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Case Report

Chondrodermatitis Nodularis Chronica Helicis et Antihelicis in a 63-Year-Old Man

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Abstract

Chondrodermatitis nodularis helicis also known as Chondrodermatitis Nodularis Chronicus Helicis (CNCH), typically presents as a tender, solitary, firm, well-demarcated, dome-shaped, erythematous or skin-colored papule/nodule on the pinna. The condition is usually unilateral, although cases of bilateral CNCH have also been described. Bilateral CNCH lesions are usually symmetrical. We herewith report a 63-year-old man with a CNCH lesion on the right antihelix and another CNCH lesion on the left helix.

Introduction

Chondrodermatitis nodularis helicis, also known as Chondrodermatitis Nodularis Chronicus Helicis (CNCH), Winkler's nodule or Winkler's disease, is a benign, inflammatory and degenerative condition that affects the skin and cartilage of the pinna, manifesting as a tender papule/nodule [1]. The condition is usually unilateral. Bilateral CNCH cases are uncommon and the lesions are usually symmetrical. We herewith report a 63-year-old man with a CNCH lesion on the right antihelix and another one on the left helix. The bilateral occurrence of CNCH lesions makes certain treatment options such as sleeping on the unaffected side infeasible.

Case Report

A 63-year-old man presented with an 18-month history of a painful nodule on his right ear. Two months later, he developed another painful nodule on his left ear. The pain was severe enough to awake him from sleep several times per night. There was no history of trauma. His past health was unremarkable. In particular, the patient did not have visual, hearing or respiratory problems. The family history was noncontributory. One of us (BB) was consulted at this stage. Physical examination revealed an erythematous dome-shaped, firm nodule measuring 6 to 7 mm in diameter with a central crust on the right antihelix (Figure 1) and a flesh-colored dome-shaped nodule measuring 2 mm on the superior pole of the left helix (Figure 2). Both nodules were tender to palpation. The rest of the physical examination was unremarkable.

A clinical diagnosis of CNCH was made. The lesion on the left ear was excised, and the lesion on the right ear was treated with liquid nitrogen followed by clobetasol cream with complete resolution of pain. Histology of the excised sample showed epidermal hyperplasia, parakeratosis and acanthosis. The underlying dermis showed fibrinoid degeneration surrounded by vascular granulation tissue (Figure 3). There was inflammation, degeneration and fibrosis of the underlying perichondrium. These findings were consistent with a diagnosis of CNCH.

Discussion

CNCH was first described by Max Winkler in 1915 who reported on eight men with painful, firm, lentil- to cherry-sized nodules with central crusting on the helix [2]. In most of the cases, the age of onset is over 40 years, with a peak between 58 and 72 years [1,3]. For lesions on the helix, the male to female is approximately 10:1 while lesions on the antihelix are more common in females [4,5].

CNCH should be differentiated from relapsing polychondritis. The latter is an autoimmune disease that causes recurrent episodes of chondritis, primarily in the auricular, nasal and laryngotracheal cartilage. Currently, the diagnosis of relapsing polychondritis requires the presence of a proven inflammation in at least two or three of the auricular, nasal or laryngotracheal cartilages or the proven inflammation in one of these cartilages plus two other signs, including ocular inflammation, vestibular dysfunction, seronegative inflammatory arthritis or hearing loss [6].

Although the exact etiology is not known, CNCH might result from local trauma, excessive use of earphones and headphones, or most likely prolonged excessive pressure such as that created by

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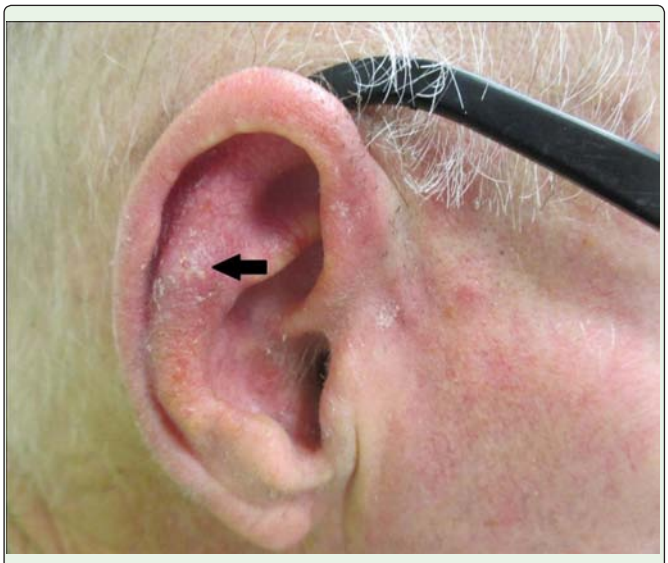

Figure 1: CNCH presenting as an erythematous, dome-shaped, firm, tender nodule on the right antihelix.

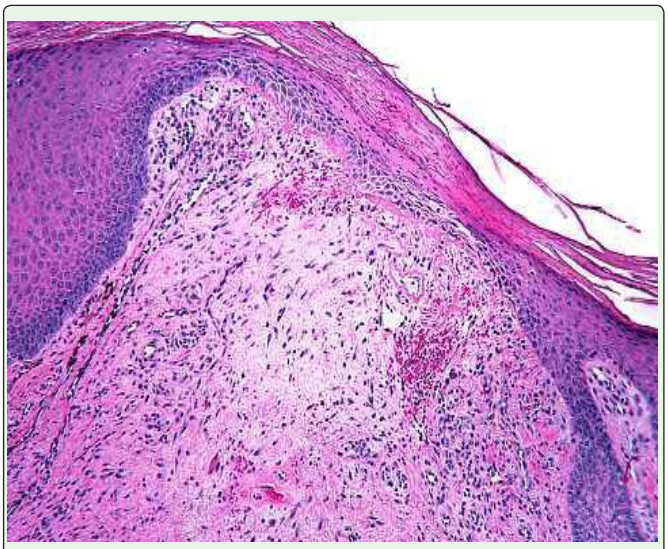

Figure 3: Histology shows eosinophilic fibrinoid material with granulation tissue flanking the zones of necrosis on both sides.

constant sleeping on one side and actinic damage, leading to arteriolar narrowing and ischemia of the auricular perichondrium, cartilage and dermis [5,7]. The ischemia may lead to necrosis of the dermis and the underlying cartilage [8]. The necrobiotic dermal collagen and sometimes the underlying cartilaginous matrix are extruded through a crater-like defect in the epidermis [5]. The auricle, especially the helix, is predisposed to this condition as the local blood supply is poor and there is little supporting subcutaneous tissue for cushioning [8]. A genetic factor might also be operative as the condition has been observed in monozygotic twins [4].

Typically, CNCH presents as a tender, solitary, firm, well-demarcated, dome-shaped, erythematous or skin-colored papule/nodule on the pinna [9]. The helix is commonly involved, followed by the antihelix, although any part of the pinna such as the antitragus, scapha and concha may also be involved [3,9]. The superior pole of the helix at the transition from the vertical to horizontal course of the ear margin is most commonly affected [3].

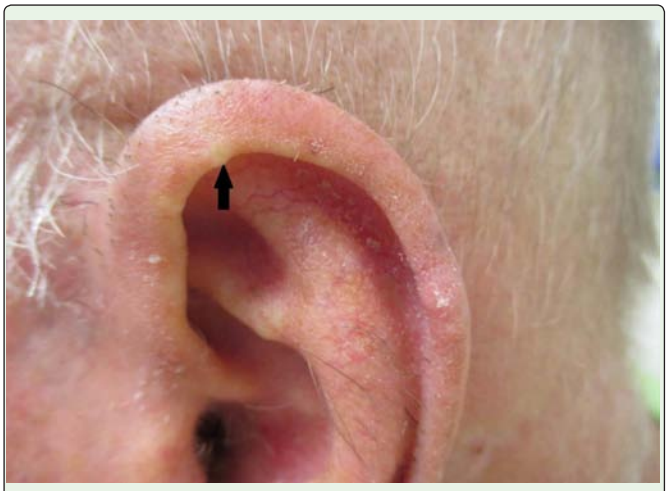

Figure 2: CNCH presenting as a flesh-colored dome-shaped nodule on the superior pole of the left helix.

The condition is usually unilateral, although bilateral cases of CNCH have also been described [4,10-18]. In 1953, Shuman and Helwig performed a systematic survey of the Armed Forces Institute of Pathology (in Washington, DC) files for all cases recorded as CNCH over a period of 15 years and found 7 cases of bilateral CNCH [7]. The authors, however, did not mention the sexes of the affected patients and locations of the lesions in their report. Four cases of bilateral CNCH were seen at the University of California Medical Center in Los Angeles, USA between 1946 and 1951, the sexes of the patients and locations of the lesions were not specified [15]. Four other patients with bilateral CNCH were seen at the Department of Surgery, Dundee Royal Infirmary in Dundee, UK between 1986 and 1989 [14]. Likewise, the sexes of the patients and locations of the lesions were not specified. In 1984, Tatnall et al reported a 74-year-old woman with CNCH on the antihelix of both ears [18]. Cannon in 1984 described a 58-year-old farmer with CNCH on the antitragus of both ears [10]. In 1988, Dean et al reported a 72-year-old woman with CNCH on the antihelix of both ears as a complication of cardiac pacemaker insertion [11]. In 2008, Chan et al described 46-year-old monozygotic male twins who simultaneously developed CNCH [4]. The first twin had the lesion on the rim of the left helix while the second twin had the lesions on both ears. In 2003, Oelzner and Elsner reported a 58-year-old woman with CNCH on the free border of the helix of both ears [16]. In 2010, Kaur et al described a 60-year-old woman with CNCH on the antihelix of both ears [12]. Recently, Khurana et al reported a man in his 40s with CNCH lesions on the free border of the helix of both ears [13]. From the reported cases, bilateral CNCH lesions were usually symmetrical. Our case is unique in which one lesion occurred on the helix while the other lesion on the antihelix.

Although the disease is benign, it can be exquisitely painful and impair sleep. Spontaneous resolution upon removal of local

pressure has been documented [19]. Affected patients should avoid the use of earphones and headphones. Measures such as the use of a doughnut or chondrodermatitis pillow or similar pressure-relieving prosthesis and sleeping on the unaffected side are often helpful. For bilateral lesions, sleeping on the unaffected side obviously is impossible. Surgical excision should be considered for persistent lesions [3,4]. Other treatment options include topical nitroglycerin, topical or intralesional corticosteroid, perilesional collagen injection, laser ablation, curettage, electrocauterization, cryotherapy, and photodynamic therapy [1,7,9].

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