Review Article

Chondrodermatitis of the ear: the painful truth

耳朵軟骨皮膚炎：痛苦之堅

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**Background:** Chondrodermatitis nodularis chronica helicis (CDH) is an inflammatory disease of the ear, typically occurring in, but not limited to, middle-aged and elderly men. Untreated disease is painful, disfiguring and sleep-depriving. Misdiagnosis and suboptimal treatment are common.

**Objectives:** To raise awareness of this disease and provide an overview of treatment modalities.

**Discussion:** Clinical presentation is specific for a painful lesion on the side a patient sleeps on. Cutaneous malignancies often occur on the ear – we stress the importance of excluding malignancy. Inappropriate management leads to recurrence or disfigurement. **Conclusion:** This is a treatable disease. Biopsy is required if there is any doubt in diagnosis.

**Keywords:** Chondrodermatitis nodularis chronica helicis, cutaneous squamous cell carcinoma, basal cell carcinoma, ear nodules

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**Introduction**

Chondrodermatitis nodularis chronica helicis (CDH) is a chronic, benign, inflammatory disease of the external ear. It most commonly occurs on the superior pole of the helix. Typically, CDH presents as a firm nodule with a central crusting, 3-20 mm in diameter (Figure 1A). Pain, especially with pressure application, is a distinguishing feature. The disease occurs more commonly on the right side. In nearly all cases, this coincides
with the side the patient preferentially sleeps on.\textsuperscript{1} However, the clinical appearance of CDH, especially when there is accompanying erythema or ulceration, often leads to misdiagnosis (Figure 1B). They can be mistaken for actinic keratoses, cutaneous squamous cell carcinoma, basal cell carcinoma, keratoacanthomas or gouty tophi (Figures 1A-1C & Figure 2). Most patients are middle-aged and elderly males,\textsuperscript{1} although relatively young men and women can also be affected. CDH is more frequently observed in Caucasians,\textsuperscript{2} although studies describing epidemiological factors are lacking. Estimates of disease prevalence are not provided in the literature, but in our experience CDH is a common disease. The disease is a source of persistent pain and often contributes to impaired sleep and reduced quality of life.

**Aetiology**

The aetiology of CDH is poorly characterised. Most hypotheses centre on repetitive injury such as trauma, thermal or chemical, to a poorly vascularised area that is lacking in a protective layer of subcutaneous tissue. Repetitive injury leads to localised ischaemia through a perichondrial vasculitis model.\textsuperscript{3} This theory was originally suggested in 1936,\textsuperscript{3} with recent histopathological examination of 16 CDH cases demonstrating arteriolar narrowing in the perichondrium, with underlying cartilage exhibiting ischaemic changes.\textsuperscript{4} Ultimately, these alterations set up a cycle of local inflammation and necrosis. Associations with systemic diseases have been made.

**Diagnosis**

The diagnosis of CDH can generally be made clinically. Small, tender papules, with or without accompanying ulceration or erythema that are located on the helix or anti-helix of the ear are usually present. A skin biopsy demonstrates distinctive features with no malignant characteristics.\textsuperscript{4} These include a central epidermal defect and overlying necrobiotic dermal changes that extend to the perichondrium.\textsuperscript{5} Surrounding the necrotic area is epidermal hyperkeratosis and dermal oedema.\textsuperscript{5} Definitive diagnosis requires a biopsy specimen extending to the perichondrium, and is necessary if diagnosis is doubtful (Figures 1A & 1B). Over time the disease may result in chronic pain and unsightly lesions typically located on the ear that is usually slept on. If untreated,
the disease can cause significant pain and disfigurement (Figure 3).

**Management**

A wide variety of management options are available for the treatment of this disease. A recent systematic review evaluated the effectiveness of several treatment modalities that have been published in the literature. Surgical techniques were the most commonly reported of the management strategies. Of 751 patients, surgical techniques achieved a cure rate of 82%. However, complications of surgery including haematoma, infection, and poor cosmetic result may make other options preferable for patients. Older patients with multiple comorbidities may desire to avoid surgery. Medical management options are effective and have minimal adverse effects.

**Conservative**

In conservative treatment approaches the aim is to minimise any further trauma to the ear. Most commonly, this involves reducing prolonged pressure to the ear, but may also include reducing sun, cold or irritant exposures. Strategies used include altering sleeping habits and using pressure-relieving headgear. Several studies demonstrate favourable results with these measures. In a systematic review, five studies assessed the efficacy of pressure relief alone in curing the disease. Between studies cure rates ranged from 16% to 87%. In the study with the highest cure rate, 13 out of 15 patients were cured after a period of one month using a home-made foam pressure-relief device applied to the ear with the lesion during sleep. This approach is affordable, simple and not associated with adverse effects. Conservative management may be used as an adjunct to medical or surgical interventions.

**Medical**

Topical nitroglycerin and corticosteroids have been used in the treatment of CDH.

**Topical nitroglycerin**

Flynn et al described cure of eight of thirteen patients with CDH using 2% topical nitroglycerin gel applied to the ear at a twice daily regimen. A further four patients in this cohort attained

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**Figure 2.** Invasive basal cell carcinoma (BCC) of the ear.

**Figure 3.** Advanced chondrodermatitis nodularis chronica helicis of the left ear.
symptomatic relief. The predominant presenting symptoms in this group included pain, followed by sleep disruption and lesion bleeding. Time to complete resolution or symptomatic resolution varied between patients ranging from two weeks to several months. Seventeen percent of patients in this study reported headache as a side-effect. The sole patient who failed to gain any benefit from the treatment did not demonstrate a response to other treatment modalities.8

**Corticosteroids**

Surprisingly, few published studies describe the use and efficacy of topical corticosteroids or steroid injections in the treatment of CDH. Beck described five patients who were cured with the application of topical 0.1% betamethasone valerate and 3% clioquinol solution applied twice daily to the affected area. The disease resolved in all patients by six weeks.

As many patients with CDH are older and already suffer from various uncomfortable or painful medical problems, the most unwelcome afflictions are extra sore points in their lives! Topical steroids are easily available and mostly inexpensive. In early cases they can result in clinical and subjective improvement, particularly if the patients are adequately monitored by the doctor. In more advanced cases well-defined foci of CDH can be treated with intra-lesional steroid injections. At times, it may be necessary to combine initial steroid injections with subsequent topical steroids. Even advanced cases can have surprisingly desirable outcomes.

**Cryotherapy and electrocauterisation**

Cryotherapy and electrocauterisation may be utilised for treating CDH. We have observed favourable results with these modalities. Follow-up examination of 78 patients demonstrated a low relapse rate of only 31% after curettage with electrodessication.10 Alternatively, cryotherapy may be tried in patients. These physical treatments are cost-effective and time-saving.

**Surgery**

Surgical treatment is the most common method described in the literature. Many surgical techniques have been outlined. Whilst surgery may provide a 'quick fix,' recurrences do occur, with various techniques exhibiting different recurrence profiles. As necrosis and inflammation of underlying cartilage is the causative and perpetuating factor in CDH formation and persistence,4 successful surgical outcomes consistently depend on excision of the perichondrium and the underlying cartilage. Furthermore, post-surgical complications such as bleeding and infection, especially in the context of older and anti-coagulated patients can be a problem. Finally, the cost of surgery and anesthaesia may be an extra burden for financially disadvantaged patients. Accordingly, we would recommend a non-surgical approach to be adopted initially.

**Conclusion**

Can we help these long-suffering and often neglected patients? Yes, we can! Various treatment options are available to ameliorate the auricular sore points due to CDH. Chronic pain is a curse which can lead to anxiety and depression. Whatever the cause, avoiding or minimising aggravating factors would be helpful in managing patients with CDH. An empathetic approach and providing adequate follow up are also crucial.

It is important that the patients with CDH do not live in fear of cancer, especially invasive squamous cell carcinoma (SCC). To this end, correct clinical and histological diagnosis is critical. For example, mistaking an infiltrating SCC in an older and immunocompromised patient for a benign CDH could be disastrous. On the other hand, overzealous surgical treatment for a limited CDH can result in disfigurement and even litigation. We may all be well advised that conservative treatment can work, but if it does not, judicious surgical intervention may save the ear!
References