

## MEIGE'S SYNDROME

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### ABSTRACT

The Meige's syndrome is characterized by the presence of bilateral, symmetrical, dystonic cramp of face muscles or muscles of middle line of body, the respiratory muscles and muscles of throat. The etiology of Meige's syndrome is uncertain. A disorder of basal ganglia function along with perhaps neurotransmitter imbalance (dopamine and acetylcholine) is likely to be the mechanism involved in the causation of this disorder. It is a rare condition and only a limited number of cases have been reported in literature. However, many patients may remain undiscovered or misdiagnosed. Our aim is to introduce a case of 68 years old man who was referred to neurology department with chief complaint of oromandibular dystonia and dysphasia. He was treated successfully with Botulinum toxin.

**KEY WORDS:** Meige's Syndrome, Oromandibular Dystonia, Botulinum Toxin.

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## INTRODUCTION

The spontaneous occurrence of blepharospasm and dystonia movements in face muscles, particularly those of the perioral and mandibular regions, has been named as Meiges's disease which was first described by Henry Meige in 1910.<sup>1</sup> It is a rare form of cranial dystonia, is characterized by bilateral involuntary activity of facial and perioral muscles, together with belpharpspasm and normal emotional and intellectual status.<sup>2</sup>

The disease typically develops between 30 and 70 years of age and is more common in women.<sup>3</sup> The usual symptoms are irritation of eyes, blinking, and stiffness of the face. Burning or a dry feeling in the mouth, dysarthria, dysphagia, and involuntary jaw movements are rare features.<sup>4</sup> Marsden in 1976 described 39 patients the idiopathic blepharospasm- oromandibular dystonia syndrome called Brueghel's syndrome.<sup>5</sup>

## CASE REPORT

A 68 years old man presented with a complaint of dystonia, dysphagia and oromandibular dystonia for a few years. Since five years ago he was noticed to have developed dysphonia and forceful speech and coarse sound. Symptoms were progressive and for the last two years dysphagia developed.

After one year the patient had head torsion to right side and also right blepharospasm. (Fig-1). Past medical history, drug history and family history of similar neurological problem were negative.

On general examination vital signs were stable. The patient was cachectic and generalized weight loss was seen. No lymphadenopathy, icterus or cervical bruits were detected. General examination was normal. In neurological exam, patient was awake and oriented; speech was forceful with coarse and high pitched sound with occasional pauses. Right sided blepharospasm and torticollis were seen.

No deviation of uvula, asymmetry in palatal movement, atrophy or fasciculation of the tongue were seen. Ocular motor activity and pupillary reactivity to light, motor, sensory and cerebellar system, gait, deep tendon reflexes and abdominal reflexes were all normal. Plantar reflexes were flexor. Dystonic neck posturing and head deviation to the right side were noted. Brain MRI was normal. Cervical MRI showed multiple chronic degenerative changes.



Fig-1: Sagittal T1-weighted MR image

(Fig-2, 3) Laboratory tests were normal. The patient was put on trihexyphenidyle and baclofen but since no response to medication was seen, he was put on injection of Botulinum toxin in Sternocleidomastoid, Masseter, Temporalis, and Trapezoid muscle. He was getting better during treatment.

## DISCUSSION

The etiology of Meige's syndrome is uncertain. It is not associated with structural central nervous system pathology. Post mortem study of a patient with Meige Syndrome for seven years revealed no morphological or microscopic abnormalities.<sup>6</sup> The similarities of the spasms to the hyperkinesia that occur in disease of the basal ganglia (e.g. Wilson's Disease, Huntington's Chorea and Von Economo's Encephalitis) and to the involuntary movements induced by levodopa and neuroleptics, suggest that a disturbance of basal ganglia function may occur in Meige Syndrome. Keane and Young<sup>6</sup> reported a case suffering from bilateral basal ganglia infarction had features similar to Meige Syndrome. Jankovic and Patel<sup>7</sup> also reported that three out of six patients with symptomatic blepharospasm had CT evidence of thalamic / upper midbrain infarctions. These findings further supported a possible extra – pyramidal basis of the disease. Tolosa and Lai<sup>8</sup>, basing on the attenuating effect of apomorphine and haloperidol (a dopamine antagonist), suggested



Fig-2: Sagittal T2-weighted MR image



Fig-3: Cervical Dystonia

a state of striatal dopamine preponderance as the pathophysiological basis of Meige Syndrome. They also found similarity of the symptoms in this disease and neuroleptic induced tardive dyskinesia. Both conditions may represent a disturbance of dopamine receptor response (hypersensitivity). In tardive dyskinesia, it is due to the long term dopamine receptor blockade and resultant hypersensitivity following prolonged use of neuroleptics. In Meige Syndrome, the cause is not apparent. The finding that physostigmine<sup>8</sup> is able to aggravate the dystonic spasms in Meige Syndrome supports this theory because in certain pathologic states, cholinomimetic drugs can activate dopamine neurons. The postulated striatal dopamine preponderance may also be the result of a disturbance of striatonigral GABA cells which have a disinhibitory influence on the nigral dopamine neurons. Fahn<sup>9</sup> reported a marked decrease in pallidal GABA concentration in the brain of a patient with "postencephalitic" dystonia. Neophytides<sup>10</sup> also reported decreased CSF GABA level in patients with Meige Syndrome. Summing up, the manifestation is probably due to an imbalance of the three principal neurotransmitters serving these areas, namely, dopamine, acetylcholine and GABA. The

medical treatment of Meige syndrome is unsuccessful. Besides the unpredictable responses to medications, the beneficial effect gained is often not persistent.<sup>11</sup> Traditionally, an antagonism between dopaminergic and cholinergic systems in the basal ganglia was hypothesized. It was based on this assumption that various medical trials were carried out on drugs that affect the dopaminergic or cholinergic pathways. Most of the dopaminergics and cholinergics were ineffective, except apomorphine, lisuride and deanol. However, response demonstrated in these drugs may not be related to their primary actions. Apomorphine is a known dopamine agonist; but at low dose, it may block dopamine transmission through its action on the presynaptic dopamine receptors.

Lisuride is a powerful serotonin agonist as well as a dopamine agonist.<sup>12,13</sup> The cholinergic action of deanol is also dubious.<sup>12,13</sup> With this in mind, Meige Syndrome seems to respond more predictably to antidopaminergics and anticholinergics in concurrence with Tolosa's proposal of a striatal dopamine preponderance. These studies were however mainly empirical and involved small sample sizes. Often treatments were withdrawn before any favourable responses were observed because of the unacceptable side effects. Besides anticholinergics and dopamine antagonists, other agents had been used. Brennan<sup>14</sup> reported a case with favourable response to Baclofen and Sodium Valproate, both having GABA enhancing properties. Merikangas<sup>15</sup> also demonstrated beneficial effects of clonazepam. Surgical approaches had been used when medical treatments failed. Keane<sup>6</sup> described a case with partial relief after serial surgical denervations of the facial nerve branch to the orbicularis oculi muscles were done. Surgery is however only reserved for those with severe intractable symptoms e.g. functional blindness due to blepharospasm. Since 1910 when this syndrome was first described by Meige, various workers had contributed to the delineation of it as a distinct disease entity. However, because of its bizarre presentation and frequent association with anxiety and

depression, they often ended in the hands of psychiatrists. The fluctuation of symptoms over time, its absence during sleep and aggravation when the patients is anxious<sup>16</sup>, often leads to a diagnosis of conversion hysteria. But as Lesser and Fahn<sup>17</sup> had pointed out, conversion hysteria very seldom presented itself as dystonia. In a retrospective study of 84 patients with dystonia, 37 cases were misdiagnosed as hysterical conversion. Only in one patient the dystonic movements were clearly part of a more general psychiatric disorder. For this reason, psychiatrists are often urged to equip themselves with adequate knowledge of neurology and to be careful with early symptoms of dystonia. Up to the present moment, the studies on the pathophysiology and treatment of Meige Syndrome are still empirical. No structural lesion has yet been identified although the pathology was thought to be in the basal ganglia. Medical treatment is still the main line of treatment, anti-cholinergics and dopamine antagonists being commonly used. No symptoms or signs have been identified to be of prognostic value. In the present case, this 68 years old man is responding to treatment with Botulinium toxin.

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