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Cerebellopontine Angle Epidermoid Cyst With a Focal White Component and Target-Like Appearance: A Case Report

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Alondra Karent Balbuena Coutiño [1], Marco A. Rodriguez Florido [2], Aura Valentina Rangel Velázquez [2], Ariadna Antoinette Fernández Chiquito [3], Lizeth Díaz Yañez [4]

1. Department of Pathology, Unidad Médica de Alta Especialidad, Hospital de Especialidades Centro Médico Nacional "Ignacio García Téllez," Instituto Mexicano del Seguro Social, Merida, MEX 2. Department of Pathology, Hospital de Especialidades "Dr. Bernardo Sepúlveda Gutiérrez," Centro Médico Nacional Siglo XXI, Instituto Mexicano del Seguro Social, Mexico City, MEX 3. Department of Neurosurgery, Hospital Regional "1ero de Octubre" Instituto de Seguridad y Servicios Sociales de los Trabajadores del Estado (ISSSTE), Mexico City, MEX 4. Department of Neurosurgery, Hospital Regional de Alta Especialidad "Bicentenario de la Independencia," Instituto de Seguridad y Servicios Sociales de los Trabajadores del Estado (ISSSTE), Tultitlan, MEX

Corresponding author: Marco A. Rodriguez Florido, mar-florido@hotmail.com

Abstract

Intracranial epidermoid cysts typically exhibit cerebrospinal fluid-like signal intensity on magnetic resonance imaging (MRI), whereas white epidermoid cysts display atypical imaging characteristics, particularly intrinsic T1 hyperintensity. We report the case of a 64-year-old woman with progressive left-sided tinnitus and hearing loss, in whom an extra-axial lesion was identified in the left cerebellopontine angle. MRI demonstrated a predominantly T1-hypointense lesion with a central punctate hyperintense focus surrounded by a hypointense rim, creating a distinctive target-like appearance. The lesion showed no significant contrast enhancement and demonstrated true diffusion restriction. Surgical resection revealed a pearly, avascular cyst containing thick, whitish keratinous material. Histopathologic examination revealed a cyst lined by keratinizing stratified squamous epithelium, containing abundant lamellated keratin, compact keratin with a mosaic-like appearance, and focal amorphous eosinophilic material, confirming the diagnosis of an epidermoid cyst. The focal T1 hyperintensity may represent a localized manifestation of the white epidermoid phenotype, although this association remains hypothetical in the absence of biochemical characterization of the cyst contents. This case expands the recognized imaging spectrum of intracranial epidermoid cysts and highlights the importance of integrating radiologic, surgical, and histopathologic findings when evaluating atypical cerebellopontine angle lesions.

Categories: Radiology, Pathology, Neurosurgery
Keywords: central nervous system cysts, cerebellopontine angle, cystic lesions, differential diagnosis, epidermal cyst, magnetic resonance imaging

Introduction

Intracranial epidermoid cysts are congenital ectodermal inclusion lesions that arise from entrapment of squamous epithelium during neural tube closure, between the third and fifth weeks of embryonic development. They are uncommon lesions, accounting for approximately 0.2% to 1.8% of all intracranial space-occupying lesions, including both non-neoplastic inclusion cysts and intracranial neoplasms. Approximately 40% to 50% occur in the cerebellopontine angle, followed by the parasellar region in approximately 30%, and the fourth ventricle in up to 18% [1].

These cysts grow slowly as a result of continuous desquamation of the epithelial lining, leading to the progressive accumulation of keratin and lipid debris within the cyst lumen rather than true neoplastic proliferation. Because of their characteristic pearly, lobulated appearance, they have traditionally been referred to as "pearly tumors" [2]. Histologically, they are lined by keratinizing stratified squamous epithelium with a well-defined granular layer and contain abundant laminated keratin within the lumen [3].

Clinical manifestations depend primarily on the location and size of the lesion. In the cerebellopontine angle, common symptoms include headache, vertigo, trigeminal neuralgia, hearing loss, tinnitus, facial nerve palsy, and cerebellar signs. Hydrocephalus, seizures, and chemical meningitis are less common [1].

On magnetic resonance imaging (MRI), classic epidermoid cysts are typically hypointense on T1-weighted images, hyperintense on T2-weighted images, and markedly hyperintense on diffusion-weighted imaging [3]. A small proportion, estimated at 1.5% to 5.6%, shows spontaneous T1 hyperintensity and is referred to as a white epidermoid cyst. This variant may exhibit variable signal intensity on T2-weighted and diffusion-weighted sequences, making radiologic recognition more challenging [4-6].

We present the case of a 64-year-old woman with an epidermoid cyst of the cerebellopontine angle

exhibiting an unusual target-like appearance on MRI, characterized by a central punctate focus of intrinsic T1 hyperintensity surrounded by a circumferential hypointense rim. We describe this distinctive imaging pattern, correlate the radiologic findings with the gross and histopathologic features, and discuss its relationship to the white epidermoid variant and its main differential diagnoses.

Case Presentation

A 64-year-old woman with an 11-year history of type 2 diabetes mellitus and systemic arterial hypertension, both apparently well controlled, was evaluated. At 13 years of age, she underwent left ear surgery with placement of a tympanic graft. In addition, 13 years before the current evaluation, she underwent subtotal resection of a presumed pituitary adenoma through a sublabial transsphenoidal approach; however, the histopathologic report was unavailable, and no subsequent follow-up had been performed.

Two years before admission, she developed progressive left-sided tinnitus, initially intermittent and later persistent, associated with ipsilateral hearing loss. One month before admission, she also developed nocturnal left frontoparietal headache. On examination, she was alert, with a Glasgow Coma Scale score of 15 and left temporal hemianopia. Audiometry showed mild sensorineural hearing loss at high frequencies in the right ear and moderate-to-profound conductive hearing loss in the left ear.

Brain MRI revealed an extra-axial lesion located in the left cerebellopontine angle. On unenhanced T1-weighted imaging, the lesion was predominantly hypointense, with signal intensity similar to that of cerebrospinal fluid, and showed a punctate hyperintense center surrounded by a hypointense halo and fine concentric lines with slightly increased signal intensity, producing a target sign (Figure *1A*). After gadolinium administration, no significant enhancement was observed; persistence of the central hyperintense focus confirmed that it represented intrinsic T1 hyperintensity rather than contrast enhancement (Figure *1B*). This target-like appearance was most evident on T1-weighted sequences. On T2-weighted imaging, the lesion was isointense to mildly heterogeneous relative to the brain parenchyma (Figure *1C*), whereas on fluid-attenuated inversion recovery (FLAIR) imaging, it remained hyperintense, without the complete suppression characteristic of cerebrospinal fluid (Figure *1D*), and demonstrated diffusion restriction (Figure *1E*, *1F*). Overall, the absence of significant enhancement and the presence of diffusion restriction favored the radiologic diagnosis of an epidermoid cyst; however, the central T1 hyperintensity and target-like pattern represented atypical findings that complicated its preoperative recognition.

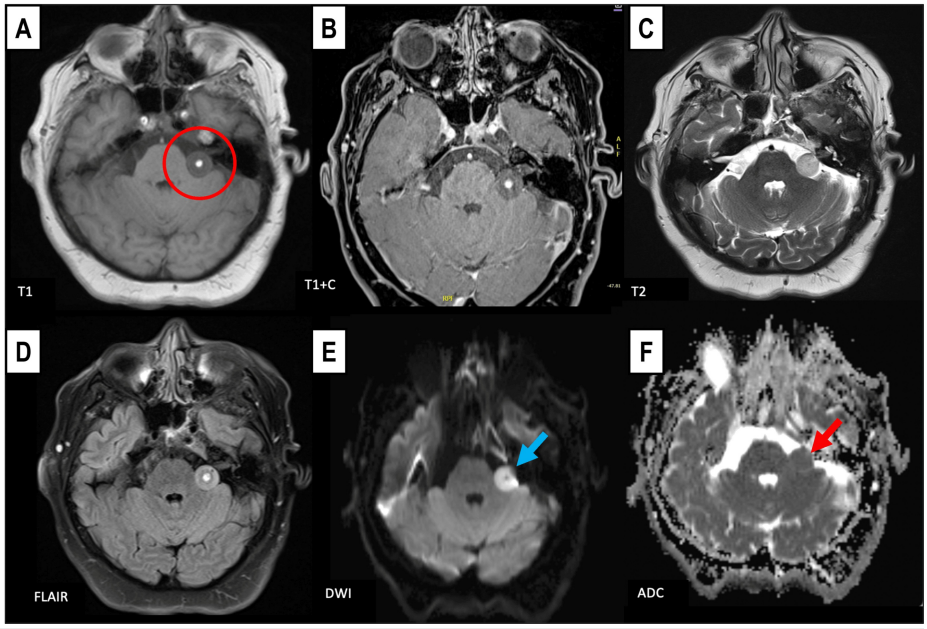


FIGURE 1: Preoperative magnetic resonance imaging of the extra-axial lesion located in the left cerebellopontine angle

(A) Axial unenhanced T1-weighted image showing a predominantly hypointense lesion with a punctate hyperintense center and a hypointense peripheral zone containing slightly hyperintense concentric laminations, producing the target sign (red circle). (B) Axial T1-weighted image after gadolinium administration (T1+C, contrast-enhanced T1-weighted imaging) showing no significant enhancement; the central hyperintense focus persists, consistent with intrinsic T1 hyperintensity. (C) Axial T2-weighted image. (D) Axial fluid-attenuated inversion recovery (FLAIR) image showing incomplete suppression of the lesion. (E) Axial diffusion-weighted imaging (DWI) showing marked hyperintensity (blue arrow). (F) Apparent diffusion coefficient (ADC) map showing corresponding decreased signal intensity (red arrow), consistent with true diffusion restriction.

A second lesion centered on the clivus, with extension into the petrous and mastoid portions of the left temporal bone, was also identified. This lesion showed low-to-intermediate signal intensity on T1-weighted imaging, no significant enhancement after gadolinium administration, and heterogeneous signal intensity on T2-weighted imaging (Figure *2*). Although diffusion restriction was also described in the radiologic assessment, this finding is not illustrated in Figure *2*. Based on its imaging characteristics, a second epidermoid cyst was considered; however, because this lesion was not surgically treated or histopathologically examined, its nature could not be confirmed and remained a presumptive radiologic diagnosis.

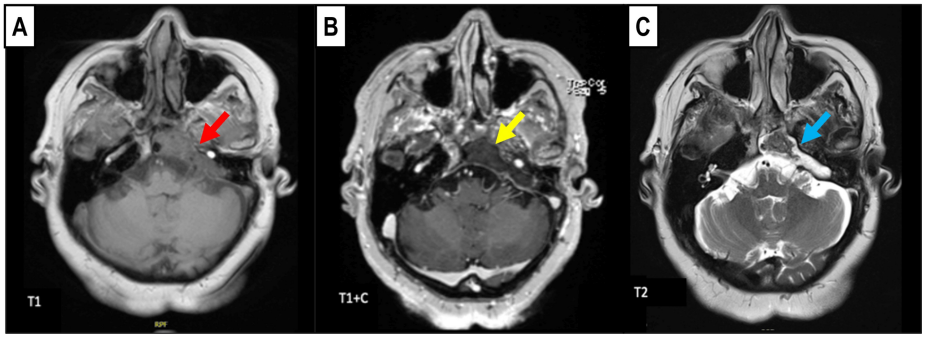


FIGURE 2: Lesion centered on the clivus, with extension into the petrous and mastoid portions of the left temporal bone, of indeterminate etiology

(A) Axial unenhanced T1-weighted image, lesion (red arrow). (B) Axial T1-weighted image after gadolinium administration (T1+C, contrast-enhanced T1-weighted imaging), showing no significant enhancement; lesion (yellow arrow). (C) Axial T2-weighted image showing heterogeneous signal intensity; lesion (blue arrow). The lesion remained a presumptive radiologic diagnosis without histopathologic confirmation; diffusion-weighted sequences are not shown in this figure.

A retrosigmoid approach was performed through a subasterional craniotomy, with resection of the cerebellopontine angle lesion under intraoperative neuromonitoring and dural reconstruction using a pericranial graft. During the procedure, a pearly, avascular lesion with easily aspirated contents was identified. The lesion showed a well-defined dissection plane and no evidence of invasion into the adjacent brain parenchyma. On intraoperative pathologic examination, an ovoid lesion with a smooth, shiny, pearly-white external surface was received. On sectioning, the wall showed concentric laminations, and the cavity was filled with thick, whitish, keratinous-appearing material (Figure *3*). These gross findings were consistent with an epidermoid cyst and correlated with the radiologic features.

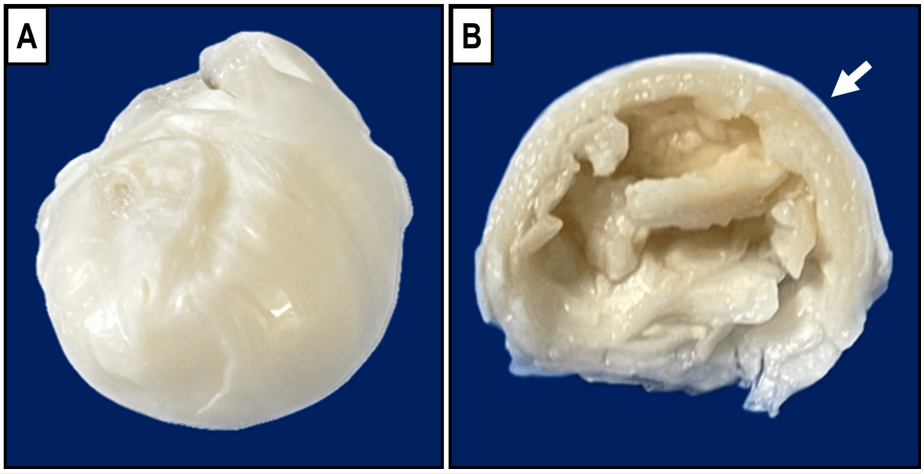


FIGURE 3: Gross findings of the surgical specimen

(A) External surface of the lesion. (B) Cut-surface appearance showing concentric laminations of the cyst wall (white arrow).

On histopathologic examination, the cyst wall was lined by keratinizing stratified squamous epithelium. Laminated keratin predominated within the cyst wall, while the lumen contained abundant keratin lamellae, with compact areas showing polygonal contours that produced a mosaic-like appearance. Focally, a gradual transition from laminated and compact keratin to amorphous eosinophilic material was identified. This latter finding may represent modification of the keratinous contents and could contribute to the lesion's atypical radiologic features (Figure *4*). No adipose tissue or hemorrhage was identified.

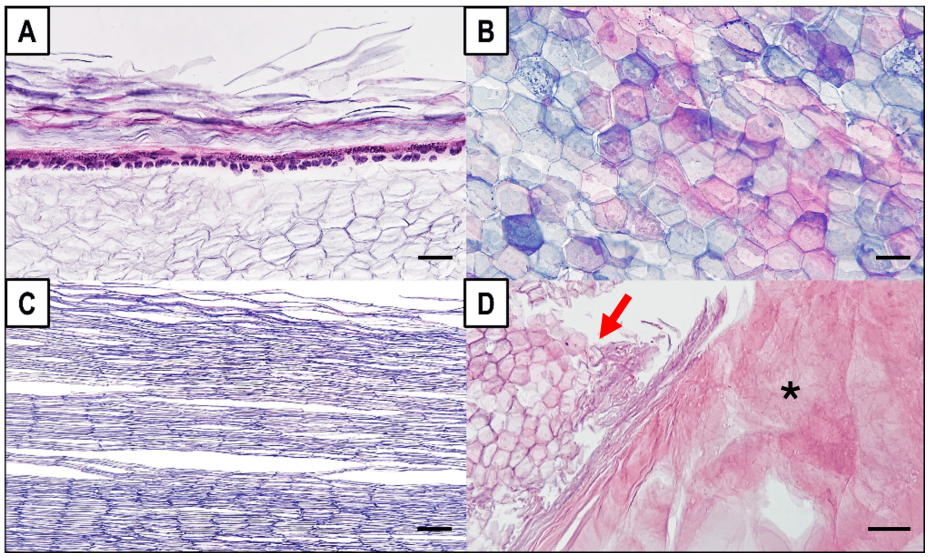


FIGURE 4: Hematoxylin and eosin-stained photomicrographs of the epidermoid cyst

(A) Keratinizing stratified squamous epithelium (40x). (B) Compact keratin with a mosaic-like appearance (40x). (C) Lamellated keratin (40x). (D) Lamellated keratin (red arrow) adjacent to amorphous eosinophilic material (asterisk) (10x). Scale bars: 20 µm in A-C and 50 µm in D.

The patient had a favorable postoperative course and was discharged without complications, with instructions for surgical wound care, analgesia, and outpatient follow-up with her treating physician at eight weeks. At follow-up, tinnitus had improved, and the neurological examination was normal. Postoperative MRI showed expected postsurgical changes, with no evidence of residual lesion at the surgical site. Annual clinical and radiological follow-up was planned with her treating physician, including surveillance of the untreated lesion involving the clivus/temporal bone.

Discussion

Target-like appearance and its relationship to the white epidermoid phenotype

The distinctive feature of this case is an unusual target-like appearance on T1-weighted MRI. To our knowledge, this specific configuration has not been clearly characterized in an intracranial epidermoid cyst. A similar pattern, known as the bull's-eye sign, has been described in testicular epidermoid cysts on ultrasonography, where it reflects the concentric arrangement of their components. Sonographically, this appearance may consist of a peripheral echogenic rim, an intermediate hypoechoic region, and a central echogenic focus corresponding to keratinous debris. On MRI, testicular epidermoid cysts may demonstrate a peripheral low-signal-intensity rim, an intermediate zone of higher signal intensity containing desquamated keratin, and a low-signal-intensity center composed of dense debris [6].

In our case, although the signal distribution does not exactly reproduce these previously described patterns, a central punctate T1-hyperintense focus is surrounded by a concentric hypointense zone, producing a well-defined target-like appearance. This unusual configuration represents the main distinctive imaging feature of the case. Furthermore, the lesion may exhibit a focal component resembling the white epidermoid phenotype, as intrinsic T1 hyperintensity is atypical of conventional epidermoid cysts.

Historically, atypical epidermoid cysts were first described by Braun et al. in 1977 as "dense epidermoid cysts" based on their hyperdense appearance on computed tomography (CT) [7]. With the subsequent development of MRI, the term "white epidermoid cyst" came to be used for epidermoid cysts exhibiting spontaneous T1 hyperintensity, although CT hyperdensity does not necessarily correlate with T1 hyperintensity [2]. White epidermoid cysts account for approximately 1.5% to 5.6% of epidermoid cysts and are characterized by intrinsic T1 hyperintensity, with variable T2 signal intensity, including hypointensity associated with the so-called shading sign [1,5].

Although most reported white epidermoid cysts demonstrate more diffuse T1 hyperintensity, no established quantitative criterion defines the extent of hyperintense signal required for this designation. Furthermore, conventional and white signal characteristics have been reported within the same lesion [8], supporting the possibility that the white phenotype may be expressed regionally. Because conventional epidermoid cysts

are typically hypointense on T1-weighted images or demonstrate signal intensity similar to cerebrospinal fluid, we interpret the punctate intrinsic T1 hyperintensity observed in our case as a possible unusually focal manifestation of the white epidermoid phenotype.

Possible pathophysiologic basis of atypical signal characteristics

The atypical signal characteristics of white epidermoid cysts have been associated with alterations in the composition and physical properties of their contents. Chemical analysis of a posterior fossa epidermoid cyst exhibiting T1 hyperintensity and T2 hypointensity demonstrated a high protein concentration, predominantly albumin, together with marked viscosity. In contrast, calcium and iron concentrations were insufficient to explain the radiologic findings [9].

In addition, these changes may develop progressively over time, as documented in a case involving the transformation of a typical epidermoid cyst into a white epidermoid cyst after 13 years of follow-up, possibly due to the progressive accumulation and degeneration of proteinaceous and lipid material within the cyst [10]. However, because no chemical analysis of the cyst contents was performed in our patient, the relationship between the target-like appearance and the biochemical composition of the cyst remains a pathophysiologic hypothesis rather than a demonstrated finding.

Differential diagnosis

The differential diagnosis of an extra-axial cerebellopontine angle lesion with a T1-hyperintense focus against a predominantly hypointense background includes dermoid cyst, lipoma, neurenteric cyst, and other lesions containing proteinaceous or hemorrhagic material. An arachnoid cyst may mimic a classic epidermoid cyst on conventional sequences, but it generally follows cerebrospinal fluid signal on FLAIR and does not demonstrate diffusion restriction. Dermoid cysts and lipomas show imaging characteristics of fat, whereas schwannomas and meningiomas typically enhance after contrast administration [11]. In this case, the absence of significant enhancement and the presence of diffusion restriction favored an epidermoid lesion, although the central T1 hyperintensity complicated its preoperative recognition and initially led to consideration of a schwannoma.

The second lesion, centered on the clivus and extending into the petrous and mastoid portions of the left temporal bone, warrants a broader differential diagnosis. Although the lack of enhancement and the reported diffusion restriction are compatible with a second epidermoid cyst, cholesteatoma must also be considered, given the lesion's location, the patient's history of left-ear surgery with tympanic membrane grafting, and her moderate-to-profound left-sided conductive hearing loss. Cholesteatoma also exhibits diffusion restriction and shares a keratinizing squamous epithelial lining with epidermoid cysts [4].

Other differential diagnoses include cholesterol granuloma, which typically exhibits hyperintensity on T1-weighted images and is therefore less consistent with the low-to-intermediate T1 signal observed in this case; chordoma; chondrosarcoma; petrous apex effusion or mucocele; and residual or recurrent disease related to the previous transsphenoidal surgery, for which histopathological findings were unavailable [10]. In the absence of histopathological confirmation, the nature of this lesion remains undetermined.

Limitations and clinical implications

The main limitation of this case is that the proposed relationship between the target-like appearance and the white epidermoid phenotype remains hypothetical. Although the focal intrinsic T1 hyperintensity suggests a possible localized expression of this phenotype, no biochemical analysis of the cyst contents was performed to investigate the underlying mechanism responsible for this imaging pattern. Furthermore, as this is a single case, the specificity and reproducibility of the target-like appearance cannot be established. Nevertheless, this case expands the recognized imaging spectrum of intracranial epidermoid cysts and suggests that focal T1 hyperintensity may represent an atypical manifestation of the white epidermoid phenotype.

Conclusions

We present a case of an epidermoid cyst of the cerebellopontine angle exhibiting an unusual target-like appearance on T1-weighted MRI, characterized by a central punctate focus of intrinsic hyperintensity surrounded by a hypointense rim. Histopathological examination revealed conventional features of an epidermoid cyst, without evidence of fat or hemorrhage. The focal T1 hyperintensity may represent a localized expression of the white epidermoid phenotype, although its underlying pathophysiological mechanism remains unproven. The observed transition from laminated keratin to amorphous eosinophilic material may provide a possible histopathological correlate; however, its relationship to the imaging pattern has not been established. This case expands the recognized imaging spectrum of intracranial epidermoid cysts and highlights the importance of considering this entity in the differential diagnosis of non-enhancing extra-axial cerebellopontine angle lesions with a target-like appearance, particularly when true diffusion restriction is present.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Alondra Karent Balbuena Coutiño, Marco A. Rodriguez Florido

Acquisition, analysis, or interpretation of data: Alondra Karent Balbuena Coutiño, Ariadna Antoinette Fernández Chiquito, Aura Valentina Rangel Velázquez, Lizeth Díaz Yañez

Drafting of the manuscript: Alondra Karent Balbuena Coutiño, Ariadna Antoinette Fernández Chiquito, Aura Valentina Rangel Velázquez, Lizeth Díaz Yañez

Critical review of the manuscript for important intellectual content: Alondra Karent Balbuena Coutiño, Marco A. Rodriguez Florido

Supervision: Marco A. Rodriguez Florido

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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