Figure 2B
Immunocytochemistry for S100 protein shows a positive staining reaction in many tumour cells, including both epithelioid and spindle cells (ABC Technique, Dako, UK) with haematoxylin counterstain, $\times 240$.

polygonal cells surrounded by an abundant basement membrane. A few poorly formed intercellular junctions were noted, but no true desmosomes were identified. No premelanosomes, keratin filaments, muscle filaments, or neurosecretory granules were present in the tumour cell cytoplasm.

Comment

Epithelioid schwannoma is a rare variant of peripheral nerve sheath tumour and usually presents as an asymptomatic mass either in superficial (dermis or subcutaneous) tissue or deep soft tissue of the extremity. Superficial

epithelioid schwannoma in the head and neck region therefore represents an uncommon variant of a rare tumour.¹⁻⁴ The majority of superficial lesions are curable if wide local excision is accomplished expeditiously.⁵ Those in deep tissues, however, tend to be highly malignant and should be treated aggressively with possible adjuvant therapy.⁵

The histological appearances of epithelioid schwannomas are variable, and in the past may have been confused with melanoma or metastatic carcinoma.⁴ The presence of epithelioid cells in a vague nodular pattern (as in this case) is characteristic.⁴ The immunocytochemical and ultrastructural features in this case are also characteristic and enable a distinction from metastatic carcinoma and melanoma.^{4,6} Epithelioid schwannomas are uncommon tumours; previous literature reports indicate that these are fully malignant neoplasms and warrant aggressive treatment.¹⁻⁶ Mitotic activity and other histological features indicating malignancy are not uniformly present; the histological appearance cannot therefore be relied upon as a predictor of biological behaviour.^{4,6}

Our case has behaved in a benign fashion with complete local excision and no evidence of recurrence or metastasis on a 2 year follow up. Accurate histological diagnosis and early complete wide excision of superficial epithelioid schwannoma should be the objectives in managing this tumour.

- 1 Bleach NR, Keen CE, Dixon JA. Superficial malignant schwannoma on the face: a case for early radical surgery. *J Laryngol Otol* 1989; **103**: 316-8.
- 2 Morgan KG, Gray C. Malignant epithelioid schwannoma of superficial soft tissue? A case report with immunohistology and electron microscopy. *Histopathology* 1985; **9**: 765-75.
- 3 Jacinto CM, Grant-Kels JM, Knibbs DR, Daman LA, Piorkowski RJ. Malignant primitive neuroectodermal tumor presenting as a scalp nodule. *Am J Dermatopathol* 1991; **13**: 63-70.
- 4 Enzinger FM, Weiss SW. *Soft tissue tumors*. 2nd ed. St Louis: Mosby, 1988: 798-801.
- 5 Alvira MM, Mandybur TI, Menefee MG. Light microscopic and ultrastructural observations of a metastasizing malignant epithelioid schwannoma. *Cancer* 1976; **38**: 1977-82.
- 6 Gosh BC, Gosh L, Huvos AG, Fortner JG. Malignant schwannoma: clinicopathologic study. *Cancer* 1973; **31**: 184-90.

British Journal of Ophthalmology 1994; **78**: 588-591

Bilateral abducens nerve lesions in unilateral type 3 Duane's retraction syndrome

Mark Mulhern, Catherine Keohane, Gerard O'Connor

Department of
Ophthalmology
M Mulhern
G O'Connor

Department of Pathology
C Keohane

Cork Regional Hospital,
Wilton, Cork, Ireland

Correspondence to:
Mr G O'Connor

Accepted for publication
7 March 1994

Duane's retraction syndrome (DRS) is a congenital ocular motility disorder of neurogenic origin but of unknown aetiology. Clinically there is deficiency or loss of abduction and adduction and there may also be palpebral fissure diminution and globe retraction on attempted adduction.

Upshooting or downshooting of the globe may occur when adduction is attempted. DRS was first described in 1887¹ and early reports suggested a myogenic disorder; however, absence of the abducens nerve and nucleus with innervation of the affected lateral rectus muscle by the

Figure 1 (A) Section of pons at the level of CN VI nucleus. On the right side the abducens nucleus is absent, and the overlying ependyma is smooth (arrows). On the left side the abducens nucleus is present (arrowheads), and elevates the overlying ependyma. Mlf=medial longitudinal fasciculus. Luxol fast blue combined with haematoxylin and eosin, $\times 120$. (B and C) Insets of areas outlined show magnified views of the sites of the abducens nuclei. In (B) large neurons are visible. The ependymal surface is in the upper right corner. In (C) there are no neuronal groups; only glial tissue and blood vessels are seen. The ependymal surface is in the upper left corner $\times 420$.

Fig 1A

Fig 1B

Fig 1C

oculomotor nerve² together with rare reports of acquired DRS in patients with brainstem pathology indicate a neurogenic basis for many cases.

Electromyography also supports a neurogenic cause. Three types of DRS are classified; (1) limited/absent abduction with relatively normal adduction, (2) limited/absent adduction with relatively normal abduction, (3) limited abduction and adduction. Clinicopathological studies are few,²⁻⁵ but include unilateral³⁻⁵ and bilateral cases.² All demonstrated absence of abducens nerves and absence or hypoplasia of abducens

nuclei with innervation of lateral rectus by the oculomotor nerve in some cases.^{2,3}

The majority of cases are sporadic, 5–10% are familial with autosomal dominant inheritance.⁶ We describe a case of familial DRS with unilateral clinical signs and bilateral abnormalities of the abducens nerve.

Case report

A 56-year-old man had a 1 month history of gradual visual loss in his left temporal field. He

Figure 2 Section of right lateral rectus muscle showing several nerve fascicles and branches (arrows). Haematoxylin and eosin, $\times 60$.

was being treated for *Pneumocystis carinii* pneumonia and had AIDS. He was aware that he and several other family members had DRS. Ophthalmic examination showed a corrected visual acuity of 6/9 in the right eye and 6/60 in the left eye. Orthoptic examination showed a type 3 DRS in the right eye – limitation of adduction and abduction with upshoot of the right eye on attempted adduction. There was no globe retraction, altered palpebral fissure, or skeletal anomaly. Left eye movements were normal. Fundus examination showed cytomegalovirus (CMV) chorioretinitis with cotton wool spots in both eyes and a macular haemorrhage in the left eye. Despite treatment the left eye vision deteriorated to counting fingers. He died as a result of *Pneumocystis carinii* pneumonia.

At autopsy the brain was mildly atrophied. The right abducens nerve was absent and the left was hypoplastic but normally located. Dissection of the external ocular muscles showed a nerve entering the right lateral rectus muscle from the right oculomotor nerve. The left lateral rectus was innervated by the hypoplastic abducens nerve and did not have any additional nerve supply. Multiple levels were cut through the pons and stained with combined Luxol fast blue/haematoxylin/eosin. Light microscopy showed almost total absence of the right abducens nucleus, but the left abducens nucleus appeared normal (Fig 1). The right trigeminal and facial nuclei appeared slightly smaller than the left. Cell counts were not performed. The right lateral rectus muscle showed numerous intramuscular nerve fibres (Fig 2), as did the left lateral rectus. Other autopsy findings not relevant to the Duane's syndrome included residual CMV retinitis and numerous elevated glial scars in the left eye, and left optic atrophy. There was no evidence of HIV encephalitis, but two small foci

of necrosis with CMV containing macrophages were found in the left caudate and in the basis pontis. The mild brain atrophy noted grossly was regarded as being the result of AIDS and CMV encephalitis and unrelated to the ocular disorder.

Comment

There are only four clinicopathological case reports of Duane's retraction syndrome²⁻⁵ illustrating unilateral and bilateral cases. As far as we are aware all four were sporadic. In our familial case, there were bilateral neuropathological abnormalities but only unilateral clinical effects. Previous studies have shown absence or hypoplasia of abducens nerves and nuclei on the clinically affected side and the detailed studies of Hotchkiss² and Miller³ also demonstrated lateral rectus innervation by oculomotor branches. Our case showed clinically normal left motility in the presence of a markedly hypoplastic abducens nerve, and innervation of the right lateral rectus by the oculomotor nerve with absent right abducens nerve and nucleus. It is possible that electromyography might have shown dysfunction in the clinically normal left eye muscles, but this was not performed.

The aetiology of DRS is not known. Because of associated skeletal, ocular, auricular, and systemic abnormalities in some sporadic cases and the occurrence of DRS in thalidomide exposed children, a teratogenic effect occurring at about 8 weeks' gestation is regarded as likely.⁶ However, this is unlikely to explain the familial cases, which presumably are the result of an inherited genetic defect giving maldevelopment of abducens nucleus and nerve. It is of interest that there were bilateral pathological changes in our case which may suggest that the sporadic and familial cases differ in their pathology.

- 1 Stilling J. *Untersuchungen über die entstehung der kurzsichtigkeit*. Wiesbaden, Germany: J F Bergmar, 1887: 13.
- 2 Hotchkiss MG, Miller NR, Clark AW, Green WR. Bilateral Duane's retraction syndrome. *Arch Ophthalmol* 1980; **98**: 870-4.
- 3 Miller NR, Kiel SM, Green WR, Clark AW. Unilateral Duane's retraction syndrome (type 1). *Arch Ophthalmol* 1982; **199**: 1468-72.
- 4 Matteuci P. I difetti consueti di abduzione con particolare riguardo alla patogenesi. *Rassegna Ital Ottalmol* 1946; **15**: 345-80.
- 5 Hickey WF, Wagoner MD. Bilateral congenital absence of the abducens nerve. *Virchows Arch Pathol Anat* 1983; **402**: 91-8.
- 6 Von Noorden G. Special forms of strabismus. In: Von Noorden GK, ed. *Burian-von-Noorden's binocular vision and ocular motility*. 3rd ed. St Louis: Mosby, 1985: 372.