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Pure ocular mucous membrane pemphigoid in a patient with axial spondyloarthritis (HLA-B27 positive)

Sir, Here we describe to the best of our knowledge the first case of pure ocular mucous membrane pemphigoid (POMMP) as an ocular manifestation of axial spondyloarthrits (SpA). A 58-year-old male veteran was seen in the ophthalmology clinic for progressive vision loss and possible bilateral symblephara. His medical history was significant for hypertension, diabetes mellitus type 2 and dyslipidemia. At the initial visit, the patient’s corrected distant visual acuity was 20/25 –1 in the right eye and 20/40 +1 in the left eye. Slit lamp examination was significant for conjunctival injection and bilateral temporal symblephara was noted on the initial visit. Differential diagnosis included POMMP, Stevens–Johnson syndrome and sequelae of adenoviral conjunctivitis. For the first 3 months the patient was treated with various topical agents, including fluorometholone drops, prednisolone suspension, tacrolium ointment as well as high-dose oral prednisone, without relief. His ocular condition had deteriorated significantly, with the formation of marked symblepharon OU with inferior fornical shortening and a small pannus of conjunctival tissue extending into the inferior aspect of the cornea on the right (Fig. 1A–D). Pathological examination of a biopsy from the conjunctiva was remarkable for subepithelial bullae on haematoxylin and eosin staining (H&E; Fig. 1E). Direct immunofluorescence revealed faint linear IgA basement membrane deposits. The patient was diagnosed with POMMP and sent to the rheumatology clinic for further diagnostic and therapeutic evaluation.

On deeper interrogation in the rheumatology clinic, a history of non-steroidal anti-inflammatory response lumbar spinal pain and unilateral right buttock pain that worsened with rest and improved with activity since his teenage years was elicited. The patient also reported a history compatible with bilateral Achilles tendon enthesisitis, plantar fasciitis and bilateral swelling of his fingers and toes in the past (possible dactylitis). There was no history of a diagnosis of psoriasis or IBD nor did the peripheral joints demonstrate active synovitis on physical examination. Investigations demonstrated a positive HLA-B27 allele and autoantibody assessment was negative for RF, anti-CCP, ANA, anti-ENA panel, anti-dsDNA and perinuclear/ cytoplasmic ANCA. Bilateral grade II sacroiliitis (Fig. 1F) was noted on radiographic examination. A diagnosis of undifferentiated axial SpA (HLA-B27 positive) was rendered based on the Assessment of SpondyloArthritis international Society (ASAS) criteria [1].

The patient was started on MTX, and the dose was titrated up to 25 mg s.c. injections once a week over 3 months, under close clinical and laboratory surveillance. Both the POMMP and the peripheral musculoskeletal symptoms of SpA have responded well to this therapy.

SpA is a group of chronic systemic inflammatory rheumatic diseases including AS, PsA, enteropathic arthritis, reactive arthritis, undifferentiated SpA and juvenile SpA. Although characterized by inflammation and new bone formation in the axial skeleton, SpA can also involve peripheral joints, entheses and other organs such as the eyes, skin, heart and gut [1, 2]. Although ocular involvement in SpA typically takes the form of acute anterior uveitis (AAU), occurring in 25–40% of patients at some point in the disease course [3, 4], rarer manifestations may include anterior scleritis, posterior scleritis and hypotony maculopathy [5].
Fig. 1 Ocular, pathological and radiological manifestation of POMMP in a patient with axial SpA.

(A) Photograph of the right eye (OD) showing a 1.4 mm pannus extending into the inferior aspect of the cornea.  
(B) Superior temporal symblepharon of the left eye (OS).  
(C) Inferior fornical shortening of the right eye (OD).  
(D) Inferior fornical shortening of the left eye (OS).  
(E) Left conjunctival biopsy (frozen tissue, H&E, 10×) showing a small strip of conjunctiva (*) separated by a cleft (△) from a fibrotic (scarred) submucosa (**).  
(F) Radiograph of the pelvis (antero-posterior) showing bilateral grade II (minimal) sacroiliitis (right more than left).
Mucous membrane pemphigoid (MMP) describes a heterogeneous group of chronic inflammatory subepithelial blistering diseases with a combination of oral, ocular, skin, genital, nasopharyngeal, esophageal and laryngeal lesions [6]. MMP limited to the eyes is known as POMMP, which begins as a chronic conjunctivitis with progressive subepithelial fibrosis and may, later in the course of the disease, lead to lasting shortening of the fornix, symblepharon, anklyoblepharon, trichiasis, entropion, obstruction of the lacrimal duct and ultimately to corneal cicatrization and keratinization [6, 7]. If left untreated, OMMP can lead to blindness. Although MMP and OMMP have been described in rheumatic patients with RA and SLE [8, 9], to the best of our knowledge, this is the first report of POMMP in a patient with axial SpA: two well-described diseases of immune dysregulation. The case highlights several points.

(i) It is worth noting that the diagnosis of SpA was delayed by about 30 years from the onset of axial symptoms in this case and was brought to the physician’s attention after the refractoriness of POMMP prompted a rheumatology referral. This is reminiscent of AAU as the only presenting symptom that draws attention to the diagnosis of SpA [3] and other HLA-B27-associated extraocular diseases [5]. Such a significant delay in SpA diagnosis is well recognized worldwide [10].

(ii) There was no involvement of mucous membranes other than the conjunctiva in the case presented.

(iii) The age of onset of POMMP in this case (58 years) is slightly below the average reported age (the seventh decade of life).

Although the concomitant occurrence of the two immune-mediated ailments by chance cannot be completely excluded, clinicians should nonetheless be aware of POMMP as a possible ocular manifestation of SpA.

**Rheumatology key message**

- Clinicians should be aware of POMMP as a possible ocular manifestation of SpA.

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**Comment on: The value of colour Doppler sonography of the knee joint: a useful tool to discriminate inflammatory from non-inflammatory disease?**

SIR, We read with great interest the article by Beitinger et al. [1], in which the authors assess the usefulness of colour Doppler ultrasonography (US) to discriminate inflammatory arthritis from non-inflammatory diseases of the knee. We would like to raise some important points regarding the methodology, as the technique can modify all the results of the study.