, but was noted to have a radio-
femoral delay. His blood pressure was 218/122 mmHg.
He had a palpable left ventricular heave and a grade 3/6
ejection systolic murmur loudest over the aortic area,
which radiated to the carotid areas and through to the back.
An electrocardiogram showed marked left ventricular
hypertrophy. A chest radiograph demonstrated rib notching.
A provisional diagnosis of coarctation of the aorta was made
and he was referred for an urgent paediatric cardiology
assessment.

Transthoracic echocardiogram showed a dilated and
hypertrophied left ventricle with mildly impaired systolic
function. There was a bicuspid aortic valve but no evidence
of left ventricular outflow tract obstruction. There was a
discrete distal coarctation of the aorta with peak Doppler
velocity of 3.4 m/s and significant antegrade diastolic tail-
ing.

He underwent cardiac catheterization. The pressures were
left ventricle 122/0–8, ascending aorta 112/77, and descend-
ing aorta 79/70. There was a very tight distal coarctation
with multiple collaterals and the luminal diameter of the
aorta narrowed to 4.5 mm versus a diameter of the ascend-
ing aorta of 10 mm.

A 26 mm/6–12 mm Jomed covered stent was delivered
over a 10 mm × 4 cm balloon via a 6 French Mullins sheath
and the coarctation was dilated to 10 mm. Repeat angiogra-
phy demonstrated good flow with a reduction in the gradient
to 15 mmHg. Repeat dilatation was performed using a 12
mm × 2 cm balloon to flare both distal and proximal ends
with a final ascending aortic pressure of 102/65 versus
descending aorta 101/66. There was good flow angiographi-
cally with abolition of gradient (Fig. 2).

There was significant reduction in blood pressure follow-
ing deployment of uncovered stent across the coarctation
segment. The lower motor neurone facial nerve palsy
resolved over the next few days.

3. Discussion

Lower motor neurone facial nerve palsy associated with
hypertension was first described in 1869 [1]. The first report
of facial nerve palsy secondary to hypertension as a result
of coarctation was in a 14-week-old infant. This child had
previous surgical correction of coarctation and presented
with a recoarctation, hypertension and facial palsy, which
resolved before the hypertension was corrected [2]. In a
review over a 10 year period of paediatric cases with severe
hypertension, six of a total of 35 cases (17%) had facial
paralysis [3]. A 14 year study quoted a lower incidence of
3%, where two out of 70 children evaluated for severe hyper-
tension had lower motor neurone facial weakness [4]. Facial
paralysis as the initial presenting feature of severe hyperten-
sion was described in three of seven children by Lloyd et al.
[3] and later in three case reports by Siegler et al. One of them
had undergone bilateral ureteral reimplantation for vesi-
coureteric reflux and the other had impaired renal function
and multiple infarcts in the brain [5].

The pathogenesis of facial palsy in hypertension is not
well described. The postulated theories are that it could be
because of oedema or haemorrhage in the facial canal [3].

Our two cases are the first reports of unidentified coarcta-
tion presenting as facial nerve palsy. The first child had a very tight coarctation and therefore presented at 11 weeks of age, the youngest reported so far. He required surgical correction. The older child had a less severe coarctation and did not present until 13 years of age. The coarctation was suitable for balloon dilatation. The recently noted association of seventh nerve palsy with 22q11-deletion [6] was not seen in our children. Hypertension and facial paralysis as the initial features of middle aortic syndrome have been recently reported [7].

4. Conclusion

Acute lower motor neurone facial paralysis is commonly idiopathic, and often termed ‘Bell’s palsy’. It is important not to assume that all presentations fall into this category as a small proportion are secondary to an underlying cause, which must be identified. Important causes to exclude are hypertension (including malignant hypertension) and ear, nose and throat conditions.

These two cases highlight the importance of checking the blood pressure in any child presenting with acute lower motor neurone facial nerve palsy. In both cases a blood pressure check was the first step towards diagnosing treatable coarctation of the aorta.

References