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Recurrent superficial vein thrombosis revealing relapsing polychondritis associated with myelodysplastic syndrome

An 83-year-old man presenting symptoms of fatigue, weight loss (6 kg in 6 months) and low-grade fever was referred to our department. The diagnosis of superficial venous thrombosis of the right forearm (Fig. 1A) was confirmed by Doppler ultrasound. Blood analysis showed inflammatory syndrome (CRP: 5.0 mg/dl) and normocytic anaemia (9.8 g/dl). Biological tests for antiphospholipid syndrome were negative. Fluorodeoxyglucose positron emission tomography and temporal artery biopsy were normal. Bone marrow aspiration revealed myelodysplastic syndrome (MDS). Six months later, the patient presented a right auricular chondritis (Fig. 1B). The diagnosis of MDS associated with relapsing polychondritis (RP) was confirmed. RP is an inflammatory condition characterized by recurrent destructive cartilaginous inflammations. Diagnosis can be challenging because chondritis might be absent at disease onset. Cutaneous manifestations of RP include recurrent superficial venous thrombosis due to skin vessel vasculitis [1], which is more frequent when RP is associated with MDS [2]. Up to 30% of RP cases are related to MDS. This patient's phenotype appears as the most severe one, with an increased mortality rate and a lower rate of response to biologics [2]. Treatment of the MDS appears as the most promising therapeutic option in this population.

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- 2 Dion J, Costedoat-Chalumeau N, Sène D *et al.* Relapsing polychondritis can be characterized by three different clinical phenotypes: analysis of a recent series of 142 patients. *Arthritis Rheumatol* 2016;68:2992–3001.

Fig. 1 Cutaneous manifestations of relapsing polychondritis.



(A) Superficial vein thrombosis: painful indurated palpable cord along a superficial vein and surrounding inflammatory oedema on the right forearm. **(B)** Right auricular chondritis (inflammation of the right auricular cartilage sparing the earlobe).