The Journal of International Advanced Otology

J Int Adv Otol 2020; 16(3): 473-5 • DOI: 10.5152/iao.2020.7854

Case Report

Mastoid Dermoid Cyst

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Cite this article as: Pepe G, Canzano F, Piccinini S, Corcione L, Falcioni M. Mastoid Dermoid Cyst. J Int Adv Otol 2020; 16(3): 473-5.

A case of mastoid dermoid cyst (DC) was presented, and differences with cases of other temporal bone DCs were analyzed. The mastoid DC was also compared with mastoid congenital cholesteatoma. We reported a case of a patient with mastoid DC, evaluating her clinical, radiological, and surgical findings. A review of the literature was performed to compare our findings with those reported. The preoperative radiological evaluation prompted us to plan a surgical approach to the lesion, suspecting the presence of a mastoid congenital cholesteatoma. The surgical findings were in line with the presence of a mastoid DC. Only two cases reported in the literature presented features that fulfilled the criteria of a true mastoid DC. A DC confined to the mastoid region is an extremely rare clinical entity, with asymptomatic and slow growth. Preoperative radiological differentiation between congenital cholesteatoma and DCs with atypical features can be difficult. However, surgical excision is the treatment of choice in both cases. Diagnosis is confirmed by the histological evaluation.

KEYWORDS: Dermoid cyst, mastoid, otologic surgical procedure, review

INTRODUCTION

Dermoid cysts (DCs) are rare ectodermal and mesodermal inclusion cysts. In the head and neck, they account for 7% of all dermoids and are more often located at the midline, in the orbit region and in oral and nasal cavities. Intracranial dermoid cysts represent less than 1% of all intracranial masses [1]. Several cases of DCs of the temporal bone have been reported in the literature but only a few cases were confined to the mastoid, with different clinical features. We reported a case of an asymptomatic mastoid DC.

CASE PRESENTATION

A 49-year-old woman presented with a history of multiple cerebral angiomas requiring serial imaging controls. Otoscopy showed a mesotympanic retraction pocket in the left ear, whereas the right ear was normal. Pure-tone audiometry showed bilateral mixed hearing loss: right ear bone conduction (BC) pure-tone average (PTA) was 40 dB and air conduction (AC) PTA was 65 dB, whereas left ear BC PTA was 33.3 dB and AC PTA had a value of 60 dB, owing to otosclerosis that was previously diagnosed and surgically treated in another hospital. No other otologic symptoms were referred.

During radiological follow-up, a mass occupying the left posterolateral mastoid area was identified, extending into the retrosigmoid bone, with evidence of growth. Magnetic resonance imaging (MRI) scan demonstrated a well-marginated intradiploic cystic lesion hypointense on T1 and hyperintense on T2 sequences with restricted diffusion on diffusion weighted imaging (DWI) performed with non–echo planar imaging technique (Figure 1). However, the anterior portion of the lesion did not show any restricted diffusion; on the contrary, it appeared hyperintense on T1 and hypointense on T2 sequences, with features suggestive of fat tissue. Temporal bone high-resolution computed tomography (CT) scan confirmed an osteolytic lesion eroding the sigmoid sinus bony plate and the mastoid cortex, without reaching the antrum and the tegmen tympanic cavity. In addition, soft tissue CT scan showed a little fatty component of the cyst lining the anterior margin (Figure 2). All radiological features were suggestive of a mastoid congenital cholesteatoma. A mastoidectomy extending to the retrosigmoid area was planned. In surgery, subcutaneous tissues were unaffected and there was no contact/extension toward the skin. The mass appeared well encapsulated and tight adherent to the sigmoid sinus wall; however, a cleavage plane was established between the sinus and the cyst. Bleeding from the origin of the superior petrosal sinus occurred, which was controlled with bone wax (B.Braun Surgical, S.A., Rubi, Spain). Following the incision of the lesion, whitish content with hair was observed (Figures 3 and 4). Complete removal of the cyst was achieved, and the sigmoid sinus was subsequently covered with bone patè. The post-operative period was uneventful. A CT scan taken 3 months post-operatively showed the reconstruction of the sigmoid sinus bony plate. Histological examination confirmed the diagnosis of DC, show-

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Submitted: 10.25.2019 • **Revision Received:** 02.23.2020 • **Accepted:** 03.05.2020
Available online at www.advancedotology.org

ing a stratified squamous epithelium with epidermal appendages (Figure 5). The patient's hearing threshold was not affected by the surgery.

DISCUSSION

Some confusion exists in the literature regarding the terminology because DC, true teratoma, and cholesteatoma have been used to describe similar lesions. According to the histological classification, dermoid cysts are composed of both ectoderm and mesoderm layers; therefore, they are described as skin-covered lesions that contain epidermal appendages and adipose matrix. Differently, true teratomas have all three germ layers (ectoderm, mesoderm, and endoderm) and cholesteatomas are only of ectodermal origin [2].

True DCs are rare and mainly reported intracranially at the midline [3, 4]. The temporal bone is an uncommon site of origin of these lesions, with most reports involving the tympanic cavity and/or the Eustachian tube. In addition, three cases of DC occupying the petrous bone apex have been reported [5-7]. Nwojo et al.[8] presented an additional case originating in the mastoid segment of the fallopian canal, causing facial paralysis in a 9-month-old child.

The tympanic cavity and the Eustachian tube are the most frequently involved regions, with 31 cases reported overall [9]. Some of these reports describe a secondary involvement of the antrum and mastoid cells. All cases of DC originating in the tympanic cavity or Eustachian tube were characterized by clinical features and symptoms such as ear discharge, presence of retrotympanic masses or aural polyps, and effusive otitis media.

According to Steel's literature review [10], only four cases of DC involving exclusively the mastoid have been described, three of them published between 1866 and 1938, with no documentation. Toynbee first described a mastoid DC discovered during an autopsy in 1866;

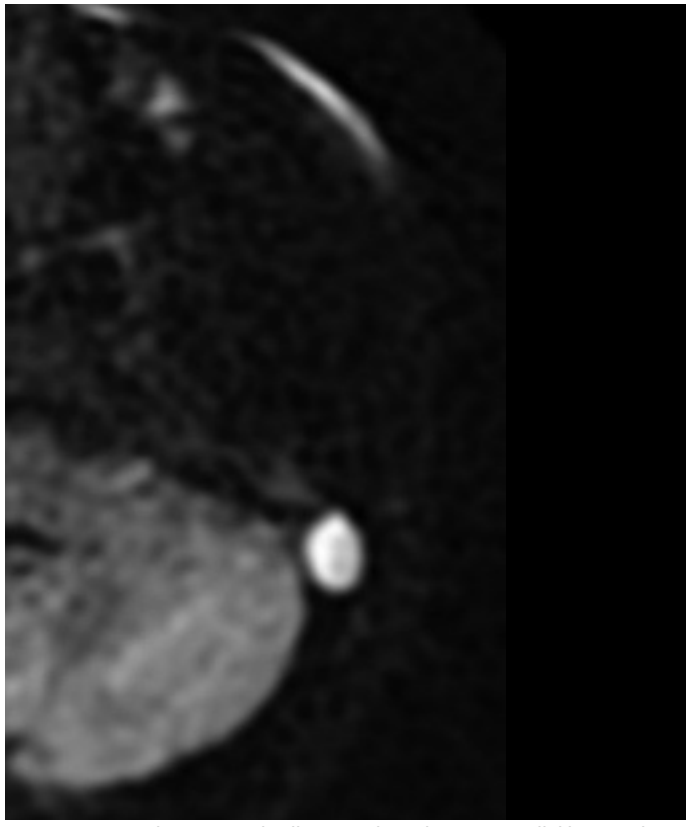

Figure 1. On axial DWI periodically rotated overlapping parallel lines with enhanced reconstruction (PROPELLER) imaging, the lesion shows evident diffusion restriction.

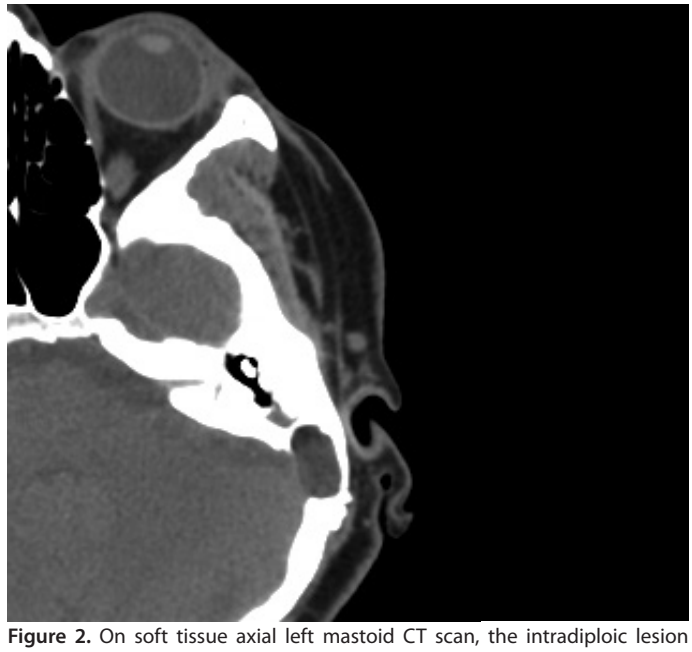

Figure 2. On soft tissue axial left mastoid CT scan, the intradiploic lesion demonstrates a fatty component anterior to a main part with cerebrospinal fluid density.

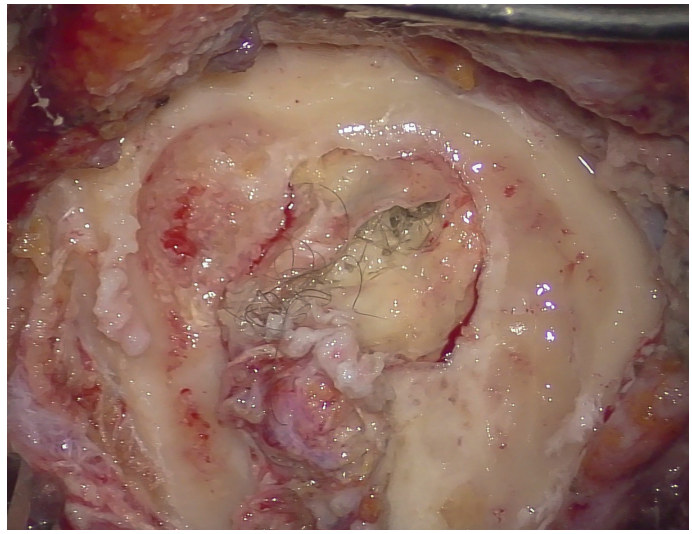

Figure 3. Once opened, the lesion showed keratin accumulation with hairs in the intraoperative view.

MAIN POINTS

- Several case of dermoid cysts (DC) of the temporal bone have been described in literature. Nevertheless, the confinement of a DC to the mastoid is a rare feature.
- The asymptomatic course of these lesions increases the difficulty encountered during the diagnosis. Furthermore, imaging studies (CT-scan and MRI) not always allow in differentiating DCs from congenital mastoid cholesteatoma.
- Surgical excision is the treatment of choice. Removing the entire cystic wall is mandatory in order to avoid recurrence of the DC.

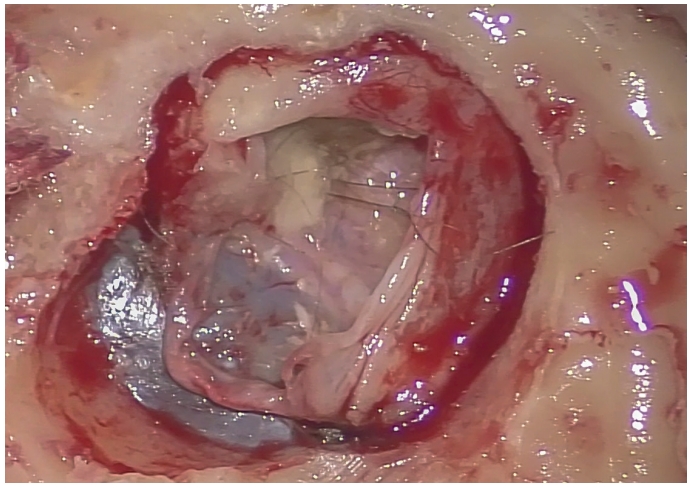

Figure 4. After debulking and partial removal of the lesion, the sigmoid sinus is clearly in view.

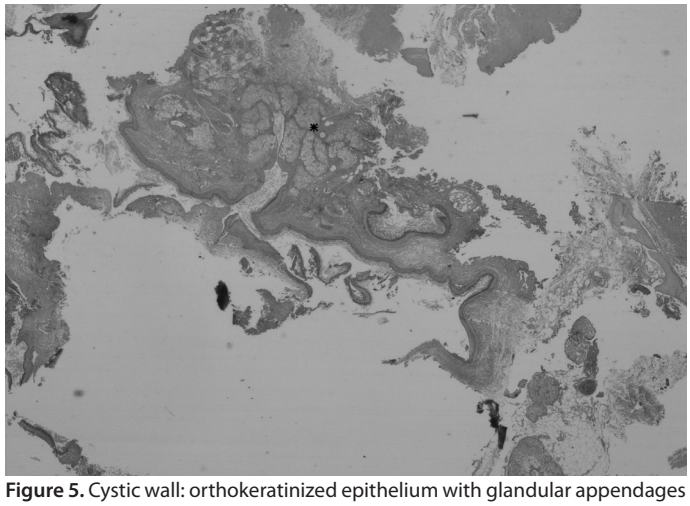

Figure 5. Cystic wall: orthokeratinized epithelium with glandular appendages (asterisk).

in 1888, Wagenhauser reported a case of recurrent abscesses in the mastoid region and a post-auricular fistula containing hairs, determined by a mastoid DC; Watanabe found a mastoid DC associated to tympanic perforation and a discharging ear [10]. Steel [10] described a case of recurrent secretory otitis media and hearing loss that led to a surgical exploration of the mastoid and middle ear, finding a defect in the mastoid bony cortex, through which a leash of hair protruded, histologically diagnosed as a DC. Of all the four cases, those described by Toynbee and Wagenhauser can be surely considered as true DCs confined to the mastoid; on the contrary, Steel and Watanabe described cases in which the patients presented symptoms not justified by an isolated mastoid lesion, but suggesting the contemporary involvement of the middle ear or a concomitant middle ear disease. Indeed, because of the anatomical features of the mastoid, dermoid cysts in this area usually grow asymptomatically until reaching a significant size, as in the case of congenital mastoid cholesteatoma, the principal affection that has to be considered in differential diagnosis [11]. Pre-operative differentiation between DC and cholesteatoma may be evident radiologically, depending on the content of the cyst, because DCs may not show a significant restriction on DWI, whereas cholesteatoma usually does [12]. However, in this case, the features were ambiguous owing to the main epidermal component of the DC. Looking at the scans retrospectively, a suspect area could be identified at the anterior extent of the cyst, where the lesion had a different component with fat features. In any case, pre-operative differentiation has no influence on the clinical management of the lesions because both equally require surgical removal.

The present case is unique because it represents the first documented case of a mastoid DC; the lesion was successfully and totally removed using a transmastoid approach. Complete surgical resection of these lesions is the treatment of choice to avoid recurrence or residue.

CONCLUSION

Through a critical review of the literature, the pure mastoid location of a DC seems to be extremely rare. The diagnostic process can be challenging because of the asymptomatic course of the lesion. The correct diagnosis should be achieved through histological confirmation; MRI features can be unclear, mimicking a congenital mastoid cholesteatoma. Surgical excision is the treatment of choice. Although uncommon, DCs need to be considered in the differential diagnosis of middle ear and mastoid lesions. These lesions are benign in nature, but they can recur unless the entire wall of the cysts is removed.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – M.F.; Design - M.F.; Supervision - M.F.; Resource – G.P., F.C., M.F.; Materials – S.P., L.C.; Data Collection and/or Processing – G.P.; F.C.; M.F.; Analysis and/or Interpretation – G.P., F.C., M.F.; Literature Search – G.P., F.C.; Writing -G.P., F.C., M.F.; Critical Reviews – M.F.

Conflict of Interest: The authors have no conflict of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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