

Intraneural perineurioma of the median nerve

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ABSTRACT

Perineuriomas present as focal intraneural peripheral nerve or isolated soft tissue lesions. In this case report, we focus on the intraneural variant of perineurioma. Intraneural perineurioma which generally presents as a slowly progressive painless focal motor neuropathy characterized slowly histologically by onion-bulb shaped whorls, was first described by Imaginario et al. (1) in 1964. The etiology of these lesions has been controversial with some authors asserting that this lesion is a reactive process associated with trauma and other proposing that the lesion is a true neoplasm (2-6). Thus the lesion has been labeled with different names including 'localized hypertrophic mononeuropathy' and 'perineurioma' (2, 7-11). The evidence relating these lesions to trauma has been scarce, and both intraneural and extraneural variants are now believed to represent clonal neoplasms associated with abnormalities of chromosome 22 (5,12). However, in recent literature the lesion continues to be referred to interchangeably as perineurioma and localized hypertrophic mononeuropathy (7,13).

We present a case of a median nerve perineurioma which has caused carpal tunnel syndrome and treated by surgical procedures. [Turk J Cancer 2009;39(4):158-160]

CASE REPORT

A forty-two year-old housewife who was right handed dominant was seen because of progressive pain in the right wrist that began two years earlier and included pain at night that routinely interrupted her sleep. She also suffered from wasting of thenar muscles and numbness in the distribution of the right median nerve. She had no known history of trauma.

On physical examination there was an elliptical firm mass on the volar aspect of his right wrist, measuring approximately 2x2 cm. The overlying skin was without abnormalities. Sensory examination showed paresthesias in the distribution of median nerve with positive Phalen and Tinel's sign.

Magnetic resonance imaging of the right wrist showed a heterogeneous mass which appeared well circumscribed. Nerve conduction velocities were consistent with severe carpal tunnel syndrome. Motor and sensory latencies were 5.8 msec and 5.2 msec, respectively.

The patient was taken to the operating room for releasing carpal tunnel and incisional biopsy of the mass. Intraoperatively we identified an abnormal mass extending approximately 2 cm within median nerve just proximal to the motor branch (Figure 1). Abnormal mass was defined

visually with the same morphological character of median nerve. Biopsy of the mass and carpal tunnel release were performed.

The tissue sections were stained hematoxylin and eosin (Figure 2) and immuno peroxidase stains for epithelial membrane antigen, S-100 and P53. Histologic examination showed multiple nodules of various size surrounded by collagen loose fibrous connective tissue linked the nodules. Within each nodule were numerous onion-bulb structures having central clear area surrounded by concentric layers of eosinophilic elongate cells having spindled nuclei. The area adjacent to the central clear area was strongly positive for S-100 protein. The next outer layer, composed of spindled eosinophilic cells, was weakly moderately positive by epithelial membrane antigen stain.

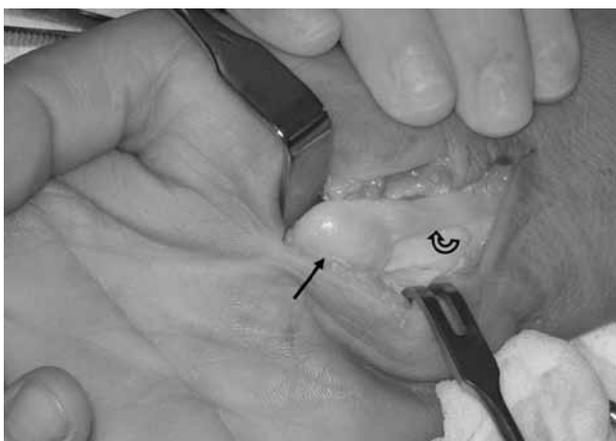


Fig 1. An intraoperative photograph revealing a 2-centimeter-diameter mass (arrow) in continuity with the median nerve (curved arrow)

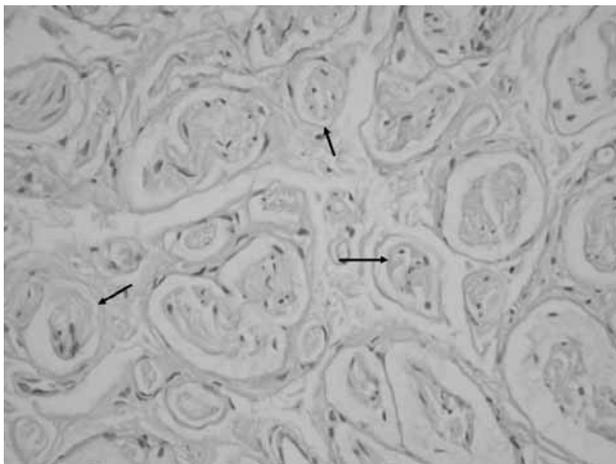


Fig 2. Photomicrograph of perineurioma showing several nerve fascicles involved by collagen separating the nerve fascicles (arrows). (Hematoxylin & eosin stain; original magnification x 50)

P53 stain was negative in all nuclei. These changes were characteristic of intraneural perineurioma.

There were no postoperative complications. Sixteen months postoperatively we have sustained clinical regression at preoperative symptoms due to the carpal tunnel syndrome. There is no clinical and radiological evidence of progression at the mass volume.

DISCUSSION

To date more than 50 cases of intraneural perineurioma have been documented by histological analysis and in most immunoreactivity has also been demonstrated with epithelial membrane antigen positivity and S-100 protein negativity (7,8, 13-16). The literature is divided as to whether this lesion is truly neoplastic or represents an inflammatory process caused by repeated trauma and resultant loss of integrity of the normal perineural barrier. Using tissue culture, cytogenetics and fluorescent in situ hybridization, Emory and colleagues (5) demonstrated that the intraneural perineurioma represents a neoplasm consistent with loss of gene on chromosome 22, similar to findings in other peripheral nerve tumors and intracranial meningiomas. Because of genetic evidence, those authors argued that the most descriptive and accurate term for this lesion is 'intraneural perineurioma' however, in their recent study, Gruen et al. (7) referred to this lesion as a localized hypertrophic neuropathy, despite their argument that is rare to find history consistent with reactive change. Authors of other recent reports have also referred to this lesion as localized hypertrophic neuropathy and perineurioma interchangeably (13).

Grossly, the affected nerve shows a characteristic fusiform enlargement with thickening and adhesions involving the perineural elements. This finding is in contrast to a schwannoma, which can be 'shelled out' from the involved nerve. Light microscopy of the lesion shows onion-bulb formation of nerve fascicles. This finding is thought to be caused by schwann cell hyperplasia (7). Results of immunohistochemistry studies of concentric are positive for epithelial membrane antigen and negative for S-100 protein. Results of central myelin sheath immunohistochemistry studies are negative for epithelial membrane antigen but positive for S-100 protein.

Because of the small number of case reports no consensus has been made about proper management of these lesions. There was no recurrence of lesions after excision. However, in the case reports in which excision of lesions performed, return of sensory nerve function was not seen, even with interpositional nerve grafts (4,14,17). Conservative management has been advocated, with incisional biopsy of lesion for diagnosis. Excision of the mass has been recommended if the lesion can be easily dissected from involved nerve (5). Additionally MR neurography

can be used to evaluate decision of intraneural microscopic resection with preservation of surrounding fascicles or a segmental resection with placement of nerve graft (13). In our patient, we performed incisional biopsy and carpal tunnel release. Furthermore, because of the rarity of this clinical entity, intraoperative diagnosis is not possible in most centers. Therefore, we believe that each case needs to be individualized until more information is available regarding this tumor.

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