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MRI of intraneural perineurioma of the brachial plexus

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Intraneural perineurioma is an uncommon benign tumor of the perineurium of peripheral nerve sheaths occurring primarily in adolescents or young adults. MRI is a valuable tool in suggesting this diagnosis and in surgical planning. We report an 18-year old female with progressive right-hand weakness, numbness, and severe atrophic changes of the hand secondary to an intraneural perineurioma involving the right brachial plexus, in whom the initial diagnosis was suggested by MRI.

Introduction

IP is a slow-growing mononeuropathy that characteristically causes slowly progressive loss of motor function, and occasional sensory loss in the distribution of the affected nerve. Histopathological examination reveals concentric whorls of perineurial cells resembling onion bulbs that stain for epithelial membrane antigen (EMA) (1). The primary imaging findings of IP on MRI are fusiform enlargement of the affected nerve or plexus, increased signal on T2-weighted images, and enhancement with gadolinium contrast material. MRI is an important tool in the presurgical evaluation of these patients; it provides precise information on the exact location and length of the involved segment and the relationship of the tumor to the nerve. Treatment includes resection with or without nerve-graft repair or end-to-end anastomosis (2).

Case report

An 18-year-old female was referred for evaluation of progressive right-hand weakness and numbness over the past year. Her illness began with a slight inability to bend her distal second digit. Eight months before presentation, she began to experience intermittent ventral-hand numbness lasting for periods of 30-40 min with handwriting changes and pain with writing. Six months before presentation, the numbness in the hand became permanent, and for the next six months, she noted that the muscles in her hand were severely atrophied. On examination, there was weakness and severe atrophic changes in all of the intrinsic muscles of the hand. Sensation was absent in the 4th and 5th digits, as well as the medial aspect of the forearm. Clinical and electrophysiological evaluation localized the lesion to the lower trunk of the brachial plexus.

The patient was referred to a neurologist for evaluation of possible thoracic outlet syndrome versus a structural abnormality of the brachial plexus. MRI of the right brachial plexus was performed with a 1.5-T MR scanner. A turbo spin-echo T1-weighted (repetition time = 550 ms, echo time = 12 ms) axial image before and after gadolinium and a conventional T2-weighted (repetition time = 3250 ms, echo time = 80 ms) image in the axial plane were obtained. Using a phase-array body coil, thin-section (4 mm), T1-weighted (repetition time = 600 ms, echo time = 14 ms) postcontrast images with fat saturation were obtained in the axial plane. Thin (4mm) T1W (repetition time = 400 ms, echo time = 8 ms) postcontrast coronal and sagittal images were also obtained. A T2-weighted short tau inversion recovery (STIR) sequence (repetition time = 4500 ms, echo time = 40 ms) was obtained in the coronal plane. Images demonstrated a 4-cm-long segment of fusiform

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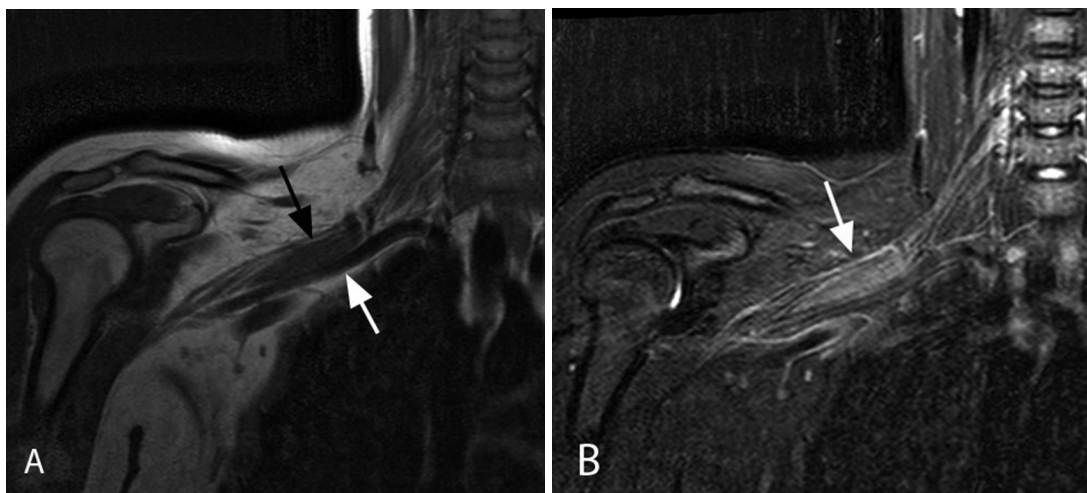


Figure 1. 18-year-old female with intraneural perineurioma. T1W image (A) demonstrates an elongated mass lesion (black arrow) superior to the subclavian artery (white arrow) in the location of the right brachial plexus divisions. STIR image (B) shows increased signal along a segmentally enlarged brachial plexus component (white arrow).

enlargement, hyperintense T2 signal (Fig. 1), and diffuse enhancement (Fig. 2) of the lower trunk of the right brachial plexus extending to the divisions (Fig. 3). The patient underwent a supraclavicular brachial plexus exposure that revealed an enlarged and fibrotic lower trunk (Fig. 4). Using intraoperative nerve stimulation of 1mA, an area that elicited no motor response was identified, and a biopsy was taken from this site, followed by external neurolysis of the

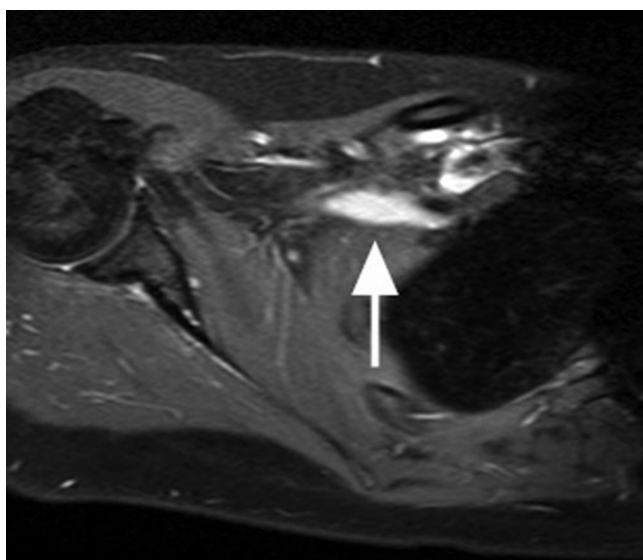


Figure 2. 18-year-old female with intraneural perineurioma. Axial, fat-saturated, gadolinium-enhanced T1-W image demonstrates homogeneous enhancement within the lesion (arrow).

lower trunk where the epineurium of the abnormal segment was dissected.

The tumor displayed foci of hypercellularity arranged in an “onion-bulb” configuration (Fig. 5). Longitudinal areas contained linear masses of connective tissue (trichrome positive). Stains for myelin (LFB-PAS) were negative, while isolated axons were identified within the lesion using a silver stain (Sevier-Munger). Immunohistochemistry of the lesion showed

prominent staining for epithelial membrane antigen (EMA), especially within the onion-bulb components (Fig. 6). S-100 and neurofilament immunohistochemical stains showed occasional linear structures, while positive-staining punctate bodies were identified in the center of the onion bulbs.

On followup evaluation one week after surgery, the patient had improved hand strength and developed a small area of localized pain over the thumb.

Discussion

We identified six cases of IP involving the brachial plexus in the literature (2-5). Only two of the reported cases include MRI images (2, 4). The age ranged from 10 to 26 years, with a mean age of 18. There was no sex predilection. The involved nerve segment size reported in the literature ranged from less than a centimeter to 18 cm (5).

Perineurioma is a benign neoplasm of the perineurium, one of the three connective tissue sheaths surrounding the axon-Schwann-cell complexes in the larger peripheral nerves. The innermost sheath, the endoneurium, surrounds each of the axon-Schwann-cell complexes. The complexes are bundled into a fascicle, which is enclosed by the perineurium, a multilayered, concentric connective tissue composed of tightly adherent epithelioid myofibroblasts (hence the positive staining for EMA). The perineurium acts as a protective barrier for spread of disease along the fascicles. The epineurium is the outermost sheath, which envelops the nerve and consists of dense connective tissue and thick collagen and elastin fibers (6).

Perineuriomas have been traditionally classified into two main types according to their location: intraneural (IP) and extraneural. Extraneural perineurioma are painless nodules found mainly in soft tissues and skin. IP is a benign neo-

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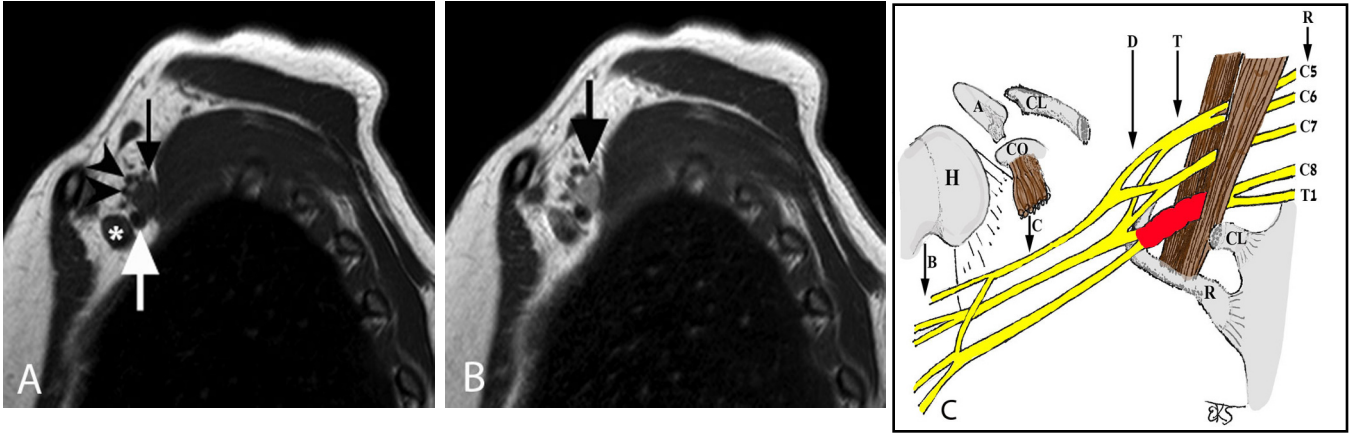


Figure 3. 18-year-old female with intraneural perineurioma. Sagittal T1-W image (A) at the level of the divisions of the plexus (arrowheads) shows the lesion (black arrow) superior to the subclavian artery (white arrow) and posterior to the subclavian vein (asterisk) involving the lower trunk. Gadolinium-enhanced coregistered T1-W image (B) shows that the lesion is enhancing (arrow). Schematic drawing of the brachial plexus with selected musculoskeletal landmarks (C). The plexus is highlighted in yellow, and the tumor is in highlighted in red. The black arrows indicate the following plexus components: R- the roots forming the plexus (C5-T1), T- the three trunks (upper, middle and lower) found on MRI at the level of the interscalene triangle (between the anterior and middle scalene muscles highlighted in brown), D - divisions (anterior and posterior) found on MRI at the lateral margins of the first rib (R), C- cords (lateral, posterior and medial) found on MRI at The level of the coracoid process (CO) and B - branches at the level of the axilla. A- acromion, CO- coracoid process, H-humerus.

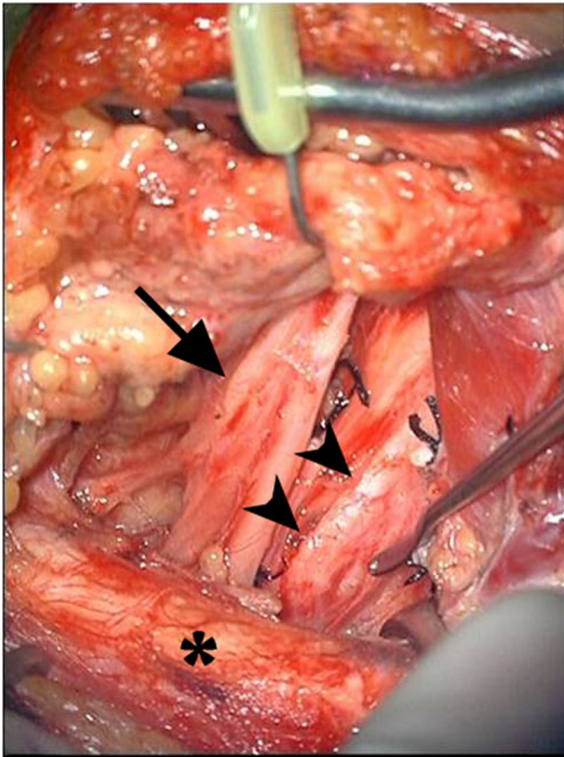


Figure 4. 18-year-old female with intraneural perineurioma. Intraoperative image shows an enlarged and fibrotic lower trunk of the brachial plexus (arrowheads) and the normal divisions of the upper trunk (arrow) located deep to the clavicle (asterisk) with the subclavian artery retracted inferomedially.

plasm composed exclusively of perineurial cells restricted to the boundaries of a nerve. Histological examination of a cross-section of the affected nerve shows irregularly enlarged hypercellular nerve fascicles containing spindled

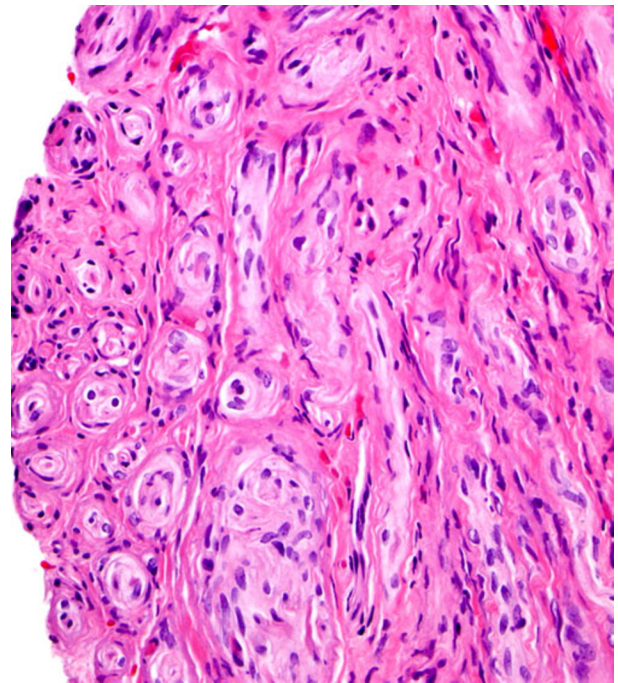


Figure 5. 18-year-old female with intraneural perineurioma. Hematoxylin-eosin (H&E) stain displays foci of hypercellularity arranged in a whorl-like formation (onion bulbs).

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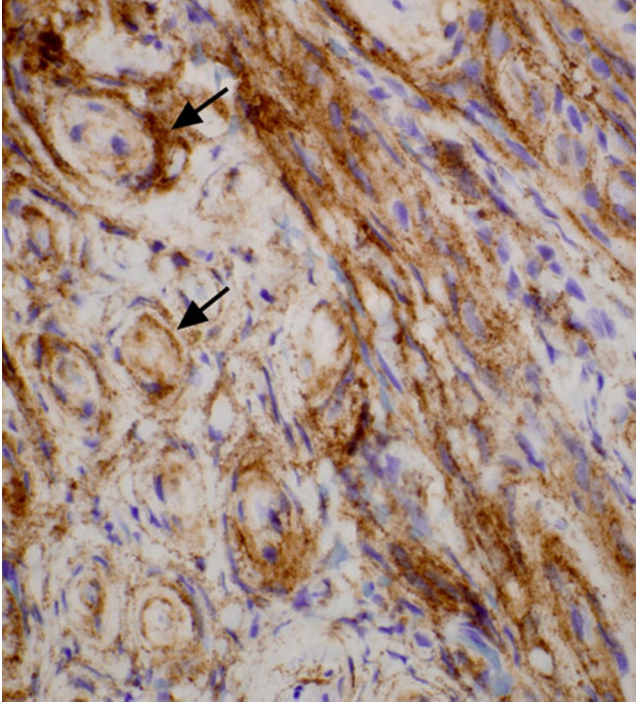


Figure 6. 18-year-old female with intraneural perineurioma. The lesion stains for epithelial membrane antigen (EMA), especially in the onion-bulb formations (arrows).

perineurial cells arranged in pseudo-onion-bulb-like whorls. These whorls stain positive for EMA, a marker of perineurial cell origin, while the center of the bulb stains positive for S-100 protein, a Schwann-cell marker (1). The mitotic rate is very low, accounting for the slow progression of mononeuropathy.

The nature of IP has been a subject of debate, and it has also been referred to in the literature as “localized hypertrophic neuropathy,” with some authors asserting that this lesion is a reactive process associated with trauma. However, the evidence relating these lesions to trauma has been scarce, and recent identification of clonal cytogenetic abnormalities in these lesions has confirmed its neoplastic nature (7). Cytogenetic studies have shown that both intraneural and extraneural perineuriomas show chromosome 22 abnormalities (8). Reported nerve sites of involvement by IP in order of decreasing frequency were the ulnar nerve (17%), median nerve (11%), peroneal nerve (9%), posterior interosseous nerve (9%), sciatic nerve (8%), radial nerve (8%), brachial plexus (8%), femoral nerve (6%), and tibial nerve (4%) (5).

Advances in MRI have improved the visualization of both normal and pathological peripheral nerves, so that it is complementary to clinical examination and electrodiagnostic studies when evaluating peripheral nerve disorders. Fat-saturated, T1W, contrast-enhanced images are most helpful in demonstrating the extension of the lesion, but the vessels are very difficult to separate from enhancing nerve fibers, and the lesion can be misinterpreted as a vascular lesion on

these images (9). Sagittal images are the most helpful, since the brachial plexus is easily identified superior to the subclavian artery and posterior to the subclavian vein.

Differential diagnosis includes neurofibroma, schwannoma, chronic inflammatory polyneuropathy (CIDP), and hereditary motor and sensory neuropathy (HMSN) (which includes Dejerine-Sottas disease and Charcot Marie Tooth disease) (9). Although the imaging features described in this case may be seen in other lesions involving the brachial plexus, the clinical presentation was atypical for schwannoma—specifically, the extensive muscle wasting. The patient had no history or stigmata to suggest neurofibromatosis. CIDP is often symmetrical and multifocal, while in our case the lesion was unilateral and solitary. And finally, in cases of HMSN, contrast enhancement is almost never present.

Treatment is controversial and attempts to preserve nerve function. Some authors advocate diagnostic biopsy followed by neurolysis instead of resection (8), while others prefer a resection with neural grafting or end-to-end anastomosis, as IP is a progressive condition that evolves inexorably to a total loss of nerve function (3).

In conclusion, the diagnosis of IP should be entertained in cases of a diffuse lesion affecting the brachial plexus when coupled with the clinical history of progressive motor/sensory disturbance in a young person.

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