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Case Report

Utility of ultrasound in the diagnosis and management of a radial nerve perineurioma in a pediatric patient

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ABSTRACT

Background: Intraneural perineuriomas are tumors originating from the perineurial cells surrounding nerve sheath fascicles. Intraneural perineuriomas represent about 1% of peripheral nerve tumors and are often misdiagnosed due to their rarity. In this case, we report a pediatric patient with a radial nerve perineurioma, in which ultrasound played a key role in diagnosis.

Case Description: We present the case of a 4-year-old male with over 6 months of progressive left upper extremity weakness found to have chronic left radial neuropathy distal to the triceps branch of the radial nerve on electromyography/nerve conduction study. Ultrasound showed a well-defined fusiform hypoechoic mass of approximately 1.5 × 0.5 × 0.8 cm, with the radial nerve noted to enter and exit the mass. Magnetic resonance imaging (MRI) did not encompass the pathology. The mass was excised with small margins, and pathology was consistent with perineurioma. The resultant 3 cm gap was repaired through nerve grafting. Surveillance MRI showed no recurrence of the perineurioma over time.

Conclusion: Ultrasound served as a necessary adjunct in the workup of upper extremity weakness despite negative MRI findings. This imaging modality should be considered if there is high clinical suspicion of a peripheral nerve lesion.

Keywords: Pediatric, Perineurioma, Peripheral nerve tumor, Peripheral nerve, Ultrasound

INTRODUCTION

Perineuriomas, rare benign tumors originating from perineurial cells surrounding nerve fascicles, are divided into two types: extraneural and intraneural. Extraneural perineuriomas, more common in older patients, occur in soft tissue and skin, whereas intraneural perineurioma is restricted to the boundaries of a peripheral nerve.[2] Similar to the onion-bulb formation of schwannomas, perineuriomas demonstrate whorled formations on pathology but are referred to as pseudo-onion bulbs due to the expression of epithelial membrane antigen (EMA) rather than S-100.

Intraneural perineuriomas represent about 1% of peripheral nerve tumors and are often misdiagnosed due to their rarity; thus, proper diagnosis requires a high level of clinical suspicion. These lesions commonly occur in young, otherwise healthy patients and have an insidious onset.[2] Although symptoms typically include mononeuropathy with predominantly motor

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symptoms, sensory impairment is possible, and there have been reports of brachial plexopathy.[2] In this case, we report a pediatric patient with a radial nerve perineurioma, in which ultrasound played a key role in diagnosis. In addition, we discuss the surgical management of this rare peripheral nerve tumor.

CASE DESCRIPTION

Case presentation

A 4-year-old male presented with over 6 months of progressively worsening left upper extremity weakness without pain. On exam, atrophy was noted along the extensor surface of the left forearm. He was unable to extend his left wrist and fingers but was otherwise neurologically intact without sensory loss. No Tinel's sign or palpable mass was present.

Electromyography/nerve conduction study findings showed a chronic left radial neuropathy distal to the triceps branch of the radial nerve. Magnetic resonance imaging (MRI) of the cervical spine and left brachial plexus were unremarkable, and MRI of the left elbow from the distal humerus through the forearm showed atrophy and increased signal intensity of forearm extensor musculature without evidence of tumor or entrapment. An ultrasound was subsequently performed due to high suspicion of a mass lesion. It identified a well-defined fusiform hypoechoic mass of approximately $1.5 \times 0.5 \times 0.8$ cm at the level of the mid-humeral shaft [Figure 1]. The radial nerve was noted to enter and exit the mass and appeared normal in caliber proximal and distal to the localized dilation.

Management

Due to progressive loss of function in the setting of a mass, surgery was offered for neurolysis and resection [Figures 2a and b]. Preoperatively, the nonpalpable mass was localized with ultrasound to plan a linear incision along the posterior aspect of the left upper extremity. Following soft tissue dissection, the mass was encountered along the radial nerve and dissected from surrounding structures. No nerve action potential was elicited between the proximal and distal poles of the dilated segment. The fusiform dilation was then excised with small margins, resulting in an approximately 3 cm gap between nerve ends. Normal-appearing fascicles were noted proximally and distally. For reconstruction, 16 cm of the sural nerve was harvested and sectioned into four segments that were secured together with fibrin glue for cable grafting. The grafts were coapted to the nerve ends and secured in place.

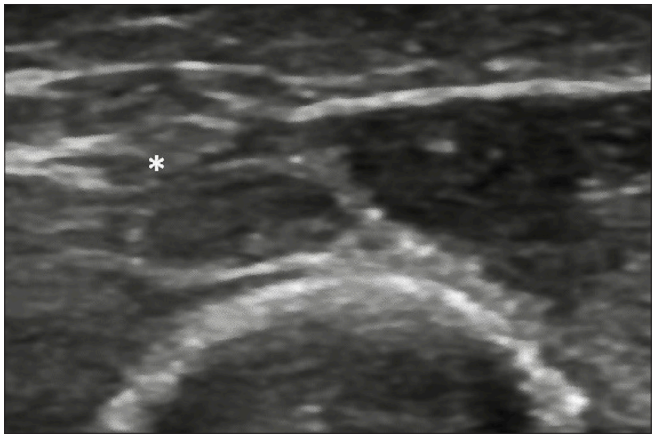

Figure 1: Diagnostic findings. Ultrasound showing a fusiform radial nerve lesion (*) at the level of the mid-humerus not visualized on magnetic resonance imaging.

Pathology and outcome

Pathology showed perineurioma with perineurial cells expressing EMA [Figures 2c and d]. The patient had persistent wrist and finger drops and underwent subsequent tendon transfers to restore extensor function. Surveillance MRI showed no recurrence of the perineurioma over time.

DISCUSSION

We present a pediatric patient with a radial nerve lesion, found to be perineurioma on pathology, which was diagnosed using ultrasound. The present report reinforces the difficulty in the diagnosis of intraneural perineurioma. The presentation of slow, progressive mononeuropathy with predominantly weakness with or without sensory disturbance is associated with a broad differential diagnosis, contributing to misdiagnosis. Additional entities to consider include entrapment neuropathy, focal inflammatory demyelination, sarcoidosis, brachial neuritis (or Parsonage–Turner syndrome), and other intraneural lesions, including hemangiomas, hemangiopericytomas, angiomyxofibromas, ganglioneuromas, and neuromuscular hamartomas.[2]

Based on the presentation, a structural lesion was suspected; however, an MRI of the elbow, a static study surveying a specific anatomic region, did not capture the lesion. Instead, the dynamic nature of ultrasound and its ability to image along the length of the nerve permitted the identification of the lesion. Additional benefits of ultrasound include its relatively low cost, noninvasiveness, lack of associated contraindications, and portability.[1] As perineurioma frequently affects young patients, the ability to perform an ultrasound at the bedside without sedation and with a caretaker present is an overlooked benefit in the pediatric population. Ultrasound also provided utility intraoperatively to aid in localization and incision planning.

Perineuriomas commonly form a fusiform dilation of a nerve rather than the globular enlargement seen in schwannomas.

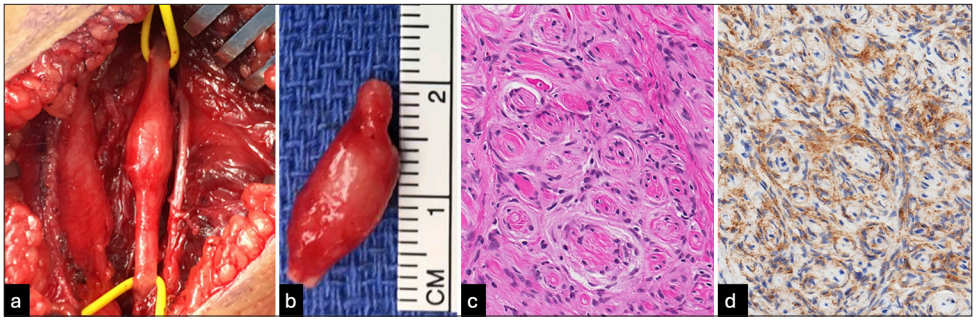

Figure 2: Intraoperative findings. (a) Fusiform dilation of the radial nerve is noted. (b) The fusiform mass was resected. (c and d) Pathology slides (hematoxylin and eosin, ×200; epithelial membrane antigen [EMA], ×200) were characteristic of perineurioma, showing concentric rings forming a pseudo-onion bulb appearance positive for EMA.

Findings can be subtle yet readily detectable on ultrasound. In a retrospective study, focal nerve pathology was detected more frequently with ultrasound compared to MRI.[4] Ultrasound was more sensitive than MRI (93% vs. 67%) with equivalent specificity (86%), and it better defined multifocal lesions.[4]

Management strategies for perineurioma are not clearly defined. This patient presented with loss of motor function and absent nerve action potential across the lesion, supporting resection with nerve grafting. This approach allows for complete removal of the lesion and an opportunity for motor improvement over time. Some surgeons perform a staged approach with fascicular nerve biopsy for diagnosis, followed by nerve reconstruction.[2,5] Others advocate that a perineurioma should not be resected routinely secondary to its slowly progressive nature and tendency to remain confined without involving new nerves.[3] Although biopsy, at a minimum, is required for diagnosis, some authors reported intraneural neurolysis as a means of preserving residual function, if present. In a recent review, neurolysis was shown to have a marginally increased rate of functional improvement over nerve resection with grafting, 41.6% versus 35.7%, respectively.[5] However, neurolysis alone without resection precludes treatment of the tumor, and while the natural history of perineuromas is not extensively characterized, several groups report the progression to complete motor loss in the given nerve.[2]

Judgment must be applied intraoperatively when balancing aggressive tumor resection with the creation of a larger gap between nerve ends, which may be associated with a lower success rate for grafting. Nerve transfer is thus a promising alternative, as it permits aggressive resection of the tumor without concern for grafting. A distal nerve transfer can be used in cases of motor weakness to provide an opportunity for reinnervation. This approach may be especially useful if the lesion spans a long segment or if significant atrophy is present.[5] Preoperative duration of motor weakness may also affect success rates of reinnervation approaches. Relatively shorter distances to target reinnervation in nerve transfers may be more favorable for patients presenting late to care, which is common in perineurioma.[2] Improved awareness of this entity may decrease the latency in diagnosis, allowing for more reconstruction options with higher reinnervation rates. Tendon transfers, as performed in our patient, may be a useful bailout strategy if nerve grafting is unsuccessful. Due to the rarity of perineurioma and its nuanced presentations, management strategies are best tailored on a case-by-case basis.

CONCLUSION

Intraneural perineuriomas are prone to misdiagnosis due to their rarity and insidious onset. These peripheral nerve tumors should be considered in young patients with slowly progressive mononeuropathy with predominant motor symptoms. Ultrasound has unique features that make it useful even in the setting of a negative MRI and may complement the diagnostic workup. It was critical in making the diagnosis in the current case.

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