Chondrodermatitis nodularis chronica helicis in a patient with systemic sclerosis associated with primary biliary cirrhosis (Reynolds syndrome): A case report
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Abstract
Chondrodermatitis nodularis chronica helicis is a rare non-neoplastic inflammatory and degenerative process of the external ear, characterized by necrobiotic changes in the dermis that extend down to the perichondrium. This condition has been occasionally reported in patients with limited cutaneous systemic sclerosis but not in those with concomitant primary biliary cirrhosis; this association is known as Reynolds syndrome. We report a 70-year-old woman diagnosed with primary biliary cirrhosis at age 47 and with limited cutaneous systemic sclerosis at age 54 who developed a painful ulcerated nodule on the helical rim of the left ear shortly after the last diagnosis. The lesion was excised because of the suspicion of malignancy, but the histopathology was consistent with chondrodermatitis nodularis chronica helicis. Although this condition is infrequent, it is necessary to know, because it may occur in patients with systemic sclerosis and be mistaken for neoplasms, such as basal cell and squamous cell carcinoma, and these patients have an increased risk for the development of skin malignancies.

Keywords: Chondrodermatitis nodularis chronica helicis, systemic sclerosis, primary biliary cirrhosis, Reynolds syndrome, skin malignancies

Introduction
Systemic sclerosis (SSc) is a multisystem connective tissue disease, the hallmarks of which are autoimmunity, inflammation, functional and structural alterations in small blood vessels, and interstitial and vascular fibrosis in the skin and internal organs (1). Primary biliary cirrhosis (PBC), a chronic cholestatic liver disease characterized by immune-mediated chronic non-suppurative cholangitis and associated with anti-mitochondrial antibodies (AMAs), is considered the most common liver disorder in patients with SSc, with an estimated prevalence of 8% and occurring mostly as the limited cutaneous variant (lcSSc) (2). The association of SSc and PBC is known as Reynolds syndrome (RS) because of its original description in the early 1970s (3).

Chondrodermatitis nodularis chronica helicis (CNCH) is a rare non-neoplastic inflammatory and degenerative process of the external ear, characterized by necrobiotic changes in the dermis that extend down to the perichondrium (4). This condition has been occasionally reported in association with SSc (5, 6) but not in patients with concomitant PBC. We report a patient with RS who developed CNCH.

Case Presentation
A 70-year-old white female was diagnosed with PBC at age 47, based on elevated levels of alkaline phosphatase (672 U/L), positive AMA test, and liver biopsy with a perportal inflammatory infiltrate and mild portal fibrosis (stage I of Scheuer’s classification). Since then, she was treated with ursodeoxycholic acid (Ursochol; Zambon, Madrid, Spain). Alkaline phosphatase levels were stable, and pruritus and signs of portal hypertension were absent. Concomitant with the diagnosis of PBC, the patient began to have episodes of Raynaud’s phenomenon and progressively developed sclerodactyly, xerostomia, xerophthalmia, and esophageal dysmotility, being diagnosed with lcSSc at age 54. The physical examination at that time showed telangiectasia on the fingers, a beaked nose, and mild perioral furrowing; capillaroscopy demonstrated giant capillary loops with minimal loss of capillaries; and serologic investigations found increased titers of antinuclear antibodies (1/320) and positive anti-centromere antibodies.

Shortly after the diagnosis of lcSSc, the patient noticed a painful nodule that involved the helical rim of the left ear (Figure 1). The nodule grew slowly over the course of several months, and she stated that the pain
was exacerbated by rubbing the telephone and supporting the head on the affected side during sleep. On physical examination, the lesion measured 10 mm, erythema was present, and a central 3-mm-diameter ulcer was covered by a crust. The right ear was normal. Because a skin malignancy was suspected, surgical resection of the lesion was performed. Histopathologic study revealed that the ulcer was composed of necrotic and granulation tissue in the dermis, with a marked mixed acute and chronic inflammatory cell infiltrate and isolated multinucleated giant cells. The epidermis surrounding the ulcer showed reactive acanthosis, and there were elastic fiber degeneration and telangiectatic vessels in the underlying dermis. The cartilage showed chondroid matrix degeneration and chronic inflammatory infiltrate that was immediately adjacent (Figure 2). These findings were consistent with CNCH. No dysplasia or malignancy was identified.

The patient experienced complete resolution of the lesion after the resection. However, she developed a recurrence in the same location 2 years after and underwent a new surgical resection. There have been no recurrences since then.

Discussion

Clinically, CNCH manifests as a single (rarely multiple or bilateral), exquisitely tender, dull erythematous nodule on the helix (less frequently on the antihelix, scapha, or concha) and varies from 3 to 10 mm. The surface of the lesion is often covered with a scale or crust over an underlying central depression. CNCH shows a marked male dominance (up to 80% of cases) and occurs typically in the elderly (average age 58-72 years) (6, 7). Some pediatric cases have rarely been reported (8). Histopathology of the lesions demonstrates dermal inflammation associated with either a central hyperkeratotic plug or ulceration and crust. Ulcer margins often show acanthosis, hyperkeratosis, and numerous dermal telangiectasias. Cartilage beneath the granulomatous and fibrotic dermis is disrupted, hemorrhagic, and even necrotic, although occasionally it can appear undamaged (4, 6, 7).

Figure 1. Nodule of chondrodermatitis nodularis chronica helicis on the helix showing erythema, superficial scaling, and a central crust

Figure 2. a, b. Photomicrograph of the nodule on the helix of the left ear (a) showing a central crateriform ulcer covered by a crust (hematoxylin and eosin; original magnification 4x). Bottom of the ulcer (b) showing granulation tissue and degenerative changes in the cartilage (hematoxylin and eosin; original magnification 20x)

The pathogenesis of CNCH has not yet been fully elucidated. The Swiss dermatologist Max Winkler was the first to describe this condition in 1915 and suspected the influence of thermal, traumatic, or chemical factors on the anatomic prominent cartilage portions of the ear (7). However, lesions often appear to arise spontaneously, and patients may not identify a triggering factor (6). In the following decades, further causes of CNCH, such as actinic damage, disorders of cornification, collagen defects, and perichondral vasculitis, were proposed (7). Only the last hypothesis has recently received support from histopathological studies, which have demonstrated arteriolar narrowing in the perichondrium as the possible cause of underlying cartilage necrosis resulting in CNCH (9). The association between CNCH and autoimmune diseases has also been observed (5, 6, 8). Newcomer et al. reported a series of 99 cases of CNCH and found autoimmune diseases in 3 of them (2 rheumatoid arthritis and 1 SSc) (8). Three additional cases of CNCH in SSc have been reported by Bottomley and Goodfield (5). The ages of the patients were 41, 54, and 47 years, respectively, and all of them were diagnosed with lcSSc, but none had concomitant PBC. No other predisposing factors were identified, except the presence of SSc (5). Our patient also developed CNCH spontaneously, but the symptoms were exacerbated by mechanical factors. These authors suggested that CNCH may be underreported in SSc, because those 3 cases were identified in a total of 23 patients (5). This condition has also been reported in juvenile dermatomyositis (8).

Factors that may increase the risk of developing CNCH in SSc include vascular pathology and autoimmunity. The most characteristic vascular finding in patients with established SSSs is bland intimal proliferation in the small- and medium-sized arteries that produce progressive luminal occlusion (1), similar to that observed in the perichondrial arteries in CNCH (9). Raynaud’s phenomena may also exacerbate the vascular occlusion and lead to ischemia in exposed areas, such as the ears (5). Antibodies to denatured type II collagen have been detected in CNCH and may contribute to its pathogenesis (10). These antibodies also occur in SSc, but their exact role in this disease is unknown (1). Skin disorders associated with PBC include xanthomas, vitiligo, melanosis, xeroderma, lichen planus, and cutaneous vasculitis (2), but CNCH has not been previously reported.

The conditions most frequently encountered with a similar clinical appearance to CNCH are basal cell and squamous cell carcinoma (7). It is important to take this into account, as patients with SSc have an increased risk of developing skin malignancies (1). Therefore, a biopsy of the lesion must be performed to rule out malignancy (4). CNCH can be treated surgically with a wedge-shaped excision or medically with more conservative therapies, including laser, pressure-relieving padding, collagen injections, and topical or intraslesional glucocorticoids (7). Recurrence after surgical excision occurs in up to 34% of patients, as in our case (6).

In conclusion, CNCH is an inflammatory and degenerative skin disorder of the external ear that may occur in patients with SSc. Although this condition is rare, it is necessary to know, because it may be confused with skin malignancies due to its clinical appearance.
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