White epidermoid cyst transformation after stereotactic radiosurgery: illustrative case

Hideki Matsumoto, MD,1 Yuki Shinya, MD, PhD,1 Satoru Miyawaki, MD, PhD,1 Masahiro Shin, MD, PhD,1,2 Satoshi Koizumi, MD,1 Dai-Suke Sato, MD,1 Munetoshi Hinata, MD, PhD,1 Masako Ikemura, MD, PhD,1 Satoshi Kiyofuji, MD, PhD,1 Taich Kin, MD, PhD,1 Mototaro Iwanaga, MD, PhD,4 Masahiro Shimizu, MD, PhD,4 Hirofumi Nakatomi, MD, PhD,1,5 and Nobuhito Saito, MD, PhD1

Departments of 1Neurosurgery and 3Pathology, The University of Tokyo Hospital, Tokyo, Japan; 2Department of Neurosurgery, Teikyo University Hospital, Tokyo, Japan; 4Department of Neurosurgery, Kanto Neurosurgical Hospital, Kumagaya, Japan; and 5Department of Neurosurgery, Kyorin University Hospital, Tokyo, Japan

BACKGROUND White epidermoid cysts (WECs) are a rare type of epidermoid cyst with atypical radiological features. The epidemiological aspects and mechanisms of their onset remain unknown. Herein, the authors report a unique case of WEC transformation from a typical epidermoid cyst after stereotactic radiosurgery (SRS), confirmed by radiological and pathological findings.

OBSERVATIONS The case involved a 78-year-old man with a history of 2 surgeries for a left cerebellopontine angle typical epidermoid cyst 23 years earlier and SRS using the CyberKnife for recurrent trigeminal neuralgia (TN) 14 years earlier. The tumor with high intensity on T1-weighted imaging, low intensity on T2-weighted imaging, without restriction on diffusion-weighted imaging had gradually enlarged after SRS. Therefore, a salvage surgery was performed via a left suboccipital craniotomy, and the intraoperative findings showed a cyst with a brown, viscous liquid component, consistent with those of WECs. Histopathologically, keratin calcification and hemorrhage were identified, leading to a diagnosis of WEC. The postoperative course was uneventful, and the TN resolved. No tumor recurrence was recorded at 2 years postoperatively.

LESSONS To the best of the authors’ knowledge, this is the first world case of WEC transformation from a typical epidermoid cyst after SRS, confirmed by radiological and pathological findings. Radiation effects could have been involved in this transformation.

https://thejns.org/doi/abs/10.3171/CASE2376

KEYWORDS white epidermoid cyst; epidermoid cyst; stereotactic radiosurgery; CyberKnife radiosurgery; radiation effects

A 78-year-old man presented with left-sided trigeminal neuralgia. Radiological examinations revealed a left cerebellopontine angle tumor. He underwent tumor removal twice via left suboccipital craniotomy, and the tumor was pathologically confirmed as a typical epidermoid cyst with a flattened keratinized, stratified squamous epithelium surrounding the keratinized cyst. Ten years after stereotactic radiosurgery (SRS), the tumor gradually enlarged. A salvage surgery via left suboccipital craniotomy was performed, and the intraoperative findings showed a brown, viscous liquid component consistent with white epidermoid cysts. Histopathologically, keratin calcification and hemorrhage were identified, leading to a diagnosis of WEC.
postoperatively, the left TN recurred, SRS using the CyberKnife (Accuray, Inc.) was performed, and the remaining tumor was irradiated with a part of the total dose (Fig. 2). After SRS, the tumor gradually increased in size (Fig. 3), and the low-intensity tumor on T1WI (Fig. 3A) changed into a high-intensity tumor (Fig. 3B).

Fourteen years after SRS, the patient presented with left TN, and tumor recurrence was noted on computed tomography (CT; Fig. 4A). The tumor demonstrated high intensity on T1WI (Fig. 4B), low intensity on T2WI (Fig. 4C), and no diffusion restriction on DWI (Fig. 4D). Therefore, we performed salvage surgery via left suboccipital craniotomy (Fig. 5A and B). The intraoperative findings showed a cyst with a brown, viscous liquid component, consistent with those of WECs (Fig. 5C and D). Histopathologically, the tumor showed keratin calcification and hemorrhage and was confirmed as a WEC (Fig. 1C and D). The postoperative course was uneventful, and the TN resolved. No tumor recurrence was confirmed 2 years postoperatively.

**FIG. 1.** Pathological findings before and after recurrence. The pathological findings of the first surgical specimen show a flattened keratinized, stratified squamous epithelium surrounding the keratinized cyst, indicating a typical epidermoid cyst (A and B). The pathological findings after the recurrence show keratin calcification and internal bleeding, indicating a WEC (C and D).

**FIG. 2.** SRS using the CyberKnife with a marginal dose of 50 Gy on the 65% isodose line (maximum dose of 76.92 Gy) was performed for trigeminal neuralgia (A and B). The remaining tumor was irradiated with a maximum dose of 23 Gy (C and D). The blue and navy lines indicate 23 and 7.7 Gy, respectively (C and D).

**FIG. 3.** Radiological changes in the WEC in a 78-year-old man during recurrence. A low-intensity lesion in the left CPA on T1WI after the initial surgery (A) and a high-intensity lesion on T1WI 10 years after the first surgery (B) are shown, which gradually increased 17 years (C) and 22 years (D) after the first surgery.

**FIG. 4.** Imaging findings of the WEC after recurrence. Preoperative imaging shows a tumor with hypodensity on CT (A), hyperintensity on T1WI (B), well-defined hypointensity on T2WI (C), and no diffusion restriction on DWI (D) in the left CPA.
Discussion

Observations

Radiologically, typical epidermoid cysts demonstrate high density on noncontrast CT, low intensity on T1WI, and high intensity on T2WI. They are usually solid and have no triglycerides. WECs are atypical because of their high protein content and viscosity, resulting in high intensity on T1WI and low intensity on T2WI. The present case demonstrated the representative radiological features before and after WEC transformation. Historically, several studies have reported the peculiar radiological features of WECs. Braun et al. first reported a high-density epidermoid cyst on CT in 1977. They reported factors causing high density on CT, including hemosiderin deposition due to hemorrhage, keratin saponification, and saponification of debris to calcium. Ishikawa et al. reported that a low-density epidermoid on CT tended to show low intensity on T1WI and high intensity on T2WI. Finally, Horowitz et al. proposed an epidermoid, which demonstrated high intensity on T1WI, as a “white epidermoid” in 1990. In addition, Ren et al. defined “atypical epidermoid cysts” other than those exhibiting low intensity on T1WI and high intensity on T2WI and examined 428 epidermoid cysts. They found 24 cases (5.6%) of “atypical epidermoid cysts,” among which 14 were not preoperatively diagnosed as epidermoid cysts. Hemosiderin deposition was observed in 21 cases (87.5%) of “atypical epidermoid cysts,” and lesions, including hematomas, were considered to have contributed to atypical imaging findings.

Histopathologically, typical epidermoid cysts are filled with soft, waxy, or flaky material rich in debris, keratin, water, and cholesterol crystals. In contrast, WECs have a relatively high viscosity and protein concentration because inflammation, infection, and keratin desquamation result in degeneration of the cyst wall and increased protein levels. The present case also suggested histopathological changes before and after the WEC transformation. As mentioned earlier, there are various reports on the pathological findings of WECs; however, the detailed pathological mechanism of the occurrence of WECs has not been fully clarified.

Furthermore, several studies have reported malignant transformation of epidermoid to squamous cell carcinoma during recurrence, and the reported incidence was 0.011%–0.045%. The exact etiology of the WEC transformation remains indeterminate in this particular case. This transformation could be attributed to surgical intervention, SRS treatment, a combination of both, or it may simply represent a natural occurrence. Although all potential explanations cannot be definitively excluded, it is evident that the WEC transformation was confirmed after radiosurgery. To the best of our knowledge, there have been no prior documented cases of a typical epidermoid cyst undergoing a transformation into a WEC following SRS, making this case unique.

Lessons

This is the first case of WEC transformation from a typical epidermoid cyst after SRS, as revealed by radiological and pathological findings. SRS using a CyberKnife for TN could influence this histopathological change. A detailed evaluation cannot be deciphered based on the present case alone, but the radiation effect might have been involved in WEC transformation in our case. The unavailability of detailed images before the initial surgery could have limited this report. Further case accumulation is required to elucidate the detailed mechanism. In conclusion, we described a case of a left CPA WEC transformed from a typical epidermoid cyst after SRS. Epidermoid cysts can transform into WECs during recurrence, and radiation could be involved in this transformation.

References


**Disclosures**
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**Author Contributions**
Conception and design: Miyawaki, Matsumoto, Shinya, Sato, Shimizu. Acquisition of data: Miyawaki, Matsumoto, Shinya, Iwanaga. Analysis and interpretation of data: Miyawaki, Matsumoto, Shinya, Ikemura, Kiyofuji. Drafting of the article: Miyawaki, Matsumoto, Shinya, Shin. Critically revising the article: Miyawaki, Shinya, Shin, Koizumi, Sato, Ikemura, Kiyofuji, Iwanaga. Reviewed submitted version of the manuscript: Miyawaki, Shinya, Koizumi, Sato, Ikemura, Kiyofuji, Kin, Nakatomi, Saito. Approved the final version of the manuscript on behalf of all authors: Miyawaki. Statistical analysis: Miyawaki. Administrative/technical/material support: Miyawaki, Hinata, Kin. Study supervision: Miyawaki, Shinya, Nakatomi.

**Correspondence**
Satoru Miyawaki: The University of Tokyo, Japan. miyawaki-tky@umin.ac.jp.