VULPIAN-BERNHARDT SYNDROME IDENTIFICATION THROUGH ELECTROMYOGRAPHIC STUDY: CASE REPORT

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Background

Vulpian-Bernhardt syndrome is a rare variation of amyotrophic lateral sclerosis (ALS) with a more prominent attack of the lower motor neuron. It presents with bilateral weakness and atrophy mostly proximally in the upper limbs, resulting in bilateral flail arms. Compared to classic ALS it progresses slower and swallowing difficulty and diaphragmatic weakness present later in the progression of the disease.

Case report

A 71 year old male presented to the NCS department with reported upper limb weakness that started 5 months prior. He reported previous chemotherapy treatments due to prostate cancer. He presented with bilateral arm weakness more prominent proximally (mostly torso and shoulder region), and very mild weakness in the lower limbs (ambulatory). Fasciculations were seen in the triceps muscles billatery. The deep tendon reflexes were absent in the upper limbs and no sensory deficits were found. The electoneurographic testing regarding the nerves: median, ulnar, peroneal and tibial was normal bilaterally. The electromyographic testing was conducted with a needle electrode and resulted in positive sharp waves, fibrilations and fasciculations at rest and reduced reqruitment in the supraspinatus, deltoid, triceps, bracoradilis, extensor capri radialis longus, abductor pollicis brevis, abductor digiti minimi, quadriceps, tibialis anterior, extensor digitorum brevis and gastrocnemius muscles bilaterally. More impacted were the upper limbs, and proximal muscles compared to distal in all four limbs.

Conclusion

It is necessary for a Physical and Rehabilitation Medicine physician to be aware of this rare variation of ALS, as its symmetrical presentation prominently affecting the upper limbs can pose a significant difficulty in the differential diagnosis.

Keywords: Vulpian-Bernhardt syndrome, electromyographic, fasciculations