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Case Report

# Wegener's Granulomatosis in Unusual Sites; A Case Report with an Emphasis on Histologic Findings

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## Abstract

Wegener's granulomatosis is an unusual pathologic lesion involving genito urinary systems in both sexes. To establish its diagnosis, we emphasize more on morphologic features than otherwise and importance of various histologic parameters encountered in this disease is analyzed extensively. Briefly, differential diagnosis is reviewed and reasons for this approach are elaborated.

**Keywords:** Wegener's granulomatosis- Morphology based diagnosis- Its necessity- Histologic differential diagnosis.



Abbreviations; WG: Wegener's granulomatosis.
C-ANCA- Cytoplasmic anti neutrophil Cytoplasmic antibodies.
P- ANCA-Perinuclear anti neutrophil Cytoplasmic antibodies.

## INTRODUCTION

Known once for its dismal prognostic outlook, Wegener's granulomatosis (W.G) is now considered as a disease with excellent remission in 75% cases extending for a period of 24 years, according to a report of N.I series [1] After the advent of C-ANCA and P-ANCA, these tests are considered absolutely essential in the diagnosis of this disease and alternate opinions contrary to the above, based on morphological grounds did not gain wide acceptance. The reasons why these tests are not absolutely essential are based upon the following: -

- A consensus international meeting [2] emphasized that the above tests have to be performed using both ELISA and I.F. techniques for accurate results.
- False positive tests are increasingly reported; particularly in other vasculitis disorders.
- During natural remission and isolated cases not falling under the category of usual clinical conditions, these tests are occasionally negative.
- According to Yi and Colby [3] these tests require expert personnel with years of experience to perform them.
- Insistence of the above authors even whom these tests are absolutely positive they have to be confirmed through biopsies.
- Savage [4] even questioned the role of ANCA and considered not essential in the light of negative ANCA tests in a significant number of patients.

In conditions prevalent in our country, morphological diagnosis is preferable because of the accuracy, ease with which it can be done, and simplicity in clinico pathologic interpretation and cost consideration. Since this disease is curable with personalized medical treatment leading to long term remission [5]; particularly when it occurs in atypical situations, to harvest the best results, it is absolutely essential for accuracy in the diagnosis. Because it involves massive outpouring of inflammatory infiltrate the liability of misdiagnosis is considerable and this article aims to pinpoint the essential features on which a confidential diagnosis can be made.

## CASE REPORT

These three cases; one from prostate and the other two from cervix, are incidental findings; hence there are no extraordinary clinical findings requiring elaboration. Cervical biopsies were from women; aged

46 and 56 years respectively and in whom a clinical diagnosis of cervical carcinoma was made. In prostatic patient, TURP was done for benign enlargement.

Since the entire lesion is necroinflammatory with or without granulomas, we tried to pick out salient histologic evidence on which reliable diagnosis could be made out. The following were considered for this analysis; (1) Parenchymal fibrinoid necrosis, (2) suppurative necrosis,(3) geographic necrosis; (4) necrosis with karyorrhexis, (5)caseation/coagulative necrosis, (6) sarcoid type of granulomas, (7) microgranulomas, loosely textured granulomas, (8) granulomas involving vessel walls, (9) granulomas in interstitium, (10) palisaded granulomas, (11) suppurative granulomas, (12) isolated giant cells; distributed singly or in clusters, (13) vasculitis and nature of the vessel, (14) eccentric or segmental thickening of vessel walls and (15) necrosis of vessel walls.

These lesions are graded: +++ when these findings were encountered easily, ++ when search was required to detect them and + when they were rare. In general, the commonest histologic finding (+++) was the presence of giant cells; fairly large with prominent dark (not pale) nuclei; arranged singly or arranged in clusters in the midst of necrotic material or inflammatory infiltrate of a nonspecific nature (figure-1aand 1b). The second feature which was less frequent (++) was the presence of fibrinoid necrosis; arranged mostly as geographic necrosis with or without inflammatory cells around them (figure-2). Palisading granulomas were not seen in our material, while other types of necrosis were occasionally present except caseous necrosis. The third feature on which a histologic diagnosis made was the presence of granulomas (figure-3). They did not show usual type of granulomas encountered in infective disorders like TB or even microgranulomas as in Crohn's disease consisting of few histiocytes and one or two giant cells, arranged in nodular fashion. These granulomas were composed of histiocytes, arranged in nodular and compactly arranged pattern or sometimes loosely. Granulomas involving vessel walls were rare when compared with granulomas in interstitium. Another feature, usually emphasized in the literature was vasculitis involving small vessels (+) (fig 4a and fig 4b). Because of heavy inflammatory infiltrate most of them were obscured. Suppurative granulomas, palisading necrosis involving vessel walls, giant cells in vessel walls, caseous necrosis, epithelioid granulomas as in TB, eosinophilic infiltrates are some of other markers not seen in our material (-).

Fig 1a and 1b –Giant cells -large with prominent dark nuclei arranged singly and in clusters in the midst of necrotic material and inflammatory infiltrate of nonspecific nature (H&E)

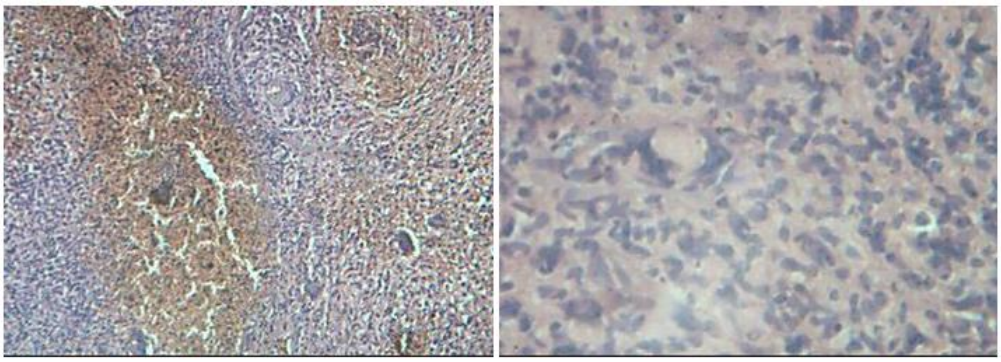
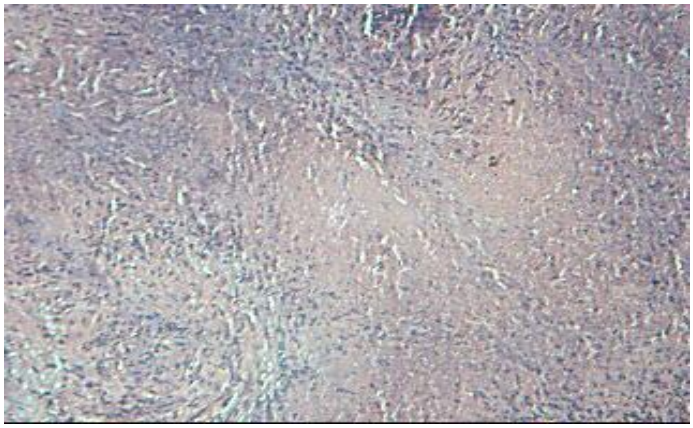


**Fig-2: Fibrinoid necrosis arranged as geographic necrosis with or without inflammatory cells around them**



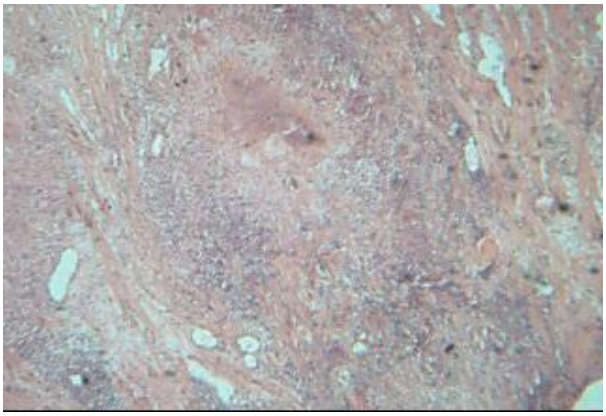


**Fig-3: Granulomas were composed of histiocytes, arranged in nodular and compactly arranged pattern**

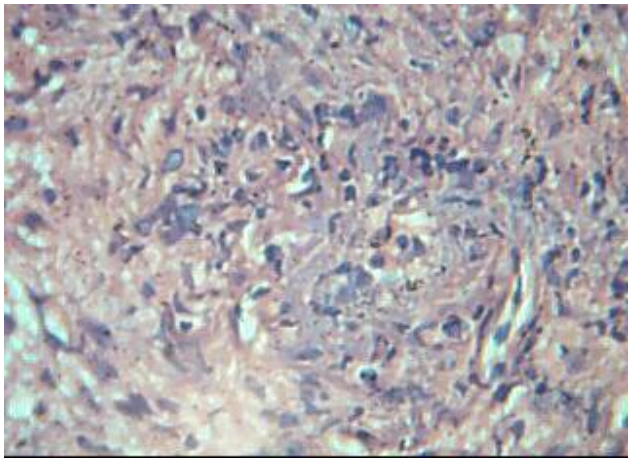


**Fig-4**

## DISCUSSION

Though W.G. was first described by Klinge in 1931, it was Wegener in a series of papers in 1936 and 1939 defined it better and later in 1954, Godman and Churg proposed for the first time classical wegener's triad [6]. Stilwell reported first case of prostatic involvement in W.G [7]. In 174 patients with W.G, only 4 patients showed involvement of prostate. In N.I. series, combination treatment with cyclophosphamide and prednisone had improved the outcome with 91% symptomatic well being, 75% with remission and 87% survival rate for 6 month to 24 years [1].

Involvement of cervix in WG was exceedingly rare in a review of 80 patients. In an analysis of 158 patients in NIAI series biopsy verified WG of cervix was noticed less than in one woman [8]. So far only in 4 women, WG was reported; aged 32, 80, 45, and 55 years respectively. All these women developed cervical manifestation of W.G when the disease relapsed and not as initial clinical manifestation [9].

In spite of extensive differential diagnosis histologically, if sufficient care is bestowed on details mentioned above, the diagnosis of W.G. becomes easier; benefiting those patients immensely. Foremost among them are infective granulomas involving these organs. Since most of the pathologists are well versed in the diagnosis of these conditions, it is superfluous to elaborate on them here. Judicious use of silver methanamine stain will solve the problem when fungal lesions are encountered. Rheumatoid granulomas show usually palisading granulomas; more over it is not difficult to separate WG from this condition clinically. In cervix, giant cell arteritis is commoner [10] than W.G. wherein giant cells are oriented around media of vessels. Churg- Strauss; a vasculitic condition may cause difficulties; since this is eosinophil based asthmatic disease diagnosis becomes easier. Other vasculitis based diseases can be ruled out on the basis of vessels involved and histological features as in micropolyangitis, periarteritis, NHL involvement of small vessels, leucoclastic vasculitis, giant cell arteritis, etc. Vasculitis is common in connective tissue disorders rather than granulomas. Nonspecific granulomatous prostatitis, because of its lobulocentric location and pus cell infiltrates due to ruptured ducts and acini need not be a consideration against W.G. Another close mimic, post-operative spindle cell tumour is less likely when clinically analyzed; palisaded granulomas are more prominent along with the presence of giant cells with phagocytosed material. Diagnosis of WG, becomes easier even in situation where WG is not common.

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