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

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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 recurrent oral aphthous ulcers, genital ulcers, and uveitis [1].

Other systemic manifestations can be found such as cutaneous or gastrointestinal 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 hematuria dipstick 4+ and Proteinuria of 3.2g/l per 24H. A skin biopsy revealed leukocytoclastic vasculitis with IgA-containing immune deposits and the patient was diagnosed as having Henoch-Schonlein Purpura (HSP).

Other findings of the clinical examination were: aphthous ulcers, 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 visual acuity 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 left eye (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 aphthous 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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Based on the results of skin biopsy and renal biopsy with the immunofluorescent study, the diagnosis of IgA vasculitis was confirmed, in association with Behçet disease according to the international criteria for Behçet disease.

Prednisolone in combination with colchicine had been prescribed by dermatologist. Because of the ocular findings, azathioprine 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.

Ophthalmic 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 autoimmune systemic vasculitis that affects small blood vessels that is identified mainly based on clinical characteristics of cutaneous palpable purpura, arthralgia/arthritis, bowel angina, and hematuria/proteinuria combined with pathohistological findings of leukocytoclastic vasculitis (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 showed all of the signs described above hence the diagnosis of HSP with renal involvement.

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

The association of IgA vasculitis and BD is extremely rare. It has been described in a limited number of articles.
Takeshi Furukawa and his team reported a similar case similar to ours of a 38 yearsoldwoman that was diagnosed as having BD and developed arthralgia, abdominal pain, purpurierecipients, and leukocytoclasticvasculitis with IgA deposits [10].

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

Higashihara et al. 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 immunopathogenesis.

Interestingly, previous studies have shown a correlation between certain human leukocyte antigens (HLA) and renal involvement 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 IgA 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 Azathioprim because of the retinal vasculitis [17]. Interestingly, previous studies have shown a correlation between inflammatory activity in intermediate and posterior uveitis, Ophthalmology 92 (1985) 467–471.

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.jamsu.2021.102446.

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