Intraneural Perineurioma: Diagnosed by CT guided biopsy, a new method of diagnosis.

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INTRODUCTION

Intraneural perineuriomas, also known as localized/telangectatic meningioma, is a benign, slow-growing tumor that presents as a slowly progressive, painless mass. It occurs in a peripheral nerve with loss of motor function and sensation, if any, sensory deficit. Intraneural perineuriomas affects young adults, most prevalently in the second decade of life (1), without a gender predilection. Some reports have found a predilection for the upper extremity (2) while others have found the upper and lower extremities are affected equally (3).

There have been few cases reported of this rare neoplasm, which is only approximately 1% of all peripheral nerve tumors (4). Given the rarity of the disease, there is a need for a deeper understanding of the natural history of the tumor (5). There are a few clinical and pathologic findings that are helpful in the diagnosis of intraneural perineuriomas. Immunohistochemical staining will play an increasingly important role in the diagnosis (4). Furthermore, it is difficult to clinically differentiate intraneural perineuriomas from neurofibromas and schwannomas, and all three conditions can present as localized intraneural masses.

Despite reasonably characteristic MRI findings of intraneural perineuriomas, such as fascicles enlargement with T1 iso- to hypointense signal and T2 hyperintense signal, and small gadolinium enhancement, imaging is still limited in providing a definitive diagnosis. Yet, clinicians will likely be the first to suspect the diagnosis.

To our knowledge, there are no reported cases of intraneural perineuriomas diagnosed by interventional radiological, percutaneous CT guided biopsy. In fact, with one exception, most intraneural perineuriomas are being diagnosed via open biopsy or surgical excision. Naouy, et al. reported in 2013 that open fascicular biopsy is the only definitive way to confirm the diagnosis (6). We present a case of intraneural perineurioma which was effectively biopsied with image guidance, and thus allowing the orthopedic oncologist a wider range of treatment options, including conservative management.

CASE REPORT

An 18-year-old right hand dominant female presented with an enlarging nontender mass of her dominant upper extremity. This was associated with worsening paraesthesia, primarily affecting her right ulnar nerve, over the last two months. She denied any weakness or loss of function. There was no family history of soft tissue tumors, neurocutaneous disorders, or neuromyopathy. Physical examination revealed a normal upper and lower extremity strength. The mass was fairly firm, ill defined, with a firm to palpable mass measuring 3x3x3 cm, applied firm pressure, and palpation, triggering spontaneous paraesthesia (patient’s left arm). Moreover, wrist flexion and crush revealed no numbness or tingling. Further evaluation of her right upper extremity revealed normal range of motion, and vascular status and demonstrated no signs of upper extremity mass. Small finger obliteration was seen with the remaining strength testing revealing no other weakness. There was no associated lymphadenopathy.

Because of the consideration that this may be a presentation of a dynamic process, laboratory tests included a CBC with differential, comprehensive metabolic panel, urinalysis, and HIV, which were all within normal limits. Electromyographic and nerve conduction studies were obtained, but were not obtained due to patient refusal. Imaging was obtained as discussed below.

Tissue would need to be available to perform a histologic diagnosis. A targeted nerve biopsy was performed with the aid of computed tomographic (CT) guidance to ensure representative tissue and minimize morbidity (Fig. 1). Magnetic Resonance Imaging (MRI) was performed at 1.5 tesla. 3D T1 weighted and fat suppressed sequences were obtained with T1, T2, T2 FLAIR, and T1 contrast enhanced. Axial and coronal images were obtained. A needle biopsy was performed under percutaneous image guidance. Image guidance was obtained through a combination of local anesthesia and venous plexus through the ulnar nerve to ensure complete excision of the lesion. The imaging findings were suggestive for a peripheral nerve sheath tumor.

DISCUSSION

MRI is the imaging modality of choice to localize and characterize peripheral nerve lesions. Some authors have suggested that ultrasound is an inexpensive and useful initial screening modality to localize the lesion to decrease the field of view for the MRI (7, 8). Ultrasound may demonstrate fascicular dilatation with increased fluid seen on color Doppler imaging which might not show any signs of enhancement and absent blood flow (6, 9). Furthermore, ultrasonographic characteristics are similar to other more common benign peripheral nerve lesions such as schwannomas and neurofibromas. Therefore, ultrasound has limited ability to definitively characterize the lesion.

Advanced cross-sectional imaging of our patient was not characteristic of a schwannoma, which often demonstrates significant hyperintensity on fluid weighted sequences and is less T2 signal in shape. Neurofibromas was a consideration as the proliferation pattern and location was felt to be more consistent with a “tomato soup” signal, which is predominantly pleomorphic; this appearance was not consistent with the pleomorphic lesions, axial chronic inflammatory demyelinating polyneuropathy (CIDP) was present as a local progressive neuropathy without significant enhancement in the lesion. T1 iso- to hyperintense lesions and T2 hyperintense enhancement are consistent with the suspicion of a schwannoma involving the peripheral nerve. Malignant peripheral nerve sheath tumours are rare and only rarely seen with concomitant (metastatic) or other radiation biopsy (10).

Intraneural perineuriomas classically present as a nodule or fascicles enlargement with proximal and distal tapering. The affected segment is hypointense to isointense on T1 weighted images and hypointense to T2 weighted images with avid, homogeneous, gadolinium enhancement (2, 11). If the morphology is more chronic, they may easily be mistaken for the lesion. Still, the MRI findings are not specific and the differential diagnosis includes other benign neural conditions such as: Schwannomas, neurofibromas, and lymphoma.

Naouy, et al. reported that initial MRI demonstrates a suspicious appearance which represent the preserved nerve fascicles (11). Moreover, the presence of fascicles is diagnostic for intraneural perineuriomas; preservation of fascicles is not specific to intraneural perineurioma as lipomatosis of the nerve also demonstrates preservation of the fascicular architecture. Others have found greater success with MRI magnification to distinguish intraneural perineuriomas from other neural masses (12).

Despite advances in recent MR magnification, radiology lacks the specificity to differentiate the precise lesion type (13). Definitive diagnosis previously required excision and now has successfully performed via minimally invasive CT guided biopsy.

CONCLUSION

Given the nonspecific clinical findings of intraneural perineuriomas, imaging has played an increasingly important role in making the diagnosis. Additionally, it has been reported that many intraneural perineuriomas are not effectively localized with a nerve conduction study (EDS), given the proximal location (6, 10), further supporting the importance of imaging in localizing the abnormality. Ultrasound has been proposed as an effective screening modality to differentiate the lesion from a soft tissue mass (12). MRI is the modality of choice for identifying lesions that may be incompletely or poorly localized with clinical signs and symptoms alone. Despite reasonably characteristic MRI findings, fascicles enlargement with T1 iso- to hypointense signal and T2 hyperintense signal, and avid gadolinium enhancement, imaging is still limited in providing a definitive diagnosis. Yet, still, radiology can be among the first to suggest the diagnosis by obtaining the imaging grade biopsy, which allows orthopedic oncology the opportunity to elicit for conservative management in the young patients.

REFERENCES