

# Histopathologic Features of Giant Cell Arteritis in an Actinic Granuloma Lesion

Zahra Asadi Kani, MD  
Farhad Malekzad, MD  
Mehdi Qeisari, MD  
Nastaran Namazi, MD

Skin Research Centre, Shahid Beheshti University of Medical Sciences, Shohada-e- Tajrish Hospital, Tehran, Iran.

Corresponding Author:  
Nastaran Namazi MD  
Skin Research Centre, Shahid Beheshti University of Medical Sciences, Shohada-e- Tajrish Hospital, Tehran, Iran.  
E-mail: nastarran22@yahoo.com

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The association between actinic granuloma and giant cell arteritis (*temporal arteritis*) has been claimed by some authors. There is a hypothesis that actinic radiation has the principal role in the etiology of both diseases in a similar way. Here, we report a case of actinic granuloma that had characteristic pathologic features of giant cell arteritis in histopathologic examination without clinical evidence of giant cell arteritis.

**Keywords:** actinic granuloma, giant cell arteritis, actinic damage, annular

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## INTRODUCTION

Actinic granuloma is an annular inflammatory reaction with a giant cell dermal infiltrate, which develops in an area of actinic elastosis.

The condition develops in the exposed 'weather-beaten' skin of the patients after the third decade, and is more common in sunny countries. Fair-skinned or freckled subjects are particularly susceptible. Disease starts insidiously as a small, pink papule, which progresses to form a smooth, slightly raised annulus of firm superficial dermal thickening. This ring measures 0.2–0.5 cm in width but may expand up to 6 cm in diameter. The center may become slightly atrophic, and variable depigmentation may occur. The lesion is usually asymptomatic.

Giant cell arteritis (*temporal arteritis*) is a granulomatous vasculitis affecting medium to large-sized elastic arteries, with a predilection for the superficial temporal and ophthalmic arteries and to a lesser extent other extracranial branches of the carotid arteries. Cutaneous manifestations

are uncommon, the most frequent is necrosis of the scalp with ulceration.

Although the etiology of temporal arteritis is unknown, actinic degeneration of the internal elastic lamina may provoke granulomatous inflammation in a reaction similar to the actinic granuloma in some cases <sup>1</sup>.

Giant cell arteritis is a rare association of actinic granuloma. Here we report a case of actinic granuloma that had histopathologic manifestations of both actinic granuloma and giant cell arteritis without overt clinical manifestation of giant cell arteritis.

## CASE REPORT

A 62-year-old man came to our dermatology clinic because of two asymptomatic depressed lesions on his face since 8 months ago. He stated that these lesions had started as a papule expanding outward gradually and leaving a depressed center. There was no antecedent history of insect bite or vesicular lesions, no signs or symptoms, and no

relevant medical or drug history.

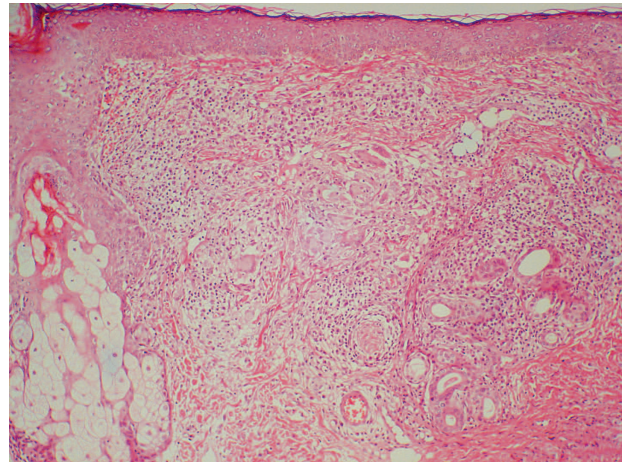
There was a sharply demarcated annular atrophic lesion without raised borders beneath the medial portion of his left eyebrow measuring 7×5 mm in diameter and another one on the bridge of the nose was 9×4 mm in diameter. There was no nodularity or palpable thickening in the left periocular region (Figure 1).

Initial diagnoses included discoid lupus erythematosus and granuloma annulare. An elliptical biopsy was taken from the border of the lesion on the bridge of the nose and histopathology showed moderately dense infiltration of mixed inflammatory cells in the dermis including lymphocytes, histiocytes and foreign body type multinucleated giant cells, some of them engulfed

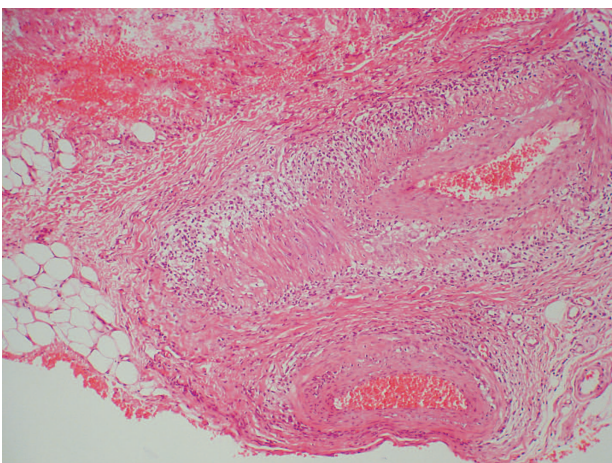
basophilic elastotic fibers and produced vague granulomatous aggregates in the vicinity of the solar elastotic changes (Figure 2). There was also an increased number of plasma cells and some eosinophils. No evidence of necrobiosis or dermal mucin was identified. These changes were in favor of actinic granuloma. In deep dermis, there were medium sized arteries with prominent infiltration of lymphocytes, some plasma cells, histiocytes and multinucleated giant cells within their walls associated with partial destruction of internal elastic lamina consistent with giant cell arteritis (Figure 3). In orcein staining, fragmentation and focal absence of internal elastic lamina was seen (Figure 4). There was also marked fragmentation and swelling of the elastic fibers in the dermis



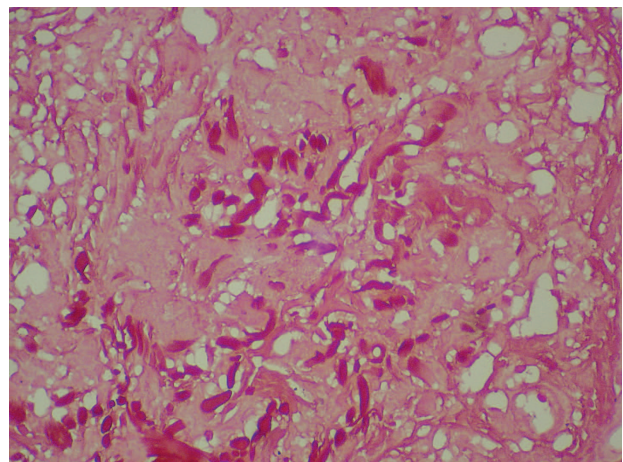
**Figure 1.** Annular atrophic lesion without raised borders beneath the medial portion of his left eyebrow.



**Figure 2.** Moderately dense infiltration of mixed inflammatory cells in the dermis including lymphocytes, histiocytes and foreign body type multinucleated giant cells, produced granulomatous aggregates in the vicinity of the solar elastotic changes (H&E\*10).



**Figure 3.** Prominent infiltration of lymphocytes, some plasma cells, histiocytes and multinucleated giant cells within the vessel walls associated with partial destruction of internal elastic lamina (H&E\*10).



**Figure 4.** Orcein staining shows fragmentation and focal absence of internal elastic lamina.

between granulomatous inflammation.

He had no history of headache, fever, malaise, visual impairment, jaw claudication, tenderness of the forehead, erythema or oedema of the skin overlying the lesions in favor of giant cell arteritis.

Routine laboratory data were within normal limits. Serologic collagen vascular tests were negative.

## DISCUSSION

Actinic granuloma is a controversial entity and has been named with different titles from the first explanation until now. The first description of actinic granuloma was made more than 30 years ago in 1971 by Wilson Jones<sup>2</sup> who described it as an atypical annular form of necrobiosis lipoidica affecting the upper face and scalp margins. Mehregan and Altman<sup>3</sup> described similar annular lesions occurring predominantly on the exposed skin of the head, neck and arms, and named these "Miescher's granuloma of the face". O'Brien<sup>4</sup>, for the first time, named it "actinic granuloma".

Whether actinic granuloma is a distinct entity or represents granuloma annulare or some other granulomatous disease that happens to occur on light-exposed skin has been the subject of controversy.

In 1979, the actinic granuloma concept was intensely criticized by Ragaz<sup>5</sup> and Ackerman who argued that the granuloma is not a primary autoimmune reaction against actinically degenerate elastic tissue but is merely a variant of the etiologically obscure granuloma annulare occurring in sun-damaged skin. Ackerman disagreed with O'Brien about the specificity of actinic granuloma and stated that elastophagocytosis may be found in any granulomatous dermatitis, ranging from suppurative granulomas secondary to rupture of follicular cysts to granuloma annulare<sup>6</sup>.

In late 1979 Hanke et al,<sup>7</sup> confirmed that the disorder is quite distinct and definitely separate from granuloma annulare; but they reclassified it on a purely morphologic basis under a new name, "annular elastolytic giant cell granuloma" (AEGCG). However, they agreed that AEGCG and actinic granuloma are "the same entity."

O'Brien and Regan in 1998<sup>8</sup> published an article regarding the actinic aetiology of actinic granuloma and temporal arteritis and proposed "the actinic

hypothesis of the granuloma" as a basic principle applying to the skin and its superficial arteries: actinic radiation ultimately and selectively injures "exposed" elastic tissue and this degenerated tissue may then become an autoimmune target or antigen. They also proposed autoimmune actinic pathogenesis for temporal arteritis.

In support of the actinic basis for actinic granuloma and temporal arteritis, there are reports describing the association of both diseases: Two cases were published by Lau, Reid, and Weedon<sup>9</sup>. And another instance was noted by O'Brien and Argyle<sup>10</sup>. Fukai et al,<sup>11</sup> reported a patient displaying generalized granuloma annulare with temporal arteritis whose lesions were presumably actinic granuloma. It should be mentioned that there has been no direct demonstration that actinic damage can induce granuloma formation, and one case report described the failure to provoke lesions after a 4-day photochallenge in a patient with pre-existing actinic granuloma<sup>12</sup>. Additionally, it has been noted that elastic fiber destruction can be observed in granuloma annulare and necrobiosis lipoidica diabetorum, demonstrating that elastin destruction may be secondary to granulomatous inflammation<sup>13</sup>.

Manifestation of actinic granuloma in our case was unusual; firstly because of the demarcated rimmed borders without elevation and secondly because the lesion beneath the eyebrow had not enough solar exposure justifying actinic damage. Although this lesion was not biopsied, the patient emphasized that both lesions had appeared simultaneously and had the same evolutionary course.

It is recommended to carry out a radial biopsy through the annulus of a lesion suspected to be actinic granuloma to see three distinct zones in the dermis; but because actinic granuloma was not in our primary differential list, we did a conventional elliptical biopsy from the border of the lesion that showed changes consistent with actinic granuloma. There were no histopathological findings of other granulomatous diseases that could induce elastorrhexis such as rosacea, acne agminata and Wegener's granulomatosis. Foreign body was not seen in the sections and there was no mucin deposition in alcian-blue staining in favor of granuloma annulare. Even though clinical presentation of the lesions was similar to atrophic scars, histopathological examination ruled it out.



Our case showed histopathological features of both giant cell arteritis coupled with typical actinic granuloma in the same lesion. It is not evident whether the presence of the features of both diseases in the same histopathological section is in favor of their association as mentioned in the literature, or one of them is a phenomenon secondary to the other one. The localization of the lesion in the ophthalmic artery domain supported temporal arteritis but the patient had no clinical manifestations of this disease at presentation and during 4 months of follow-up. However, we recommended long-term follow-up after fully explaining the symptoms and signs of temporal arteritis to the patient.

If the association between the two diseases is established, screening and follow-up of all actinic granuloma patients to detect progression to temporal arteritis is then prudent and an actinic aetiology could be proposed for giant cell arteritis. On the other hand, if giant cell arteritis is proved to be a histopathological feature secondary to actinic granuloma or a coincidental finding, there is no concern regarding this serious disease.

We can propose a hypothesis that actinic radiation can provoke autoimmune granulomatous inflammation in relation to actinically damaged and degenerated ("elastotic") elastic tissue of the skin and its vessels. For dermatologists and rheumatologists, this is a significant problem that deserves to be addressed and resolved.

However, further studies are recommended to estimate the prevalence of histopathological features of giant cell arteritis in actinic granuloma lesions.

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