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Dancing eye syndrome: a case presentation

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Abstract

Aim: **To describe a case of a 14-month-old girl who presented to the eye clinic with unusual bouts of horizontal nystagmoid movements.**
Methods: **The orthoptic findings are presented, along with the investigation results, underlying pathology, treatment and long-term prognosis.**
Results: **A thoracic neuroblastoma was identified on chest X-ray and subsequent CT scan. After resection of the tumour the initial presenting sign of intermittent opsoclonus persisted.**
Conclusions: **This case shows the presentation of an infant with dancing eye syndrome (DES), secondary to a thoracic neuroblastoma. Following the surgical resection of the primary tumour the intermittent opsoclonus remained. The ocular motility slowly returned to normal but some neurological sequelae persisted.**

Key words: Ataxia, Dancing eye syndrome (DES), Opsoclonus-myoclonus, Thoracic neuroblastoma

Introduction

Dancing eye syndrome (DES) is characterised by jerky eye movements that are chaotic and often correspond with jerky movements which develop in the limbs, especially the arms. It is also known as opsoclonus-myoclonus, referring to the jerky movements of the eyes and limbs respectively. Opsoclonus-myoclonus was described as early as 1927 but it was not until 1968 that the association between it and neuroblastoma was reported.[1]

Case report

A 14-month-old girl presented to the eye department, having been referred by her health visitor. The orthoptist was the first clinician the baby saw at the hospital.

The infant's mother reported noticing that her daughter had 'wobbly eyes' from 2 months of age, and that the movements were intermittent and especially obvious when the baby was interested and concentrating on an object. There was no previous ocular history. The baby had been born at full term by Caesarean section, weighing 7 lb 10 oz (3.5 kg). The only family history of note was that her mother was mildly myopic. The infant's general health was reported to be good, but her mother commented that the baby could be 'unsteady and wobbly' on occasions.

On initial orthoptic examination the infant's visual acuity was recorded as 6/30 both eyes open using Cardiff acuity cards. Her corneal reflections were initially nasal and symmetrical but cover testing revealed an intermittent alternating esotropia followed by short bursts of rapid nystagmoid jerks, which had the appearance of back-to-back saccades. Her ocular movements were full. Further nystagmoid movements were initiated by OKN, swinging baby test and interest in the Lang stereotest. Examination by a consultant ophthalmologist confirmed the orthoptic findings. The history of clumsiness was noted. Pupil reactions were normal. Fundus examination and refraction were also unremarkable. Urgent referral to a consultant paediatrician was advised.

The paediatrician noted that the baby had been sitting independently from 6 months, could stand only with support and was not yet walking due to poor balance. When she tried to grasp objects very tremulous movements of her trunk and arms were noted. Urgent brain MRI, abdominal ultrasound and urinary VMA/HVA were ordered. The findings were unremarkable. The MRI report did not show any focal intracranial abnormalities. The ventricles were reported as being symmetrical and no abnormality was found in the brainstem or the posterial fossa.

Three months later the baby was admitted to hospital for 10 days following a 'flu-like illness. The baby lost motor skills and had become very floppy. The combination of opsoclonus, myoclonus and ataxia made the most likely diagnosis one of an occult neuroblastoma. An urgent abdominal ultrasound and chest X-ray were performed. A posterior mediastinal mass was revealed. A further chest CT scan showed no calcification within the mass, consistent with a neurogenic tumour. Oral prednisolone was started and the child referred urgently to a tertiary unit, where it was decided to surgically excise the tumour. She was continued on oral steroids (prednisolone) which were later phased out over the subsequent 4 years.

The patient was followed up in the orthoptic department and discharged after 3 years with normal findings. Six years later, however, she remained under paediatric follow-up and there was still some ataxia with motor difficulties and mild developmental delay.

Discussion

The cause of the opsoclonus (in neuroblastoma) is not known but it is thought to be an auto-immune one. As

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'nearly 100% of children with neuroblastoma associated with opsoclonus-myoclonus survive'[1] this has further added to the speculation that it occurs as a result of an auto-immune process rather than metastases. Opsoclonus-myoclonus occurs in 2–3% of patients with neuroblastoma, but neuroblastoma is found in as many as 50% of children who present with opsoclonus-myoclonus.[1] Brunklaus *et al*.[2] conclude that CT/MRI imaging of the chest and the abdomen is the most accurate test to detect occult neuroblastoma. This is especially pertinent in this case as initial MRI of the brain did not reveal any pathology but a later chest X-ray and ultrasound clearly illustrated the thoracic mass. The neuroblastomas are 'almost all small with no associated metastases',[1] which can therefore make detection more difficult in the initial investigations. Metabolic studies have been used as part of tumour detection investigations, but had poor sensitivity, 'reflecting the low metabolic activity of these tumours'.[2]

The literature is clear that despite good survival rates for treated neuroblastoma, the neurological sequelae and their effects can be debilitating. In Pohl *et al*.'s cohort of 54 patients with DES from all causes a 'persisting disability was found in 88%'.[3] They comment that the 'neurological legacy of DES is often evident well into childhood'.[3] Mitchell *et al*. reviewed records of 17 children with DES who had had neuroblastomas resected and found 'cognitive development and adaptive behaviour were delayed or abnormal in nearly all'.[4]

The infant in this case report was monitored in the orthoptic department for several years following resection of the neuroblastoma. It appeared that at the time of her final orthoptic examination no abnormality was found. However, in common with the majority of patients with DES there were persistent disabilities. At discharge the only evidence that alluded to her previous pathology was a slight tremor, which was noted when she pointed to the missing segments on TNO testing.

Conclusion

DES is rare, but affects children more frequently than adults. It is a disease of infancy with a mean age of onset before the second birthday.[5] The patient in this case presented at 14 months old, within this age mean. A child can present in various ways, with varying degrees of ataxia and myoclonus. The orthoptist plays a vital role in the initial assessment of such children and in their ongoing management.

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