A rare entity: Benign osteonecrosis of the temporal bone

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Abstract
Objective: To investigate the factors that may contribute to the rare disease process of benign osteonecrosis of the temporal bone and to differentiate this poorly understood disease from other disorders.

Methods: A retrospective chart review using tertiary academic medical center electronic medical records (EMR) for otolaryngology was performed in August 2021. The study included data from January 1, 2014, to June 30, 2020. The patients were identified by a series of predetermined criteria. Their demographic information and clinical course were qualitatively compared. Patients with a history of radiation to the head or neck or who had other otolaryngologic comorbidities were excluded from the study. Inclusion/exclusion criteria were used to filter patients and a thorough chart review was conducted among the patients identified.

Results: A total of three patients were identified that met criteria for the study upon thorough chart review. Out of these patients, all were male, fell within the age range of 67–78 years old and had a history of hypertension. Two out of three had diabetes. In all cases, the plan included supportive care and regular follow up appointments.

Conclusion: Benign osteonecrosis of the temporal bone is a rare disease that is difficult to diagnose and even more complicated to document. There is no specific ICD-10 code for this disease making it exceedingly difficult to identify patients with a simple EMR search. Several previously hypothesized risk factors were studied in this review. The limited patient population makes it difficult to confidently identify risks. In the patients studied, the commonalities included that they were all men, above the age of 67 when diagnosed, and all had a history of hypertension. To fully understand this rare entity further research must be conducted and more patients must be studied to aid with the identification of risk factors and clinical diagnosis of this condition.

Level of Evidence: 4.

Keywords
benign osteonecrosis of the temporal bone, chronic otitis externa, ear fullness, external auditory canal cholesteatoma
1 | INTRODUCTION

Benign osteonecrosis of the temporal bone (BONTB) is a rare condition of which the specific etiology or etiologies are uncertain. Patients typically present with unspecific symptoms such as a sensation of ear fullness, decreased hearing, otalgia, or often no symptoms at all.\(^2\) More rarely a patient may present with tinnitus.\(^2\) The clinical presentation triggers a broad differential with cerumen impaction, cholesteatoma, and otitis externa at the top of the list of possibilities. Like other types of osteonecrosis, the hypothesized presentation is a result of a compromised blood supply to the bone, resulting in avascularity and bone deterioration.\(^3\) Specifically, in BONTB, the blood supply to the tympanic plate of the ear canal is thought to be disrupted. Previous articles acknowledge a correlation between the use of bisphosphonates, advanced age, adult-onset diabetes mellitus and trauma with the occurrence of this condition. In the past decade, there have been very few published documents discussing BONTB. There is a retrospective case study from the period of 1988–1997, however they did not exclude cases caused by radiation or infection.\(^1\) The goal of this case series is to report more current information that focuses solely on patients with a diagnosis of BONTB that have not encountered radiation or infection, and to identify other potential factors leading to the presentation of this rare entity. The study is focused on the presence of predisposing illnesses, medication use, hearing aid usage, and age to create a demographic picture of the patients susceptible to this condition. The purpose of this research is to highlight the rareness of this condition and to create a better understanding of the disease's presentation to aid recognition and diagnosis. In doing so, providers can avoid inadvertent expenditure on extensive workups including imaging, biopsies and surgeries that are not warranted based on the known, expected clinical course.

This research is an extension of previous work on BONTB coauthored by the senior author detailing an overview of the condition's presentation, diagnosis, and treatment.\(^2\) This research offers a retrospective chart review with specific patient presentations, predisposing conditions, and treatment courses.

2 | METHODS

This study was exempt by the local institutional review board (submission number 21-0152). A retrospective chart review was conducted on all patients who met the inclusion criteria and were diagnosed between January 1, 2014 and June 30, 2020. The patients were identified through UTMB (University of Texas Medical Branch) Otolaryngology clinics and UTMB Epic EMR. Demographics, medical history, social history, physical examination, imaging, assessment, and plan of the patients identified were analyzed in the review.

BONTB does not have a specific ICD-10 code. Therefore, to narrow the search results, the ICD-10 code for otitis externa and osteonecrosis in addition to certain CPT codes for CT scans of the temporal bone were used in combination (see Table 1). The reasoning for this is that CT scans are not a routine order for otitis externa, however it is the primary diagnostic tool for BONTB. Anyone without a CT scan was not a candidate for a diagnosis of BONTB. In addition to these parameters, the inclusion and exclusion criteria in Table 1 were used to identify the patients of interest. The search resulted in 80 patients. At this point, the 80 charts underwent detailed review for documentation of a diagnosis of BONTB. After reviewing HPI's, physical exams, assessments, plans, and CT scan findings, three patients were determined to have this uncommon diagnosis. The demographic information and factors of interest are reported in Table 2.

3 | RESULTS

Out of 80 potential patients, three were found to have a diagnosis of BONTB. The small number of data points highlights the rarity of the disease; however, it creates a challenge for identifying risk factors. The data in Table 2, indicates an age range of diagnosis of 67–78 years old with a mean of 74.3 years. All patients identified were male and two out of three had elevated HbA1c values. Two out of the three patients smoked tobacco. Two of the patients identified as Caucasian but one had an ethnicity of Hispanic/Latino, and the other did not. None of the patients studied were on any medications of interest.

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**Table 1: Inclusion and exclusion criteria**

| Included | Excluded |
|----------|----------|
| **Target population** | Adults 18–95 years | Patients <18 or >95 years, Texas Department of Corrections patients |
| Time | January 2010–June 2020 | Before January 2010 and after June 2020 |
| Settings | Patients seen in otolaryngology clinics | Patients not seen in otolaryngology clinics |
| Otolaryngology history | ICD-10 code of otitis externa (H60), osteonecrosis (M87) | History of radiation therapy to head or neck or other otolaryngology comorbidities |
| ICD-10 codes | H60.90 otitis externa BN2F02Z-CT scan of temporal bone | None |
| CPT codes | 380.1—otitis externa 70480—CT of temporal bone w/o contrast 70482—CT temporal bone with and w/o contrast | 77338—radiation to head/neck 797.4—hearing aid use |

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including bisphosphonates, kinase inhibitors, or cisplatin. None of the patients had ear trauma or surgery. A common comorbidity in all three patients was chronic hypertension. Overall, the most noteworthy demographic information seemed to come from the age of the patients and the fact that all patients were male. The age range identified is similar to the age range found in a past case series where the median age was 62 years old. However, the aforementioned study had a much wider age range with the youngest person being diagnosed at only 18 years old. Based on the patients identified in this review, the disease tends to be diagnosed in the elderly population. Table 3 describes the presenting symptoms, binocular microscopy findings, and CT findings from each patient’s chart at diagnosis. Figure 1 shows the erosive changes to the external auditory canal that were visualized on non-contrast CT scan of the temporal bone in one of the patients studied.
DISCUSSION

BONTB is an uncommon and thus underdiagnosed condition. Recent literature has focused primarily on the relationship between the use of bisphosphonates and the development of BONTB. Bisphosphonate use is one of the only known exposure risks that has been researched.\(^4\) Previously, it was hypothesized that diabetes mellitus was a risk for development of BONTB due to the associated vascular impairment, however more recent case studies did not find this association to be supported.\(^5\) It has been found that vascular interventions appear to be successful in the preservation of bone integrity and treatment of this condition.\(^5\) A more recent journal article, published in 2019, the only case of its kind, related the use of sorafenib for metastatic renal cell cancer to the development of avascular necrosis of the external auditory canal.\(^6\) The theorized risk factors discussed in these articles were explicitly studied in this chart review. Of the options, only diabetes and hypertension seem to have a possible connection with the occurrence of this disease process. In this case study, none of the patients identified with BONTB had a history of bisphosphonate, kinase inhibitor, or cisplatin use.

This condition presents a challenge in that it does not have a specific ICD-10 code. It is identified clinically then diagnosed with confirmatory imaging. Clinical presentation commonly includes a persistent sensation of ear fullness, decreased hearing, pain, or tinnitus.\(^1,2\) The symptomatic presentation observed is thought to be due to the subsequent development of another otologic condition resulting from the osteonecrosis present.\(^7\) A true cholesteatoma of the EAC was cited in a case report to be the “result rather than the cause of this pathological process.”\(^7\) Other differential diagnoses are much more likely, however, in cases where the patient has ongoing otological issues or failure to heal, it is important to perform binocular microscopy and obtain a non-contrast CT scan of the temporal bone to assess for possible BONTB (see Figure 1). This chart review brings to attention several potential risk factors. The first of which is elderly age. All patients identified in the study were above the age of 65 years old. This observation is consistent with the fact that osteonecrosis occurs more commonly in the older patient. Furthermore, all three patients were male, and all had a medical history significant for hypertension. Although this study is not large enough to confidently identify risk factors, it is helpful to understand similar patterns in the presentation of this disease.

The treatment options for BONTB are few. In most cases the disease appears self-limiting and managed only with follow-up appointments and symptomatic management. In the patients reviewed in this study, the management was strictly supportive with consistent follow-up appointments. However, a case report from 1997 discusses the successful resolution of the disease process following the use of hyperbaric oxygen therapy in a particularly extensive case of necrosis.\(^8\) This is a potential treatment option for more advanced cases of osteonecrosis, however, based on the prognosis of the disease, intervention is not commonly necessary. Surgical intervention has also proved to be a viable alternative for medical management.\(^3\)

CONCLUSION

BONTB is a rare disease that is difficult to diagnose and even more complicated to document. There is no specific ICD-10 code for this disease making it exceedingly difficult to identify patients with a simple EMR search. The commonalities in the patients studied included that they were all men, above the age of 67 when diagnosed, and had a history of hypertension. This condition should be on the list of differentials in elderly patients, particularly males, presenting with complaints of persistent ear fullness that does not fit the diagnostic criteria for another condition. A CT exam of the temporal bone showing benign bone erosion without other associated pathology may be confirmatory. To fully understand this rare entity further research must be conducted and more patients must be studied to aid with the identification of risk factors and clinical diagnosis of this condition.

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CONFLICT OF INTEREST

The authors declare no potential conflict of interest.

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