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Case Report

CASE REPORT

Recurrent auricular inflammation caused by Kimura's disease: reminiscent of the early phase of relapsing polychondritis?

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Abstract

Recurrent auricular inflammation with pain and swelling is due to the composition and environmental exposures experienced by the constituent parts of the ear. A painful, swollen ear may suggest acute perichondritis, subperichondrial hematoma or relapsing polychondritis (RP). Here, we report a case of a 51-year-old Asian man who had an approximately 2-year history of recurrent auricular swelling and was referred for suspected RP. Biopsy of the lesion revealed the formation of lymphoid follicles with a dense infiltrate of lymphocytes, histiocytes and eosinophils. His serum IgE level was 12 040 U/mL (normal range 0–358). These findings suggest that the patient had Kimura's disease (KD). Physicians should be aware of KD as a potential cause of recurrent auricular inflammation.

INTRODUCTION

Lesions of the ear reflect the composition and environmental exposures experienced by the constituent parts of the ear, namely, the skin, subcutaneous tissues and cartilage of the external ear and the mucosa, ossicles, nerves, muscles and blood vessels of the middle ear and mastoid. Lesions of the external ear in elderly individuals predominantly consist of bumps originating from the skin and cartilage. With few exceptions, they are similar to lesions of the skin and soft tissues in other parts of the body and particularly reflect the exposure of the skin and underlying cartilage to actinic injury. A painful, swollen ear may suggest acute perichondritis, subperichondrial hematoma or relapsing polychondritis (RP), although a definite diagnosis of the lesion may be difficult to achieve based on clinical symptoms due to various environmental exposures and components of the ear [1].

Perichondritis is a diffuse infiltrate of inflammatory or neoplastic leukocytes in the skin of the external ear [1, 2]. Acute myeloid leukaemia, B- and T-cell lymphomas and acquired immunodeficiency disease-related non-Hodgkin's lymphoma can present as auricular perichondritis [1]. Recurrent or bilateral auricular swelling should prompt a rheumatologic workup for RP [2].

RP is a rare immune-mediated disease of unknown aetiology that causes recurrent inflammation of the cartilage in the body. The most common feature of RP at the time of presentation is unilateral or bilateral inflammation of the ear, which is observed in approximately 43% of RP patients. Auricular inflammation can persist from days to weeks [1–6].

Kimura's disease (KD) is also a rare, chronic inflammatory disorder of unknown aetiology. Clinically, it presents as solitary or multiple subcutaneous nodules, predominantly in the head and neck region, typically in the pre-auricular region, forehead

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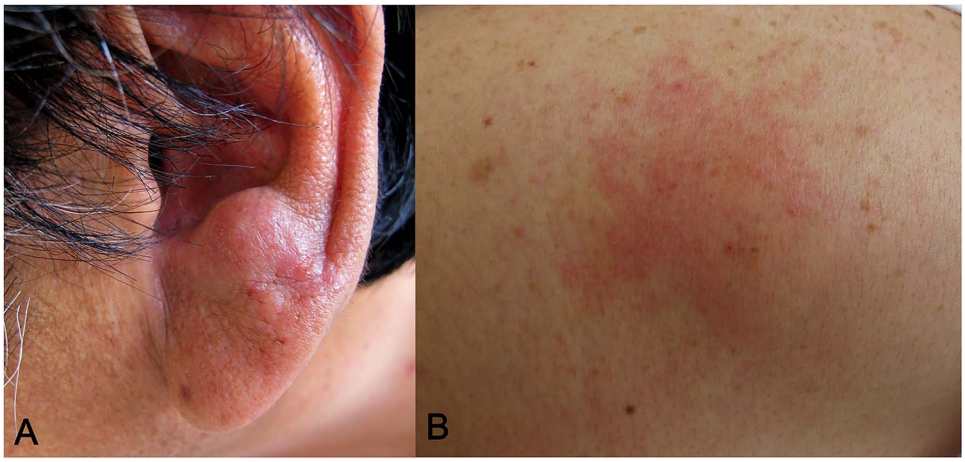

Figure 1: Auricular inflammation of the patient's left ear with redness and swelling of the pinna (A). Notably, the ear lobe, which does not contain cartilage, was free from inflammation. Urticarial erythema and papules were observed on the left upper trunk (B).

and scalp. This rare disease is endemic in parts of Asia (Japan, China, Indonesia, etc.). Although KD has been reported in Europe and America, the predominant population is Asian. It mainly affects young Asian men (median age 32 years; male:female ratio 3.5–6.0:1) [7, 8].

Here, we report a case of a patient with recurrent auricular inflammation that was suspected of being an early phase of RP but was eventually diagnosed as KD.

CASE REPORT

A 51-year-old Asian man with an approximately 2-year history of recurrent left auricular swelling was referred to our division with suspected RP. His relevant medical history included hypertension and chronic eczema. He was presented with pain and swelling of the left ear (Fig. 1A). The lesion, which did not include the ear lobe, was reminiscent of RP.

The patient also presented with urticarial erythema and papules on the right upper trunk (Fig. 1B). He was treated with 300 mg/day oral cefteram pivoxil and 120 mg/day oral loxoprofen sodium for the ear inflammation and 5 mg/day oral levocetirizine hydrochloride and topical corticosteroid (0.05% betamethasone butyrate propionate) for the skin lesions. Both the ear inflammation and the skin lesions improved with the medication.

Approximately 3 months later, the pain and swelling of the left ear along with the stiffness of the post-auricular area had worsened. A contrast-enhanced CT scan revealed a diffuse, enhanced tumour of the soft tissue in the left parotid region (Fig. 2) and enlargement of the lymph nodes near the caudal portion of the tumour. A punch biopsy of the ear lesion showed a dense infiltrate of lymphocytes, histiocytes and eosinophils, with the formation of lymphoid follicles mainly in the subcutaneous tissue (Fig. 3). His serum IgE level was 12 040 U/mL (normal range 0–358 IU/mL). Based on this evidence, the patient was diagnosed with KD.

In addition to continuous treatment with oral levocetirizine and topical corticosteroids, the subcutaneous tumour was gradually reduced by local injections of triamcinolone acetonide.

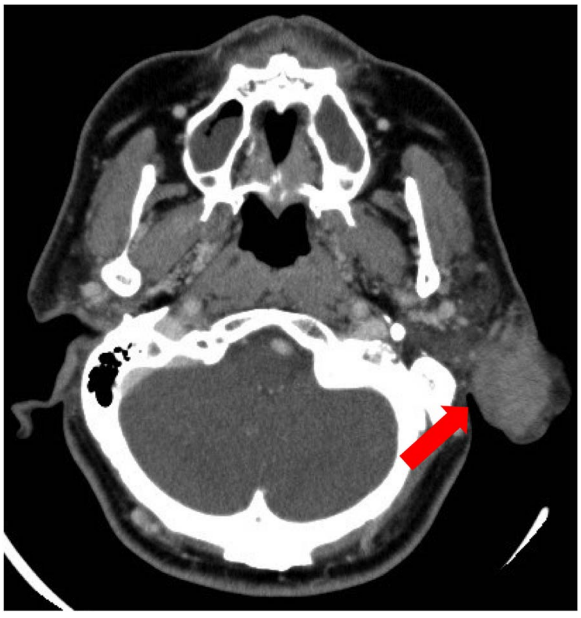

Figure 2: Contrast-enhanced CT showing tumors with diffuse enhancement in the left subaural region (arrow).

DISCUSSION

Currently, the diagnosis of RP relies mostly on the criteria established by Michet *et al.* [6], which require the presence of proven inflammation in at least two of three critical areas (auricular, nasal or laryngotracheal cartilage) or proven inflammation in the cartilage in one of these areas plus two other signs, including ocular inflammation, vestibular dysfunction, seronegative inflammatory arthritis or hearing loss [3, 5, 6]. However, establishing a diagnosis of RP is still challenging because these common symptoms are often absent in the early stages of the disease, resulting in a typical delay in diagnosis of nearly 3 years [3, 5]. In fact, an RP patient only exhibiting inflammation of the laryngotracheal cartilage has been reported [9].

The skin is involved in 20–30% of patients with primary RP [3]. Urticarial erythema and papules on the shoulders, neck and upper trunk are characteristic in RP patients before the onset

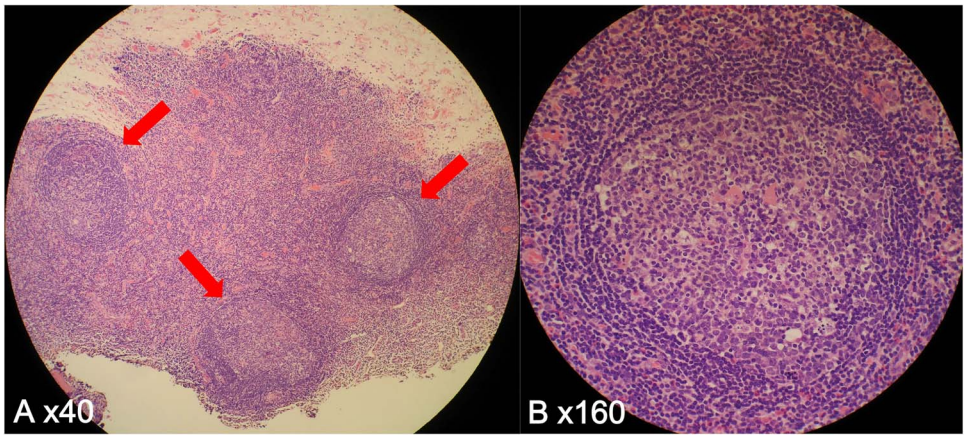

Figure 3: Histopathology of the subaural tumor showing lymphoid follicles (A; arrows) with infiltration of lymphocytes, histiocytes and many eosinophils around the lymphoid follicle (B).

of RP chondritis [10]. Therefore, it may be difficult to clinically distinguish between the early phase of RP and KD with auricular inflammation and urticarial erythema and papules. Pathological analysis was helpful for diagnosing this patient.

In terms of treatment options for RP, non-steroidal anti-inflammatory drugs are generally used for mild auricular or nasal chondritis. Colchicine and dapsone can also be used. Systemic corticosteroids are required for acute exacerbations of RP. Immunosuppression may be necessary for patients who are refractory to corticosteroids, require corticosteroid-sparing therapy or cannot tolerate corticosteroids. Cyclosporine, methotrexate and azathioprine have all shown benefit in patients with RP. Furthermore, tumour necrosis factor-α antagonists such as infliximab, etanercept and adalimumab show some efficacy [10].

Multiple treatment methods also have been proposed for KD, including surgical excision, corticosteroids and radiotherapy. The preferred initial treatment for localized KD may be surgical excision, while surgical excision with postoperative low-dose radiation therapy, radiation therapy alone or systemic corticosteroids have been proposed [8, 11, 12].

In conclusion, KD may be considered as a differential diagnosis for recurrent auricular inflammation.

CONFLICT OF INTEREST STATEMENT

None to report.

CONSENT

Written consent received.

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