Neurilemmoma of Extremities: MR Findings

Ki Bum Kim, M.D., Kyung Jin Suh, M.D., Duck Sik Kang, M.D.

Department of Radiology, Kyungbuk National University College of Medicine

Abstract

Six patients with twenty histologically proven neurilemmomas of the extremities were studied using magnetic resonance (MR) imaging.

The size, number, signal intensity on spin-echo T1WI (TR 500-650ms/TE 14-25ms) and gradient-echo (TR 200-600ms/TE 14-20ms; flip angle 25-30°) image, enhancement pattern, detectability of nerve of origin, nerve-lesion relationship, and presence of a capsule were analyzed.

The masses ranged from 1 to 12 cm in longitudinal diameter and originated from the median nerve, ulnar nerve, sciatic nerve, radial nerve, and tibial nerve. All the nerve tracts except for those of 5 lesions, which could not be detected due to their small diameter, were visualized as low intensity tubular structures. All visible nerve tracts were situated along the periphery of the lesion and this finding was considered to be specific for neurilemmoma. All neurilemmomas were isointense with the surrounding muscle on spin-echo T1WI and hyperintense on gradient-echo image. After a Gd-DTPA injection, all masses showed moderate or marked enhancement and more prominent inhomogeneity than that on nonenhanced scan. In 19 out of 20 lesions (95%), a low signal intensity capsule surrounding the masses could be seen. Four of the six patients showed multiple masses, which was unusual as neurilemmoma usually arises as a solitary mass.

In conclusion, the MR findings, especially the eccentric location of the mass lesion from the nerve of origin and the presence of a capsule, were useful in making a diagnosis of neurilemmoma of the extremity and that multiple neurilemmomas were not uncommon.

Index Words: Soft tissue, neoplasms, 40.37
Soft tissue mass, MR studies, 40.1214
Neoplasm, MR studies

INTRODUCTION

Neurilemmoma is a common benign tumor arising from the neural sheaths of the peripheral motor, sensory and cranial nerves. Although there have been many reports dealing with MR findings of spinal or cranial neurilemmoma (1-7), only few literature has reported MR features of the neurilemmoma of extremities (8).

We reviewed 6 patients with histologically verified peripheral neurilemmoma and attempted to find some specific features of this soft tissue tumor. We evaluated the following MR parameters: signal intensity on spin-echo T1WI and gradient-echo image, detectability of the nerve of origin, nerve-lesion relationship, presence of a capsule, and number of lesions.

MATERIALS AND METHODS

A total of 20 lesions in 6 patients were examin-
ed over a 10 month period. No evidence of von Recklinghausen's disease such as skin manifestation of family history was identified in any of the patients. The patients ranged from 24 to 64 years of age; four were men and two were women. All the examinations were performed with a 0.5T superconducting MR system (Max, GE medical systems, Milwaukee, WI, U.S.A). A 10mm thickness axial, coronal, and sagittal scan were routinely used, and other parameters such as matrix size, field of view, and type of surface coil varied according to size, site, and depth of the lesion. In all cases a spin-echo T1 weighted sequence (TR 500-650ms/TE 14-25ms) and gradient-echo (TR 200-600ms/TE 14-20ms; flip angle 25-30°) sequence were performed. Gd-DTPA(0.1mmol/kg) enhanced T1WI were obtained in each patients. Signal intensity on the spin-echo T1WI and gradient-echo image were evaluated and expressed as hypointense, isointense, or hyperintense with muscle intensity. The visibility of the nerve of origin was also determined, as well as its relationship with the mass. The presence of a capsule was determined on the MR imagings by the recognition of a low signal intensity rim along the borders of the lesion.

RESULT

The masses originated from the median nerve, femoral nerve, ulnar nerve, sciatic nerve, radial nerve, or tibial nerve, and most commonly

<table>
<thead>
<tr>
<th>Table 1. Nerve of Origin and Number of the Lesion</th>
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<tbody>
<tr>
<td>Originated Nerves</td>
</tr>
<tr>
<td>Median nerve</td>
</tr>
<tr>
<td>Femoral nerve</td>
</tr>
<tr>
<td>Ulnar nerve</td>
</tr>
<tr>
<td>Sciatic nerve</td>
</tr>
<tr>
<td>Radial nerve</td>
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<tr>
<td>Tibial nerve</td>
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<tr>
<td>Total</td>
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Fig. 1. Case 1, A 44-year-old woman with multiple neurilemmomas of the median nerve. T1-weighted (600/20) (a) and gradient-echo (300/20; flip angle 30°) coronal images. Six of eight lesions are visualized (the other two lie distal to the carpal tunnel) as beaded appearance along the course of the median nerve (arrows in b), which is displaced by the masses. The nodules show a homogenous intermediate signal intensity on T1WI and high signal intensity on gradient-echo image. A photomicrograph (c) of the tumor shows it to be composed of mainly Antoni A tissue.
originated from the median nerve (Table 1). A summary of our findings is presented in Table 2. Four of the six patients showed multiple masses, up to eight lesions (Fig. 1), which originated from single or multiple nerves of only one extremity. All the neurilemmomas were isointense with surrounding muscles on spin-echo T1WI and relatively hyperintense on gradient-echo image (Fig. 2).

Signal intensity was homogenous in 90% (18/20) and 80% (16/20) on spin-echo T1WI and gradient-echo image, respectively. Following intravenous administration of Gd-DTPA, all the tumors revealed moderate to marked enhancement and signal intensity was homogenous in 60% (12/20). The size of the tumors ranged from 1 to 12 cm in longitudinal diameter with a mean of 3 cm. When the lesion was more than 3 cm in longitudinal diameter, inhomogeneity was prominent in those cases with cystic degeneration (Fig. 3). A low density capsule (Fig. 3a and 4c) surrounding the mass could be seen in 19 out of 20 lesions (95%). The nerve of origin, visualized as a low intensity tubular structure (Fig. 3b),

Fig. 2. Case 2, A 55-year-old man with two neurilemmomas of the sciatic nerve and one neurilemmoma of the femoral nerve. Precontrast (left of a) and postcontrast (right of a) coronal T1-weighted image (650/25) of the right thigh reveal two well-defined solid lesions with a capsule (larger arrows in right of a) along the course of the sciatic nerve (arrows in right of b), which displaced toward the pihhery. In a precontrast image the lesion is relatively homogenous, but a postcontrast image shows several nonenhanced cystic foci (thin arrows in right of a). Contiguous two coronal gradient-echo images (600/20; flip angle 30°) (b) show homogenous hyperintense femoral (left of b) and sciatic lesions. The lesion of the femoral nerve is the only one with no apparent capsule in this study.
Fig. 3. Case 4, A 57-year-old man with cystic neurilemmoma of ulnar nerve. Precontrast coronal T1-weighted image (350/14) (a) shows a large inhomogenous insointense mass with a thick capsule (arrows in a). Gadolinium enhanced sagittal T1WI (b) shows mostly cystic degeneration with only a small peripheral irregularly enhanced solid portion. The displaced ulnar nerve is well visualized (arrows in b). A photograph (c) of the mass shows it to be composed of mainly Antoni B tissue.

Fig. 4. Case 6, A 64-year-old woman with a neurilemmoma of the femoral nerve. Sagittal T1WI (500/25) (a) and gradient-echo image (600/20; flip angle 30°) (b) of right thigh reveal a 12 x 6 x 5cm, well-encapsulated inhomogenous and intermediate signal intensity (a) and a homogenous high signal intensity mass. The nerve of origin is located along the posterior margin of the mass (arrow in a). Axial postcontrast T1WI (c) shows a thick low density capsule (arrows) surrounding the inhomogenous mass with multiple cystic components.
Table 2. MR Findings of 20 Neurilemmomas

<table>
<thead>
<tr>
<th>Case/Age (yrs)/Sex</th>
<th>Nerve of Origin</th>
<th>No. of Lesion</th>
<th>Size in Lesion (cm)</th>
<th>T1WI</th>
<th>MR Imaging T2WI</th>
<th>Gd-enhancement of Nerve</th>
<th>Position of Nerve</th>
<th>Presence of Capsule</th>
</tr>
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<tbody>
<tr>
<td>1/44/F</td>
<td>Median N</td>
<td>8</td>
<td>1</td>
<td>Iso (Hs)</td>
<td>Hyper (Hs)</td>
<td>Mark. (Hs)</td>
<td>Eccentric</td>
<td>Yes</td>
</tr>
<tr>
<td>2/51/M</td>
<td>Sciatic N</td>
<td>2</td>
<td>5, 3</td>
<td>Iso (H, H)</td>
<td>Hyper (H,H)</td>
<td>Mark. (I, I)</td>
<td>Eccentric</td>
<td>Yes</td>
</tr>
<tr>
<td>3/60/F</td>
<td>Femoral N</td>
<td>1</td>
<td>3</td>
<td>Iso (H)</td>
<td>Hyper (H)</td>
<td>Mark. (I)</td>
<td>Undetectable</td>
<td>No</td>
</tr>
<tr>
<td>5/24/M</td>
<td>Ulnar N</td>
<td>2</td>
<td>2, 1</td>
<td>Iso (H,H)</td>
<td>Hyper (H,I)</td>
<td>Mark. (I, H)</td>
<td>Eccentric</td>
<td>Yes</td>
</tr>
<tr>
<td>4/57/M</td>
<td>Ulnar N</td>
<td>6</td>
<td>1.5</td>
<td>Iso (H)</td>
<td>Hyper (I)</td>
<td>Marginal (I)</td>
<td>Eccentric</td>
<td>Yes</td>
</tr>
<tr>
<td>5/24/M</td>
<td>Femoral N</td>
<td>1</td>
<td>1.2</td>
<td>Iso (H)</td>
<td>Hyper (H)</td>
<td>Mark. (H)</td>
<td>Undetectable</td>
<td>Yes</td>
</tr>
<tr>
<td>5/64/F</td>
<td>Femoral N</td>
<td>1</td>
<td>12</td>
<td>Iso (I)</td>
<td>Hyper (H)</td>
<td>Mark. (I)</td>
<td>Eccentric</td>
<td>Yes</td>
</tr>
</tbody>
</table>

Note.- Iso = isointense; Hyper = hyperintense; Mod. = moderate enhancement; Mark. = marked enhancement; H = homogenous; I = inhomogenous; Hs = all of 8 lesions are homogenous.

was detected in 15 lesions, but was unidentifiable in 5. In all 15 lesions with detectable nerve origin, the nerves were located peripherally around the mass.

On microscopic examination, all the tumors showed typical features of neurilemmoma composed of compact cellular areas called Antoni A regions and loosely arranged hypocellular areas known as Antoni B region. However, no relationship could be established between the signal intensity on spin-echo T1WI and gradient-echo image and the histologic type of the neurilemmomas.

**DISCUSSION**

Neurilemmomas arise from the neural sheaths of the peripheral motor, sensory, and cranial nerves, with the exception of the optic and olfactory nerves, which lack Schwann cell sheaths and are part of the central nerve system (9). They occur in young and middle age adults, but no age group is exempted (10). Women are affected twice as often as men (11). The patients in this study ranged from 24 to 64 years of age; 4 were men and 2 were women. Neurilemmomas are usually solitary and painless (12), but when large they can produce pressure symptoms of paresthesia or local tenderness. In this study, however, 4 out of 6 patients showed multiple lesions and were associated with Tinel's sign or tenderness. Occasionally a patient might have multiple neurilemmomas which frequently are associated with von Recklinghausen's disease, however in this study no case with skin manifestation or family history was identified.

The tumors occurred anywhere in the soft tissue or in the viscera, but the more common location included the head and neck, especially the lateral aspect of the neck, the extremities, trunk, mediastinum, and retroperitoneum (13).

Grossly, neurilemmomas commonly are fusiform, round, or oval masses that are sharply circumscribed and encapsulated (9,10). Neurilemmomas are usually less than 5cm in diameter, however can be as large as 20cm. In this study the size of the tumors ranged from 1 to 12cm in longitudinal diameter and the mean diameter was 3cm. Larger lesions had cystic and hemorrhagic foci (Fig. 3). It was possible to dissect the tumor from the nerve of origin, since the nerve of origin coursed along the periphery of the tumor, flattened along the capsule but not encased within the substance of the tumor.

The histologic appearance of neurilemmoma alternates between compact cellular areas, which
have historically been called Antoni A regions, and loosely arranged hypocellular areas known as Antoni B regions (11,14). Although all cases of this study, like those described in the literature (10,14), revealed both Antoni A and B areas within each of the tumors, small tumors less than 1cm in diameter were composed of almost entirely type A areas (Fig. 1c) and larger tumors tended to have the Antoni B component (Fig. 3c).

Although a neurilemmoma is typically hypointense or isointense on spin-echo T1WI and hyperintense on gradient-echo image (2,4), few reports in the literature have described the MR features of neurilemmoma of the extremities (8). All of the 20 tumors studied were isointense with the muscle on spin-echo T1WI and hyperintense on gradient-echo image. Eighteen (90%) out of 20 neurilemmomas showed a homogenous signal intensity on spin-echo T1WI, and on gradient-echo image 16 (80%) were homogenous. On Gd-DTPA enhanced image, only 13 lesions (65%) were homogenous. The causes of inhomogeneity were cystic degeneration, hemorrhagic necrosis, and fibrosis (4,13). Neurilemmomas are frequently encapsulated whereas neurofibroma, which is one of the most common peripheral nerve tumors, is usually not (8). The MR detection of a capsule, visualized as a low intensity rim along the margin of the tumors, could therefore be used as a criterion to differentiate neurilemmoma from neurofibroma (8). This feature was found in 19 out of 20 lesions (95%). Neurilemmomas were located along the nerve trunks which could be visualized in 15 of 20 lesions (75%) on 1cm thickness contiguous sections. Normal nerves appeared as low intensity tubular structures in all pulse sequences. Blood vessels may have a similar appearance but they could be distinguished from nerves by the appreciation of flow phenomena within the lumen, such as even-echo rephasing.

In conclusion, combination of MR findings such as peripheral location of the nerve of origin, the presence of a capsule, and inhomogenous signal intensity primarily due to cystic degeneration are helpful in making a diagnosis of neurilemmoma. Even when the lesions are multiple, the possibility of neurilemmoma still exists.

REFERENCES

4. 박길선, 장기현, 한문희 등. 척추 신경 종과 수막종의 자기공명영상 소견. 대한방사선 의학회지 1991; 27(3) : 337-342
10. Abell MR, Hart WR, Olson JR. Tumors of the peripheral nerves system. Hum. pathol. 1970; 1:503-504
12. Alvira M, Mandybur T, Meneic G. Light
발리조직학적으로 확진된 사지 신경초종 20개의 자기공명영상 소견을 종양의 크기, 갯수, T1과 T2 강조영상에서의 신호강도, 조영증강소견, 종양발생신경의 발견 유무, 종양과 신경과의 위치 관계, 그리고 피막의 존재 유무를 중심으로 분석하였다.

신경초종의 크기는 1-12cm 이였고 종양이 발생한 신경은 15개(75%)에서 발견할 수 있었다. 이들 신경은 모두 예에서 종양의 주변부를 따라 주행하였고, 이 소견은 신경초종의 매우 특징적 소견으로 생각되었다. 모든 종양들은 T1 강조영상에서는 근육과 동일한 신호 강도를 나타냈으며 gradient echo 영상에서는 고신호 강도를 나타냈다. 조영제 주입후 모든 종양들은 종래도 내지 현저한 조영증강을 보였으며 조영제보다 불규칙하게 나타나는 경우가 많았다. 종양을 둘러싼 피막은 19예(95%)에서 발견되어 피막의 존재유무가 신경초종을 진단하는데 중요한 소견이라 생각되었다. 총 6명의 환자중 4명에서 다발성 종양을 나타냈으며 이는 신경초종이 흔히 단발성이라는 대부분의 보고들과 차이를 보였다.

결론적으로 이러한 자기공명영상 소견들은 종양발생신경이 종괴 주변부에 위치하는 것과 피막의 존재등은 사지의 신경초종을 진단하는데 유용한 소견으로 생각되어 비록 다발성인 경우라도 이러한 소견이 관찰되면 신경초종을 의심해야 한다고 생각된다.