Dystonia is a syndrome of sustained muscle contractions, frequently causing repetitive movements or abnormal postures. They are classified by age of onset, by aetiology and by distribution. Younger the age of onset, the more likely that the dystonia will become severe and involve multiple parts of the body (Marsden et al., 1976). Approximately one third of all patients with dystonia have symptomatic dystonia (Fahn et al., 1987). In primary or idiopathic dystonia the only neurological abnormality is the presence of dystonia, and a familial pattern may be seen. The importance of personal examination of family members for the presence of dystonia has been emphasized (Zeman and Dyken, 1968). Focal dystonias are often considered to be a milder expression of torsion dystonia as compared to generalized dystonia. The initial picture is at times bizarre and emotional factors are known to aggravate the condition. Hence they tend to be often misdiagnosed as primary psychiatric illness (Lesser and Fahn, 1978). We report an adolescent with oromandibular dystonia leading to severe self mutilation and discuss some of the issues in classification and assessment.

**CASE REPORT**

RM, a 15 year old boy was referred to our institute with a provisional diagnosis of a psychosis. Ten days before the psychiatric examination, he had started complaining that his tongue was twisting and getting caught between his teeth. He became markedly restless and was constantly crying for help. Sleep was reduced and he could eat very little. He was easily irritated and would often become abusive and assaultive. He was seen spitting out pieces of bitten tongue.

RM was the product of a non-consanguinous marriage. Birth and early development was normal. Two years ago he had left home without information and was traced only about a month back. He could tell that the dystonic movements had started about one and a half years back. Initially, the most troublesome feature was pursing of lips leading to repeated bites. Gradually the movements involved other oromandibular muscles. He had been constantly moving in and out of various hospitals but no further details could be obtained.

On examination, he showed features of intense anxiety. He complained of his tongue getting bitten by his teeth and that he had severe difficulty in swallowing. There was some problem in understanding his speech. Local examination revealed a punched out healed wound on the lower lip, and a severely mutilated tongue. Almost the entire anterior third of the tongue was missing. Fresh tongue bites were seen amidst areas of heaped granulomatous tissue. Dystonic movements of the tongue, palate, pharynx and larynx were obvious. These were particularly provoked by attempts at swallowing though they were also present during rest. He could take only small quantities of fluids by putting himself in certain specific position in the bed. Saliva was retained in the buccal cavity in order to avoid action dystonia. The jaw would open abruptly with retraction of the lips alternating with severe grimacing movements. There was some overflow of these movements to the chest and

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the cervical region but it was hard to say what proportion of this extended dystonia was the result of compensatory attempts engaged in swallowing. No other neurological abnormality was found. General systemic examination was also unremarkable.

Routine investigations including X-ray skull were normal. Slit lamp examination was negative. Serum ceruloplasmin was within normal range. Though no family history of movement disorder was found, while interviewing the mother it was observed that she had repetitive sustained contractions of zygomaticus muscles along with mild contractions of right orbicularis oculi, leading to a narrowing of the palpebral fissure.

Initial treatment with benzodiazepines was not of much help. Then he was put on gradually increasing dose of trihexyphenidyl (THP). Troublesome constipation and blurring of vision along with mild confusion appeared at 20 mg per day doses. Pimozide was added in gradually increasing doses. He did show significant symptomatic improvement with a combination of 20 mg THP and 18 mg Pimozide. As obvious dystonic movements decreased, he showed significant psychosomatic overlay and attention seeking behaviour which was managed through behaviour therapy.

DISCUSSION

Dystonias are rather infrequently seen in clinical practice. In the present case the intense anxiety, disturbances in vegetative functions and self mutilation were initially misinterpreted as suggestive of a psychosis.

This patient had no other neurological abnormality besides dystonia. As the systematic examination and results of routine investigations were also within the normal range, further attempts were made only to rule out Wilson's disease, the only cause of symptomatic dystonia amenable to specific therapy. The diagnosis of a primary dystonia may be easier if there is a positive family history. There is at present no consensus on what constitutes a full syndrome, an incomplete form, or forme fruste of dystonia. In the light of the present case we suggest that the mother be regarded as a forme fruste of dystonia. Further, focal dystonia may only have "incomplete distribution" and may not really represent an "incomplete form" of dystonia.

The Fahn-Marsden scale (1987) for primary torsion dystonia is designed to offer a broad range of scores to quantify generalized dystonia and is not very effective for rating focal dystonia. For the eyes, mouth and neck the product of provoking factor and severity factor is multiplied by 0.5 to down weight the scores for those regions, because "their involvement seems to add less to the overall disability" (Burke et al., 1985). This seems to be unjustified as focal dystonia may also be markedly disabling, as illustrated by the index case. Clearly there is a need to improve on the existing scales for proper assessment of focal dystonias.

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