Multiple Plexiform Neurofibroma of the Hand Misdiagnosed as Ganglion Cyst

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A healthy 28-year-old man presented to our clinic, complaining of a slowly growing mass in the thenar eminence of his right dominant hand.

On examination, a 1 × 1 cm mass was palpable and sensitive to touch. Sensation and motor function were normal, with no numbness or paresthesia. Ultrasound showed a 2 × 1 × 1 cm cystic lesion, which was diagnosed as a ganglion cyst. Magnetic resonance imaging (MRI) demonstrated a 2 × 1 × 1 cm lesion, protruding from the carpal canal and partially adherent to the median nerve. The lesion showed a low-to-medium signal on the T1-weighted image, only peripherally enhanced by gadolinium, and a medium-to-high signal on the T2-weighted image. Again, the lesion was diagnosed as a ganglion cyst.

During the investigation period, the patient observed a new subcutaneous mass on the ulnar aspect of the middle and distal phalanges of the index finger. This mass was also sensitive to touch and appeared superficially similar to the previous lesion.

Due to the additional lesion and irregular location of the original mass, the diagnosis of a ganglion cyst was questioned, and the patient was operated for exploration and excision of both lesions. The carpal tunnel was released for proximal isolation of the median nerve. A segmental thickening of a branch of the palmar cutaneous branch of the median nerve was found in addition to a second smaller enlarged mass comprising the distal palmar segment of the ulnar digital nerve of the index finger (► **Fig. 1**). Both were merged with the proximal nerve, and separation of the mass from the nerve was unsuccessful, thus both were excised. Histologically, the lesions exhibited a wavy pattern of mixed Schwann cells and fibroblasts, and bundles of collagen, with positive S-100 staining, consistent with plexiform neurofibroma. There was no mitotic activity, atypia, or pleomorphism.

Two years later, palmar sensation was normal, and ulnar-sided sensation of the index finger was decreased. Motor

function was unaffected. Notably, there were no signs of café au lait spots on physical examination, there is a lack of family history of neurofibromatosis, and genetic tests for neurofibromatosis were negative. The patient declined any further intervention for his hypoesthetic index.

Plexiform neurofibromas are peripheral nerve sheath tumors composed of diffuse, elongated fibromas wrapped around multiple fascicles, producing a distorted and thickened nervous structure. While considered pathognomonic of neurofibromatosis type 1, unrelated tumors have been described.¹ Segmental neurofibromatosis, a rare form in which manifestations are limited to one area of the body due to somatic mosaicism, may partially explain these sporadic cases. Most cases are present since birth, but grow slowly and go unnoticed for years. Hand involvement is uncommon,² especially involvement of several branches of a single nerve.

Clinically and radiologically, neurofibromas are indistinguishable from neurilemmomas, and both may be misdiagnosed as another soft tissue tumor, such as a giant cell tumor of tendon sheath, or a ganglion cyst. Accounts in the literature show a high degree of initial incorrect diagnosis, as high as 96%,³ often due to lack of signs associated with nervous lesions, such as Tinel's sign or local numbness. Additionally, peripheral nerve sheath tumors and ganglion cysts share similar cystic appearances on ultrasound, and exhibit low-to-medium T1 and medium-to-high T2 signals on MRI.⁴ During surgery, ganglion cysts will be found tangent to the nerve, neurilemmomas will surround the nerve fibers and be easily detached, and a neurofibroma will often be inseparable from the nerve fascicles and require en block resection.

This patient presented with multiple plexiform neurofibromas arising from the median nerve in the palm, with no signs of neurofibromatosis. We recommend a high index of suspicion when approaching supposed ganglion cysts in unusual locations.

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Fig. 1 The tumor of the palmar cutaneous branch. The superficial arterial arch is tagged with a vessel loop (A). The ulnar digital nerve of the second finger infiltrated by the tumor (B).

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