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Case Report

Co-occurrence of Behçet disease with Ig A vasculitis revealed by ophtalmic examination: A case report

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ABSTRACT

Introduction: Renal involvment of Behçet disease is not usual and Ig A vasculitis complicated by Behçet disease is extremely rare.
Case presentation: We report a case of Henoch Scholein purpura and nephritis associated with Behçet disease revealed by ophthalmic examination.
Clinical discussion: Recurrent oral ulcer is not a manifestation of Ig A vasculitis which doesn't affect large or middle sized arteries. If the patient had'nt been referred to our department for ophthalmic examination, we may have failed to make the diagnosis and treat coexisting Behçet disease.
Conclusion: The link of HSP and BD has yet to be fully understood, and more researchisdefinitelyneeded.

1. Introduction

Behçet disease (BD) is a vasculitic disorder caused by chronic inflammation that can affect vessels of all sizes. It is characterized by a recurrentoral aphthousulcers, genitalulcers, and uveitis [1].

Othersystemic manifestations can be found such as cutaneous or gastro intestinal symptoms, neurological disease, and arthritis.

The renal involvement, however, is not very common in this affection. Here we report the association of Henoch-Schonlein purpura (HSP) with nephritis in BD.

This case report has been reported in line with the SCARE Criteria [2] .

2. Presentation of case

A 56-year-old male, with no medical history, was initially referred to the dermatology department for purpuric "gloves and socks" syndrome (Fig. 1) with hematuriadipstick 4+ and Proteinuria of 3,2g/l per 24H. A skin biopsyrevealedaleukocytoclastic vasculitis with IgA-containing immune deposits and the patient was diagnosed as having Henoch-Schonlein Purpura (HSP).

Otherfindings of the clinical examination were: aphthousulcers, arthritis of the knee and elbow, abdominal pain, and a blurred vision. Therefore he was referred to our ophthalmology department for better investigation of ocular manifestations.

The corrected visualacuity in both eyes was 3/10, the cornea was clear, the anterior chamber was deep and quiet, there were vitritis of the right eye (RE) **diagnosed with minimal vitreous haze (posterior pole clearly visible)**, and **Vitreous cells 0.5+** of the lefteye (LE), **according to the National Institutes of Health (NIH) classification** [3]. The fundus examination of the right eye showed signs of Retinal vasculitis and hemorrhages in the superotemporal area (Fig. 2). Fluoresce in angiography revealed retinal capillary hypoperfusion in that area with capillary leakage related to retinal occlusive vasculitis (Fig. 3). The exam of the LE was normal.

A macular OCT was performed and the results had not shown any further complications.

A skin pathergic test was then performed and it was positive.

Because of the association of aphtous ulcer, retinal occlusive vasculitis and positive skin pathergic test, **the diagnosis of Behçet disease was made, based on The International criteria for Behçet disease** [4].

The patient also underwent a renal biopsy showing glomerulonephritis with granular deposits of Ig A and C3 in the mesangium in the immunofluorescent studies. He also benefited from a study of the human leukocyte antigen (HLA) types.

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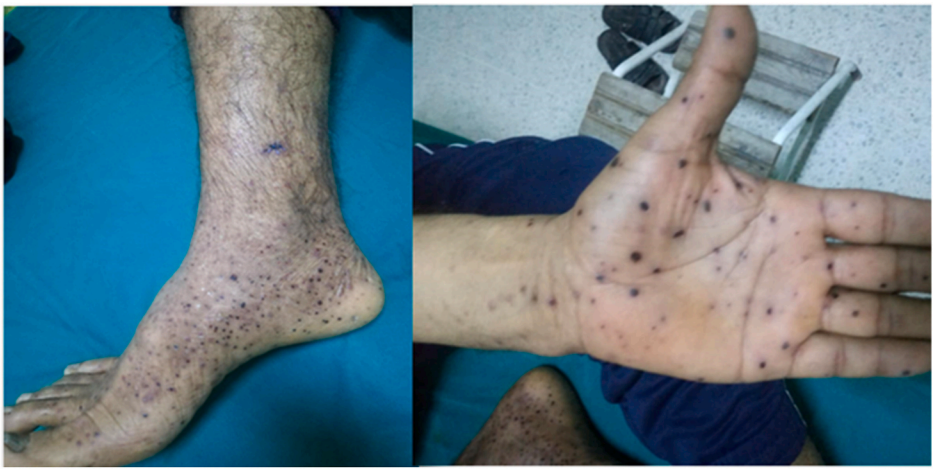

Fig. 1. Photography of the foot and hand of our patient showing purpuric "gloves and socks" syndrome.

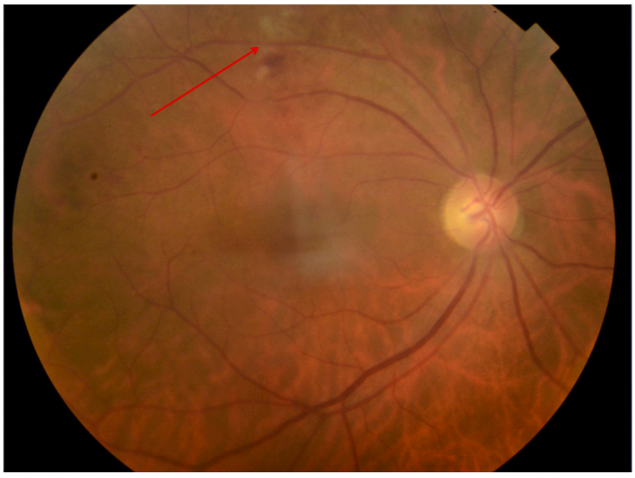

Fig. 2. Fundus photography of the right eye of our patient showing retinal hemorrhage and retinal ischemic oedema in the supero-temporal area (red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Based on the results of skin biopsy and renal biopsy with the immunofluorescent study, the diagnosis of Ig A vasculatis was confirmed, in association with Behçet disease according to the internatioal criteria for Behçet disease.

Prednisolone in combination with colchicine had been prescribed by dermatologist. Because of the ocular findings, azathioprime 200 mg per day was added.

After treatment, the joint manifestations and abdominal pain improved within 7 days, the purpura disappeared in 1 month and the proteinuria in 6 weeks.

Ophtalmic examination after 4 weeks of treatment showed visual acuity of 7/10, improvement of the vitritis and vasculitis on fundus examination.

This case report has been reported in line with the SCARE Criteria [2].

3. Discussion

HSP was first described by Dr. William Heberden in 1801. It is an auto immune systemic vasculit is that affects small bloodvessels that is identified mainly based on clinical characteristics of cutaneous palpable purpura, arthralgia/arthritis, bowelangina, and hematuria/proteinuria combined with pathohistological findings of leukocytoclasticvasculitis (LCV) and IgA-immune deposits in vessel walls [5].It is much more frequent in children than adults [6]and it appears to be more severe in the latter situation. Our patient show cased all of the signs described above hence the diagnose of HSP with renal involvement.

Renal involvment in Behçet disease is not common. The first occurrence of proteinuria and hematuria was described in 1963 [7]. Thereafter, Behçet disease with renal involvment was described in many reports ([8,9]).

The association of Ig A vasculitis and BD is extremely rare. It has been described in a limited number of articles.

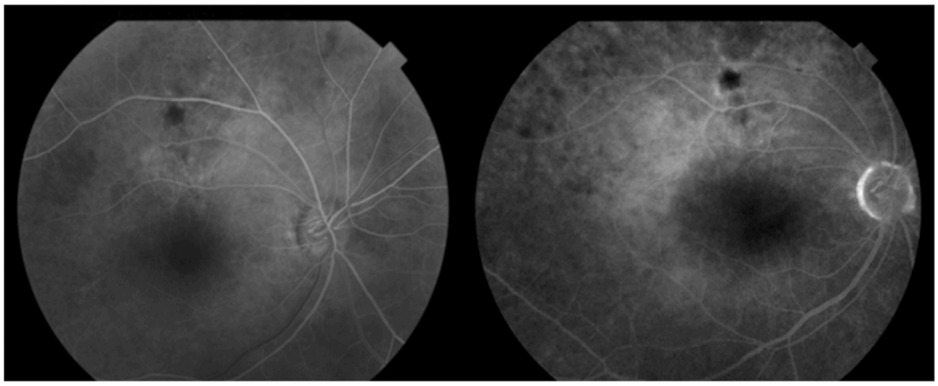

Fig. 3. Fluorescein angiography of the right eye of our patient showing hypoflorescence caused by retinal hemorrhage, capillary non perfusion and Capillary leakage in the supero-temporal area.

Takeshi Furukawa and his team reported a similar case similar to ours of a 38 yearsoldwomanthatwasdiagnosed as having BD and developed arthralgia, abdominal pain, purpuriceruptions, and leukocytoclasticvasculitis with IgA deposits [10].

Other cases of Behçet disease complicating Ig A vasculitis, have been also reported [11–13].

Higashihara et al. a reported a case of Behçet disease revealed by pathergy reaction induced by renal biopsy, which was masked by the presence of IgA vasculitis, and elucidated the etiology of the unexplainable symptoms [14].

Levinsky et al. [15] found high levels of IgA-containing immune complexes in serum of patients with Behcet' disease that could explain the coexistence of these two conditions as they share the same background of auto immunepathogenesis.

Interestingly, previous studies have shown a correlation between certain human leukocyte antigens (HLA) and renal involvment in Behçet disease [16]. Most of patients with nephropathy have a negative finding for HLA B 51. However, HLA-A2, A11, and B35 are associated with Ig A vasculitis [17].

The HLA types of blood lymphocytes in our patient were A3, B35 and DR 4 which confirm the findings of the last cited studies.

In addition to prednisolone and colchicine prescribed by dermatologist in our case, treatment included Azathioprime because of the retinal vasculitis confirmed by ophthalmological examination. Uveitis associated with Behçet disease is an absolute indication for immunosuppressive therapy, which has contributed to the improvement of the visual prognosis [18].

4. Conclusion

Despite the progress that has been done in recent years to determine the exact pathogenesis, the exact cause of HSP and BD and the link between them are yet to be fully understood, and more research is definitely needed.

Please state any conflicts of interest

No conflicts of interests

Please state any sources of funding for your research

None

Sources of finding

None.

Consent

Authors obtained consent of the patient.

Ethical approval

This is not a research study.

Author contribution

Mhamdi Tasnim and Ben Abdesslem Nadia have written the case report.

Registration of research studies

This is not a research study.

Name of the registry:

Unique Identifying number or registration ID:

Hyperlink to your specific registration (must be publicly accessible and will be checked):

Guarantor

Dr Ben Abdesslem Nadia: the corresponding author.

Declaration of competing interest

No conflicts of interests.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2021.102446.

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