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Antiphospholipid Syndrome and Relapsing polychondritis: an unusual association

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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, LAscreen/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 β 2-glycoprotein I antibody (ELISA kit, Phadia, EliA β2 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


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