IRIS AperTO

UNIVERSITÀ DEGLI STUDI DI TORINO

AperTO - Archivio Istituzionale Open Access dell'Università di Torino

Antiphospholipid syndrome and relapsing polychondritis: an unusual association.

This is the author's manuscript

Original Citation:

Availability:

This version is available http://hdl.handle.net/2318/95150 since

Published version:

DOI:10.1177/0961203311409270

Terms of use:

Open Access

Anyone can freely access the full text of works made available as "Open Access". Works made available under a Creative Commons license can be used according to the terms and conditions of said license. Use of all other works requires consent of the right holder (author or publisher) if not exempted from copyright protection by the applicable law.

(Article begins on next page)

12 October 2024

UNIVERSITÀ DEGLI STUDI DI TORINO

This is an author version of the contribution published on:

Antiphospholipid Syndrome and Relapsing polychondritis: an unusual association,

October 2011 , doi:10.1177/0961203311409270

Savino Sciascia, Mario Bazzan, Simone Baldovino, Antonella Vaccarino, Daniela Rossi, Alessandra Russo, Dario Roccatello, Lupus, October 2011 vol. 20 no. 12, pp 1336-1337

The definitive version is available at:
http://lup.sagepub.com/

Title page

Antiphospholipid Syndrome and Relapsing polychondritis: an unusual association

Savino Sciascia1, Mario Bazzan2, Simone Baldovino1, Antonella Vaccarino2, Daniela Rossi1, Alessandra Russo1, Dario Roccatello1

1 Dipartimento di Malattie Rare, Immunologiche, Ematologiche ed Immunoematologiche, Centro di Ricerche di Immunopatologia e Documentazione su Malattie Rare (CMID) Struttura Complessa a Direzione Universitaria di Immunologia Clinica Ospedale Torino Nord Emergenza San G. Bosco ed Università di Torino, Italy

2 Dipartimento di Malattie Rare, Immunologiche, Ematologiche ed Immunoematologiche, Struttura Semplice Dipartimentale di Ematologia, Ospedale Torino Nord Emergenza San G. Bosco

Address for correspondence:

Savino Sciascia, Centro di Ricerche di Immunopatologia e Documentazione su Malattie Rare (CMID), Struttura Complessa a Direzione Universitaria di Immunologia Clinica, Ospedale Torino Nord Emergenza San G. Bosco ed Università di Torino,

Piazza del Donatore di Sangue 3, 10154, Torino, Italy

Email: savino.sciascia@unito.it

phone 00390112402056

fax 00390112402052

Sir,

relapsing polychondritis (RPC) is an immune-mediated condition associated with inflammation in cartilaginous structures and other tissues throughout the body, particularly the ears, nose, eyes, joints, and respiratory tract. Approximately one-third of RPC cases occur in association with other diseases, such as systemic vasculitis, connective tissue disorders, or myelodysplastic syndromes.

We report the case of a 73-year-old man with antiphospholipid syndrome (APS) associated with relapsing polychondritis.

He came to our attention with a history of ultrasound-proven, recurrent thrombophlebitis (since 2009, two episodes involving popliteal veins, one involving the cephalic vein).
Coagulation screening was performed and lupus anticoagulant (LA) resulted positive (testing was repeated 16 weeks later; testing to confirm lupus anticoagulant positivity was performed 1 week after discontinuing vitamin K antagonists, when the international normalized ratio was less than 1.5. Bridging therapy after vitamin K antagonist discontinuation to low-molecular-weight heparin was carried out). LA measurement included three tests: dilute Russell's viper venom time (dRVVT, Hemosil, LA-screen/confirm, Instrumentation Laboratory, Lexington,USA), partial thromboplastin time-LA (PTT-LA, Diagnostica Stago, Asnieres,France), silica clotting time (SCT, HemosIL™, DiaPharma Group, Inc. Ohio, USA). If PTT-LA was prolonged, the hexagonal phospholipid neutralization test was performed as confirmation (STACLOT-LA, Diagnostica STAGO, Asnières,France).

Anticardiolipin antibodies (ELISA kit, Phadia, EliA Cardiolipin IgG/IgM) and anti ß 2-glycoprotein I antibody (ELISA kit, Phadia, EliA ß2 Gliocoprotein I IgG/IgM were searched for twice 16 weeks apart and resulted negative.

Thus, a diagnosis of Antiphospholipid Syndrome was made according to Myakis et al. (1) , and long term anticoagulation therapy was started.

During follow up the patient presented with persistent fever (up to 38.5 °C), bilateral external ear inflammation, nasal involvement with crusting and cartilage flogosis and diffuse polyarthralgia. During the same time period, he was taken to the Emergency Room with ocular inflammation and he was discharged with a diagnosis of scleritis/episcleritis that was successfully treated with steroids. His laboratory findings revealed elevated erythrocyte sedimentation rate (up to 100 mm/h) and high C-reactive protein levels (12.1 mg/dl). ANA, ENA and ANCA were negative. Blood tests revealed normocytic and normochromic anaemia (Hb 10.2 g/dl), which is consistent with anaemia occurring in chronic diseases. Considering the patient's age and the ongoing anticoagulation therapy, bone marrow biopsy to rule out myelodysplastic syndromes was postponed and haematologic follow up was planned.

Due to the presence of bilateral auricular chondritis, non-erosive, sero-negative inflammatory polyarthritis, nasal chondritis and ocular inflammation (scleritis/episcleritis), a diagnosis of RPC was made on the basis of McAdam's criteria (2). Positive histology of the involved cartilage was not required for inclusion, since the clinical setting itself was diagnostic for RPC (3). Therapy with 10 mg/daily prednisone was started, leading to immediate improvement of the auricular lesions and a rapid decrease of the sign/symptoms of inflammation. During 15 months of follow-up we observed no recurrences with a maintenance dose of 5 mg/daily prednisone.

Some studies have speculated on the possible association between antiphospholipid antibodies and RPC. However, no convincing evidence regarding the possible association between RPC and APS has been found (3). When APS occurs in RPC it seems to be more closely related to associated conditions like Systemic Lupus Erythematosus. (4)

In our case, no signs or symptoms (except arthralgia) were suggestive of Lupus-like conditions, and RPC occurred in a pre-existing condition of Primary APS.

To date, the co-existence of APS and RPC has not been convincingly reported in English language scientific literature. The present case supports the possible association between these two rare conditions.

References

1. Miyakis S, Lockshin MD, Atsumi T, et al. International consensus statement on an update of the classification criteria for definite antiphospholipid syndrome (APS). J Thromb Haemost. 2006; 4: 295-306.

2. McAdam LP, O'Hanlan MA, Bluestone R, Pearson CM. Relapsing polychondritis: prospective study of 23 patients and a review of the literature. Medicine (Balt) 1976; 55: 193-215.

3. Zeuner M, Straub RH, Rauh G, Albert ED, Schölmerich J, Lang B. Relapsing polychondritis: clinical and immunogenetic analysis of 62 patients. J Rheumatol. 1997; 24: 96-101.

4. Zeuner M, Straub RH, Schlosser U, et al. Anti-phospholipid-antibodies in patients with relapsing polychondritis. Lupus. 1998;7(1):12-4.

This research received no specific grants from any funding agencies in the public, commercial, or not-for-profit sectors.