Br Ir Orthopt J 2005; **2:** 40–42

Pilocytic astrocytoma of the brainstem

CLAIRE MACINTOSH DBO(D)

Orthoptic Department, Oxford Eye Hospital, Radcliffe Infirmary, Oxford

Abstract

Aim: **To describe a case of pilocytic astrocytoma arising in the medulla, which presented as a sudden-onset esotropia.**
Method: **A case is documented with history, neurological signs, orthoptic findings and management. Pilocytic astrocytoma is discussed in relation to the literature.**
Results: **A small esotropia in a healthy 2-year-old, presenting to a vision screening clinic, was the sole initial sign of a brainstem pilocytic astrocytoma. There was subsequent diagnosis of a mild VIth nerve palsy and development of other neurological signs including gaze-evoked nystagmus and ataxia.**
Conclusion: **Pilocytic astrocytoma is a neuroepithelial tumour of the central nervous system, of low grade, occurring mainly in children and young adults and most commonly in the cerebellum. Pilocytic astrocytoma of the brainstem is much less common and carries a worse prognosis owing to the difficulty of achieving complete surgical resection. This is a rare condition, but orthoptists must remain aware that they may be the first point of referral in such cases and recognise signs indicative of urgent investigation.**

Key words: Medulla, Pilocytic astrocytoma, VIth nerve palsy

Introduction

Brain tumours have an annual incidence in children of 2–3 per 100 000 and, although uncommon, are the most common solid tumours in childhood.[1] Pilocytic astrocytoma has been described as the most common childhood brain tumour.[2]

Astrocytomas are a form of glial tumour (glioma) and take their origin from astrocytes. Gliomas are the commonest form of primary intracranial tumour; all are infiltrative and therefore complete surgical resection is difficult to achieve, there being a liability for recurrence. Astrocytoma is one of the commonest gliomas and accounts for 10% of all primary central nervous system (CNS) tumours in adults, but the figure increases to 40–50% in children.[3]

CNS tumours are classified according to their cell of origin. The World Health Organization (WHO) grades tumours on a numerical scale of I to IV, with IV being the most aggressive. Pilocytic (hair-like) astrocytoma is classed as grade I[4] and is characterised by a distinctive histological pattern of elongated cells with long fibrillary processes, creating a compact, spongy tumour. It most commonly occurs in the cerebellum, but other sites of predilection are the optic chiasm, third ventricle, hypothalamus, brainstem and optic nerve.[4]

Correspondence and offprint requests to: Claire Macintosh, Orthoptic Department, Oxford Eye Hospital, Radcliffe Infirmary, Oxford Radcliffe Hospitals NHS Trust, Woodstock Road, Oxford OX2 6HE. e-mail: Claire.macintosh@orh.nhs.uk

Case report

A 26-month-old boy was seen at the local vision screening clinic by the orthoptist because of a suspected right convergent squint, onset 3 months previously, noticed by his parents. There was a positive family history of squint. There were no other concerns. His general heath was good, he had no allergies and was not taking any medication. He had been born at full term by caesarean section with a birth weight of 3.6 kg.

A small right esotropia with grossly full ocular movements was found and referral made to the local eye hospital.

Orthoptic and ophthalmic assessment

When the patient was seen at the eye clinic 6 weeks later, the esotropia had increased to a moderate angle and measured 40$^{\Delta}$. The boy was reluctant to look to right gaze and there was a suspected right lateral rectus underaction of −1. Some horizontal jerky nystagmus was observed on right and left gaze. Vision assessment with Kay's logMAR was right 0.700, left 0.400. His mother had noticed him tilting his head to the right and remarked that he seemed to be falling over a lot.

Cycloplegic refraction revealed a very small hypermetropic error, which was not prescribed. Examinations of the fundi and media were normal with no evidence of papilloedema. At this time the ophthalmic opinion was that the lateral rectus weakness was debatable, especially in view of the difficulty in obtaining a good evaluation of right gaze; the nystagmus was considered to be minimal and could be end point, and the episodes of 'falling over' could be of normal frequency for a 2-year-old. A 1-month review was arranged.

On review 1 month later, although he remained a bright, active child, the problems with balance had become worse and an unsteady gait was noticed in clinic. The esotropia and suspected lateral rectus weakness

were unchanged, but in addition there was a moderate head posture of head tilt to the right, an unsteadiness of fixation and slight horizontal gaze-evoked nystagmus on right gaze, with an intermittent downbeat element. Visual fields to confrontation appeared full.

Referral was made to the paediatric neurologist. Magnetic resonance imaging (MRI) showed a mass within the medulla.

Neurosurgical management

Three months following initial presentation, the boy underwent a surgical exploration with de-bulking of the tumour. The brainstem was found to be very swollen, and a light grey, soft tumour was found in the medulla. The tumour was described as benign but in a very malignant position, owing to the difficulty of achieving complete resection. After surgery, chemotherapy was considered the most appropriate mode of management.

Chemotherapy management

Chemotherapy was commenced 5 months after surgery using the UKCCSG (United Kingdom Children's Cancer Speciality Group) low-grade astrocytoma protocol. This consisted of vincristine weekly for a 10 week period and carboplatin every 3 weeks. Provided there is evidence of tumour shrinkage, this is followed with vincristine and carboplatin at 4 weekly intervals for a total of 53 weeks. To date, this child has completed his course of treatment and MRI has shown eradication of the tumour. There will be long-term continuing review and follow-up.

Discussion

Presenting symptoms of a brain tumour are those of local neuronal damage or distortion.[1] They include epilepsy, intellectual deterioration, symptoms of impaired consciousness, headache, and focal neurological deficit such as hemiparesis, dysphasia, hemianopia, vomiting, visual disturbance and cranial nerve palsy.

In this case, the presenting sign was an esotropia due to a mild VIth nerve palsy. Hopkin[1] states that cranial nerve palsy is a rare presenting sign, occurring in only 2% of brain tumours, but rising to 11% by time of presentation to the neurosurgeon. However, Bannister[3] states that diplopia due to VIth nerve palsy is usually the first symptom of a tumour arising in the medulla. He also describes some degree of ataxia of the limbs to be common.

Cranial nerve palsy in brain tumour is caused either by the nerve being stretched through direct pressure of the expanding tumour, or by stretching of the nerve by displacement from a more distant lesion. There may also be ocular motor dysfunction due to raised intracranial pressure and secondary hydrocephalus.[5]

In the case presented, the tumour was in the medulla and it is likely that the VIth nerve was stretched by direct pressure, as it exited the brainstem at the junction of the pons and the medulla. The ensuing signs of poor balance, unsteadiness of fixation and gaze-evoked nystagmus with a downbeat element are all signs of a cerebellar lesion,[3] but could also apply to a medullary tumour owing to its close proximity to the cerebellum. It was felt that the adoption of a head tilt was due to medullary involvement, rather than an ocular origin of the lesion.

The literature reports most pilocytic astrocytomas to occur in the cerebellum,[4,6,7] where they are considered to have a good prognosis, as they are well circumscribed and can be surgically excised without recurrence, often resulting in complete cure.[1] The case reported here, of pilocytic astrocytoma of the brainstem, is much less common. Bowers *et al.*[2] identified four cases in the brainstem among a series of 20, and Fernandez *et al.*[7] reported 16 in a total of 80 cases. The latter authors concluded that pilocytic astrocytoma in the brainstem carries a less good prognosis than at other sites, owing to difficulty of achieving complete resection, and they found that 42–45% of partially resected pilocytic astrocytomas recur.

Only two cases of pilocytic astrocytoma presenting with VIth nerve palsy have been reported in the literature, both of which were in young adults. Shaya *et al.*[10] described a man aged 22 years who presented with diplopia of 3 months' duration as a result of brainstem pilocytic astrocytoma. Kuroiwa *et al.*[11] described a woman aged 27 years with diplopia and cerebellar symptoms who was found to have a pilocytic astrocytoma on the dorsal side of the medulla.

Chemotherapy is regarded as playing a stabilising role, with positive objective responses to treatment,[8] and it may delay the need for radiotherapy, which is often reserved until progressive disease is well documented. At 2 years of age our patient was considered too young for stereotactic radiosurgery, but favourable results have been reported with this mode of treatment.[9]

Orthoptic and ophthalmology follow-up of this child continue. He is now aged 3 years 8 months and treatment of the tumour has been successful to date. There remains a very slight residual limitation of right abduction, the esotropia has reduced over time to 8^{Δ} for near and distance, and vision has remained equal without the need for occlusion. There may be potential for regaining some binocularity, but as yet subjective responses to binocular vision tests are unreliable.

Comments

Pilocytic astrocytoma is an astrocytic tumour of low grade, occurring mainly in children and young adults and most commonly in the cerebellum. The child reported presented at 2 years of age with an esotropia and was found to have a pilocytic astrocytoma in the medulla. The orthoptist was the first point of referral, the patient being considered a routine vision screening referral with no other concerns. The later confirmation of a mild VIth nerve palsy and onset of other signs including abnormal head posture, nystagmus and ataxia were the hallmark of this case not being a typical esotropia of childhood with a family history of squint, but one requiring urgent investigation.

References

1. Hopkin A. *Clinical Neurology. A Modern Approach.* Oxford: Oxford University Press, 1993: 272.
2. Bowers DC, Krause TP, Aronson LJ, Barzi A, Burger PC, Carson B, Weingart JD, Wharam MD, Melhem ER, Cohen KJ. Second surgery for recurrent pilocytic astrocytoma in children. *Paediatr Neurosurg* 2001; **34:** 229–234.
3. Bannister R. Intracranial tumours. In: *Brain and Bannister's Clinical Neurology*, 7th edn. Oxford: Oxford University Press, 1992.
4. Graham D, Bell J, Ironside J. *Colour Atlas and Text of Neuropathology.* London: Mosby-Wolfe, 1995: 168–170.
5. Leigh R, Zee D. *The Neurology of Eye Movement.* Philadelphia: FA Davis, 1987: 218.
6. Nelson J, Parisi J, Schochet S. *Principles and Practise of Neuropathology.* St Louis: Mosby, 1993: 216.
7. Fernandez C, Figarella-Branger D, Girard N, Bouvier-Labit C, Paz Paredes A, Lena G. Pilocytic astrocytomas in children: prognostic factors – a retrospective study of 80 cases. *Neurosurgery* 2003; **53:** 544–553.
8. Packer RJ, Lange, B, Ater J, Nicholson HS, Allen J, Walker R, Prados M, Jakacki R, Reaman G, Needles MN. Carboplatin and vincristine for recurrent and newly diagnosed low-grade gliomas of childhood. *J Clin Oncol* 1993; **11:** 850–856.
9. Hadjipanas CG, Kondziolka D, Gardner P, Niranjan A, Dagam S, Flickinger JC, Lunsford LD. Stereotactic radiosurgery for pilocytic astrocytomas when multimodal therapy is necessary. *J Neurosurg* 2002; **97:** 56–64.
10. Shaya MR, Fowler MR, Nanda A. Pilocytic astrocytoma presenting as an intrinsic brainstem tumour: a case report and review of the literature. *J La State Med Soc* 2004; **156:** 33–36.
11. Kuroiwa T, Ohta T, Tsutsumi A. Malignant pilocytic astrocytoma in the medulla oblongata: case report. *Brain Tumour Pathol* 1999; **16:** 81–85.