EMG showed absent left sural sensory potential, fasciculation and evidence of chronic partial denervation in the arms. The diagnosis was confirmed with an abnormal CAG repeat of 44 within the XLBSN gene.

EMG of the anal sphincter showed evidence of reinnervation (Large motor units above 6mv).

The Sphincter EMG suggests the pathological role of Onufrowicz nucleus (ON). To our knowledge, this is the first reported case of Kennedy's disease genotype with bladder involvement.

174

BLADDER INVOLVEMENT IN KENNEDY'S DISEASE: A RARE ASSOCIATION

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Kennedy's disease, or spinal and bulbar muscular atrophy (SBMA), is an inherited X-linked degenerative disorder characterised by progressive proximal limb weakness, bulbar weakness, fasciculation, androgen insensitivity and characteristic electromyography findings.

A 44 year old Caucasian police officer presented with asymmetric sensory symptoms of pins and needles in both legs. In addition, he had dysphagia and history suggestive of intrinsic hand muscle weakness and lower urinary tract symptoms (LUTS) for 1 year. He had surgery for bilateral gynaecomastia. His maternal grandfather had been investigated for suspected multiple sclerosis and two brothers had surgery for gynaecomastia.

Examination revealed bifacial weakness and bilateral partial ptosis. There were fasciculation in the muscles of the shoulder girdle and upper limb with wasting of the small muscles of the hands bilaterally. In the lower limbs, there was wasting of the quadriceps muscles, clawing of toes and weakness of hip flexion.