A Case of Winkelmann Granuloma

Kyoung-Ae Jang, M.D., Jee-Ho Choi, M.D., Kyung-Jeh Sung, M.D., Kee-Chan Moon, M.D., and Jai-Kyoung Koh, M.D.

Department of Dermatology, Asan Medical Center, College of Medicine, University of Ulsan, Seoul, Korea

We report a case of Winkelmann granuloma in a 63-year-old man. Histopathological findings of the biopsy specimens from the lesions of the ear, finger and iliac crest area were compatible with Winkelmann granuloma. Winkelmann granuloma is a rare disorder showing an association with systemic immunoreactive disorders. Although our patient did not have any definite systemic disease, he had characteristic clinical and histopathological findings of Winkelmann granuloma, arthralgia, an elevated erythrocyte sedimentation rate, positivity to the rheumatoid factor and antinuclear antibodies. Therefore, we believed that he was strongly suspected to have an unclassifiable systemic immunoreactive disease. (Ann Dermatol 10(3) 163–166, 1998).

Key Words: Winkelmann granuloma, Systemic immunoreactive disorder

Dicken and Winkelmann1 observed “the cutaneous, necrotizing, palisading granuloma” termed as “Churg-Strauss granuloma” in 1978 during a histopathological study of skin specimens from seven patients. Since then, lesions showing similar histopathological findings have been described as various nomenclatures. Wilmoth and Perniciaro2 proposed the unifying name “cutaneous extravascular necrotizing granuloma of Winkelmann (Winkelmann granuloma)” in 1996.

We report a case of Winkelmann granuloma involving the ear lobes which are an unusual location of this distinct histopathological condition.

CASE REPORT

A 63-year-old man had a 4-year history of asymptomatic papules and nodules on both his ear lobes, fingers, and the iliac crest area. A physical examination revealed multiple, yellow-red papules and nodules (Fig. 1-3). A histopathological exami-

nation of a papule from the right ear rim showed a palisading granuloma with central necrobiosis composed of neutrophils, neutrophilic debris, and necrobiotic collagens within the entire dermis (Fig. 4-6). The central necrobiotic area was surrounded by histiocytes. The epidermis was acanthotic and capillary proliferations and dilatations were noticed in the superficial and deep dermis. The two skin biopsy specimens from the lesions of his finger and iliac crest area showed similar histopathological pictures as in the lesion of the right ear rim. His past medical history showed that he had been taking analgesics and systemic corticosteroids for arthralgia of both knees for many years. He began suffering from arthralgia 6 years ago, which did not fulfill the “classic” rheumatoid arthritis criteria based on an American Rheumatism Association (ARA) criteria. Laboratory studies revealed the elevated erythrocyte sedimentation rate (60 mm/hr), the presence of rheumatoid factor (1:224), and positive antinuclear antibodies (1:160, homogenous pattern). The results of the following studies were within normal limits or negative; a blood cell count, urinalysis, liver function tests, immunoglobulins, complement levels, uric acid, anti-double strand DNA, extractable nuclear antigen (ENA) series, and antineutrophil cytoplasmic antibodies (ANCA). We concluded that this case was most likely Winkelmann granuloma.
Fig. 1. Several discrete, yellow-red papules on the right ear.

Fig. 2. A relatively well-defined, 5 × 4 cm size, brownish subcutaneous nodule on the left iliac crest area.

Fig. 3. Several reddish papules on the right hand.

The patient's lesions were removed by shaving and CO₂ laser treatment was administered. However, all the lesions were recurred within 3 months of treatment. The patient is now being treated with hydroxychloroquine, 200mg, twice a day and a

Fig. 4. Two palisading granulomas with central necrobirosis in the dermis (H&E, ×12.5).

Fig. 5. Central necrobirosis surrounded by numerous neutrophils, neutrophilic debris, and necrobiotic collagen (H&E, ×100).
course of intralesional triamcinolone injections. Unfortunately, there has been no marked improvement.

**DISCUSSION**

In Winkelmann granuloma, an association with allergic granulomatosis, other systemic diseases including lupus erythematosus, Wegener’s granulomatosis, rheumatoid arthritis, subacute bacterial endocarditis, chronic active hepatitis, inflammatory bowel disease, and Takayasu’s arteritis has been reported. A group of diseases characterized by cutaneous papules and nodules where histopathological examinations reveal a palisading granuloma composed of necrobiotic collagens, neutrophils and neutrophilic debris, has been reported under the various nomenclatures, such as Churg-Strauss granuloma, cutaneous extravascular necrotizing granuloma, rheumatoid papules, superficial ulcerating rheumatoid necrobiosis, interstitial granulomatous dermatitis, rheumatoid neutrophilic dermatitis, palisaded neutrophilic and granulomatous dermatitis of immune complex disease, and cutaneous extravascular necrotizing granuloma of Winkelmann (Winkelmann granuloma). We prefer the term of “Winkelmann granuloma”, because “Winkelmann granuloma” is a simple term to use and Winkelmann, who was the first to describe this unique disorder, was deserved to represent this disorder. All these diseases were associated with an underlying illness. The clinical features of Winkelmann granuloma are multiple discrete, skin-colored to erythematous or violaceous papulo-nodules located predominantly on the elbows or the distal upper extremities and fingers. The involvement of the ear lobe has been rarely reported. Our patient had the skin lesions on the ears, fingers, and iliac crest areas. Granuloma annulare, rheumatoid nodules, pyoderma gangrenosum, infectious granuloma, and erythema elevatum diutinum should all be included in the differential diagnosis of Winkelmann granuloma. However, the histopathological findings showing palisading granuloma composed of neutrophils and neutrophilic debris with necrobiotic collagen cannot be seen in any other dermatosis except Winkelmann granuloma. Although rheumatoid neutrophilic dermatitis is sometimes considered as a different disease entity from Winkelmann granuloma, we thought it the same dermatosis because of a similar clinical setting and histopathological findings. Until recently radical treatment has not been settled, thus various therapeutic regimens, such as systemic corticosteroids, penicillamine, hydroxychloroquine, melphalan, antihistamines, intralesional steroid injections and systemic antibiotics have been tried. Usually the systemic and the cutaneous disease show the parallel course, with the appearance or reappearance of Winkelmann granulomas coincidental with an exacerbation of an underlying disorder. Because our patient’s skin lesions were worsened despite treatment with systemic corticosteroids for arthralgia, we started to treat some lesions by shaving and a CO₂ laser, but all those lesions recurred. We are now in the process of trying intralesional steroids and hydroxychloroquine, 200 mg, twice a day. Although he did not have any definite systemic disease, he had characteristic clinical and histopathological findings of Winkelmann granuloma, arthralgia, an elevated erythrocyte sedimentation rate, positivity to the rheumatoid factor and antinuclear antibodies. Therefore, we believed that there was a strong possibility the patient had an unclassifiable systemic immunoreactive disease. Although he did not meet the ARA criteria, rheumatoid arthritis might have been the main underlying disorder.

**REFERENCES**

1. Dicken CH, Winkelmann RK: The Churg-Strauss granuloma: cutaneous, necrotizing, palisading gran-


