

Monomelic Amyotrophy: A Case Study

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Abstract

Monomelic amyotrophy of a single upper limb termed “brachial monomelic amyotrophy” (BMMA) is a benign lower motor neuron disorder in the young, with male preponderance, insidious onset of atrophy and weakness, electromyographic evidence of neurogenic pattern without conduction block, slow progression followed by a stationary course.

Keywords: motor neuron disease, neuro rehabilitation, pyramidal signs

1. Introduction

Monomelic amyotrophy (MMA) is a benign motor neuron disorder (MND) in the young with male predominance [1, 2]. The disease is characterized by insidious onset of weakness and wasting restricted to a single limb which usually progresses for a few years followed by spontaneous arrest and lack of sensory, bulbar, and pyramidal signs [2]. MMA has been reported chiefly from Asian countries, although there has been scattered cases reported from other regions [3, 4]. But as yet there is no convincing evidence to explain the unique geographic distribution and predilection to Asian. The precise cause for this disorder is yet unknown [2]. Laboratory testing is frequently normal or nonspecific except for electrophysiological studies which typically demonstrated reduced compound muscle action potential (CMAP) amplitudes, and features consistent with acute and chronic denervation in affected muscle [5-8].

MMA is commonly misdiagnosed and should be considered in differential diagnosis in patients presenting with amyotrophy involving lower and upper extremity behind other diagnosis. In this case report, we described the features of a 21 year-old man with MMA of the upper limb.

2. Discussion

The patient was apparently asymptomatic about 8-9 months back when the wasting of the left upper limb around the shoulder associated with the mild weakness was noticed. There was no radiating pain features at the neck, no sensory features as well as no bladder and bowel involvement.

On evaluation his vitals were normal, was conscious, oriented. The cranial nerve examination was normal and the examination of the motor system revealed wasting of the deltoid and supraspinatus muscle on the left side. He had mild weakness of the shoulder abductors as well. Reflexes were all 2+.

On investigations when a magnetic resonance imaging was done for the cervical spine it revealed diffuse disc bulge at C-5/6 with bilateral foraminal disc/osteophyte complexes compressing both existing C 6 roots [Left>Right]. When an electromyography test was conducted focal modest lower motor neuron changes comprising large motor unit potentials and fasciculation's were noted in the left C8/T1 segmental muscles were noted as well as slight changes in general. Other investigation did not reveal any significant finding.

3. Treatment Options

A Physiotherapy management program was planned with Short term goals such as to reduce Pain, Reduce tenderness, maintain the Cardio Vascular Fitness, Improve the Strength and make him perform his activities of daily living.

Exercises especially for the triceps and biceps were taught and made to perform; with it shoulder stabilization exercises were also made to perform. It basically included the muscle strengthening exercises and training in hand coordination and a home exercise program included exercise regimes and proper rest. Other modalities used were the intermittent cervical traction unit and interferential current modality.

4. Prognosis

As we know in such condition after reaching a plateau level the symptoms go down. Hence in this case as well the disability was slight later on and the use of cervical collar was of great help to the patient as well as orthotic pillow use was also beneficial.

5. Conclusion

In Conclusion we should consider the diagnosis of the Monomelic amyotrophy as the most important part and therefore an early diagnosis is important. The use of electromyography investigations also proves to be of great help. With physiotherapeutic rehabilitation and use of medicine the effect of the disease can be reduced.

6. References

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