

# Horner's Syndrome, An Unusual Manifestation of Multiple Sclerosis

COLIN R. BAMFORD, MICHAEL S. SMITH, WILLIAM A. SIBLEY

**SUMMARY:** *The prevalence of Horner's syndrome in the MS patient population is 2 - 3%. Using pharmacologic testing in three patients the lesions were localized to the central or intermediate sympathetic neurons. It is likely that demyelination of the central sympathetic neuron accounts for this probable manifestation of MS.*

**RÉSUMÉ:** *La prévalence du syndrome de Horner dans une population souffrant de sclérose en plaques (SEP) varie entre deux et trois pourcent. Grâce à des tests pharmacologiques il nous fut possible de localiser la lésion aux neurones sympathiques centraux ou intermédiaires. Il est probable qu'une démyélinisation du neurone sympathique central explique cette manifestation de la SEP.*

It seems remarkable that multiple sclerosis does not commonly cause a Horner's syndrome for this syndrome is a manifestation of many different disease processes which affect the caudal brainstem and cervical spinal cord, as does multiple sclerosis. Yet, a careful review of the literature detailing the clinical presentation of multiple sclerosis failed to reveal any mention of this syndrome.

On the other hand, a review of the literature relating to the pathogenesis of Horner's syndrome covering 216 cases (Giles and Henderson, 1958) listed two cases of the syndrome attributable to multiple sclerosis; a prevalence of less than 1%.

Although the majority of cases of Horner's syndrome are idiopathic, benign and non-progressive or are due to overt carcinoma (Younge, 1979), it is reasonable to attempt to identify the cause of the syndrome in all patients. The occurrence of Horner's syndrome in the following patients caused us to reappraise their clinical presentations to exclude the possibility of some other condition masquerading as multiple sclerosis, and also prompted, in two of the cases, a search for a second unrelated disease process.

From January 1976 until August 1979 between 125 and 130 patients have been registered with the University of Arizona Health Sciences Center MS Clinic as having a diagnosis of either probable or clinically definite MS. The patients are interviewed monthly by an experienced nurse practitioner and are neurologically examined every three months, or whenever their disease courses appear to alter. There is approximately a 10% turnover of the patient population per year due to attrition and a further 10% due to death or dementia.

**CASE 1:** R.D. is a 42 year old white female, who developed weak legs when she was 30 and this resolved after 4 months. When she was 31 she had an attack of vertigo which lasted 4 days. At 32 she developed double vision which lasted 2 months. Between the ages of 32 and 36 she had transient episodes of dizziness, double vision, numbness of the right arm, and a Bell's palsy, all of which recovered. When she was 38 she had deafness of the right ear which resolved within 1 month. When she was 39 she had an episode of urinary retention, and at 40 she complained of a Lhermitte's phenomenon. When she was 41 she complained of drooping of the right eyelid, fluttering of the right eye, and buzzing in the right ear.

On neurologic examination her visual acuity was normal, her cranial nerves were normal except for ptosis and miosis of the right eye.

Motor function testing revealed normal strength and tone. The deep tendon reflexes of the right arm were hyperactive (3+), those of the left arm were normal (2+), the right knee jerk was normal (2+), the left knee jerk hyperactive (3+), and the ankle jerks were absent bilaterally.

Sensory examination was normal to light touch, pain, vibration, and position testing. Cerebellar function testing was normal. Fourteen months later the patient still complained of ptosis of the right eye towards evening. In the dark the diameter of the left pupil was 7 m.m., and of the right pupil 6 m.m. On exposure to light the right pupil decreased to 3 m.m., the left to 4 m.m. Two drops of hydroxyamphetamine 1% were instilled into each conjunctival sac, both pupils dilated to 8 m.m. in the dark and contracted to 6 m.m. in strong light.

At the age of 41, a pattern reversal visual evoked response was obtained which revealed an abnormally prolonged latency of the left eye (latency to P2 patient: Control :: 128 : 110 msec.).

A diagnosis of clinically definite multiple sclerosis was made at this time and the previous Horner's syndrome was attributed to this.

Reprint requests to: Dr. Colin R. Bamford, University of Arizona, Health Sciences Center, Tucson, Arizona U.S.A. 85724.

From the Department of Neurology, Arizona Health Sciences Center, University of Arizona.

**CASE 2:** H. B. a 57 year old white male developed transient weakness of the legs when he was 21 and this recurred over the next 4 years. When he was 25 he complained of a staggering gait and numbness of the arms. When he was 31 he began using a cane because he was slowly developing permanent leg weakness. At about that time he developed blurred vision of the right eye which persisted. When he was 51 he developed paralysis of the legs which lasted 3 months. At the age of 56 he developed drooping of the right eye.

When he was 57 a neurologic examination revealed a visual acuity on the right of 20/400, on the left of 20/20. The cranial nerves were intact except for ptosis and miosis of the right eye. In ambient light, the diameter of the right pupil was 3 m.m., the left pupil 4 m.m. With intense light the right pupil constricted to 2 m.m., the left to 2½ m.m. Two drops of 1% hydroxyamphetamine 1% were placed in each conjunctival sac and both pupils dilated to 5 m.m.

On motor function testing there was slight weakness of the right hip flexor, moderate weakness of the left hip flexor and ankle extensor. There was a spastic left leg and slightly increased tone of the right leg. Deep tendon reflexes were hyperactive in the upper extremities (3+) and sustained clonus was obtained in both lower limbs.

Sensory function was normal. Coordination was intact. Chest x-ray was normal. A diagnosis of clinically definite multiple sclerosis was made.

**CASE 3:** C. C. is a 40 year old white male who developed poor vision of the right eye

and an unsteady gait when he was 30 years old. The poor vision recovered in 2 days, the gait returned to normal in 6 weeks. When he was 34 he developed leg weakness which progressively deteriorated. When he was 35 he had an episode of incontinence. When he was 36 he became confined to a wheelchair, developed weakness of the arms, and then developed incoordination of the arms. At the age of 40 he noted drooping of the right eyelid. On his physical examination the right optic disk was pale and the visual acuity of that eye was 20/400. The visual acuity on the left was 20/30. He had a right Horner's syndrome, a mild right facial weakness, and a nasal and scanning speech. On motor function testing he was paraplegic with a spastic right leg. Sensory function testing was intact except for absent vibration over all 4 extremities and absent position sense at both great toes. Coordination of the arms was significantly impaired. He had an external catheter in situ.

In ambient light, the right pupil was 4 m.m. and the left pupil was 6 m.m. With intense light, both pupils constricted to 3 m.m. Two drops of 1% hydroxyamphetamine were instilled into each eye and after thirty minutes the left pupil had dilated to 9 m.m. and the right pupil had dilated to 8 m.m.

A CT scan demonstrated moderate cerebral atrophy and an unremarkable posterior fossa.

A diagnosis of clinically definite multiple sclerosis had previously been made.

## DISCUSSION

Horner's syndrome is not a common

accompaniment of MS, .64% of the MS clinic patient population develops a Horner's syndrome each year, and the prevalence of the syndrome in the current MS patient population is 2.3%.

The dilatation of the miotic pupils by 1% hydroxyamphetamine in all 3 cases restricts their lesions to the 1st (central) and 2nd (intermediate) order sympathetic neurons. No attempt was made to pharmacologically distinguish between these 2 neurons, as the testing available to do this is somewhat unreliable (Thompson 1977).

Since the prevalence of Horner's syndrome in our MS clinic is significantly higher than in the general population, and there is a reasonable probability that these affected patients have a first order neuron lesion, it would be logical to conclude that Horner's syndrome is a manifestation of MS.

## REFERENCES

- GILES, C.L., HENDERSON, J.W. (1958). Horner's syndrome: an analysis of 216 cases. *Am. J. Ophth.* 46: 289-296.
- THOMPSON, H.S. (1977). Diagnosing Horner's syndrome. *Trans. Am. Acad. Ophth.* 83: 840-842.
- YOUNGE, B.R. (1979). 5th Annual Rocky Mountain Neuro-Ophthalmology Course.