An Unexpected Etiology of Rhizomelic Pseudo-Polyartharthritis (Polymyalgia Rheumatica) in the Elderly

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Abstract

Introduction: Rhizomelic pseudo-polyartharthritis or polymyalgia rheumatica (PMR) is a common chronic inflammatory rheumatism in the elderly. Its etiologies are by far dominated by giant cell arteritis and cancers. Other exceptional etiologies can be seen, and present a real diagnostic challenge for clinicians.

We report an unusual case of PMR caused by primary hypothyroidism.

Case Report: A 68-year-old patient, with no notable pathological history, was hospitalized for exploration of a PMR non-responding to corticosteroids prescribed by the family doctor. The somatic examination was without abnormalities. Extensive investigations have eliminated possible associated giant cell arteritis, connective tissue diseases, necrotizing vasculitis, infections, osteomalacia, cancer, and haematological malignancies. Thyroid assessment revealed primary hypothyroidism with fT4 at 4.24 Pmol/l and TSH at 54.16 μmol/ml. Under thyroxine, the evolution was favorable with disappearance of any rhizomelic symptomatology.

Conclusion: This observation reports an exceptional and often overlooked etiology of PMR, which is easy to find and treat. Its knowledge by clinicians makes it possible, in certain cases, to avoid several invasive explorations as well as an unjustified corticosteroid treatment that is not devoid of side effects, sometimes severe in frail elderly subjects.

Keywords: Rhizomelic pseudo-polyartharthritis, polymyalgia rheumatica, hypothyroidism, elderly.

Introduction

Rhizomelic pseudo-polyartharthritis or even better polymyalgia rheumatica (PMR) represents the most common chronic inflammatory rheumatism in the elderly, with a prevalence estimated at 1.7% for men and 2.7% for women [1]. It can be isolated called “pure PMR”, associated with giant cell arteritis, or represents the mode of onset of several other highly polymorphic conditions such as: cancers, hematological malignancies, rheumatoid arthritis, inflammatory myositis, necrotizing vasculitis, osteomalacia, joint chondocalcinosis, severe infections, spondylarthopathies, RS3PE syndrome, degenerative neuropathies, and some endocrine disorders [2-4].

As a result, PMR remains, to this day, an affection described as “enigmatic” [5], and often represents a real diagnostic and therapeutic challenge for clinicians [1,5].

Hypothyroidism remains an exceptional cause of PMR [6]. It is often overlooked and neglected, especially in the elderly when a PMR may be the first manifestation of hypothyroidism, even in the absence of specific clinical signs of this thyroid dysfunction [3].

We report an original observation of PMR revealing primary hypothyroidism in the elderly.

Case Report

A 68-year-old female, with no notable pathological history, was hospitalized for exploration of a PMR that had been evolving for two months and not improved by ambulatory treatment.

She was initially seen by her family doctor for joint pain of both shoulders of inflammatory type. Prescribed non-steroidal anti-inflammatory drugs and analgesics had no effect. The initial biological assessment
revealed an erythrocyte sedimentation rate at 80 mm/H1, thus the diagnosis of PMR was suspected and oral corticosteroid therapy at a dose of 17.5 mg/day was initiated. No improvement was noted after three weeks of treatment and the patient was referred to us for diagnostic investigation and adapted treatment.

The anamnesis reveals that the complaints are inflammatory pain with obvious stiffness and limitation of active movements of both shoulders, evolving for two months, and associated with asthenia, and fever. No cephalic signs or recent visual disturbances are reported.

Apart from the provoked pain and the active limitation of the shoulders, the somatic examination was without abnormalities; in particular, there were no cutaneous signs of vasculitis or connective tissue disease, and the superficial temporal arteries were well perceived on both sides with no inflammatory signs.

Biology showed normocytic anemia at 10.5g/dl and erythrocyte sedimentation rate at 70mm/H1. The other basic bioassays were without abnormalities (C-reactive protein, platelets, leukocytes, serum protein electrophoresis, creatinine kinase, lactate dehydrogenase, liver enzymes, calcium, phosphorus, fasting blood glucose, alkaline phosphatase, and urine analysis).

Extensive investigations for an underlying etiology of this PMR were negative, eliminating in particular giant cell arteritis, cancer, hematological malignancies, necrotizing vasculitis, infection, systemic lupus erythematosus, rheumatoid arthritis, polymyositis, and osteomalacia (anti-nuclear antibodies, anti-native DNA antibodies, anti-soluble nuclear antigen antibodies, ANCA, latex and Waaler-rose reactions, ophthalmologic examination with fundus, tumor markers, chest X-ray (fig1) abdomino-pelvic ultrasound, Thoraco-abdomino-pelvic CT (fig 2 and 3), cardiac echocardiography, gastroduodenalfibroscopy, colonoscopy, immnuoelectrophoresis of serum and urinary proteins, and biopsy of the temporal artery). The patient was treated with oral corticosteroids at the dose of 20 mg/day but without any improvement.

![Normal chest X-ray](image)
The subsequent thyroid assessment indicated a fairly deep primary hypothyroidism with fT4 at 4.24 Pmol/l and TSH at 54.16 μmol/ml (Nl 0.5-5 μmol/ml). Corticosteroid therapy was discontinued, and the patient was put on thyroxine in progressive doses up to 125 μg/d normalizing her TSH to 3.2 μmol/ml. The subsequent evolution was favorable, with disappearance of any rhizomelic symptomatology and normalization of its hemogram.

**Discussion**

The diagnosis of PMR is based primarily on clinical [5,7] because of the absence of specific biomarkers [1,5,7]. It is classically presented as an acute inflammatory pain of proximal muscles and large joints, predominating in scapular and pelvicgirdle, and associated with morning stiffness, general malaise, and fever [1-3]. In 50% of cases, it can be associated with peripheral manifestations such as asymmetric and non-erosive seronegative polyarthritis, synovitis, flexor tenosynovitis, carpal tunnel syndrome, and distal edema [8].

More recently, radiological investigations (joint ultrasonography and MRI) were recognized as being very helpful in the diagnosis of this condition, particularly by highlighting bilateral sub-acromial bursitis and/or deltoid bursitis, which are characterized by their high sensitivity and specificity: sensitivity=92.9%, specificity=99.1%, and PPV=98.1% [7.9].

Thus, several diagnostic criteria have been validated to facilitate the positive diagnosis of this condition [10]; the most used are those developed jointly by the EULAR and the ACR in 2012, which introduce the above-mentioned specific radiological signs [11].
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Given the disparity of possible etiologies, an underlying cause should always be sought before retaining the diagnosis of isolated or "pure" PMR because of the very different therapeutic and prognostic implications; indeed in the series of 200 cases of PMR of Ceccato F et al, 8% (16/200) of patients expressed an underlying cause during the prolonged follow-up [2]. Similarly, in the Gonzalez-Gay MA et al, series of 208 patients with the initial diagnosis of apparently isolated PMR, 11% (23/208) had a final diagnosis adjusted to another underlying pathology [12].

Hypothyroidism remains an exceptional and often neglected cause of PMR [3,6]: indeed, only one patient among the 200 with PMR in the series of Cerrato F et al, had a causal hypothyroidism (frequency = 0.5%) [2], a single patient in the series of 13 PMR of Mastaglia FL et al [13], and also a single case in the series of 41 patients with PMR of Paice E et al [14].

Although the exact pathogenic mechanism of PMR is not yet well understood [15], it seems that the association between PMR and dysthyroidism is far from being a mere coincidence. Indeed, there was a significantly higher frequency of hypothyroidism during PMR, compared to the general population: 4.9-11.1% [16, 17]. For some authors this could be explained by the frequency of comorbidities in elderly subjects and the particularly high frequencies of these two affections (PMR and hypothyroidism) in the population over 60 years [18,19].

However, the particular frequency of Hashimoto’s thyroiditis at the origin of the hypothyroidism associated with PMR, the simultaneous occurrence of PMR and hypothyroidism in the same patient [20] as well as the possibility of transient hypothyroidism during the initial phase of PMR [21] suggests a particular causal link [16,17,20,21]. Both conditions share a complex and multifactorial pathogenesis involving age-related immunological alterations, both innate and adaptive immunity activation, a genetically predisposed terrain, and possible triggering infection [15].

This causal link is comforted by the study by Singh U et al, where it has been shown that autoimmune thyroid diseases can produce high levels of rheumatoid factor and anti-CCP2 autoantibodies, and lead to joint manifestations sometimes difficult to diagnose [22].

It is thus recommended to evoke and screen for hypothyroidism in front of any PMR, and particularly if there is no or bad response to initial corticosteroid therapy [2,14].

**CONCLUSION**

PMR is the most common inflammatory rheumatism of the elderly. Multiple etiologies and the often polymorphic clinic, as well as the absence of specific biomarkers, make his diagnosis a real challenge for the clinician, especially on primary care institutions.

Hypothyroidism represents an exceptional and often neglected cause of PMR which deserves to be known, because of its therapeutic and prognostic implications different from that of an isolated PMR, and sometimes serious.

It is thus recommended to evoke and screen hypothyroidism in front of any PMR, especially if there is no or bad response to the initial corticosteroid treatment.

**ABBREVIATIONS**

- Anti-CCP: anti-cyclic citrullinated peptide antibodies
- ACR: American College of Rheumatology
- ANCA: Antineutrophil cytoplasmic antibodies
- EULAR: European League Against Rheumatism
- fT4: free tetraiodothyronine
- MRI: Magnetic resonance imaging
- PPV: Positive predictive value
- RS3PE: Remitting seronegative symmetrical synovitis with pitting edema
- TSH: Thyroid Stimulating Hormone

**REFERENCES**


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