CASE REPORT

Schwannoma Presenting as a Scalp Mass: A Case Report with Magnetic Resonance Imaging Findings

Jong Seo Park[1], Jungyoon Moon[1], Soo Ick Cho[1], Je-Ho Mun[1,2]

[1]Department of Dermatology, Seoul National University College of Medicine, [2]Institute of Human-Environment Interface Biology, Seoul National University, Seoul, Korea

A schwannoma can develop anywhere along the course of nerves. However, a schwannoma presenting as a scalp nodule is rare. Here, we present a rare case of schwannoma on the scalp with a review of magnetic resonance imaging (MRI) findings, which was initially misdiagnosed as an epidermal cyst or vascular malformation despite various radiologic examinations. Recognition of characteristic MRI features of schwannomas, such as low signal margin, target, entering-and-exiting-nerve, and fascicular signs, may result in an accurate diagnosis and proper management of tumors. In this report, we summarized differential characteristics of a schwannoma with an epidermal cyst and a lipoma. **(Ann Dermatol 32(1) 64~68, 2020)**

-Keywords-
Magnetic resonance imaging, Nerve sheath neoplasms, Neurilemmoma, Scalp, Skin neoplasm

INTRODUCTION

A schwannoma is a benign neoplasm of the nerve sheath derived from Schwann cells[1]. We present a rare case of schwannoma on the scalp that was difficult to diagnose despite various preoperative radiologic examinations.

CASE REPORT

A 47-year-old woman who visited our clinic presented with a 35-year history of a subcutaneous mass protruding from the left occipital scalp (Fig. 1A). She underwent surgical excision 35 years ago, and she remembered that the diagnosis was lipoma. The patient reported that she initially had no symptoms but started experiencing pain two years previously, which gradually worsened. The patient underwent ultrasonography and computed tomography (CT) of the brain at a local hospital. Multiple low echoic lesions were observed. The radiologist suggested that the tumor could be an epidermal cyst. Therefore, she was referred to our clinic for surgical removal. We received the patient's consent form about publishing all photographic materials.

We performed further radiologic examination using magnetic resonance imaging (MRI) before surgery. MRI revealed a heterogeneously enhancing mass with multilocular lesions extending to the posterior neck space. In addition, a heterogeneous hyperintense signal on T2-weighted images, isointense signal on T1-weighted images, and internal microhemorrhages were observed. The radiologist suggested that the tumor could be a type of vascular tumor, such as a vascular malformation.

Because of these inconsistent preoperative radiologic findings, exploratory surgery was performed for accurate diagnosis and treatment. We made a zigzag incision on the scalp and elevated the flaps for sufficient visual field to remove the mass. We successfully excised the tumor (Fig. 1B, C). The specimen was a multilobulated mass of measuring $7.0 \times 2.7 \times 1.0$ cm^3. Histologic examinations revealed

Fig. 1. (A) Clinical photography of subcutaneous protruding mass on the left occipital scalp before surgery. (B, C) Successful removal by exploratory surgery. We made a zigzag-line incision and elevated the flaps for a visual field to remove the entire mass. (D) Clinical photography of scalp 12 months after surgery. There was no recurrence of the mass until 12 months follow-up.
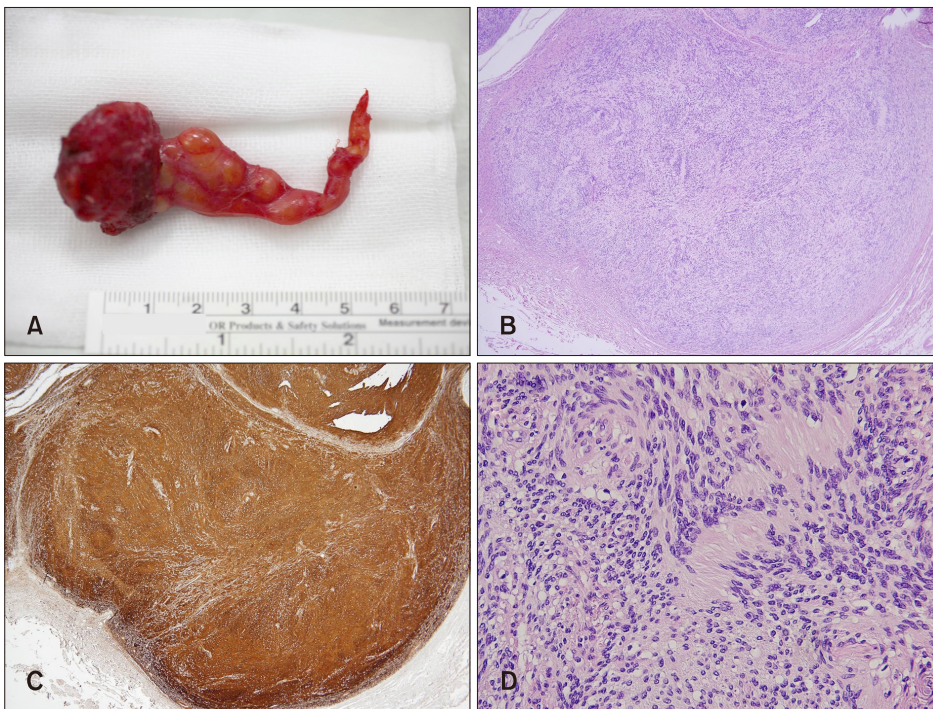

Fig. 2. (A) Gross specimen of multilobulated mass measuring $7.0 \times 2.7 \times 1.0$ cm^3. (B) Histologic examination shows an encapsulated, well-circumscribed mass that included alternating Antoni A and Antoni B (hematoxylin and eosin [H&E], ×40). (C) Tumor cells are positive with S-100 (×40). (D) Acellular areas lying between opposing rows of parallel nuclei (verocay body) are seen (H&E, ×400).

an encapsulated, well-circumscribed mass that included alternating hypercellular (Antoni A) and hypocellular (Antoni B) areas of myxoid stroma. Immunohistochemically, the lesion was positive for S-100 protein and vimentin and negative for epithelial membrane antigen and cluster of differentiation 34 (Fig. 2). Finally, the tumor was diagnosed as a schwannoma. After the surgery, the patient was free of pain and had no neurologic complications. In addition, the patient satisfied with cosmetic outcome since there was no conspicuous alopecia. There was no recurrence at 12 months follow-up (Fig. 1D).

DISCUSSION

A schwannoma is one of the most common benign nerve sheath tumors[2]. Reports have indicated that 25%~45% of extracranial schwannomas develop on the head and neck[3]. However, schwannoma development on the scalp is rare[4-6]. According to a study analyzing 22 patients with extracranial non-vestibular head and neck schwannomas, 76% were unilateral neck masses[7].

A pathologic examination is the gold standard for diagnosing a schwannoma. However, a preoperative radiologic examination is helpful. According to the literature, the use-

fulness of CT is limited for schwannomas as it only correctly diagnosed 14% of cases[8]. In CT scans, schwannomas may show low to intermediate attenuated cystic masses; therefore, it cannot easily be differentiated from other common scalp cystic lesions. MRI is a more accurate imaging tool for schwannoma as 20 out of 25 cases (80%) were diagnosed as schwannoma[8].

We retrospectively reviewed the MRI findings and found the following characteristic features of benign nerve sheath tumors that can also be seen in schwannomas (Fig. 3). First, "low signal margin" can be seen as a low signal rim surrounding the mass. It corresponds to the epineurium covering the schwannoma[9]. Second, "target sign" can be seen on T2-weighted images as a peripheral high signal rim and central low signal intensity within the mass. This pattern corresponded histologically to peripheral myxomatous and central fibrous tissues with high collagen content[10]. A previous report suggested that the target sign is more commonly seen in a neurofibroma (58%), which is also one of the common benign nerve sheath tumors, but can also be detected in a schwannoma (15%)[11]. Third,

"entering-and-exiting-nerve sign" can be seen as a high signal situated longitudinally to a fusiform mass on T2-weighted images[9,11]. Finally, "fascicular sign" can be seen as multiple small ring-like structures within the lesion with peripheral higher signal intensity. Fascicular appearances are significantly suggestive of schwannomas (63%) than neurofibromas (25%)[12]. These features can be helpful clues in diagnosing schwannomas.

In South Korea as well as in other countries, dermatologists deal with various skin tumors, and cases of surgical treatments by dermatosurgeons increase. Therefore, it is necessary to make a proper diagnosis through preoperative clinical and radiologic examinations because it may influence treatment options or surgical methods. Clinical findings are often insufficient to distinguish schwannoma from other common tumors. Therefore, it is necessary for dermatosurgeons to understand the typical imaging findings of common scalp tumors. Commonly benign tumors of the scalp that require a differential diagnosis from schwannoma include epidermal cyst and lipoma. Differential clinical, histologic, and radiologic characteristics of schwan-
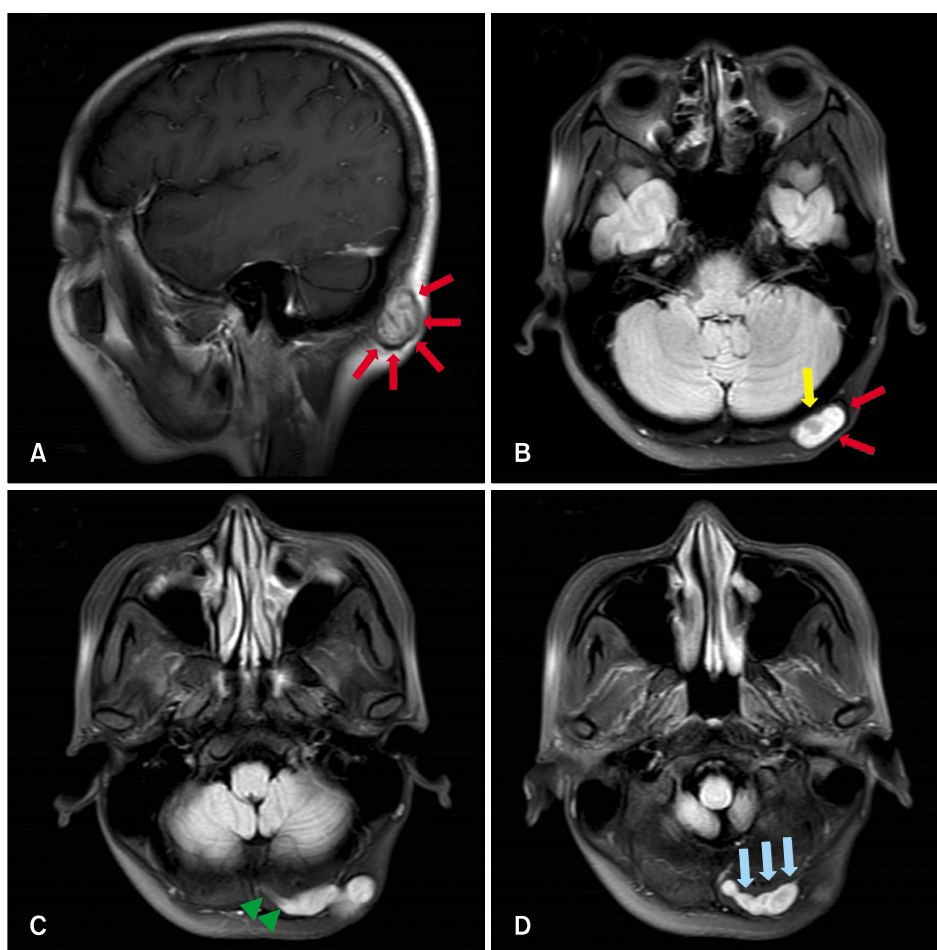

Fig. 3. Magnetic resonance imaging findings. (A) A T1-weighted sagittal image, (B~D) T2-weighted transverse images, and several characteristic features of a schwannoma: low signal margin (red arrows), target sign (yellow arrow), entering-and-exiting-nerve sign (green arrowheads), and fascicular sign (blue arrows).

Table 1. Differential characteristics of schwannoma and epidermal cyst and lipoma

Characteristic	Epidermal cyst	Lipoma	Schwannoma
Clinical[13]	Dermal or subcutaneous mobile nodules with a central punctum Foul smelling cheesy debris Rupture can occur	Painless, slowly enlarging mass involving the subcutaneous tissue	Soft, asymptomatic, dermal, or subcutaneous papules or nodules
Histologic[13]	Stratified, squamous lining with an intact granular layer Cysts contain central, eosinophilic, keratinaceous debris	Circumscribed mass surrounded by a thin fibrous capsule Composed of lobules of mature white adipose tissue divided by fibrous septa	Well-encapsulated mass with spindled, elongated, and wavy appearance cells High cellular areas with verocay bodies (Antoni A) alternate with hypocellular areas (Antoni B)
Imaging US[14]	Well-circumscribed, oval-shaped, hypoechoic masses with occasional linear anechoic and/or echogenic reflections[15] Posterior acoustic enhancement[16] Variable findings on Doppler flow depend on the phases (increase in the periphery during the inflamed and ruptured phases)[16]	Well-circumscribed mass that can have variable echogenicity (59% isoechoic, 26% hyperechoic, 15% hypoechoic) No posterior acoustic enhancement No or minimal color Doppler flow	Well-circumscribed hypoechoic mass Posterior acoustic enhancement Internal vascular flow on color Doppler imaging
CT	Low-to-intermediate-attenuated cystic mass	Low-attenuated cystic mass	Low-to-intermediate-attenuated cystic mass
MRI	Intermediate to high T2 signal mass[15] Occasional low signal debris and thin peripheral enhancement with no central enhancement after the administration of intravenous contrast material[15]	Fat signal intensity acquired with any pulse sequence[17] No enhancement after the administration of intravenous contrast material[17]	Isointense-to-low T1 signal, heterogeneously high T2 signal mass Avid enhancement after the administration of intravenous contrast material[18] Several characteristic MR findings: low signal margin, target sign, entering-and-exiting-nerve sign, and fascicular sign

US: ultrasonography, CT: computed tomography, MRI: magnetic resonance imaging.

noma with epidermal cyst and lipoma are summarized in Table 1[13-18]. Imaging studies can be helpful in the differentiation of benign tumors from malignant soft tissue tumors in head and neck areas such as liposarcoma, fibrosarcoma, malignant fibrous histiocytoma, rhabdomyosarcoma, etc. Large size (generally ≥5 cm), extracompartmental extension, poorly defined margins, broad interface with underlying fascia, intratumoral hemorrhage or necrosis, invasion of bone or neurovascular structures, heterogenoues MRI signal intensity, high signal intensity on T2-weighted MRI images and marked, primarily peripheral enhancement are suggestive imaging features of malignant soft tissue tumors in the head and neck[19,20].

The surgical approach of cutaneous schwannoma needs more consideration because complications such as neurologic deficits by peripheral nerve damage can develop. If neurologic complication of surgery is present, observation or partial removal is recommended. In our case, the patient did not report any neurologic complication after surgery.

Here, we report a rare case of scalp schwannoma with a review of MRI findings. Adequate understanding of char-

acteristic MRI findings of schwannoma will be helpful for clinicians to diagnose and manage this tumor properly.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

ORCID

Jong Seo Park, https://orcid.org/0000-0002-9662-0970
Jungyoon Moon, https://orcid.org/0000-0002-7575-0063
Soo Ick Cho, https://orcid.org/0000-0003-3414-9869
Je-Ho Mun, https://orcid.org/0000-0002-0734-2850

REFERENCES

1. Noh S, Do JE, Park JM, Jee H, Oh SH. Cutaneous schwannoma presented as a pedunculated protruding mass. Ann Dermatol 2011;23(Suppl 2):S264-S266.
2. Zhang H, Cai C, Wang S, Liu H, Ye Y, Chen X. Extracranial head and neck schwannomas: a clinical analysis of 33 pa-

tients. Laryngoscope 2007;117:278-281.

3. Colreavy MP, Lacy PD, Hughes J, Bouchier-Hayes D, Brennan P, O'Dwyer AJ, et al. Head and neck schwannomas--a 10 year review. J Laryngol Otol 2000;114:119-124.

4. Asuquo ME, Nwagbara VI, Akpan SO, Otobo FO, Umeh J, Ebughe G, et al. Recurrent benign schwannoma of the scalp: Case report. Int J Surg Case Rep 2013;4:65-67.

5. Mohan Kh, Manjunath H. Cutaneous schwannoma masquerading as trichilemmal cyst over scalp in a young male. Indian J Dermatol 2013;58:407.

6. Kim YC, Park HJ, Cinn YW, Vandersteen DP. Benign glandular schwannoma. Br J Dermatol 2001;145:834-837.

7. Kang GC, Soo KC, Lim DT. Extracranial non-vestibular head and neck schwannomas: a ten-year experience. Ann Acad Med Singapore 2007;36:233-238.

8. Yasumatsu R, Nakashima T, Miyazaki R, Segawa Y, Komune S. Diagnosis and management of extracranial head and neck schwannomas: a review of 27 cases. Int J Otolaryngol 2013; 2013:973045.

9. Salunke AA, Chen Y, Tan JH, Chen X, Foo TL, Gartner LE, et al. Intramuscular schwannoma: clinical and magnetic resonance imaging features. Singapore Med J 2015;56:555-557.

10. Varma DG, Moulopoulos A, Sara AS, Leeds N, Kumar R, Kim EE, et al. MR imaging of extracranial nerve sheath tumors. J Comput Assist Tomogr 1992;16:448-453.

11. Petscavage-Thomas JM, Walker EA, Logie CI, Clarke LE, Duryea DM, Murphey MD. Soft-tissue myxomatous lesions: review of salient imaging features with pathologic comparison. Radiographics 2014;34:964-980.

12. Jee WH, Oh SN, McCauley T, Ryu KN, Suh JS, Lee JH, et al. Extraaxial neurofibromas versus neurilemmomas: discrimination with MRI. AJR Am J Roentgenol 2004;183:629-633.

13. Wolff K, Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ. Fitzpatrick's dermatology in general medicine. 8th ed. New York: McGraw-Hill, 2012:1333-1334, 1476-1477, 1489-1490.

14. DiDomenico P, Middleton W. Sonographic evaluation of palpable superficial masses. Radiol Clin North Am 2014;52: 1295-1305.

15. Kim HK, Kim SM, Lee SH, Racadio JM, Shin MJ. Subcutaneous epidermal inclusion cysts: ultrasound (US) and MR imaging findings. Skeletal Radiol 2011;40:1415-1419.

16. Wortsman X. Common applications of dermatologic sonography. J Ultrasound Med 2012;31:97-111.

17. Beaman FD, Kransdorf MJ, Andrews TR, Murphey MD, Arcara LK, Keeling JH. Superficial soft-tissue masses: analysis, diagnosis, and differential considerations. Radiographics 2007; 27:509-523.

18. Skolnik AD, Loevner LA, Sampathu DM, Newman JG, Lee JY, Bagley LJ, et al. Cranial nerve schwannomas: diagnostic imaging approach. radiographics 2016;36:1463-1477.

19. Razek AA, Huang BY. Soft tissue tumors of the head and neck: imaging-based review of the WHO classification. Radiographics 2011;31:1923-54.

20. De Schepper AM, Bloem JL. Soft tissue tumors: grading, staging, and tissue-specific diagnosis. Top Magn Reson Imaging 2007; 18:431-444.