Solitary Myxoid Neurofibroma of the Palm

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Dear sir

A 21-year-old female presented with a four-month history of a gradually-enlarging mass of her non-dominant left palm. She denied pain at rest, but noted some discomfort when firm pressure was applied to the area. She also noted mild tingling in the middle and ring fingers. The patient had no history of trauma or prior surgery to the area. Past medical history was unremarkable.

On physical exam there was a nodular mass palpable at the base of the middle and ring fingers at the level of the distal palmar crease, measuring approximately one x one centimeters (cm). Sensation to light touch and capillary refill were normal throughout. She had full wrist and digital range of motion. An MRI of the left hand demonstrated a homogenous cylindrical mass measuring $1.9 \times 0.7 \times 0.8$ cm, that was hypo- and hyper-intense on T1- and T2-weighted imaging, respectively.

The patient underwent surgical excision under tourniquet control, with intravenous sedation and local anesthetic. The mass was seen tenting the common sensory nerve to the ring and middle fingers. The nerve was mobilized and neurolysed from the underlying mass, which was subsequently excised from the surrounding soft tissues. After hemostasis of feeder vessels was achieved, the skin was closed. Gross examination of the specimen revealed a pale yellow well-demarcated,

elongated mass measuring $2.0 \times 0.7 \times 0.7$ cm. (Fig. 1). Histologic analysis demonstrated a benign myxoid spindle cell neoplasm consistent with myxoid neurofibroma (MN). The lesion was negative for atypia and malignancy (negative S-100, CD34 and CD57 staining). At her first follow-up visit, the patient noted improvement in her tingling symptoms.

Neurofibromas are benign, heterogeneous tumors arising from the connective tissue of peripheral nerve sheath [1]. While multiple neurofibromas are characteristic of Neurofibromatosis 1 (NF1), solitary lesions can present as a number of different variants. Myxoid Neurofibroma is a relatively uncommon variant of solitary neurofibroma characterized histologically by abundant myxoid matrix and small spindle cells with variably tapering nuclei, condensed chromatin, inconspicuous nucleoli and scant cytoplasm [1]. MN typically occurs in younger adults with a predilection to the face, shoulders, arms and periungual regions [1, 2].

Though benign, MN can pose clinical complications as they enlarge and compress nearby structures. There is one report of a “giant” MN in the forearm of an elderly male that eroded into the radius and ulna, resulting in limited elbow motion and wrist drop, ultimately necessitating above-elbow amputation [3]. Though such sequelae are not typical of MN, they illustrate the potential for problems resulting from encroachment into adjacent structures. In our presented case, the patient did have mild clinical signs of compression of the sensory nerve which was found to be displaced by the tumor intraoperatively. Unchecked progression of the mass possibly may have lead to more profound long-term sequelae. As in our case, the mainstay of surgical treatment is standard excision.

The differential diagnosis of MN includes intramuscular myxoma, nerve sheath myxoma (NSM) and myxoid dermatofibrosarcoma protuberans (DFSP) [1, 4]. The distinction is made largely from histopathological and immunohistochemical analysis. MN stains positively for S-100, though

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Fig. 1 Gross specimen in its entirety after surgical excision

intramuscular myxoma does not. MN typically has collagen bundles within the myxoid stroma, is not mitotic and lacks a lobular structure [4]. In addition, there are often nerve fibers or

axons present within the tumor. NSM on the other hand, commonly demonstrates a multi-lobulated architecture and no collagen bundles within the stroma and may show mitoses [4]. Finally, myxoid DFSP is often poorly-circumscribed with infiltrative margins and a storiform architecture and will stain negative for S-100 and positive for CD-34.

Conflicts of interest All authors note no conflicts of interest.

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