Clinico-Pathological Analysis of Cutaneous Vasculitis

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Abstract

Background: To assess the clinical presentation and histological pattern of cutaneous vasculitis

Methods: In this descriptive study patients with a clinical diagnosis of cutaneous vasculitis were included. Gross examination of the specimens was carried out and recorded. The biopsies were processed in an automatic tissue processor. The paraffin embedding was done in the embedding station. The sections were cut (3-5µm) and stained for haematoxylin and eosin. After the slides were prepared, they were observed under the microscope. All cases regardless of age and gender were included in our study.

Results: A total of 37 cases (20 males and 17 females) with age range of 8-80 years were studied. The majority of the patients presented with ulcer of the skin, followed by purpura, nodules, rash and plaques. On histopathology, the patients predominantly had leukocytoclastic vasculitis (27%), followed by cryoglobulinemic vasculitis (10.8%), panniculitis with vasculitis, and lymphocytic vasculitis. A few cases of Churg Strauss Syndrome, Capillaritis and drug induced vasculitis were also seen.

Conclusion: Cutaneous vasculitis predominantly presents as ulceration and leukocytoclastic vasculitis is the main histological type. Biopsy definitely helps the exact characterization of vasculitis for further management of the cases.

Key Words: Capillaritis, Granulomatous vasculitis, Leukocytoclastic vasculitis

Introduction

Vasculitis can present in many ways and can be challenging for the treating physician. Always the diagnosis cannot be made solely on the basis of clinical presentation and biopsy correlation is very important. Vasculitis is defined as inflammation and necrosis of the blood vessel walls which may occur independently or be a consequence of certain primary disorders. A more specific condition, cutaneous vasculitis preferably involves small and medium sized vessels of the dermis and subcutaneous tissue. It may be a localized, self limited process or may spread to involve multiple organs. Vasculitis predominantly involving small vessels include cutaneous small vessel vasculitis, cryoglobulinemic vasculitis, urticarial vasculitis, Henoch-Schonlein purpura and vasculitis associated with malignancy. The medium sized vessels are mainly involved by polyarteritis nodosa, microscopic polyangiitis, Wegener’s granulomatosis, Churg-Strauss Syndrome and drug induced, neither may be found in both small and medium sized vessels. Superficial lesions of cutaneous vasculitis commonly manifest as infiltrated erythema or palpable purpura whereas lesions affecting deeper regions appear as nodular erythema, livedoracemosa, deep ulcers or digital gangrene. A careful clinical history is taken and close examination is required to rule out any treatable etiology such as systemic disease, drugs, malignancy or infection.

The diagnosis of cutaneous vasculitis cannot be made solely on the basis of clinical presentation. When there is a suspicion of vasculitis a biopsy is required. This is followed by histopathological examination which yields information such as the distribution of vasculitis along with the type of vessel (small/medium) affected. Further investigations include immunofluorescence to detect immune complexes and serological investigations to detect antineutrophil cytoplasmic antibodies (ANCA). Depending on the extent of disease, treatment can range from general measures such as leg elevation to use of anti-B-cell antibody rituximab.

Patients and Methods

This study was a descriptive cross sectional survey carried out in the Department of Histopathology, Army Medical College, Rawalpindi from January 2013 to April 2015. Skin biopsy specimens in formalin accompanied by their requisition forms bearing the clinical details and provisional diagnosis were received mainly from Military Hospital and Combined Military Hospital as well as some of the civil hospitals of the surrounding area. Gross examination of the specimens was carried out and recorded. The biopsies were processed in an automatic tissue processor. The paraffin embedding was done in the embedding
station. The sections were cut (3-5µm) and stained for haematoxylin and eosin. After the slides were prepared, they were observed under the microscope. All cases regardless of age and gender were included in our study.

Results

A total of 37 cases of cutaneous vasculitis were reported during the study period. Only 3 (8.1%) of the patients were less than 20 years of age. The highest frequency of cutaneous vasculitis was among patients between the ages of 21 and 40 years (37.8%), followed by the 41 to 60 years age group with 13 (35.1%) patients. Seven (19%) patients were over 60 years of age. Males were more frequently affected (54%). Ulcer was found to be the most common clinical presentation (29.7%) (Table 1). On histopathological examination, Majority of the cases, 10 (27%) had leukocytoclastic vasculitis (Figure 1). This was followed by non specific vasculitis, 7 (18.9%) cases, cryoglobulinemic vasculitis, 4 (10.8%) cases, panniculitis with vasculitis and lymphocytic vasculitis in 3 (8.1%) patients each. Furthermore, urticarial vasculitis and polyarteritis nodosa were diagnosed in 2 (5.4%) patients (Table 1).

Table 1: Cutaneous Vasculitis- Gross Lesion

<table>
<thead>
<tr>
<th>Gross lesion</th>
<th>No(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ulcer</td>
<td>11 (29.7)</td>
</tr>
<tr>
<td>Purpura</td>
<td>8 (21.6)</td>
</tr>
<tr>
<td>Nodules</td>
<td>6 (16.2)</td>
</tr>
<tr>
<td>Rash</td>
<td>6 (16.2)</td>
</tr>
<tr>
<td>Plaques</td>
<td>5 (13.5)</td>
</tr>
</tbody>
</table>

Discussion

Cutaneous vasculitis is inflammation of the blood vessels leading to vessel wall destruction along with hemorrhage and ischemia. Its incidence ranges from 15.4 to 29.7 cases per million per year. However, as this condition is fairly uncommon in our setup, it sometimes manages to elude dermatologists. In this study, 37 cases of cutaneous vasculitis were reported over a period of more than 2 years. Similar results were shown in a study in Spain.5 Another study in Germany identified 642 primary systemic vasculitidies over 5 years.6 Adults between the age groups of 41 to 60 years (37.8%) were the most commonly affected age group. On the other hand, Blanco et. al. reported Henoch Schonlein Purpura, a type of cutaneous vasculitis to be more common in children with an average age of about 7 years than in adults whereas a study in Europe showed that Wegener’s Granulomatosis, microscopic polyangiitis and Churg Strauss Syndrome are more common in the older, 65 to 70 year old age group.7,8 Males were more commonly affected by cutaneous vasculitis in the present study. Similarly, Trapani et. reported a male to female ratio of 1.8:1. In a study by Lane et al. 61.1% of the patients were male. 10 Ulcers were seen to be the most common clinical presentation in the present study. However, the
disease more commonly presents as palpable purpura or infiltrated erythema, indicating involvement of the dermal small vessel vasculitis. Leukocytoclastic vasculitis (27%) was undoubtedly the most frequent finding. A study in Minnesota showed its incidence to be 4.5 per 100,000 person years. It manifests as palpable purpura and is diagnosed on histopathology based on the findings of neutrophilic infiltrate and nuclear debris around vessels, endothelial swelling and RBC extravasation. Cryoglobulinemic vasculitis is characterized by presence of immunoglobulins that precipitate below 37°C and re-dissolve on re-warming.

In Taiwan, 114 cases of the condition were diagnosed over a period of 10 years and almost the same was observed in the present study. Panniculitis, that is, tender subcutaneous nodules were seen to be associated with vasculitis (8.1%) was seen to be present. A study in Barcelona isolated 91 cases of panniculitis with vasculitis. Lymphocytic vasculitis was also identified in 8.1% of the patients. About the same frequency (9%) was reported in Albany Medical Center. Urticarial vasculitis (5.4%) affects 5-10% patients affected by chronic urticaria. Its rare nature is further supported by the fact that in Spain, only 15 cases were isolated over 10 years which was in accordance with our findings as well. Polyarteritis nodosa (5.4%) is a systemic necrotizing vasculitis. In France, 348 patients were diagnosed with it between 1963 and 2005. Granulomatous inflammation with vasculitis (2.7%) is used to describe cutaneous vasculitic lesions of Wegener’s granulomatosis and Churg Strauss Syndrome. One patient was specifically diagnosed for Churg Strauss Syndrome as well. In Portugal, only 4 patients were admitted with the disease in the Internal Medicine Department of a tertiary care hospital over a 24 year period. Lastly, drug induced vasculitis, an inflammatory vasculopathy was also found in one patient. It can be caused by virtually any drug and accounts for approximately 3% of all vasculitidies.

**Conclusion**

Cutaneous vasculitis is not a common condition worldwide. Despite this, it can be life threatening. It predominantly presents as ulceration and leukocytoclastic vasculitis is the main histological type. Biopsy helps to delineate the exact characterization of vasculitis for further management of the cases.

**References**


