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Case report

"Anterior interosseous nerve syndrome (Kiloh Nevin Syndrome) revealing Gantzer muscle and simultaneous myasthenia gravis"

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ABSTRACT

There hasn't been a previous case report of the anterior interosseous nerve injury secondary to the presence of the muscle of Gantzer in a patient with myasthenia gravis in literature before. The anterior interosseous nerve compressive syndrome, also known as Kiloh-Nevin syndrome, is a rare disorder comprising less than 1% of all upper limb neuropathies. Establishing the etiology of anterior interosseous nerve compressive syndrome is challenging because of the lack of specific clinical findings or testing. Herein is the case of a 46 years-old male presented with left eye ptosis, ophthalmoparesis, diplopia, and right-hand weakness. On physical examination, the Pinch Grip test was positive. Electromyography studies showed neurogenic atrophy in the muscles innervated by the anterior interosseous nerve, as well as a pathological decrement of the muscle action potential of more than 10% on repetitive nerve stimulation. Concluding that the presence of the Gantzer muscle caused anterior interosseous nerve compressive syndrome was mainly a diagnosis of exclusion, after careful consideration of other possible etiologies including carpal tunnel syndrome, cervical radiculopathy, and Parsonage-Turner Syndrome. Even though anterior interosseous nerve compressive syndrome is very rare, clinical suspicion ought to arise in the presence of weak radial flexor digitorum profundus and flexor pollicis longus muscles. This case highlights the importance of a thorough medical history, a meticulous physical examination, and par-

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ticularly the significance of electromyography studies in diagnosing different neuropathological entities. When appropriate, these steps offer information crucial to the differential diagnosis and eventual surgical management, assisting physicians in making informed and accurate treatment decisions.

Introduction

Anterior interosseous nerve syndrome or AINS is a rare, well-defined neuropathy affecting the anterior interosseous nerve (AIN). This AINS is characterized by diffuse pain in the forearm and cubital fossa, weakness of the index finger and thumb, and a positive Pinch Grip test [1]. By definition, a pure motor neuropathy, and patients do not present with any sensory changes or complaints [2]. The prevalence of AINS is estimated to be around 1% of all upper extremities palsies [1,2].

The AIN is the terminal motor branch of the median nerve. It innervates the deep muscles of the forearm, including flexor pollicis longus (FPL) muscle, the radial aspect of the flexor digitorum profundus (FDP) muscle, and the pronator quadratus muscle [10]. The AINS represents an isolated palsy of these 3 muscles. [10] Pathophysiology of AINS remains poorly understood despite the hypothesis of spontaneous (neuritis, compression neuropathy) and traumatic factors having been proposed as a possible etiology [1–3]. AINS may also occur due to neuritis secondary to a viral illness [1,3,4], entrapment or compression of the AIN [3,5–8], traumatic events including supracondylar fractures, penetrating wounds, venipuncture, and cast fixation. [2,9]

Patients present with motor deficits alone, with weakness or loss of function of FPL and FDP to the index and occasionally middle fingers, the pronator quadratus of the distal forearm, impaired flexion of the interphalangeal joint of the thumb and the distal interphalangeal joint of the index finger [1–3]. Consequently, on physical examination, they are unable to make the “Ok” sign and have a positive Pinch Grip test (holding the sheet of paper between an extended index finger and thumb, due to not being able to hold it with their fingertips) [1–3]. In time, patients may develop difficulties forming a fist, buttoning their shirts, sewing, turning on their car keys, as well as deteriorating handwriting [2,11]. Differential diagnoses to consider stenosing tenosynovitis, flexor tendon adherence or adhesion, flexor tendon rupture, and brachial plexus neuritis [1,10].

Suspicion of AINS ought to arise based on the clinical findings during the physical examination, principally a clinical presentation with motor deficits and a complete lack of sensory symptoms. Electrodiagnostic studies are critical in assessing the severity of neuropathy, distinguishing a compression neuropathy from neuritis, and assisting in the differential diagnosis [1–3]. Imaging modalities such as MRI reveal increased signal intensity in some or all AIN-innervated muscles [2,3].

Treatment of AINS remains controversial. Nevertheless, most authors and literature recommend adopting a conservative approach for a period extending from 8 to 12 weeks [2,3,12]. This conservative management consists of a period of rest, avoidance of exacerbating activities, physiotherapy informed by the severity of pain and accompanying symptoms, as well as the use of medication (NSAID, analgesics) [2,3,13]. If there are no clinical or electrophysiological improvements and no signs of spontaneous recovery for more than 12 weeks, surgical exploration is called for with neurolysis and surgical decompression offering excellent outcomes, particularly in the light of an identifiable, space-occupying mass [2,3,13]. Rigorous dissection and visualization of AIN are required to identify the sites of compression, in addition to careful hemostasis to prevent a postoperative hematoma [3].

Case report

A 46 years old male presented with a 9-year history of left eye ptosis, ophthalmoparesis, diplopia and right hand weakness. His symptoms had progressively worsened over the past 2 years. His neurological examination was significant for left eye ptosis and ophthalmoparesis, as well as weakness of the distal thumb phalanx, middle index finger phalanx, and right hand pronation. The patient was unable to perform the “Ok” sign using his thumb and index finger (Fig. 1).

High clinical suspicion arose for Myasthenia Gravis based on the patient’s clinical presentation and Osserman’s score of 85 points. The diagnosis was subsequently established after a positive Tensilon test, electromyography (EMG) studies showing a pathological decrement of the muscle action potential of more than 10% on repetitive nerve stimulation between the 1st and 4th response (Fig. 2), and blood tests confirming the presence of anti-acetylcholine antibodies.

EMG of the muscles of the forearm showed neurogenic atrophy in the muscles innervated by the AIN, more evidence showing pathological repetitive stimulation decreases (> 10%) in different territories, thus revealing the concomitant presence of both conditions, Myasthenia Gravis and a nerve compression syndrome.

Further imaging modalities revealed the presence of this anatomical variant compressing the AIN, thereby causing the right-hand weakness. MRI of the forearm showed the accessory head of FLP muscle, the Gantzer muscle (Fig. 3). Surgical exploration allowed for a definitive diagnosis, displaying the nerve compression caused by the Gantzer muscle, in addition to enabling the complete surgical treatment of the condition (Fig. 2).

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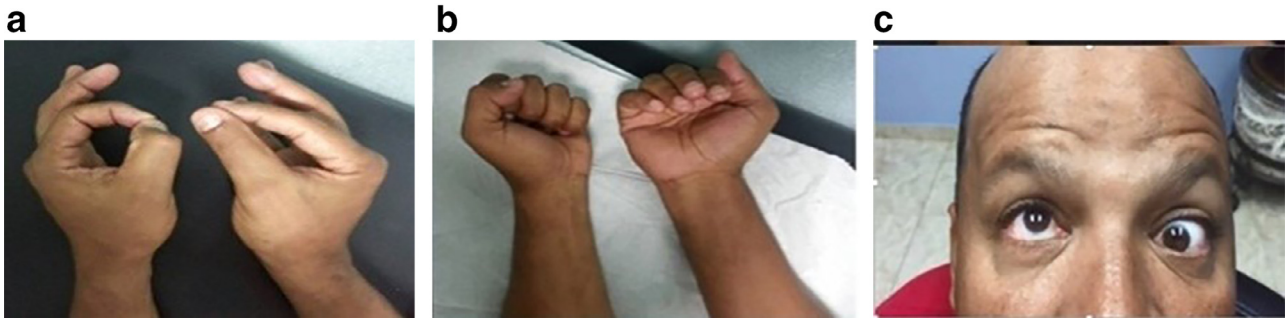

Fig. 1 – (A) canʹ demonstrate the "Ok" sign with the right hand (B) limitation in the right hand closure (C) limitation in vertical left gaze (D) left ptosis.

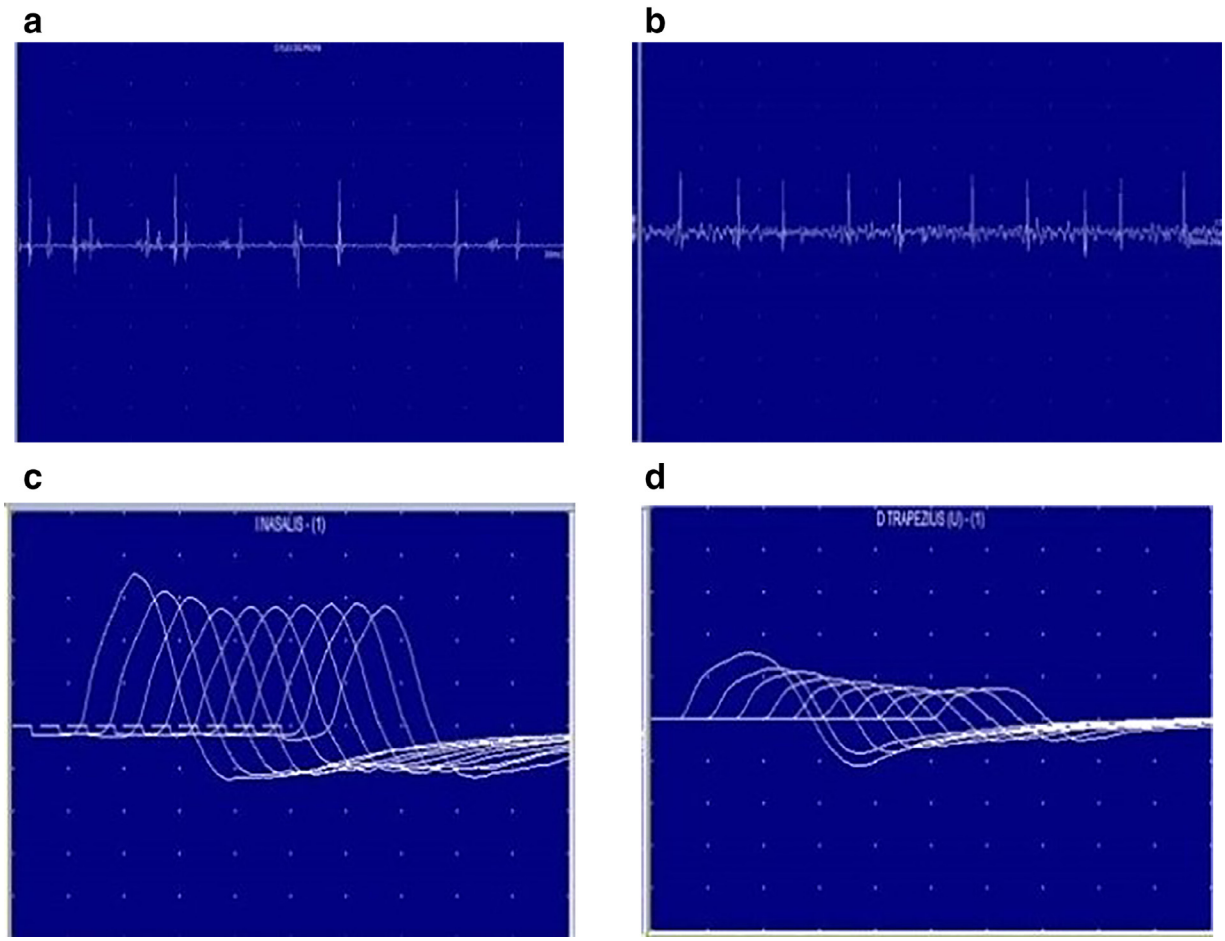

Fig. 2 – (A) Digitorium deep flexor muscle (neurogenic atrophy) (B) Pollicis longus flexor right muscle (C) Pathological decrement of 22 % between 1st and 4th response of nasalis muscle (D) Pathological decrement of 37% between the 1st and 4th response of couple spinal nerve and trapezius muscle

sory head of FLP muscle, the Gantzer muscle (Fig. 3). Surgical exploration allowed for a definitive diagnosis, displaying the nerve compression caused by the Gantzer muscle, in addition to enabling the complete surgical treatment of the condition.

Pre-treatment and post-treatment changes

Ultimately, it was concluded that weakness of the external ocular muscles responsible for the ocular manifestations re-

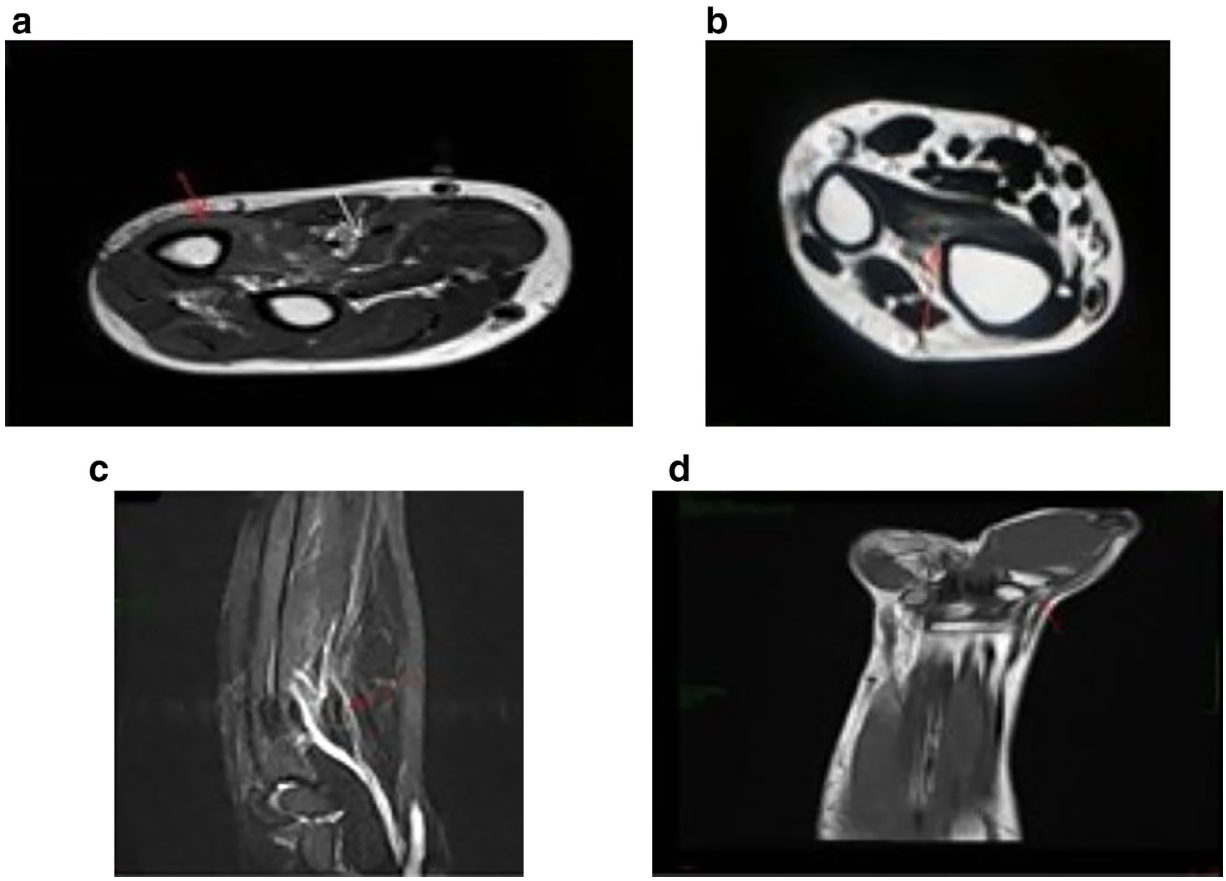

Fig. 3 – (A) Cut Axial T2 image. Increased signal (edema; red arrow) deep flexor digitorum muscle is evident; white arrow shows accessory muscle belly or Gantzer muscle (B) Axial T2 Court: shows edema square pronator (C) Sagittal STIR: Evidence of accessory Pollicis head flexor Longus (Gantzer muscle) at the interosseous membrane (red arrow) (D) Coronal T1: sample insertion point accessory muscle (Gantzer) in the carpus.

sulted from the underlying Myasthenia Gravis, whereas the compression of the AIN prompted the right-hand weakness. The patient was started on pyridostigmine and prednisone for his myasthenia with significant improvement, in addition to undergoing a surgical decompression of the nerve resulting in full recovery (Fig. 4)

Discussion

Determining the precise etiology of the AINS poses a severe challenge. Numerous explanations have been put forward as potential etiological factors, generally classified as spontaneous or traumatic [1–3,10]. Kiloh and Nevin initially proposed AINS arising secondary to transient neuritis as a potential cause, whereas Fearn and Goodfellow suggested it was a compressive neuropathy, with both theories remaining accepted [14–16]. Traumatic events are also a well-recognized etiology [2,9]. As the AIN runs deeply along the interosseous membrane, isolated lesions to it are rare [17]. However, these same structures can at times become sources of compression [17]. Moreover, the AIN is prone to entrapment by anatomical variants such as the Gantzer muscle, as was seen in our patient [2,18].

The Gantzer's muscle is an accessory head of the flexor FPL muscle or the FDP muscle, first described by Gantzer in 1813. [19] The incidence is reported to be approximately 43%-68% [16].

According to data from a number of studies, the origin of the Gantzer muscle may be the medial epicondyle of humerus, the ulnar coronoid process, and the deep surface of the flexor digitorum superficialis [11,16]. There have also been reports of an origin in the flexion-pronator muscle group [20]. The insertion point has been mostly reported in various levels of the FPL tendon [11,16]. There have been reports of an insertion in the FDP as well as the presence of two or more tendons arising from the Gantzer muscle [16,20–22].

Several studies show Gantzer muscle is more frequently bilaterally than unilaterally [16,23–25]. From an embryologic perspective, the flexor muscle group develops during the seventh week of gestation, from the flexor mass of the arm buds that are eventually divided to give rise to the superficial and deep muscle layers [11,26]. It has been hypothesized that

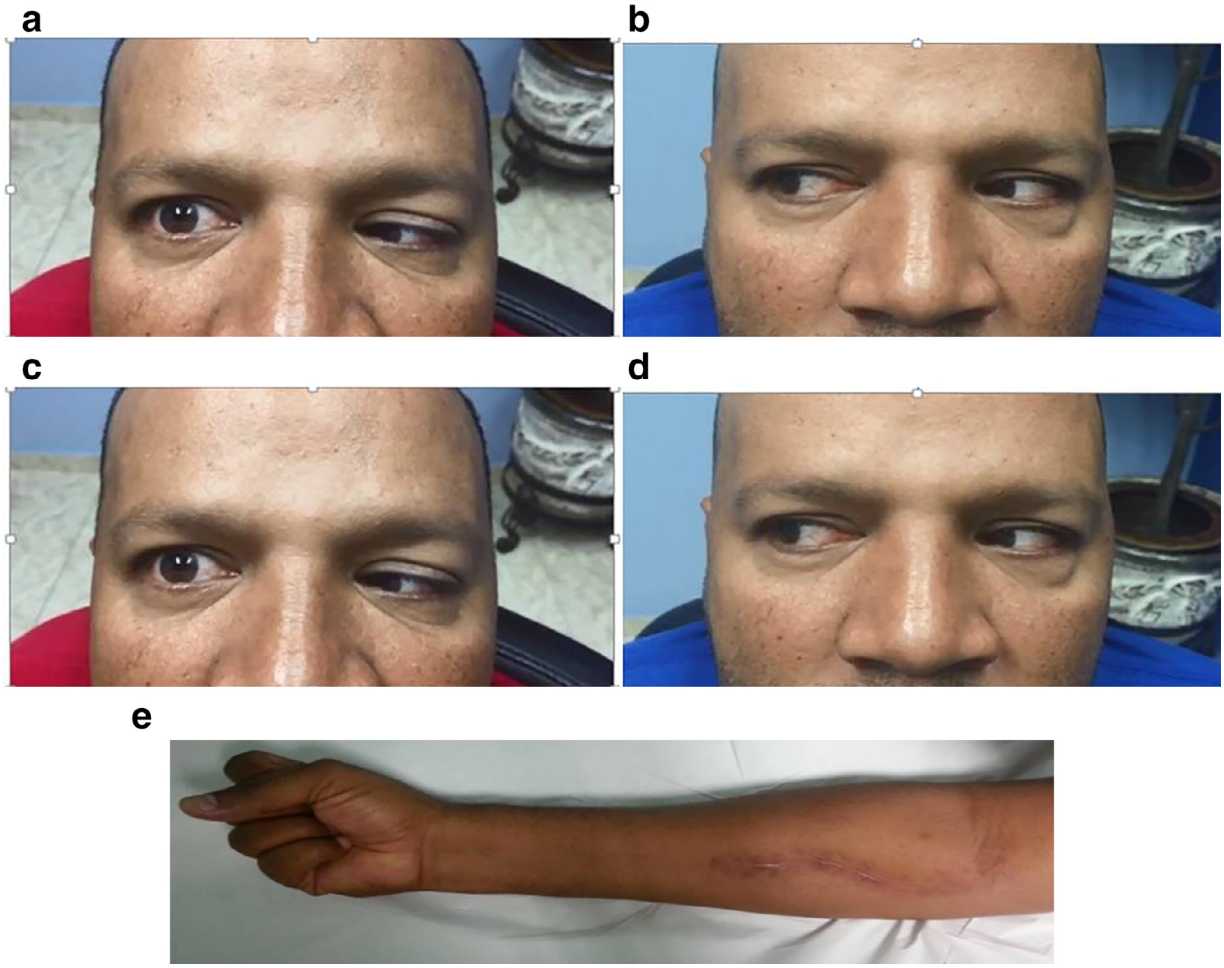

Fig. 4 – (A,B) pre-treatment: left eye ptosis and limited right eye movement when is turned toward right side. Right lateral gaze palsy (C,D) post treatment: Right lateral gaze is normal and improvement of ptosis (E) The patient can close the right hand correctly after the surgical decompression of anterior interosseous nerve, surgical scar is evident.

incomplete delamination during the embryological development may be responsible for the presence of this accessory muscle [11].

Gantzer muscle is predominantly innervated by AIN alone, as documented by a large number of studies. However, cases of a double innervation from the AIN and the median nerve as well as innervation from the median nerve have also been reported [16,22]. The relationship between the AIN and the Gantzer muscle has been extensively studied, with reports documenting the location of AIN both anteriorly and posteriorly of the Gantzer muscle [11,16,25]. A hypertrophic Gantzer muscle situated anteriorly to the AIN may result in compression and entrapment of the nerve, causing either a complete or incomplete AINS [8,11,16].

It is worth noting that our patient presented with both an AINS and myasthenia gravis. The concomitant presence of these two distinct neurological disorders, to the best of our knowledge, has not been reported before.

Conclusion

AINS or Kiloh Nevin syndrome, secondary to an anatomical variant muscle such as the Gantzer muscle, is a rare disorder that poses a great diagnostic challenge. Knowledge of the structural and possible morphological variations of the muscles of the forearm can help identify other possible unusual causes of nerve compression injuries. This case report underscores the importance of a thorough medical history, a meticulous physical examination, and electrodiagnostic studies like EMG in guiding the diagnosis of diverse neurological disorders. These steps offer information crucial to the differential diagnosis, aiding physicians in making informed decisions regarding the best treatment course on a case to case basis. To the best of our knowledge, this is the first report of the occurrence of an injury to the AIN secondary to the presence of the Gantzer muscle in a patient with an accompanying underlying neurological condition such as Myasthenia Gravis.

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