Congenital Crocodile Tears with Duane’s Syndrome - Congenital Cranial Dysinnervation Syndrome

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This 9 year old boy presented with history of copious lacrimation from birth while taking feeds (Fig. 1). It continued after the child started taking solid foods. There had been no lacrimation from either eye when crying and parents were unaware of any eye movement disorder. On examination, the vision was normal. No facial asymmetry was noted in repose (Fig. 2). There was marked limitation of abduction and retraction of globe, narrowing of palpebral fissure on attempted adduction in each eye (Figs. 3 and 4). Adduction and convergence were impaired. MRI brain (High T2 sequence) with cranial nerves screening showed that both abducens nerves were not seen and oculomotor nerve exited from upper pons (Fig. 5). With the above findings, the diagnosis of congenital crocodile tears with Duane’s syndrome type 1 was made.

In Duane’s type 1, abduction of the affected eye is limited, whereas adduction is normal or nearly so. The palpebral fissure narrows and the eyeball retracts into orbit when the affected eye attempts to adduct. The exact cause of this syndrome is not known. Teratogenic effects of thalidomide has also been suggested.

The most plausible explanation for this syndrome would be a defect in the normal development of cells forming abducens nucleus and superior lacrimal nucleus in the brainstem.1 They lie in close proximity during embryogenesis. As a consequence of this, aberrant innervation of lateral rectus by oculomotor nerve and lacrimal gland by fibres from salivary nucleus occurs. The absence of abducens nuclei and the aberrant course of oculomotor nerves noted on MRI brain would give credence to our hypothesis.2 This combination of congenital cranial dysinnervation syndrome is relatively rare.

References


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