RESEARCH PAPER | Medical Science | Volume : 5 | Issue : 3 | March 2015 | ISSN - 2249-555X

Holocord Syrinx Presenting as Bilateral Ptosis.a Case Report

KEYWORDS: syringomyelia,ptosis,horner's syndrome

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ABSTRACT *INTRODUCTION*

Horner's syndrome,along with weakness and atrophy of hand as well as anaesthesia to pain and temperature is a well recognized finding in a case of syringomyelia.Few cases have been published as horner's syndrome as a presenting feature of chairy-1 malformation with syringomyelia.We report a case of 22yr female presenting with bilateral ptosis probably part of syringomyelia with wekness and atrophy of both upper limbs with out any sensory findings such as loss of pain and temperature sensation having chiary-1 malformation and syringomyelia on imaging.

Case report-

We report a case of 22year female patient with chiary-1 malformation and syrinx upto 9th thoracic vertebra level.who presented with bilateral ptosis along with weakness and wasting of both small muscles of hand and forearm .Typical neurological signs and symptoms of syringomyelia like anaesthesia to pain and temperature was absent in this case. Though horner's syndrome is described but only bilateral ptosis without anhydrosis and miosis is a rare finding

Conclusion-possibility of a syringomyelia could be thought in patients presenting with only ocular findings like progressive ptosis with upper limb wasting even if in the absence of classical sensory manifestation of syrinx.

Case report;
A 22 Year female presented with insidius onset slowly progressive weakness of both upper limbs.Initially she marked difficulty in holding article in her grip like glass of water,difficulty in holding broomstick,mixing dal with rice,writing in paper. The weakness was present similarly and simultaneously in B/L upper limbs. There was no difficulty in doing overhead activity.There was no weakness in lower limbs/neck muscle/truncal muscle. Gradually she marked progressive wasting & hollowing of interdigital space, thenar,hypothenar eminence and thinning of forearm.

There was no wasting of lower limbs.After 2yrs of progressive hand wasting she marked progressive drooping of both eye lids. There was no h/o double vision,proptosis,fluctuation.

She also having neck pain for last one year.The pain is dull aching,constant,localised pain without radiation or postural variation or restriction of neck movements or without aggravation by coughing. There was no h/o-stiffness,Lower limb/truncal weakness,Paresthesia in LL,face,trunk ,Fluctuation of weakness,Fatiguability ,Diplopia/blurring of vision/eye pain/proptosis ,Bladder/bowel problems,Fasciculations ,Incordination of UL/LL.,Dysphagia,nasal regurgitation of liquid,Trauma ,prolonged fever,seizure,visual problem. On examination found to have- B/L ptosis without pupillary involvement ,weakness and atrophy of distal UL with absent reflex (bicep,tricep,supinator),exaggerated knee jerk, b/l flexor plantar ,without sensory deficit,cerebellar signs. The routine investigations were normal including complete blood counts,electrolytes,liver and renal function test,lipid profile,HIV,HEPATITIS-B,C,A,VDRL.CPK 57U/L. Nerve conduction study revealed no abnormality other than decreased CMAP amplitude due to atrophy in upper limbs.EMG revealed no spontaneous activity,reduced recruitment pattern(neurogenic)at wasted muscles and normal recruitment at other muscles with normal phase,amplitude,duration MUAP.The RNS study was normal.MRI showed chairy-1 malformation with syringe-hydromyelia from C2-D9 segment.

DISCUSSION;
Syringomyelia is the development of a fluid-filled cavity or syrinx within the spinal cord. Hydromyelia is a dilatation of the central canal by cerebrospinal fluid (CSF) and may be included within the definition of syringomyelia. Estimated prevalence of the disease is about 8.4 cases per 100,000 people and occurs more frequently in men than in women. The disease usually appears in the third or fourth decade of life, with a mean age of onset of 30

Volume : 5 | Issue : 3 | March 2015 | ISSN - 2249-555X

years. Syringomyelia usually progresses slowly; the course may extend over many years. The condition may have a more acute course, especially when the brain stem is affected (i.e., syringobulbia). Syringomyelia usually involves the cervical area. Symptomatic presentation depends primarily on the location of the lesion within the neuraxis. Syrinx interrupts the decussating spinothalamic fibers that mediate pain and temperature sensibility, resulting in loss of these sensations, while light touch, vibration, and position senses are preserved (dissociated sensory loss).When the cavity enlarges to involve the posterior columns, position and vibration senses in the feet are lost; astereognosis may be noted in the hands.Pain and temperature sensation may be impaired in either or both arms, or in a shawllike distribution across the shoulders and upper torso anteriorly and posteriorly.Dysesthetic pain, a common complaint in syringomyelia, usually involves the neck and shoulders, but may follow a radicular distribution in the arms or trunk. The discomfort, which is sometimes experienced early in the course of the disease, generally is deep and aching and can be severe.These all sensory presentations are absent in our case. Syrinx extension into the anterior horns of the spinal cord damages motor neurons (lower motor neuron) and causes diffuse muscle atrophy that begins in the hands and progresses proximally to include the forearms and shoulder girdles,which is present in our case. Horner syndrome may appear, reflecting damage to the sympathetic neurons in the intermediolateral cell column.In our case only ptosis is present while anhydrosis and miosis are absent.Again ptosis in our case is bilateral which is a rare finding. Painless ulcers of the hands are frequent which is absent in our case.

CONCLUSION

A bilateral ptosis without other features of horner's syndrome, with wasting of both hand muscles without any sensory symptoms and signs could be a presenting feature of holo cord syrinx.