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Case Report

A Rare case of Ulnar Nerve Intraneural Perineurioma in an elderly gentleman ☆,☆☆

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ABSTRACT

Intraneural perineurioma (IPN) is a rare benign neoplasm of the peripheral nerve sheath, most commonly affecting the sciatic nerve of adolescents or young adults. We present a rare case of perineurioma in a 67-year-old man with an infiltrative clinical presentation affecting his ulnar nerve. Complete excision required *en-bloc* resection of the ulnar neurovascular bundle from mid-forearm to mid-palm with sural nerve and saphenous vein grafts used to reconstruct the ulnar nerve and artery, respectively. Despite recurrence from a previously localized excision, there has been no recurrence to date following *en-bloc* resection. The available literature on intraneural perineurioma is reviewed in light of this case report. IPN is an important entity because of its ability to mimic neural malignancies, and this case challenges the assertion that it is a tumour that occurs strictly in the young.

Background

Peripheral nerve tumours are predominantly benign, and while they can cause sensory or motor deficits secondary to compression, it is more common that they are associated with minimal

☆ The corresponding author is not a recipient of a research scholarship.
☆☆ We have included four figures in this case report.

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nerve dysfunction. Benign lesions may be removed from the affected nerve through careful surgery if surveillance is not appropriate and is associated with low recurrence rates and minimal intraoperative nerve injury depending on the nature of the lesion. Malignant peripheral nerve tumours are associated with greater nerve dysfunction due to direct infiltration, including both sensory and motor deficits in mixed nerves. Imaging, nerve conduction studies (with electromyography as appropriate) and nerve biopsy are all useful adjuncts to help the treating clinician distinguish between benign and malignant lesions. En-bloc excision of malignant lesions is the mainstay of surgical treatment, but it causes complete interruption of nerve function and often necessitates reconstruction of the entire segment of the affected nerve using nerve grafts. The nerve may need to be resected along with associated arteries and veins as a neurovascular bundle resection with interposition vascular grafts as appropriate. Early recurrence is a prominent feature of malignant lesions that have only had marginal excision.

Perineurioma is an uncommon benign peripheral nerve sheath tumour that almost exclusively presents in early to mid-adulthood. While initially considered to be the result of a reactive process, molecular analysis has recategorized these lesions as neoplastic with a BCR locus deletion (22q11) present in 75% of nuclei.[1] Perineuriomas are subdivided into intraneural and extraneural forms on pathology.[2]

Extraneural perineurioma is not associated with a peripheral nerve and is found mainly in the skin and soft tissues.[3] Intraneural perineurioma is a benign tumour of perineurial cells within nerves, making it more difficult to distinguish from malignant nerve tumours. IPN have been described in case reports involving single and multiple peripheral nerves including sciatic, femoral, peroneal, tibial, brachial plexus, ulnar, median, radial, facial, mandibular dental, oculomotor, tongue, C8/T1, and jugulocarotid region of the neck and laryngeal.[3-7] Until recently, the diagnosis of intraneural perineuriomas has been complicated by the inability to differentiate between lesions of Schwann cells (forming real onion bulb whorls) and perineural cells (forming pseudo-onion bulbs). This has contributed to misdiagnosis and the use of multiple different names for intraneural perineurioma in the literature including localized hypertrophic mononeuropathy (LHM), interstitial hypertrophic neuropathy, pseudo-onion bulb neuropathy, intraneural neurofibroma, hypertrophic neurofibrosis, and hypertrophic interstitial neuritis. The advent of epithelial membrane antigen (EMA) immuno-cytochemistry provided the ability to distinguish between Schwann cell and perineural cell origins, thereby permitting appropriate diagnosis of intraneural perineuriomas.[8, 9] While the literature describes a slight predilection for IPN in females over males, the most recent systematic review by Uerschels et al., 2020, of 22 articles including case series and individual case reports included ten ulnar nerve lesions (13%, n=77) and average age 22.5 years (5-37) with a slight predilection for males (6:4). The largest case series of 32 patients described four ulnar nerves in young females aged between 13 and 35 years.[8] To our knowledge, there have been no reported cases of ulnar nerve IPN in elderly male patients.

Clinical Case

A 67-year-old right-hand dominant man was referred to our centre by an orthopaedic colleague for the proposed resection of ulnar nerve mass and reconstruction with a sural nerve graft. Prior to the presentation, he noticed pain and sensory loss in little and ring fingers as well as weakness in grip. Eleven months prior, he had surgery to remove the tumour from his ulnar nerve under frozen section control with cubital tunnel and Guyon's canal releases. The histopathology showed benign lipomatosis of ulnar nerve. The patient's symptoms did not improve and the mass recurred. The examination has revealed mild ulnar claw, severe intrinsic muscle wasting, and a soft fluctuant non-tender mass over a well-healed scar at the wrist (Figure 1 and Figure 2). He had no sensation in ulnar nerve distribution. His grip strength was preserved in keeping with the level of the lesion, but he had almost no intrinsic function with 1/5 power in the 1st dorsal interosseous and 0/5 in abductor digiti minimi.

Investigations

MRI confirmed a hyperintense dilated ulnar nerve segment of 11 cm from the distal forearm to hand just beyond Guyon's canal (Figure 3).

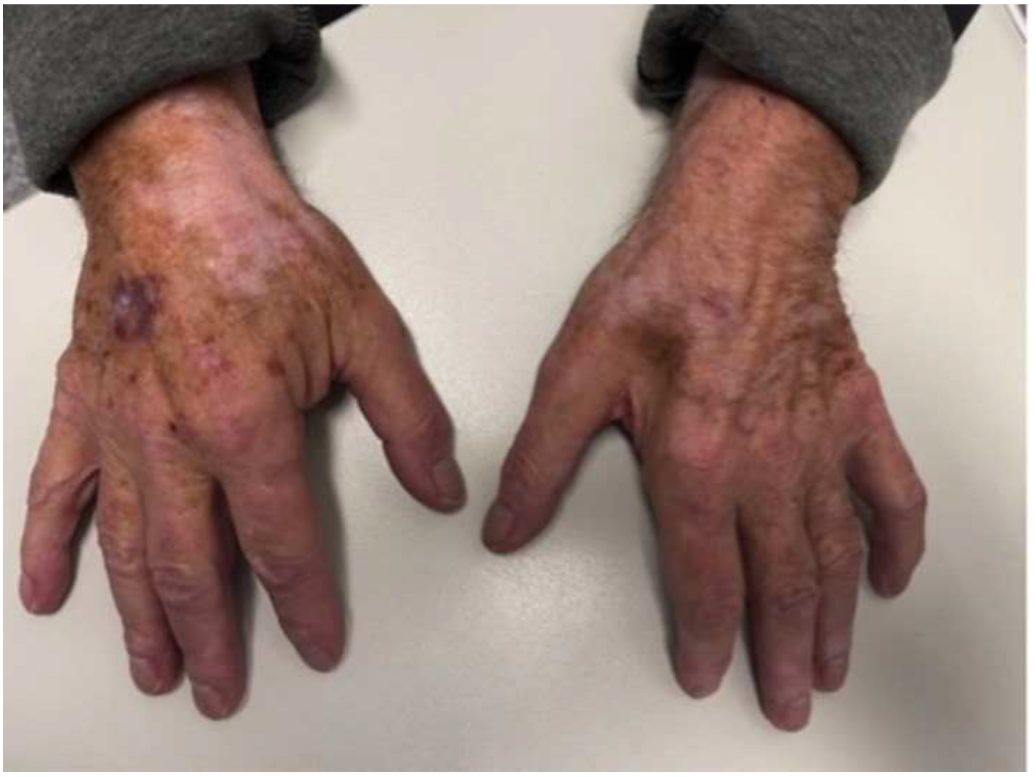

Figure 1. Severe intrinsic muscle wasting of the left hand

Nerve conduction studies confirmed neurophysiological evidence of a severe left ulnar neuropathy at the level of the wrist and forearm, with only two motor units in the first dorsal interosseous and none in abductor digiti minimi.

Presumed Diagnosis

The clinical picture of early recurrence and worsening of sensory function in an older patient was clinically consistent with a malignant or locally aggressive tumour affecting the peripheral nerve.

Surgical technique

He underwent excision of ulnar nerve mass. Intraoperatively, it was found that the tumour mass enveloped the ulnar artery, which had to be sacrificed leaving a 13 cm defect in both nerve and artery from distal forearm to proximal hand. His reconstruction included left sural nerve graft of 32 cm and reverse saphenous venous graft of 15 cm. We used single cable of sural nerve for sensory branches of ring and little fingers and single cable for motor branch. Ulnar nerve coaptation was with 8/0 nylon at both proximal and distal ends. We repaired artery using 8/0 nylon. A protective dorsal splint was applied for two weeks, followed by the on-going management with splintage and hand therapy to counteract mild ulnar clawing.

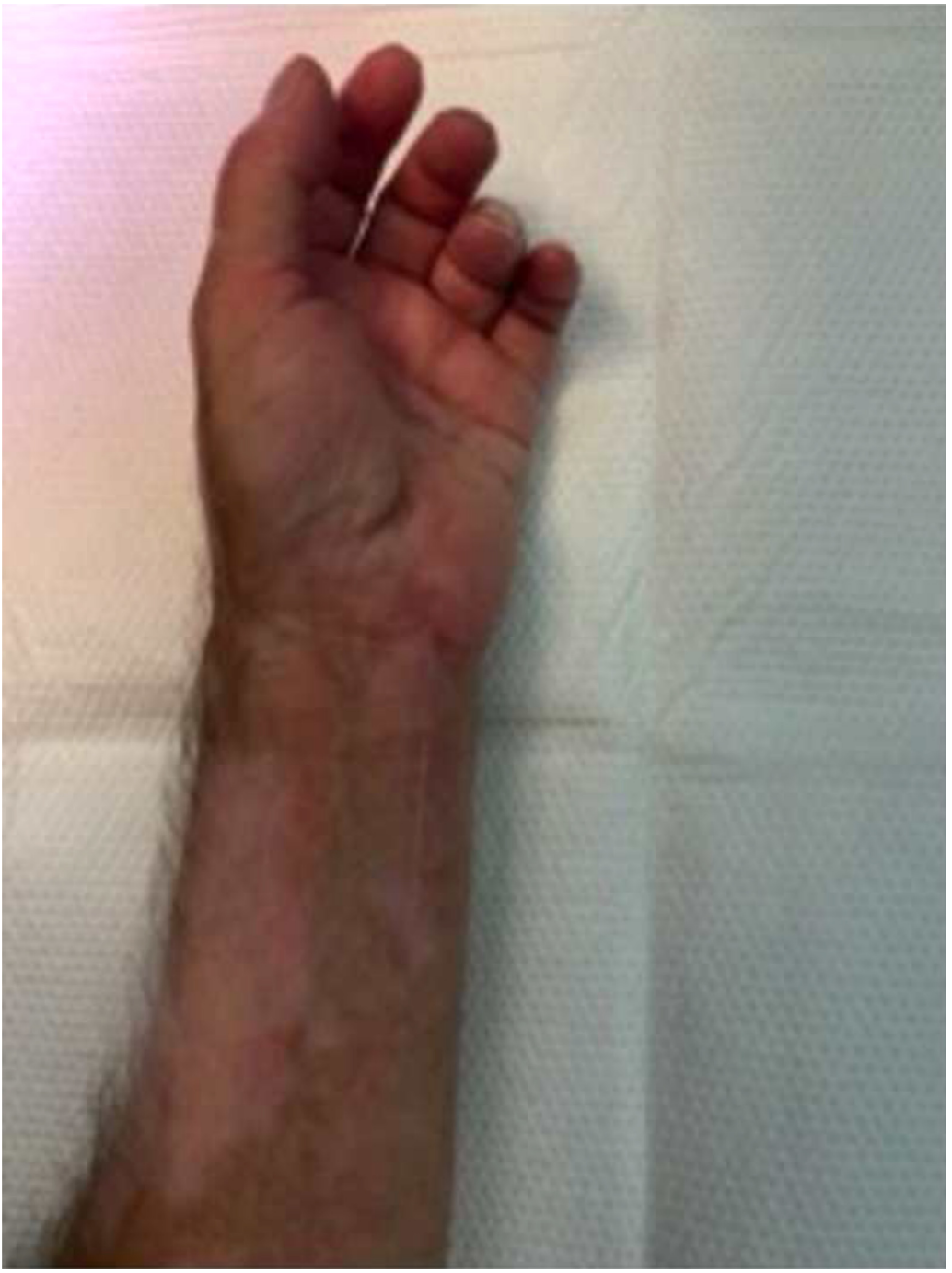

Figure 2. Ulnar clawing

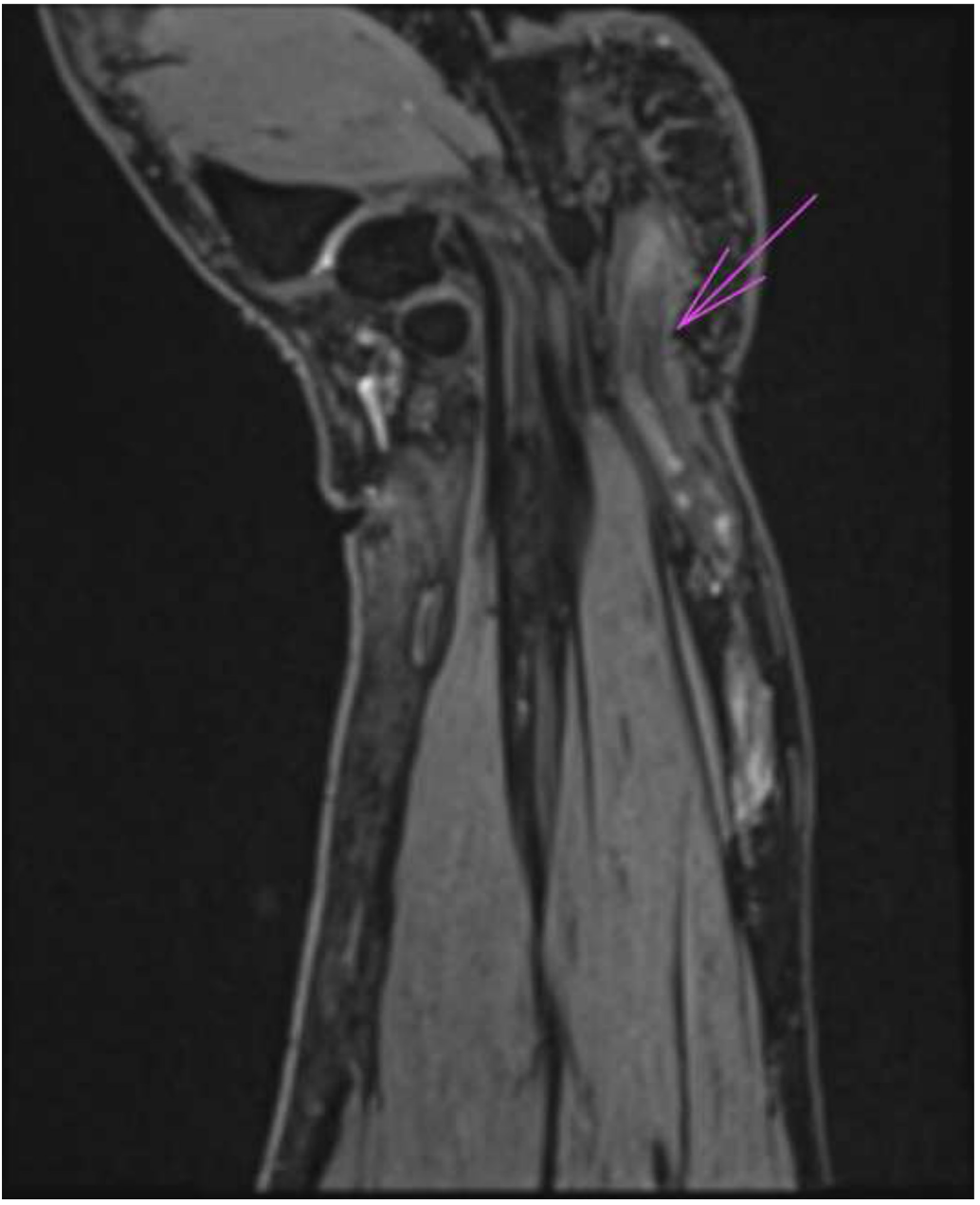

Figure 3. MRI of upper limb showing thickened ulnar nerve passing through Guyon's canal (arrow)

Histopathological diagnosis

Both frozen section and resection specimen confirmed the diagnosis of fibrolipomatous hamartoma of nerve. However, the involvement of ulnar artery and our concern for malignancy prompted us to seek a secondary opinion from a quaternary cancer centre, which confirmed a diagnosis of IPN.

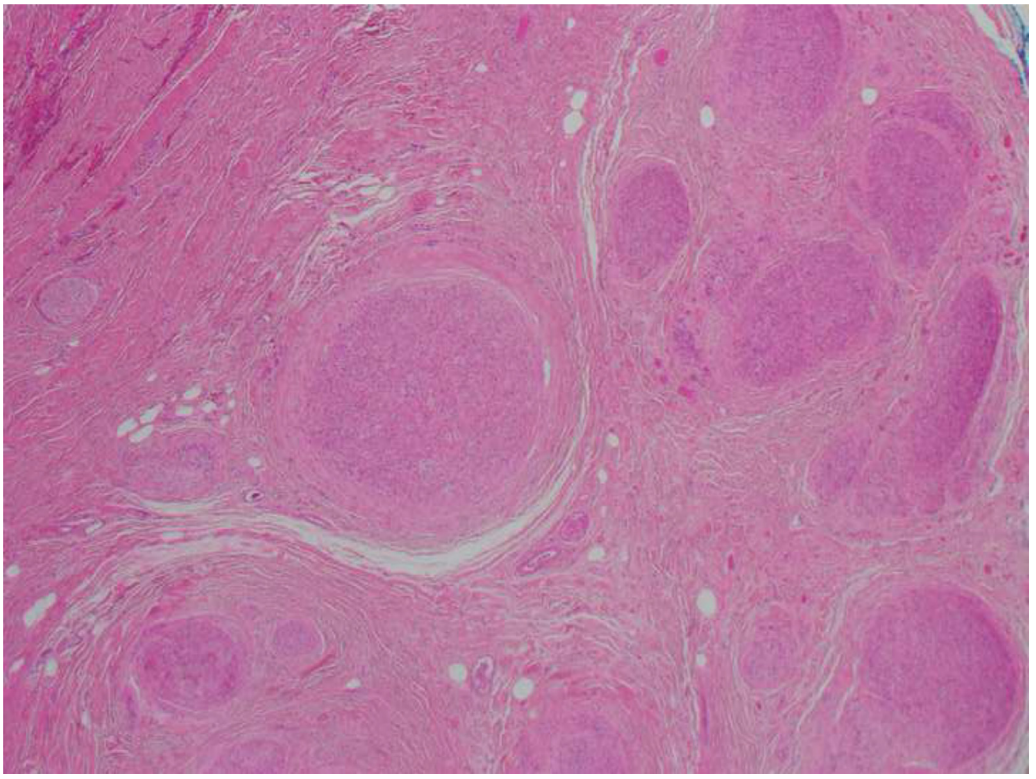

Figure 4. Enlarged nerve fascicles surrounded by perineural cells arranged in pseudo-onion bulb formation

Histopathology showed multiple neural bundles with an expanded perineurium (Figure 4). Some of the nerve bundles had an onion bulb appearance with the outer cells EMA positive and inner axons S100 positive.

Discussion

IPN is a rare benign tumour of peripheral nerve sheath originating from perineural cells. The perineurium is mesenchyme-derived and surrounds nerve fascicles in concentric layers modulating traction forces, regulating the endoneurial pressure, and being critical in the blood-nerve barrier.[9]

IPNs typically present in adolescence or early adult life as painless mononeuropathy with progressive weakness. Our case demonstrates even a more rare presentation of ulnar nerve IPN in an elderly patient, which was never previously described in the literature to our best knowledge. By the time, these lesions are referred to a surgeon that they are often thought to be compressive lesions and are often commonly misdiagnosed.

Imaging with ultrasound is recommended as a first-line modality where entrapment neuropathy or peripheral nerve tumour is suspect to localize and diagnose a lesion.[9] MRI shows a typically fusiform swelling of nerve fascicles with isointensity on T1 and hyperintensity on T2 with frequently a 'target sign' showing hyperintense periphery compared to the centre of lesion.[2]

Modern treatment of perineuriomas remains controversial without a general consensus. Surgery is recommended in patients with well-localized lesions and good history of progressive mononeuropathy. The authors of the second-largest case series recommend excision of lesion with interposition nerve graft if there is intraoperative evidence of no or poor action potentials.[10] They reported motor improvement in seven out of ten patients with grafts, and only some improvement in two out of four

patients without grafts. Uerschels et al. recommended that extensive nerve resection with grafting should only be considered in distal lesions without muscle atrophy, as it may worsen disability and pain in proximal lesions.[2] As an alternative, Sachanandani et al. recommended distal nerve transfer to decrease regeneration length and eliminate secondary operative site.[9]

However, according to the systematic review of all surgical treatment options, the neurological outcome did not differ.[2] The authors have suggested that there may a shift from total resection towards a wait-and-see approach following the targeted fascicular biopsy.

Conclusion

Intraneural perineurioma is a rare benign peripheral nerve tumour that is capable of mimicking the clinical appearance of malignant nerve tumours. Existing wisdom states that these lesions occur only in young and have a predilection for females. The gender disparity has been challenged by a recent meta-analysis of existing case series and we present the first case of intraneural perineurioma in an elderly patient to demonstrate that it can also occur in the elderly. It is anticipated that IPN will be found more often in older patients as our population's age, as knowledge of these lesions becomes more widespread and as improved diagnostic techniques become more mainstream.

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Ethics

Not required. This report was published with the consent and permission of the patient involved.

Conflict of interest statement

None.

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