European Journal of Ophthalmology / Vol. 13 no. 2, 2003 / pp. 207-208

SHORT COMMUNICATION

Case report

Divergent strabismus fixus - A case report

T.K. SHARMA, J.S. GREWAL, M. MacDONALD

Birmingham & Midland Eye Centre, City Hospital, Birmingham - UK

Strabismus fixus is a rare condition and usually is of convergent type in which one or both eyes are anchored in a position of extreme adduction. Convergent type strabismus fixus is considered to be a congenital disorder (1) and a part of congenital extraocular muscle fibrosis syndrome (1). Villasecca (2) and Martinez (4) described an acquired type of strabismus fixus. Hayashi et al (3) reported that progressive esotropia could develop into the acquired type of convergent strabismus fixus. There are very few reports of divergent strabismus fixus in the literature. It may or may not be accompanied by ptosis or generalized extraocular muscle fibrosis. In our report, a case of divergent type strabismus fixus is described and discussed. (Eur J Ophthalmol 2003; 13: 207-8)

KEY WORDS. *Strabismus, Divergent, Fixus, Extraocular, Fibrosis*

Accepted: October 7, 2002

Case report

A sixty-six year old woman presented with complaints of poor vision and marked deviation of the eyes outwards causing great discomfort and disability as she was unable to look straight ahead. She had mental retardation and poor coordination since childhood for which she was investigated at the National Hospital for Nervous Diseases in London where a diagnosis of cerebellar degeneration was made. Review of past medical records showed she had divergent squint since the age of six years and that she was seen with similar complaints in 1971 when she was found to have uncorrected high myopia with astigmatism (OD and OS). Corrected visual acuity was 3/60 OD and 4/60 OS. There was a very large bilateral divergent squint. There was marked limitation of adduction in both eyes and both eyes were in extreme abducted position. The patient was able to adduct the right eye up to the midline. There was no ptosis and vertical movements were normal.

She was again seen in 1995 in view of severe disability caused by extreme exotropia. The ocular examination revealed similar findings. Lateral rectus muscles were recessed to 7mm bilaterally. There was a small initial improvement in ocular motility, which lasted for 2 months.

She was referred to us in July 1999. She had to adopt an extreme head posture in order to see straight ahead as her eyes were so grossly divergent. Visual acuities were 3/60 OD and 4/60 OS. There was extreme exotropia in both eyes with marked limitation of adduction. A forced duction test revealed marked restriction for all horizontal movements in both eyes. There was marked fibrosis of the conjunctiva, Tenon's and the lateral rectus. A left lateral rectus tenotomy with conjunctival recession and conjunctival autografting to the outer fornix was performed along with a left medial rectus resection of 6 mm in an attempt to improve the primary position of the left eye. There was a slight improvement in head posture but none in eye movements two months postoperatively.

DISCUSSION

The pathogenesis of strabismus fixus remains poorly understood. The convergent type is less uncommon than divergent type of strabismus fixus. Some authors (2, 5) believe it to be a part of extraocular

1120-6721/207-02$01.00/0

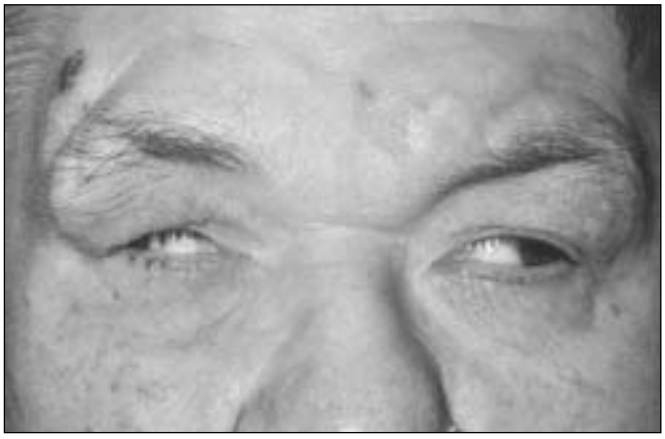

Fig. 1 - *Patient in primary gaze, showing marked bilateral divergent exotropia.*

muscle fibrosis syndrome which arises from dysfunction of oculomotor, trochlear and abducens nerve and the muscles they innervate causing incomitant strabismus with or without ptosis. Wang et al (1) claim to have identified the genetic loci for both types of strabismus fixus. Cases described in their series had bilateral ptosis with restrictive ophthalmoplegia in all gazes. Hayashi et al (3) reported that they have photographic evidence of progression of esotropia into strabismus fixus and suggested early surgical correction of esotropia to avoid severe disability. Awan (6) reported craniofacial anomalies with the divergent type of strabismus fixus. We did not find any such associations in our case.

CONCLUSIONS

Our case had normal vertical gaze and no ptosis. It is unlikely to be of the congenital/genetic type described by Wang et al (1) as it lacks most of the features reported by them. It could be explained by a neurological abnormality causing defective innervation of both medial recti causing exotropia and fibrosis. It could also be a progressive exotropia developing into strabismus fixus. High myopic astigmatism causing poor vision may have accentuated /or caused exodeviation in early childhood which developed over 37 years into strabismus fixus. Surgical treatment in our case was offered at the age of 61. As Hayashi et al (3) suggested, surgery should be offered early in cases with extreme degrees of deviation.

Reprint requests to:
Tarun K. Sharma, MD
Birmingham & Midland Eye Centre
City Hospital, Dudley Road, Winson Green
Birmingham B18 7QU, UK
tks99sus@yahoo.com

REFERENCES

1. Wang SM, Zwaan J, Mullaney PB, et al. Congenital fibrosis of the extraocular muscles type 2, an inherited exotropic strabismus fixus, maps to distal 11q13. Am J Hum Genet 1998; 63: 517-25.
2. Villasecca A. Strabismus fixus. Am J Ohthalmol 1959; 48: 51.
3. Hayashi T, Maruo T. Acquired progressive esotropia and acquired strabismus fixus. Nippon Ganka Gakkai Zasshi 1999; 103: 604-11.
4. Martinez L. A case of fixed strabismus. Am J Ophthalmol 1948; 31: 80.
5. Gillies WE, Brooks AM, Scott M, McKelvie P. A case of divergent strabismus fixus. Binoc Vis Strabismus Q 1999; 14: 40-4.
6. Awan KJ. Strabismus fixus divergens and associated craniofacial anomalies. Can J Ophthalmol 1976; 11: 186-7.