Chondrodermatitis nodularis chronica helicis - a review

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Abstract

Chondrodermatitis nodularis chronica helicis (CNCH) is a condition characterized by spontaneously evolving tender single or multiple nodules over the apex of helix or antihelix of the external ear. Various factors have been proposed to be the causative agents and different medical /surgical modalities of treatment have been devised. We review this disease in detail and present one such case which was managed on conservative lines.

Key words
Chondrodermatitis nodularis chronica helicis (CNCH), Winkler's disease, pressure, nodule, excision, recurrence.

Introduction

Chondrodermatitis nodularis chronica helicis (CNCH) is a cutaneous lesion of the external ear characterized by spontaneously evolving tender nodule over the apex of helix or antihelix. It was first described by Winkler in 1915 and independently reported by Foerster in 1918. Multiple theories have been proposed to explain the etiology and pathogenesis of CNCH from time to time over the last century. Several treatment options have been reported in the literature ranging from simple modifications of habits to complete excision of the lesion. In this article, we review the various aspects related to CNCH in the light of literature derived from Medline/Pubmed and present one such case which was managed conservatively.

Case presentation

A 39-year-old male, Egyptian, pharmacist by profession, presented with painful swelling over the right ear of 3 months duration. The pain in the swelling would awaken him from sleep during night. There was no history of trauma or any medical/surgical disorder of significance. He denied using any gadgets or bands over his affected ear. On examination, the patient was well built and healthy. Local examination revealed a single, tender nodule over apex of helix of right pinna, 5mm x 5mm x 2mm, skin colored with erythematous rim, minimally crusted surface, firm, and without any discharge or evidence of infection (Figure 1 – A, B). Left ear was normal. There was no lymphadenopathy. The patient was diagnosed as CNCH on the basis of history and examination. Patient was counseled and explained pros and cons of conservative as well as surgical management. Conservative approach was adopted in form of application of custom made ear padding with a hole to relieve pressure over the lesion. The patient became pain free after 2
weeks and remained so even after 6 months of follow up.

However, the nodule did not decrease in size but the patient was satisfied with pain free status.

**Synonyms**

Chondrodermatitis nodularis helicis (or antihelices – depending on site of lesion), chondrodermatitis nodularis chronica helicis, ear pressure sore, painful nodule of the ear, Winkler's disease

**Pathophysiology**

The exact cause of CNCH is still unknown. Winkler\(^1\) who first described this disease suggested that the lesion was due to degenerative changes of the cartilage of ear and these changes acted as an inflammatory effect to the skin. However, most authorities believe the lesion to be a result of prolonged and excessive pressure.\(^2\) The peculiar anatomic features of the external ear (pinna) act as predisposing factors for development of this lesion. The pinna has relatively little subcutaneous tissue for insulation and padding, and only small dermal blood vessels supply the skin and cartilaginous framework. Trauma, cold, actinic damage, or focal pressure probably initiates the disease leading to inflammation and subsequent necrosis. The anatomic features of the ear, as listed above, retard adequate healing and lead to secondary perichondritis.

The focal pressure can be generated between the head and the firm pillow particularly at night if a person sleeps predominantly on one side. It can also be precipitated by gadgets like tight headgear, hair band, cell phone\(^3\) or Bluetooth ear devices.

Santa Cruz in 1980 had found the natural history and histology to be similar to a group of disorders characterized by transepithelial elimination known as the perforating dermatoses.\(^4\) Upile et al.\(^5\) in 2009 designed a detailed histopathological review of 16 confirmed cases of CNCH. The study revealed arteriolar narrowing in perichondrium region of pinna most remote from arterial blood supply, i.e. helix. This perichondrial arterial narrowing was believed to be responsible for ischemic changes and death of the metabolically active underlying cartilage with subsequent necrosis and extrusion.

Although CNCH is regarded as an idiopathic disorder in general with no systemic associations, exceptions to this have been reported in literature and cases may occasionally be associated with autoimmune or connective-tissue disorders,\(^6-8\) including autoimmune thyroiditis, lupus erythematosus, dermatomyositis, and scleroderma. Such cases may be more prevalent in pediatric age group or young adult female patients. A possible hereditary factor has been suggested in the pathogenesis by Chan et al.\(^9\) who reported this disease in middle-aged male monozygotic twins who simultaneously developed CNCH.
Epidemiology

Frequency/Site

The exact incidence/prevalence of CNCH is not known. Right side is predominantly involved and lesion is usually unilateral though bilateral cases are rarely reported in literature.\(^9\)

Morbidity

Spontaneous resolution is the exception and has been reported in few series. Moncrieff and Sassoon reported resolution in 87% by conservative management.\(^{10}\) Remissions may occur, but the disease usually continues unless adequately treated.

Race

CNCH occurs in persons of all races though it is most commonly reported in fair-skinned individuals with severely sun-damaged skin.

Gender

Although CNCH mostly occurs in men, 10-35% of cases involve women.\(^4,11\) Some studies, however, report equal incidence in both genders.\(^{12}\)

Age

CNCH can occur in patients of any age group but generally affects middle-aged to older individuals. Age at onset is similar in both genders. Cases are however reported in pediatric age group.\(^{13,14}\)

Clinical presentation

History

The classical presentation is the spontaneous appearance of tender nodule on the helix or antihelix. The nodule enlarges rapidly to its maximum dimensions and then stabilizes. The patients are often awakened at night when they roll over and pillow comes in contact with the lesion. There may be occasional episodes of intense pain without any contact of any kind.

Physical examination

There may be single (usually) or multiple dome-shaped, skin-colored or erythematous popular excrescences at the apex of the helix or antihelix. These lesions are tender, firm, well demarcated, and ranging in size from 3-20 mm. An adherent scale/crust over the horny lesion is characteristic. Removal of this scale/crust often reveals a small channel. The lesion may ulcerate and the ulcer will have raised, rolled edge with minimal granulation at the base.

Differential diagnosis

The differential diagnosis includes benign or malignant cutaneous tumors, such as basal cell carcinoma, keratoacanthoma, squamous cell carcinoma actinic keratosis, atypical fibroxanthoma, cystic chondromalacia, elastotic nodules of the ears, Merkel cell carcinoma, perforating dermatoses, gouty tophi, verruca vulgaris, molluscum contagiosum, cutaneous horn and pseudocyst of the auricle. These conditions are usually painless, even when ulcerated in contrast to CNH where pain is the clinically distinctive feature.\(^{15}\) Biopsy may be required only if patients with CNH have chronic solar damage and a history/suspicion of skin cancer.
Histopathology

The histopathological analyses of excised specimen reveal necrotic debris enveloped by a layer of pseudoepitheliomatous hyperplastic epidermis. In ulcerative lesions, the ulcer base may be covered with granulation tissue. There may be foci of fibrinoid necrosis. There are no dermal adnexa at the lesion and vascular proliferation may resemble glomus tumor.

Cribier et al. submitted 37 cases of clinically typical CNCH to conventional microscopy after surgical excision and analyzed with immunohistochemistry using S-100 and neurofilament antibodies. Another set of 25 tumors of the ear were taken as controls and studied with similar techniques. Nerve hyperplasia was present in CNCH, but was often masked by intense vascular and inflammatory reactions. This finding of neural hyperplasia was proposed as the possible explanation of pain and tenderness of CNCH.

Management

Ever since the initial description of CNCH by Winkler some hundred years back, a wide range of techniques have been devised and proposed for tackling this lesion.

1. Medical/less invasive treatment

Some of the commonly employed medical /less invasive treatment modalities include:

a) Conservative approach

The conservative approach relies on proper counseling of the patient and steps to relieve or eliminate pressure at the site of the lesion. This involves changing sleep habits or discontinuation of pressure inducing gadgets like Bluetooth ear piece. Moncrieff and Sassoon in 2003 presented a series of 15 patients who were treated conservatively by application of using a home-made, pressure-relieving prosthesis. 87% cases got relief over a period of one month and avoided invasive management. Similarly Kuen-Spiegl et al. in 2011 reported a significant improvement in form of pain relief and reduction in size of lesion with the use of a self-made bandage of foam plastic. Sanu et al. designed a doughnut shaped pillow to relieve pressure in cases of CNCH and found the treatment to be cost effective. Topical antibiotics are advised if there are features of infection in ulcerated lesions.

b) Steroids

Insoluble corticosteroid injections (2.5 to 40 mg/ml triamcinolone acetonide) directly into the nodule at weekly intervals twice to three times or applied topically has been reported in literature with equivocal responses. Some cases have been cured by the use of prednisolone crystal suspension or twice-daily application of 0.1% betamethasone valerate for three to six weeks.

c) Collagen injection

Subcutaneous injection of reticulated collagen implants around the nodule to create a cushion between the skin and perichondrium has been reported by Greenbaum. This collagen cushion relieves the pressure on the lesion and reduces pain effectively.

d) Cryotherapy

There are reports of successful use of cryotherapy in literature. Sanel reported a case in 2010 where the hyperkeratotic lesion of left ear in a 32-year-old female resolved completely
Two cycles of therapy with a 3-week interval between treatments were used. Pain was relieved and the lesion completely healed without any scar. No recurrence was observed at 1-year follow up.

d) Laser therapy

Laser therapy has been reported as effective for pain relief and nodule resolution with a lower rate of recurrences and post-procedure complications. The cosmetic outcome and patient satisfaction is also good. The distinct advantage of laser therapy is immediate pain relief after the procedure.

e) Photodynamic therapy

Chondrodermatitis nodularis helicis has been successfully treated with photodynamic therapy in recent years. Pellegrino et al. in 2011 reported the successful use of a 635 nm light source over a period of 20 minutes (70J/cm²) in two cases after application of cream containing 20% 5-aminolevulinic acid (5-ALA) and occlusion for 3 hours. Pain relief and decrease in size of lesions was achieved within few weeks. Gilaberte described 5 patients with CDNH who were successfully treated with methyl aminolevulinate photodynamic therapy (PDT).

2. Surgical treatment

Surgical treatment is one of the mainstays of management. There are multiple procedures proposed in literature which can be conducted under local anesthesia. In all these procedures, conservation of normal tissue is important for optimal esthetic outcome. These include:

a) Curettage

The dissection is done with sharp curets. The necrotic cartilage of the lesions is soft and curetted out easily. The endpoint is reached when the curet is resisted by firm, elastic cartilage. Curettage can also be combined with electrocauterization. Since the skin is gradually expanded over the nodule, the resultant defect does not normally heal with any contour deformity.

b) Punch and graft

Rajan et al. in 2007 reported a novel punch and graft technique for management of CDNH. A punch biopsy instrument is used, the diameter of which is such that the lesion is encompassed by the punch. The punch is applied perpendicular to the skin surface and advanced until a deep punch of the underlying diseased cartilage is excised. A full thickness skin graft is harvested from posterior auricular area after ensuring a reasonable skin color match. The same size punch tool is used to harvest the full-thickness skin graft. The donor site is closed with interrupted sutures whereas the graft is sutured over the recipient defect after proper preparation of the graft. Rajan et al. reported a cure rate of 83% with good cosmetic results.

c) Excision

Excision of the lesion is the definitive treatment if the diseased portion is removed completely. There is a difference of opinion regarding the extent of excision needed to eradicate the disease without compromising on aesthetics or increasing the incidence of recurrence. Lawrence demonstrated that only cartilage needs to be removed after raising a flap or by making a longitudinal incision over the lesion. Dreiman proposed full wedge excision of the lesion and reconstruction with chondrocutaneous helical rim advancement flap of Antia and Buch.
Affleck, however, strongly proposes that full wedge excision of CDNH is often unnecessary and should be avoided in favor of more conservative excision.28

Prognosis

The prognosis for patients with CNCH is excellent. However, long-term morbidity is common and recurrences can occur. Recurrence rate is dependent on treatment modality. Recurrence is around 10% for surgical excision and 31% for curettage and electrocauterization.11 Munnoch et al.12 however, reported recurrence free series with minimal skin and extensive cartilage excision.

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References


