Stone deaf: the petrified ear—case report and review of the literature

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Of the many causes of conductive hearing impairment, few are as rare as the petrified ear. Petrified ears describe auricular cartilage hardening, due usually to ectopic calcification or, less commonly, ossification. The process can affect the auricle either completely or partially without any visible external change. It is an uncommon clinical entity, identified mainly in dermatology texts and unreported in the adult radiology literature. Only 12 histologically proven cases have been reported in the English-language literature of auricular ossification. We report a case of idiopathic bilateral auricular calcification presenting with a conductive hearing impairment in an adult female. The diagnosis was made on imaging alone. We discuss the systemic causes associated with this unusual clinical entity and review the literature on “petrified ears.”

Introduction

Deafness is secondary to either a conductive or a sensorineural cause. Conductive hearing impairment, a common abnormality, is due to blockage of the external auditory meatus, or damage to the ear drum or the ossicular chain. Sensorineural deafness is due to disease either of the cochlea or of the eighth cranial nerve and its connections in the brain stem (1). There are other causes, but few as rare as petrification of the auricle.

The pinna consists of a single thin plate of yellow cartilage and elastic tissue (which gives it shape and pliability). The elastic tissue surrounds the cartilage cells like a sheath when seen under electron microscopy (2). The yellow cartilage is similar in structure to the cartilages of the nasal ala. It is then covered by a thin layer named the integument (containing sebaceous glands), and connected to the auditory meatus by ligaments and a few muscular fibers (3). The ligaments of the pinna consist of two sets, one set that connects the pinna to the side of the head and another set that connects it to various parts of the cartilage (3).

The auricular cartilage may be either calcified or ossified. In our institution, 51 patients were imaged with temporal bone CTs in a single year (2007). Of these patients, 12 had a focus or more extensive auricular calcification. This is an observational point; thus, the incidence or prevalence of focal or total auricular calcification cannot be truly known without a larger study.

Calcification is the deposition of calcium or salt-related compounds, while ossification describes the conversion of soft tissues to bone by deposition of calcium and phosphorus (4). This distinction can only be made histologically, as the soft tissue is the same density as bone on radiography. However, there appears to be no advantage in distinguishing between calcification and ossification, as the condition itself is benign unless it is associated with inner- or middle-ear developmental abnormality. The only other known case in the radiological literature is in the pediatric literature, where petrified ears have been associated with skeletal dysplasia and ectopic calcification elsewhere. Further investigations lead to a genetic analysis and a diagnosis of Keutel syndrome (5). However, in a number of cases ossification occurred with no related precipitating cause or event (6, 7).

The condition is usually asymptomatic, but can present for a variety of reasons: pain (8), incidental finding (9,10), or painless auricular nodule interfering with sleep (11).
Gradual stiffening of the appendage can be frequently re-called if explicitly questioned for.

The petrified ear is rarely seen in clinical practice, with an unknown incidence rate as there is very little in the medical literature. The petrified auricle results in partial or total rigidity of the external pinnae. It is said usually to affect the upper pole or one or both pinnae (11) and mainly to affect men (4, 6, 11-17). The condition has been reported to affect females (8, 18, 19).

We report and illustrate the imaging features in a 73-year-old female who presented with conductive hearing loss due to osteomeatal stenosis secondary to auricular calcification. We review the etiology and discuss the literature of this unique condition.

Case report

A 73-year-old female with an unremarkable past medical history and no family history presented to the nurse-led ENT clinic with conductive hearing impairment. The staff were unable to pass an otoscope through the left meatus but managed to dewax with difficulty. It was noted that the right meatus was also narrowed. She was subsequently seen in the ENT clinic.

Clinical examination, while difficult, confirmed the findings in the nurse clinic. The otoscope could not be passed into the left external auditory canal, and the right external meatus was found to be tight with possibly a normal tympanic membrane. The auricular anatomy and overlying skin was normal. There were no associated symptoms apart from the hearing impairment. A referral was made for CT examination of the temporal bones to elucidate a cause.

High-resolution CT of the temporal bones performed with reformatted views showed bilateral auricular cartilage calcification and normal soft-tissue density of the overlying subcutaneous tissues (Figs. 1A, 1B, 2A, and 2B). Multiplanar reconstruction was consistent with the clinical presentation of a markedly narrowed external meatus. This was due to both the auricular cartilage calcification and the overlying soft tissue. Normal anatomy and normal density of the inner and middle ear (including the ossicles) were demonstrated on CT. The findings were consistent with the diagnoses of petrified ears as described in the dermatology literature.

There was no previous history of any hearing problems or history of any ear abnormalities, but the patient always had narrow canals. Routine biochemical measurements including glucose, calcium, albumin, and phosphorus were normal, and no evidence of chondrocalcinosis appeared on plain radiographs in the major joints.

Discussion

There are many causes of conductive hearing impairment, but none as rare as petrification of the auricular cartilage. Calcification or ossification of the auricular cartilage is known as “petrified ears.” Bochdalek first described petrified ears in 1866 (10, 13). The condition is characterized by development of partial or total rigidity of the auricle without any visible external change. The rigidity is due either to calcification or ossification of an auricle, and a variety of causes has been reported (Table). The process may involve one or both pinnae, with sparing of the earlobes. Shape and contour do not change. Some may experience discomfort if the ears are compressed, and ulceration can rarely occur (10).

The etiology is usually related to ectopic calcification (13), whereas true auricular ossification (in which the normal cartilaginous structures are replaced by bone) is less common. True auricular ossification with histological confirmation has been reported in only 12 prior case reports in the literature. Ten of these 12 cases were men (average age = 55, range 17 to 77) and two were women (average age = 44, range 34 to 53). Of these 12 cases, four were associated with cold injury (4, 11, 13, 14), two with perichondritis (6,
Two with Addison’s disease (16), one with trauma (12), and three were of unknown cause (9, 17, 19).

Ectopic calcification may be classified as primary (dystrophic) or secondary (metabolic). Petrified ears can occur with dystrophic calcification; this condition arises when there is deposition of calcium into damaged or post-inflammatory soft tissues in the presence of normal calcium metabolism. This situation can be found in systemic disorders such as diabetes mellitus and ochronosis. Metabolic calcification occurs when there is disruption of calcium and phosphorus metabolism, as is the case in chronic renal failure or Addison’s disease (8, 20). The exact mechanism is unclear, especially in endocrinopathies where the elevated serum calcium is not constant (20).

Again, ossification may be classified as primary or secondary. Primary ossification occurs de novo; causes of primary ectopic ossification include Albright hereditary osteodystrophy, fibrodysplasia ossificans progressiva, and osseous heteroplasia (9). Secondary ossification can occur after mechanical trauma, and previous authors have reported frostbite/hypothermia as the most common cause (4). There has been a case report of petrified auricles due to cold exposure without frank frostbite (14). With endocrine disorders such as Addison’s disease, approximately 24 cases have been described (15, 21); in addition, there may be an association with hypothyroidism and Addison’s disease, in a case where two members of a family were affected (20).

As the condition is so rare, information on treatment is limited, and treatment of the underlying metabolic cause does not alter or reverse the auricular changes. Unless there is discomfort due to local pressure effects, particularly related to sleep, treatment is not necessary. Cases have been described where surgical intervention has been employed to relieve the discomfort (12, 17).

Keane et al. (10) report a case of asymptomatic auricular ossification found in a man with concomitant gastric carcinoma, suggesting that the ossification may be a marker for underlying systemic disease.

These clinical features described were consistent with our case. However, the auricular calcification was demonstrated on CT of the temporal bones (Figs. 1A, 1B, 2A, and 2B), whereas within the dermatology literature, the calcification was demonstrated well on a lateral skull and Caldwell view radiograph (22). Radiographic technique was also able to determine ossification (histologically proven) (13), as it demonstrated the trabecular bone pattern.

No biopsy was performed in our case, as there was no clinical indication to do so.

The petrified auricle is a rare clinical entity and very little is known about it; however, there is a loose connection with a number of systemic causes that should be investigated to ensure subclinical disease is not missed.

Table: Causes of auricular calcification

| Primary | Secondary |
|---------|-----------|
| Albright’s hereditary osteodystrophy | Trauma |
| congenital plaque-like osteomatosi | mechanical trauma |
| neoplasms (for example, naevi of the face, Pilomatrixoma, chondroid syringoma) | frostbite—most common cause (4) |
| osseous heteroplasia | |
| myositis ossificans progressiva | |
| | collagen vascular disease (in particular, CREST syndrome—limited systemic sclerosis) |
| | childhood dermatomyositis (13) |
cold exposure (10, 11)
acrobatic manipulation (12)

**Endocrine / Metabolic**
Addison’s disease (15, 16)
hypothyroidism
hyperthyroidism
scleroderma
acromegaly
prolactinoma
diabetes mellitus
hypopituitarism (23)
renal failure (9)
ochronosis (24)

**Inflammatory**
polyarteritis nodosa
Von Meyenburg’s disease (systemic chondromalacia)
condritis
perichondritis (6)
relapsing polychondritis

**Iatrogenic**
radiation therapy (25)

**Infection**
syphilitic perichondritis

**Other**
hypertension (13)
insect bite
sarcoidosis (13)

**References**
1. Edwards C, Bouchier I, Haslett C, Chilvers E. Davidson’s principles and practice of medicine. 17th ed. Churchill Livingstone; 1995.
2. Cox R, Little K. An electron microscope study of elastic tissue. Proc R Soc Lond B. 1961 Nov;1;155:232–242.
3. Drake RL, Gray H, Vogl W, Mitchell AWM. Gray's atlas of anatomy. Churchill Livingstone; 2008.
4. DiBartolomeo JR. The petrified auricle: comments on ossification, calcification and exostoses of the external ear. Laryngoscope. 1985 May;95(5):566–576. [PubMed]
5. Parmar H, Blaser S, Unger S, Yoo S, Papsin B. Petrified ears in a patient with Kieutel syndrome: temporal bone CT findings. Pediatr Radiol. 2006 Mar.;36(3):241–243. [PubMed]
6. Knapp H. Ossification of the auricle in consequence of perichondritis seropurulent. Arch Otol. 1890 Jul; 12;19:45–50.
7. Kewlaquir L. Ectopic ossification. Am J Phys Med. 1977 Jun; 12;265:876–877.
8. Chiu J, Chu P. Calcified auricles in a patient receiving hemodialysis. Mayo Clin. Proc. 2006 Aug;81(8):1095. [PubMed]
9. High WA, Larson M, Hoang MP. Idiopathic bilateral auricular ossificans: a case report and review of the literature. Arch. Pathol. Lab. Med. 2004 Dec.;128(12):1432–1434. [PubMed]
10. Keane FM, Muller B, Murphy GM. Petrified ears. Clin. Exp. Dermatol. 1997 Sep.;22(2):242–243. [PubMed]
11. Yeatman JM, Varigos GA. Auricular ossification. Australas. J. Dermatol. 1998 Nov;39(4):268–270. [PubMed]
12. Lari AA, al-Rabah N, Dashi H. Acrobatic ears: a cause of petrified auricles. Br J Plast Surg. 1989 Oct;42(6):719–721. [PubMed]
13. Stites PC, Boyd AS, Sic J. Auricular ossificans (ectopic ossification of the auricle). J Am Acad Dermatol. 2003 Jul;49(1):142–144. [PubMed]
14. Lautenschlager S, Itin PH, Rufl T. The petrified ear. Dermatology (Basel). 1999;189(4):435–436. [PubMed]
15. Cohen AM, Talmi YP, Flug M, Kalmanovitz M, Zohar Y, et al. X-ray microanalysis of ossified auricles in Addison's disease. Calcif Tissue Int. 1991 Feb.;48(2):88–92. [PubMed]
16. Cohen AM, Talmi YP, Flug S, Bar Ziv J, Zohar Y, Djaldetti M. Ossification of the auricle in Addison's disease. J Laryngol Otol. 1989 Sep.;103(9):865–866. [PubMed]
17. Lister GD. Ossification in the elastic cartilage of the ear. Br J Surg. 1969 May;56(5):399–400. [PubMed]
18. Gordon EJ, Perlman AW, Schechter N. Diffuse inflammation of cartilage; a case report of a hitherto unreported entity. J Bone Joint Surg Am. 1948 Oct.;30A(4):944–956. [PubMed]
19. Scherrer F. Calcification and ossification of the external ears. Ann Otol. 1932 Jun; 12;41:867–885.
20. Thomson MA, Biggs P, Berth-Jones J. Familial petrified ears associated with endocrinopathy. J Am Acad Dermatol. 2007 Nov.;57(5 Suppl:S121–2. [PubMed]
21. Jarvis JL, Jenkins D, Sosman MC, Thorn GW. Roentgenologic observations in Addison's disease; a review of 120 cases. Radiology. 1954 Jan.;62(1):16–29. [PubMed]
22. Clarke JT, Clarke LE, Miller J. Petrified ears: calcification of the auricular cartilage. J Am Acad Dermatol. 2004 Nov.;51(5):799–800. [PubMed]
23. Barkan A, Glantz I. Calcification of auricular cartilages in patients with hypopituitarism. J Clin. Endocrinol. Metab. 1982 Aug;55(2):354–357. [PubMed]
24. Pomeranz MM, Friedmann L, T UNIXK IS. Roentgen findings in alkaptonuric onchronsis. Radiology 1941; 37:295–303.
25. Williams JP, Dixon G, Fowler GW, Pribram HF, Sargent EN, Turner AF. Calcification of the auricular cartilage and basal ganglia secondary to radiation therapy. Bull Los Angeles Neurol Soc. 1973 Jan.;38(1):33–36. [PubMed]