Case report

Annular Elastolytic Giant Cell Granuloma - A Rare Case with Systemic Involvement

Das S[1], Chowdhury J[2], Patra S[3], Achar A[4]

Abstract:

Annular elastolytic giant cell granuloma (AEGCG) is a rare granulomatous dermatosis characterized by loss of elastic fibers and elastophagocytosis by multinucleated giant cells. It is characterized by annular plaques that are similar to those observed in granuloma annulare but that specifically appear in sun-exposed skin and occurs more commonly in females than males. There have been reported cases of AEGCG associated with diabetes mellitus, systemic sarcoidosis, cutaneous amyloidosis, molluscum contagiosum, squamous cell carcinoma of the lung and cutaneous T-cell lymphoma. We report a case of AEGCG in both sun-exposed as well as covered areas of a middle aged lady with hepatic nodules and Barret's esophagus.

Key words: annular elastolytic granuloma; liver nodules; Barret's oesophagus

Bangladesh Journal of Medical Science Vol. 15 No. 03 July'16. Page : 488-491

Introduction: The term annular elastolytic giant cell granuloma (AEGCG) was first proposed by Hanke et al[1] to include similar clinical cases that had been diagnosed as atypical necrobiosis lipoidica of the face and scalp, Miescher's granuloma of the face[2], and actinic granuloma.

Annular elastolytic giant cell granuloma (AEGCG) is a rare granulomatous skin disease, which is characterized histologically by an absence of elastic fibres, due to elastophagocytosis by multinucleated giant cells in the dermis[3, 4] The majority of AEGCG cases have been identified in middle-aged, Caucasian women. The most common clinical presentation consists of annular plaques or patches often with elevated borders and central atrophy [4], Clinically, AEGCG presents as multiple, large, annular plaques with a raised, erythematous border and central atrophy. The lesions are mostly located on sun-exposed areas such as the face and neck, but they are also seen on nonexposed skin although rare reports of a papular variant of AEGCG exist [5, 6]

Case report:

A forty year old female patient was referred to us from medicine department with multiple asymptomatic scaly skin lesions. She noticed these lesions since last five years. It initially started on her back and gradually appeared on face and neck. On examination multiple well defined erythematous, plaques mainly distributed over the sun exposed parts were found. The predominant sites of the lesions are face, both upper & lower back. [Figure 1]The lesion on the back showed central atrophy. Lymph nodes were not palpable. There was no mucosal involvement.

She had a history of pain abdomen for the past five years with occasional dysphagia. For that initial investigation was done from medicine department. Ultasonography of whole abdomen revealed presence of multiple nodules in the liver and retroperitoneal lymphadenopathy. CT abdomen showed presence of multiple small hypodense poorly enhancing lesions throughout

1. Dr. Suchibrata Das, Assistant Professor, Department of Dermatology, Venereology and Leprosy, NRS Medical College.
2. Dr. Joyeeta Chowdhury, RMO Cum Clinical tutor, Department of Dermatology, Venereology and Leprosy, NRS Medical College.
3. Dr Sangita Patra, RMO Cum Clinical tutor, Department of Gynaecology and Obstetrics, Medical College and Hospital, Kolkata, 88, College Street, Kolkata 700072.
4. Dr. Arun Achar, Associate Professor, Department of Dermatology, Venereology and Leprosy, NRS Medical College.

Corresponds to: Dr. Suchibrata Das, Assistant Professor, Department of Dermatology, Venereology and Leprosy, NRS Medical College, 138 AJC Bose Road, Kolkata 700014, West Bengal, India.
E-mail: suchibratadas@yahoo.com

Figure 1: Erythematous plaque on back and neck

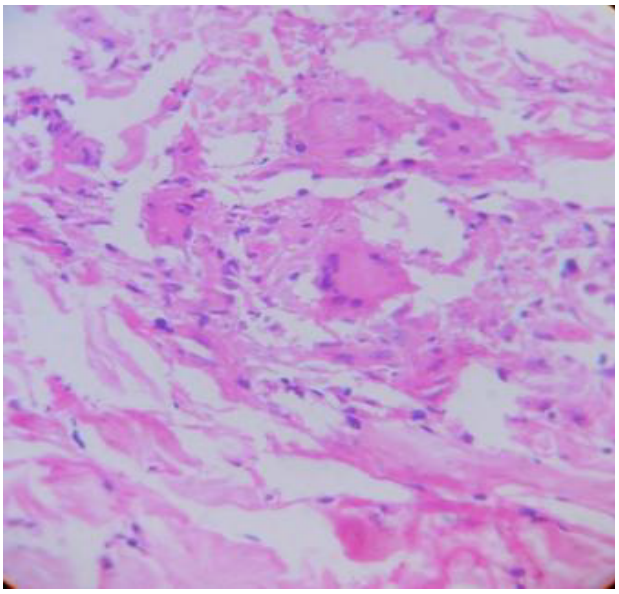

Figure 2- Histopathology of skin lesion showed dermal histiocyte aggregates, granulomas & giant cells centered around a degenerated collagen with absence of mucin [400x]

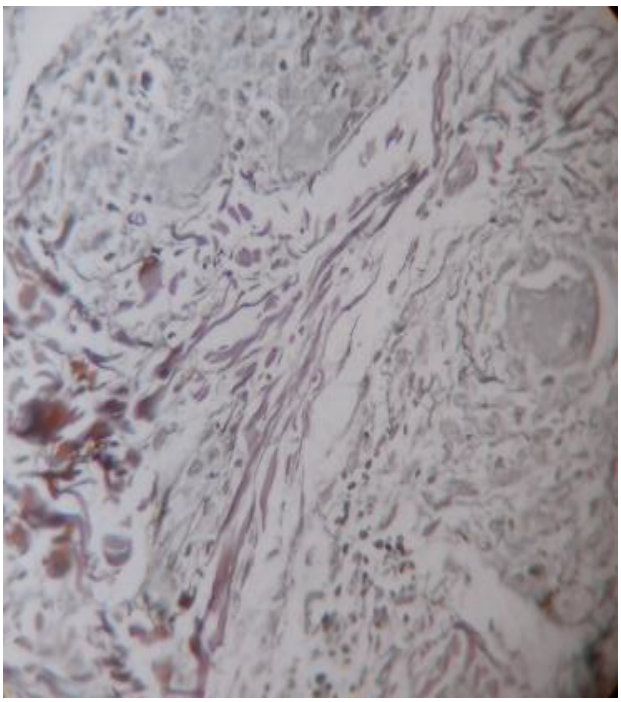

Figure 3- Von-Gieson stain shows loss of elastic fibres

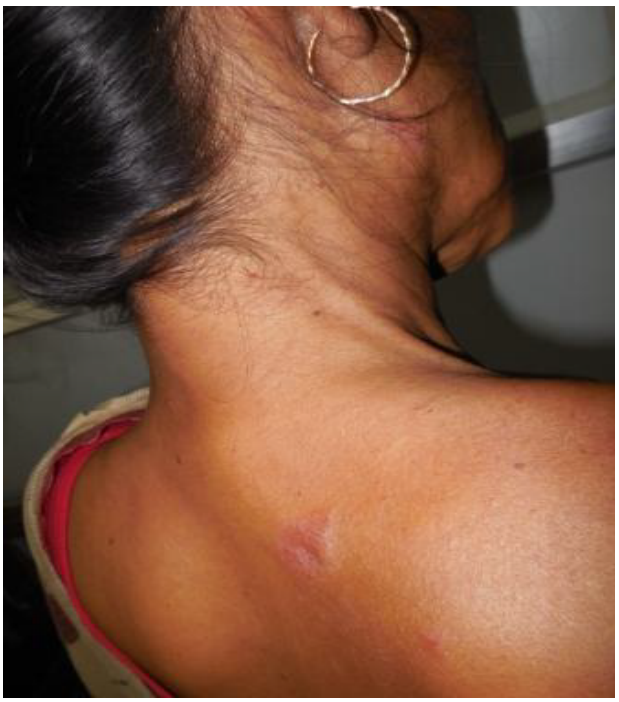

Figure 4- Healing plaques with atrophy after PUVASOL therapy

the liver & splenic parenchyma associated with retroperitoneal lymphadenopathy. Ultrasonography guided Fine needle aspiration of the liver nodule & retroperitoneal lymph nodes show presence of non-specific granulomatous lesion with elastolysis. For dysphagia, upper GI Endoscopy and biopsy was done and it was suggestive of Barret's oesophagus, but the biopsy of squamocolumnar junction remains negative for dysplasia. Colonoscopy was normal. Chest x-ray was normal. Mantoux test was negative. She was treated as abdominal TB with CAT-1. After completion of antitubercular drugs, a CT scan of whole abdomen was done and it showed reduction of the size of the retroperitoneal lymph

nodes. But the hepatic nodules and skin lesions were unchanged.

We advised all routine investigations along with skin biopsy. Apart from mild anaemia & raised erythrocyte sedimentation rate complete haemogram report was within normal limit. Alkaline phosphatase was raised in liver function test. Serum calcium and angiotensin converting enzyme I levels were also normal. Histopathology of skin lesion showed dermal histiocyte aggregates, granulomas & giant cells centered around a degenerated collagen with absence of mucin i.e. annular elasolytic giant cell granuloma [Figure-2].Von-Geison (elastin) stain showed absence of elastin [Figure-3].

We put the patient on PUVASOL and topical steroids and after one month all the lesions regressed remarkably with atrophy [Figure-4].

Discussion-Annular elastolytic giant cell granuloma(AEGCG) is catagorised as non-diabetis- associated- necrobiasis lipiodica of face. It is currently unclear whether they simply represent variant of Granuloma Annulare occurring on sun-damaged skin or are distinct disease. There are two patterns. One is a single, asymptomatic, atrophic-appearing, yellow thin plaque on forehead, other is multiple, upper extremity and trunk lesions occurring mainly on sun exposed areas predominantly in women. A papular variant is also described. The histopathologic features are best demonstrated by a biopsy of the elevated edge of the plaque[7].

They are characterized by granulomatous infiltrates with multinucleated giant cells in the upper and mid dermis, loss and fragmentation of elastic fibers, and elastophagocytosis by giant cells, without necrobiosis or mucin deposition.

These features help to distinguish AEGCG from granuloma annulare and necrobiosis lipoidica, which are the main disorders in the histological differential diagnosis[7]. Association with temporal arteritis is reported[8]. There have been reported cases of AEGCG associated with diabetes, sarcoidosis, and hematological malignancies [9, 10].

Although the etiology and pathogenesis of AEGCG are not fully understood it is believed that solar radiation, heat, or other factors lead to the alteration of elastic fibers precipitating an immune response and granuloma formation [11]. Several authors suggest that AEGCG and mid-dermal elastolysis may represent different stages of the clinical spectrum of dermal elastolysis. After an elastophagocytosis inflammatory phase by multinucleated giant cell is followed by a phase of loss of elastic fibers in the reticular dermis [12, 13].

The clinical features and histopathology of our patient were compatible with AEGCG. But it is reported as- 1. It affected both sunexposed and sun protected areas.2. It was associated with abdominal TB. 3. It was associated with Barret's esophagus. 4. The patient had hepatic nodules that showed non-specific granulomas with elastolysis, similar to the skin lesions.

The differential diagnosis of this case includes granuloma annulare, sarcoidosis, necrotic xanthogranuloma. Palisading granuloma with prominent necrobiosis , characteristic of granuloma annulare was absent in this case. Multinucleated giant cells were prominent in this patient but are not usually seen in GA.

Extensive hyaline necrobiosis, granulomatous infiltrate with foam cells and Touton-type giant cells,

and paraproteinemia, which are usually observed in necrobiotic xanthogranulom [14, 15] were absent in this patient.

Although an association of AEGCG with sarcoidosis has been reported, the elastolytic granulomas of the liver in this patient were different from those of sarcoidosis which is characterized by epithelial tubercles without caseation necrosis in.

Conflict of interest: None

References-

1. Hanke CW, Bailin PL, Roenigk HH Jr. Annular elastolytic giant cell granuloma. J An Acad Dermatol 1979; 1:4l 3- 21.
2. Doming GB, Wilson-Jones E. Atypical (annular) necrobiosis lipoidica of the face and scalp. Dermatologica 1967; 135:11-26.
3. Morita K, et al. Papular elastolytic giant cell granuloma: a clinical variant of annular elastolytic giant cell granuloma or generalized granuloma annulare? Eur J Dermatol 1999; 9: 647 PMID:10586135
4. Goncalves RR, et al. Annular elastolytic giant cell granuloma--case report. An Bras Dermatol. 2011; 86: S69-71 PMID:22068775
5. Ishibashi A, Yokoyama A, Hirano K. Annular elastolytic giant cell granuloma occurring in covered areas. Dermatologica 1987; 174: 293PMID:3622881
6. Kato H, et al. Annular elastolytic giant cell granuloma: an unusual case with papular lesions. J Dermatol1991; 18: 667 PMID:1800533
7. Meadows KP, O'Reilly MA, Harris RM, Petersen MJ. Erythematous annular plaques in a necklace distribution. Arch Dermatol. 2001;137:1647-1652. PMID:11735722.
8. Shoimer I, Wismer J. Annular elastolytic giant cell granuloma associated with temporal arteritis leading to blindness. J Cutan Med Surg. 2011 Sep-Oct;15(5):293-7. PMID:21962191
9.. Djilali-Bouzina F, Grange F, Krzisch S, Schnebelen MP, Grosshans E, Guillaume JC. Annular elastolytic giant cell granuloma. Ann Dermatol Venereol. 2010 Aug-Sep;137(8-9):536-40. PMID:20804898
10. Rongioletti F, Baldari M, Burlando M, Parodi A. Papular elastolytic giant cell granuloma: report of a case associated with monoclonal gammopathy and responsive to topical tacrolimus. Clin Exp Dermatol. 2010 Mar;35(2):145-8 PMID:19508563
11. Ventura F, Vilarihno C, da Luz Duarte M, Pardal F, Brito C. Two cases of annular elastolytic giant cell granuloma: Different response to the treatment. Dermatol Online J. 2010 Mar 15; 16(3):11. PMID:20233568
12. Müller FB, Groth W. Annular elastolytic giant cell granuloma: a prodromal stage of mid-dermal elastolysis? Br J Dermatol. 2007 Jun;156(6):1377-9. PMID:17459032
13. Hohenleutner S, Wlotzke U, Landthaler M, Stolz W. Elastolysis of the mid-dermis and annular elastolytic giant cell granuloma: different stages in the clinical spectrum of dermal elastolysis? Case report and review of the literature. Hautarzt. 1997 Jan;48(1):45-50. PMID:9132388
I4. Lever WF, Schaumburg-Lever G. Histopathology of the skin. 6th ed. Philadelphia: JB Lippincott Co, 1983:399-400.
15. Kossard S, Winkelmann RK. Necrobiotic xanthogranuloma with paraproteinemia. J Am Acad Dermatol 1980;3:257-70. PMID:7451693