

## MRI of Intraneuronal Perineurioma: Review of 27 Cases with Histopathologic Correlation

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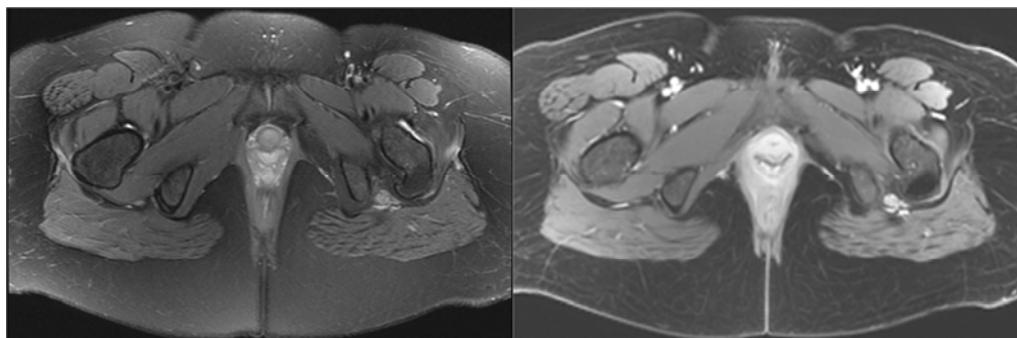
**Background:** Intraneuronal perineurioma is a rare benign peripheral nerve sheath tumor. It is characterized pathologically by whorls of perineurial cells that demonstrate reactivity for Epithelial Membrane Antigen (EMA) and are negative for S-100 (Schwann cell marker) on immunochemistry, and demonstrates pseudo-onion bulb formation on electron micrograph. Intraneuronal perineuriomas generally present in children and young adults with a slowly progressive motor predominant mononeuropathy or plexopathy.<sup>1-3</sup> In the radiological, clinical and histopathological setting, differentiation from localized hypertrophic neuropathies such as focal chronic inflammatory demyelinating polyneuropathy (CIDP) may pose a diagnostic difficulty. Peripheral nerve MRI demonstrates characteristic findings that help it to be readily differentiated from other neurogenic entities, including CIDP. Diagnosis is critical as subsequent treatments are significantly different.<sup>1,4</sup>

**Objective:** The aim of this study was to review the MR imaging features of intraneuronal perineurioma in pathologically proven cases and demonstrate how these imaging features can help to differentiate intraneuronal perineurioma from other hypertrophic neuropathies, such as CIDP, eliminating the need for unnecessary nerve biopsy in select cases.

**Materials and Methods:** Twenty-seven cases of pathologically proven intraneuronal perineurioma were evaluated. Detailed evaluation included patient demographics, lesion size, lesion location, denervation changes and MR signal characteristics including post-gadolinium enhancement characteristics.

**Results:** The study consisted of 19 males and 8 females, with mean age at time of evaluation of 24 years (min: 2 max: 54). The sciatic nerve and/or its branches were most commonly involved, seen in 17 cases (63%), followed by the brachial plexus seen in 5 cases (19%). Mean lesion length was 12 cm (min: 1, max: 46). All (100%) lesions demonstrated fusiform nerve enlargement, T1 isointensity and T2 hyperintensity. The majority (78%, 21/27) demonstrated avid post-gadolinium enhancement. In four cases (15%) imaging was done without contrast. Two lesions (7%) demonstrated mild to moderate enhancement. The majority (67%, 18/27) of lesions demonstrated denervation changes in the respective musculature with 72% (13/18) of patients having moderate or advanced denervation changes.

**Conclusion:** Intraneuronal perineurioma demonstrates characteristic features on MRI including fusiform nerve enlargement, T2 hyperintensity, avid post-gadolinium enhancement, and denervation changes that help to distinguish this rare entity from other neurogenic entities, including hypertrophic neuropathies such as focal CIDP. These findings may eliminate unnecessary nerve biopsy in patients clinically suspected of having perineurioma.



Axial T2-fat saturated (left) and post-gadolinium SPGR images (right) demonstrate increased T2 hyperintensity, diffuse enlargement and avid enhancement of the left sciatic nerve at the level of the ischial tuberosity in a 27 year old female with pathology proven intraneuronal perineurioma.

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