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Case Report

Vascular Purpura: A Cohort Study of 73 Patients

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Abstract

Purpura are red-brown lesions caused by localized haemorrhage into the skin. Vascular causes of purpura may be the result of inflammation, vascular fragility, or microvascular occlusion leading to local ischaemia, and it is most commonly associated with small vessel disease affecting the superficial vessels of the dermis.

This article provides the experience of an internal medicine department about purpura with vascular causes.

Case Presentation: It was a retrospective study including 73 patients with palpable purpura hospitalized in the internal medicine department of the Hedi Chaker University Hospital of Sfax during the period between 1996 and 2020.

Our population consisted of 27 men (36.9%) and 46 women (63%). The average age of the patients was 43.7 years with extremes ranging from 14 to 90 years. Systemic diseases were the most frequent aetiology, found in 26 cases (35.6%), Henoch-Schoenlein purpura (HSP) was the most frequent systemic disease found (16 cases), 7 patients were diagnosed with Sjögren's syndrome (SS). Other autoimmune diseases were found: systemic lupus erythematosus (2 cases), eosinophilic granulomatosis with polyangiitis (1 case).

An infectious cause was found in 12 cases (16.4%): hepatitis (7 cases), post streptococcal (2 cases), post-meningococcal (1 case), cytomegalovirus infection (1 case) and parvovirus B19 (1 case). In 20 cases (27.4%), the aetiology of purpura was unknown.

Skin biopsies were performed in 49 patients, showed leukocytoclastic vasculitis in 32 cases (43.8%).

In terms of therapy, bed rest was recommended for all patients. Colchicine was prescribed in 6 cases. Corticosteroid therapy was prescribed in 20 patients, associated in 3 cases with immunosuppressive treatment. The evolution of PV was favorable in the majority of cases with regression of lesions in 47 cases (64.3%).

Keywords: Purpura; Adult; Systemic Diseases; Palpable Purpura;, Henoch-Schonlein Purpura; Vasculitis with IgA; Sjogren Syndrome

Abbreviations

HSP: Henoch-Schonlein Purpura; PP: Palpable Purpura; SS: Sjogren Syndrome

Introduction

Purpura are red-brown lesions caused by localized haemorrhage into the skin. They can be divided by size into petechiae (1-2 mm in diameter) and ecchymoses (>1 cm).

Purpura can usually be attributed to a problem with either platelets or blood vessels. Platelet disorders may be quantitative (thrombocytopenia) or qualitative as the result of an acquired or inherited defect in platelet function.

Vascular causes of purpura may be the result of inflammation, vascular fragility, or microvascular occlusion leading to local ischaemia, and it is most commonly associated with small vessel

disease affecting the superficial vessels of the dermis, Lesions are often palpable and may have prominent surrounding blanching erythema or preceding urticarial rash [1]. Palpable purpura (PP) is a situation frequently observed in internal medicine, however if the diagnosis is most often easy, the etiological investigation can be difficult, imposing a specific attitude.

This article provides the experience of an internal medicine department

- It was a retrospective study including 73 patients with PP hospitalized in the internal medicine department of the Hedi Chaker University Hospital of Sfax during the period between 1996 and 2020.
- Our population consisted of 27 men (36.9%) and 46 women (63%). The average age of the patients was 43.7 years with extremes ranging from 14 to 90 years.
- The purpuric lesions had the following characteristics: petechial and infiltrated in 51 cases (69.8%), petechial and ecchymotic in 6 cases (8.2%), petechial and necrotic in 6 cases (8.2%), necrotic and ecchymotic in 10 cases (13.6%). It was localized in the lower limbs in 33 cases (45.2%) and diffuse in 30 cases (41%).
- The onset of purpra was acute in 34 cases (46.5%), subacute in 11 cases (15%), chronic in 7 cases (9.5%) and recurrent in 19 cases (28.7%). The etiology of PP was found in 60 cases (82.1%).
- Vasculitis may be secondary to infections (bacteria, parasites, fungi, viruses), inflammatory diseases (systemic lupus erythematosus, Sjogren's syndrome, rheumatoid arthritis, Bechet's disease, and inflammatory bowel disease), drugs (chemicals and foods), and malignancies (lymphoproliferative disorders or solid tumors), or may be idiopathic [2].
- In our study, systemic diseases were the most frequent etiology, found in 26 cases (35.6%). This percentage was 55% and 41% respectively in another series including 101 and 58 patients each [3,4].
- Henoch-Schonlein purpura (HSP) or vasculitis with IgA was the most frequent systemic disease found in our work (16 cases), in fact, it is a vasculitis affecting small vessels (capillaries, venules, or arterioles) with IgA-dominant immune deposits typically involving the skin, gut, glomeruli, and joints. It is most common in children with prevalence as high as 1.5 per 1000 but can also occur at any age. The classic tetrad of HSP includes palpable purpura, joint pain, gastrointestinal complaints, and renal involvement. A survey from 2016 analysed clinical symptoms in 260 adults with HSP, at diagnosis, 100% of patients presented with purpura [5].

Sjögren syndrome (SS) is a chronic autoimmune inflammatory disease that involves primarily the exocrine glands resulting in their functional impairment. The most common clinical presentation of primary SS associated cutaneous vasculitis is palpable and non-palpable purpura occurring on the lower extremities [6], in our work, Sjögren syndrome was found in 7 cases.

Other autoimmune diseases were also diagnosed: systemic lupus erythematosus (2 cases), eosinophilic granulomatosis with polyangiitis (1 case).

An infectious cause was found in 12 cases (16.4%): hepatitis (7 cases), post streptococcal (2 cases), post-meningococcal (1 case), cytomegalovirus infection (1 case) and parvovirus B19 (1 case). However, in 20 cases (27.4%), the purpura was of unknown aetiology.

Skin biopsies are the gold standard for diagnosing any cutaneous vasculitis [5]. It was performed in 49 patients, showed leukocytoclastic vasculitis in 32 cases (43.8%). Leukocytoclastic vasculitis is the usual histological finding in cases of small vessel vasculitis with a variety of causes, and the disease is only classified as cutaneous leukocytoclastic angiitis if no systemic involvement is found [7].

In terms of therapy, bed rest was recommended for all patients. Colchicine was prescribed in 6 cases. Corticosteroid therapy was prescribed in 20 patients, associated in 3 cases with immunosuppressive treatment. The evolution of PV was favourable in the majority of cases with regression of lesions in 47 cases (64.3%).

Conclusion

Purpura can be the revealing symptom of many diseases, whose lack of recognition and late diagnosis can be life-threatening for patients. The frequency of different vascular causes of purpura is still poorly defined in the literature. However systemic diseases and infectious causes should be eliminated as the first line.

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