Review paper

Reumatologia 2025; 63, 2: 182–190
DOI: https://doi.org/10.5114/reum/200191

When can calcium pyrophosphate deposition disease be considered a polymyalgia rheumatica mimicking disease?

Ciro Manzo[1], Paolo Falsetti[2], Alberto Castagna[3], Marco Isetta[4], Edoardo Conticini[2]

[1]Rheumatologic Outpatient Clinic, Internal and Geriatric Medicine Department, Azienda Sanitaria Locale Napoli 3 sud, Sant'Agnello, Naples, Italy
[2]Rheumatology Unit, Department of Medical Sciences, Surgery and Neurosciences, Azienda Ospedaliero-Universitaria Senese, Università degli studi di Siena, Italy
[3]Department of Primary Care, Health District of Soverato, Azienda Sanitaria Provinciale Catanzaro, Italy
[4]Central and North West London NHS Trust, England

Abstract

Introduction: Similarly to polymyalgia rheumatica (PMR), calcium pyrophosphate deposition (CPPD) disease is common among older people. Calcium pyrophosphate deposition can present in several forms, including proximal manifestations associated with raised inflammatory markers. Consequently, CPPD disease may be diagnosed as PMR. Recently, a European Alliance of Associations for Rheumatology and American College of Rheumatology (EULAR/ACR) collaborative initiative proposed new classification criteria for symptomatic CPPD disease. This review paper aimed to discuss when CPPD disease could be considered a PMR-mimicking disease in the light of these criteria.

Material and methods: We performed a non-systematic literature search on PubMed, regardless of the language. Abstracts submitted at conferences or from non-peer-reviewed sources were not included.

Results: The prevalence of CPPD among patients categorized as having PMR supported the inclusion of CPPD among the PMR-like diseases. However, CPPD disease was not diagnosed among the 169 subjects in the non-PMR comparison group in the 2012 EULAR/ACR classification proposal for PMR. According to the 2023 EULAR/ACR study design for symptomatic CPPD, within the 148 definite mimickers forming the derivation cohort, 6 were affected by PMR; only one was affected by PMR within the 162 definite mimickers forming the validation cohort. Finally, in all the studies on this topic, no patient with PMR and CPPD was reported to have a late diagnosis of giant cell arteritis, at least within the term of follow-up of each study.

Conclusions: The relationship between PMR and CPPD should be reviewed in light of the 2023 EULAR/ACR classification criteria for symptomatic CPPD disease. Applying these 2023 criteria, we were able to identify three possible scenarios in patients categorized as having PMR according to the 2012 EULAR/ACR criteria: 1) polymyalgic manifestations in patients with already diagnosed CPPD disease (PMR/CPPD or pseudo-PMR CPPD pattern); 2) polymyalgic manifestations categorized as PMR in patients with concurrent diagnosed CPPD disease (symptomatic CPPD with overlapping PMR); 3) polymyalgic manifestations categorized as PMR in patients with undiagnosed chronic CPPD disease (PMR with concurrent undiagnosed CPPD). Further studies are additionally required to confirm the possibility that the PMR/CPPD subset may be a non-vasculitic pattern of disease.

Key words: polymyalgia rheumatica, classification criteria, calcium pyrophosphate deposition disease, crowned dens syndrome.

Introduction

Polymyalgia rheumatica (PMR) is one of the most common inflammatory rheumatic diseases among older people, especially in the 70 to 80 age group [1–3]. Its diagnosis is based on recognition of a clinical syndrome: typically, PMR patients complain of a sudden-onset bilateral pain in the shoulder and pelvic girdles limiting all

Address for correspondence
Ciro Manzo, Rheumatologic Outpatient Clinic, Internal and Geriatric Medicine Department, Azienda Sanitaria Locale Napoli 3 sud, vale dei Pini 1, 80065, Sant'Agnello, Naples, Italy, e-mail: manzoreumatologo@libero.it
Submitted: 23.09.2025; **Accepted:** 16.01.2025

self-care activities of daily living involving these parts of the body, associated with morning stiffness lasting more than 45 min. Neck ache may also be present [4, 5]. At present, no specific laboratory tests are available. High C-reactive protein (CRP) concentrations and/or erythrocyte sedimentation rate (ESR) are required criteria in the 2012 classification criteria proposed by a European Alliance of Associations for Rheumatology and American College of Rheumatology (EULAR/ACR) collaborative initiative [6].

As similar manifestations may be present in other conditions or diseases, differentiating between PMR and what looks like PMR ("PMR mimickers or PMR-like disease") may not be a straightforward challenge in everyday clinical practice [7–9].

Calcium pyrophosphate deposition (CPPD) disease is a clinically heterogeneous condition that can present in several forms, including crowned dens syndrome (CDS) and proximal manifestations associated with raised inflammatory markers [10, 11]. Similarly to PMR, CPPD disease is common among older people, its prevalence being estimated at > 4% in the 70 to 80 age group, and very rare below 50 years of age [12]. Consequently, it is possible that PMR and CPPD may be associated by chance in the same patient.

Recently, a EULAR/ACR collaborative initiative proposed classification criteria for symptomatic CPPD disease.

The objective of this article was to discuss when CPPD disease could be considered a PMR-mimicking disease in the light of these criteria.

Material and methods

We performed a non-systematic literature search on PubMed with the following search terms: "polymyalgia rheumatica" AND "calcium pyrophosphate deposition disease" OR "chondrocalcinosis" OR "crown dens syndrome" OR "ultrasonography", both MESH headings and free text (in each language in which they were written). Each paper's reference list was scanned for additional publications meeting this study's aim. When papers reported data partially presented in previous articles, we referred to the most recent published data. Abstracts submitted at conferences or from non-peer-reviewed sources were not included.

Results

In Table I, we list the main studies and reports that we found in published literature. In some studies, the percentages of CPPD among patients categorized as having PMR supported the inclusion of CPPD among the PMR-like diseases, excluding the possibility that it was mere coincidence. On the other hand, it is worth highlighting that CPPD disease was not diagnosed within 169 subjects in the non-PMR comparison group in the above-mentioned 2012 EULAR/ACR study [6].

Discussion

Polymyalgia rheumatica and CPPD are two diseases of the elderly population, especially in the 70 to 80 age group. Consequently, it is possible that PMR and CPPD may be associated by chance in the same patient. Their pathogenesis is significantly different. Specifically, CPPD belongs to the group of crystal-related arthropathies, and its clinical manifestations are a consequence of activation of the NLRP3 inflammasome in response to the pathological presence of CPP crystals inside joints [10]. On the other hand, the pathogenesis of PMR is not at all linear, and some working hypotheses are awaiting confirmation [24, 25]. The evidence that PMR and CPPD occur almost exclusively in individuals aged over 50 may indicate that age-related immune modifications in genetically predisposed subjects contribute to development of these diseases [26, 27]. Proximal involvement is reported in patients with chronic pyrophosphate arthropathy.

The possibility that CPPD disease may be a PMR-mimicking disease was first discussed by Dieppe et al. [13] in a 1982 clinical and radiological study of 105 patients diagnosed with CPPD. Among 92 consecutive CPPD patients with chronic joint disease, they found 8 patients who had been treated with glucocorticosteroids (GCs) for presumed PMR. As chronic GC therapy may predispose to crystal deposition, the authors were unable to distinguish whether pyrophosphate arthropathy had occurred with polymyalgic symptoms (so-called "CPPD/PMR") or if it was favoured by GCs.

In a 2005 prospective study, Pego-Reigosa et al. [15] involved 36 patients with CPPD and polymyalgic manifestations (5 met McCarthy's revised criteria for definite CPPD, and 31 for probable CPPD), and pointed out that presence of tibiofemoral osteoarthritis, tendinous calcifications, and ankle arthritis should have raised suspicion for CPPD in patients with polymyalgic manifestations. For these patients, they suggested a "pseudo-PMR/CPPD" pattern, distinct from the other CPPD disease patterns previously described in the literature. According to their suggestion, CPPD must be included among the rheumatic diseases, with which PMR can be confused.

Other studies agree to propose a PMR/CPPD subset with some different characteristics compared with pure PMR: older females, lower levels of acute inflammatory markers at onset, higher frequency of peripheral arthritis, and lower exudation on bursal sites of shoulders. A possible overlap between PMR and CPPD disease was

Table I. Published studies on PMR and CPPD

First author	Type	Number of patients	PMR and CPPD diagnosis	Length of follow-up	Imaging	Presence of definition of subset/ subgroup/cluster	Significant characteristics of subset/subgroup/cluster	Suggested PMR-like condition
Dieppe et al. [13]	Prospective monocentric	8 PMR/CPPD over 105 CPPD	n.r.	5 years of GC therapy	Rx	CPPD with concurrent PMR	CPPD/PMR: advanced age	Coexistence of PMR and CPPD for chance association or long-term GC therapy
Aouba et al. [14]	Case series	5 CDS PMR-like	Clinical	Max. 14 months	Rx and CT	CDS-CPPD with PMR-like presentation	Older, CC in Rx and CT, CDS in atlanto-axial CT. Responsive to NSAIDs and/or colchicine	Yes, different diagnosis and therapy between CPPD and PMR
Pego-Reigosa et al. [15]	Prospective monocentric	118 PMR – 36/118 (31%) CPPD/ PMR	PMR Chuang and Healey CPPD Mc Carty	1 year	Radiologic imaging and SFA	CPPD with concurrent PMR	CPPD/PMR: 31%, older, more frequent peripheral arthritis, more advanced knee OA, more frequent tendinous calcifications and ankle and wrist arthritis Shorter GC course and disease duration (not signif.)	Yes, shorter GC course and disease duration (not signif.)
Salaffi et al. [16]	Case series	2 PMR-like over 25 CPPD with CDS	Mc Carty for CPPD		T	CDS-CPPD with PMR-like presentation	Older, CC in Rx and CT, CDS in atlanto-axial CT	Yes, different diagnosis between CPPD and PMR
Yanai et al. [17]	Case series	1 CPPD over 10 PMR	SFA		SFA Rx	CPPD with PMR-like presentation	Older, higher CRP, prompt response to NSAIDs	Yes, different diagnosis and therapy between CPPD and PMR
Siau et al. [18]	Case report	1 PMR-like CPPD	Clinical	n.r.	CT	CDS-CPPD with PMR-like presentation	Higher CRP, prompt response to NSAIDs and GC	Yes, different diagnosis between CPPD and PMR
Ceccato et al. [9]	Retrospective multicentric	200 PMR – 16 (8%) had other diagnosis in follow-ups – 2 had CDS	Chuang criteria for PMR	1 year	Radiologic imaging	CPPD with concurrent PMR	PMR/CPPD: 1%, calcifications at C1–C2, typical chondrocalcinosis, good response to NSAID	Yes, other diagnosis with different therapy and better outcome
Falsetti et al. [19]	Prospective monocentric	61 PMR at onset – 9/61 (15%) PMR/CPPD	Bird criteria	1 year	MSK US	PMR with US diagnosis of other conditions	PMR/CPPD: 15%, more frequent females, higher frequency of knee menisci calcifications and tendinous calcaneal calcifications. Lower ESR, CRP, PLT vs. others (not signif.)	Yes, suggestion for lower dosage of GC and different course

Table I. Cont.

First author	Type	Number of patients	PMR and CPPD diagnosis	Length of follow-up	Imaging	Presence of definition of subset/ subgroup/cluster	Significant characteristics of subset/subgroup/cluster	Suggested PMR-like condition
Oka et al. [20]	Case series and review	72 CDS published cases 7 (19.4%) PMR-like			CT	CDS-CPPD with PMR-like presentation	Higher CRP (mean 12.6 mg/dl), frequent fever (80.4%), prompt response to only NSAIDs 67.5%	Yes, different diagnosis and therapy between CPPD and PMR
Manzo et al. [21]	Prospective monocentric	134 PMR (by GP) 41 PMR 93 not PMR	Healey	18 months	Not specified	PMR with diagnosis of other conditions	PMR/CPPD: 11%	Yes, but authors consider the possible overlap between the two diseases
Ottaviani et al. [22]	Prospective monocentric	94 Sy PMR – 52 PMR diagnosis – 25/52 (48%) PMR/ CPPD	ACR/EULAR 2012 crit. and McCarty/ Zhang ACR/ EULAR recommend.	n.r.	US and SFA	Yes: PMR patients with concurrent diagnosis of CPPD (on US and synovial fluid analysis)	PMR/CPPD: 48%, more frequent older females, with humeral head erosions, synovitis and calcifications of AC joint. Lesser frequency of SAD bursitis	Yes, PMR/CPPD is considered other condition, suggesting shorter courses of GC
Conticini et al. [23]	Retrospective multicentric	204 PMR 31 CPPD over 104 evaluated by MSUS (22%)	Bird 2012 ACR EULAR	Range 12–60 months	MSUS and CDUS vs. only clinical	Yes: PMR with early or late diagnostic shift in other diagnosis	PMR/CPPD: 22%, patients with more frequent peripheral synovitis, lesser frequent flares, lesser dependence on GC, more frequent use of DMARDs	Yes, PMR/CPPD is considered other condition, suggesting different management. Only PMR with MSUS at onset can have a change on diagnosis

AC – acromion-clavicular, ACR – American College of Rheumatology, CDUS – color duplex ultrasound, CPPD – calcium pyrophosphate deposition, CRP – C-reactive protein, CT – computed tomography, DMARDs – disease-modyfing antirheumatic drugs, ESR – erythrocyte sedimentation rate, EULAR – European Alliance of Associations for Rheumatology, GC – glucocorticosteroid, MSUS – musculoskeletal ultrasound, n.r. – not reported, NSAID – nonsteroidal anti-inflammatory drugs, PLT – platelet count, PMR – polymyalgia rheumatica, SAD – subacromial deltoid, US – ultrasound.

not excluded in a 2018 monocentric cohort study performed in a primary care setting [21].

More recently, Ottaviani et al. [22] found a surprisingly high percentage of CPPD disease (48.07%) in patients considered to have PMR according to the 2012 EULAR/ACR criteria for PMR. In their observational study, CPPD of the acromion-clavicular (AC) joint had the best ratio of sensitivity to specificity (sensitivity: 85.2%; specificity: 97.1%). Consequently, they proposed ultrasonographic (US) assessment of the AC joints as a more effective way to distinguish CPPD from true PMR.

Crowned dens syndrome can be a clinical-radiological manifestation for CPPD disease. It occurs with acute or subacute upper-neck pain limiting rotation of the cervical spine. Usually, fever and elevated inflammatory markers are present. The presence of calcification around the dental process of the second cervical vertebra (the "crowned dens") on cervical computed tomography (CT) is almost pathognomonic [28, 29]. Crowned dens syndrome is estimated to represent about 5% of acute presentations of CPPD disease, and it is therefore considered rare. However, in some countries (such as Japan) it is not uncommon, being diagnosed within 1 day of its presentation during hospitalization [30].

Crowned dens syndrome clinical manifestations can mimic meningitis, infectious spondylodiscitis, or septic arthritis of the facet joint. Is CDS a potential PMR mimicker? Painful stiffness of the pelvic girdle is always absent in CDS patients, suggesting that this is a discriminating manifestation in clinical practice. According to the 2012 EULAR/ACR classification criteria, patients aged 50 years or older presenting with bilateral shoulder pain and elevated CRP and/or ESR can be classified as having PMR in the presence of morning stiffness > 45 min and new hip pain, in the absence of peripheral synovitis or positive rheumatoid arthritis (RA) serology [6]. Therefore, the possibility that CDS may be a PMR-like disease seems more like speculative reasoning than actual diagnostic uncertainty.

Additionally, in their 2004 case series, Aouba et al. [14] reported on 4 patients first diagnosed with PMR whose final diagnosis was CDS. In these patients, GCs were ineffective, and a fast improvement followed treatment with nonsteroidal anti-inflammatory drugs (NSAIDs) and colchicine.

Finally, an interesting aspect should be underlined regarding the prognosis and clinical course of patients with PMR and CPPD. In fact, in all the studies on this topic, no patient with PMR/CPPD has had a late diagnosis of GCA (at least within the term of follow-up of each study) [15, 23]. On the other hand, patients with pure PMR not infrequently experience late GCA (16–47% within 2–24 months of PMR diagnosis). This remarkable divergence in clinical course raises the question of whether PMR/CPPD may constitute a peculiar non-vasculitic subset.

Ultrasonographic assessment is included as an option in the 2012 classification criteria for PMR, where US criteria (Table II) increased the specificity (from 81.5 to 91.3%). Additionally, US serves as a valuable tool for excluding alternative diagnoses such as non-RA shoulder conditions and subjects without shoulder conditions. Ultrasonography has proved helpful in the diagnosis of CPPD [31]. In 2023, an international multidisciplinary working group developed a framework for diagnosing CPPD based on imaging modalities, including US [32]. Typical US findings were considered as follows: 1) crystal deposition must be found in the fibrocartilage or hyaline cartilage to qualify as CPP deposition; 2) crystal deposition must be described as hyperechoic deposits with variable shape and size and not creating posterior shadowing; 3) hyperechoic deposits may also be visualized on US in the synovial membrane, joint capsule, or tendons, and 4) in dynamic scanning, the deposits situated within hyaline cartilages move solidly in a synchronous way and in the same direction of joint movement, whereas CPP deposits of the synovial membrane and joint capsule move in opposition to joint movement, as cartilage slides under the capsule and synovial membrane. Basic calcium phosphate (BCP) crystals – mostly partially carbonate substituted hydroxyapatite crystals – are associated with BCP deposition (BCPD) disease, in tendons (calcific tendinopathy of the shoulder and hip), bursae, or joints, where they can lead to a severe destructive arthropathy associated with osteoarthritis (mostly in large joints such as the shoulders, where the pathology is called Milwaukee shoulder syndrome) [33]. In BCP arthropathy, a recent review concluded that there are limited data on the utility of US in differentiating the crystals of BCP and CPP [34], whereas in the 2023 EULAR recommendations on imaging for diagnosis of crystal-induced arthropathies, Recommendation 5 states that in the diagnostic assessment of BCPD, imaging is necessary, and conventional radiology or US is the recom-

Table II. US criteria proposed by the 2012 EULAR/ACR collaborative initiative [6]

• At least one shoulder with subdeltoid bursitis and/or biceps tenosynovitis and/or glenohumeral synovitis and at least one hip with synovitis and/or trochanteric bursitis
• Both shoulders with subdeltoid bursitis, biceps tenosynovitis, or glenohumeral synovitis

mended modality [35]. In any case, a risk of misidentification should always be taken into account in clinical practice for some consequences: as an example, the response to local intra-articular GCS may be impressive in patients with CPPD disease, whereas it should be used with caution in patients with BCD disease because of the risk of disrupting the hydroxyapatite deposit or seeding further calcification.

Finally, new classification criteria for CPPD disease have been recently proposed. In the 1960s, Ryan and McCarty first proposed diagnostic criteria for CPPD disease. According to these criteria, the definite diagnosis was when CPP crystals were found (presence of both typical linear or punctate calcification on radiography and findings consistent with CPP crystals on synovial fluid polarised light microscopy) [36].

In 2023, a EULAR/ACR collaborative initiative proposed classification criteria for symptomatic CPPD disease [37]. These criteria would apply to CPPD disease as a whole, which meant that separate classification criteria for each clinical presentation were not within the aim of this initiative. The identification of CPP crystals in synovial fluid from a symptomatic joint was sufficient for classification as CPPD as long as the exclusion criteria were not met (e.g. another condition did not explain the entire presentation). Crowned dens syndrome was another sufficient criterion. Attribution of symptoms to CPPD disease can be challenging, particularly in patients with coexisting conditions. Those patients for whom all symptoms are better explained by another coexisting condition cannot be categorized as having CPPD disease.

In the absence of the aforementioned two sufficient criteria, patients could be categorized by scoring some remaining imaging and clinical criteria through 8 domains and several levels: the face validity of a threshold score of > 56 was assessed. In other words, according to the 2023 EULAR/ACR criteria, a patient without sufficient criteria may be equally classified as having CPPD disease if the sum of points according to the criteria presented in Table III is > 56 points.

Among the 148 definite mimickers forming the derivation cohort, 6 were affected by PMR. The classification criteria demonstrated high sensitivity and specificity in an independent validation cohort. Among the 162 definite mimickers forming the validation cohort, only 1 was affected by PMR [37].

Given the above data, when should we consider CPPD disease as a differential diagnosis in a patient with polymyalgic manifestations categorized as having PMR according to the 2012 EULAR/ACR criteria? Schematically, there are 3 possible scenarios that may occur in clinical practice.

Polymyalgic manifestations categorized as PMR in a patient with already diagnosed CPPD disease: PMR/CPPD or pseudo-PMR CPPD pattern.

Proximal involvement is reported as common in patients with chronic pyrophosphate arthropathy. In these patients, US assessment may be useful for differentiating true PMR from chronic CPPD disease with polymyalgic manifestations. Additionally, peripheral joints are commonly involved in chronic CPPD disease. In contrast, absence of peripheral synovitis is one of the 2012 classification criteria for PMR. In these patients with pseudo-PMR CPPD pattern, oral GCs can be used. However, GCs rarely give the rapid improvement that the clinician usually observes in a patient with true, isolated PMR. NSAIDs associated with colchicine are conversely more effective [38].

A previous diagnosis of CPPD disease does not exclude a true PMR: PMR with concurrent symptomatic CPPD disease. In this case, the therapeutic recommendations for PMR should be used. However, CPPD disease must also be treated if widespread and massive, to avoid its complications [39].

Polymyalgic manifestations categorized as PMR in patients with undiagnosed chronic CPPD disease.

In this case, the critical point is whether the polymyalgic manifestations can be explained by PMR or by CPPD disease. Ultrasonography assessment may be crucial for a correct diagnosis, and consequently for a proper therapeutic approach.

Conclusions

The possibility that CPPD disease and PMR may be present in the same patient is not negligible, especially if the patient is aged over 70 years. On the other hand, CPPD disease is often included in the list of common PMR-mimicking diseases, because CPPD patients with marked proximal involvement and elevated acute inflammatory markers may be categorized as having PMR.

The relationship between PMR and CPPD disease should be reviewed in the light of the 2023 EULAR/ACR classification criteria for symptomatic CPPD disease. Specifically, presence of CPP crystals in synovial fluid and/or a score > 56 points (using additional weighted criteria taking into account clinical features, associated metabolic disorders, and results of laboratory and imaging investigations) can classify patients as having CPPD disease with polymyalgic manifestations. In these patients, a diagnosis of PMR-like CPPD is therefore possible. On the other hand, when these criteria for symptomatic CPPD are absent (no CPP crystals and/or score < 56 points), CPPD should be considered a coincidental finding in patients with PMR (that is, PMR without symptomatic CPPD).

Table III. Domains, levels and points in the 2023 EULAR/ACR classification criteria for CPPD disease [37]

Domains and levels		Points
A	Age at onset of joint symptoms (pain, swelling, and/or tenderness)	
	≤ 60 years	0
	> 60 years	4
B	Time course and symptoms of inflammatory arthritis	
	No persistent or typical inflammatory arthritis	0
	Persistent inflammatory arthritis	9
	1 typical acute arthritis episode	12
	More than 1 typical acute arthritis episode	16
C	Sites of typical episode(s) of inflammatory arthritis in peripheral joints	
	1st MTPJ	–6
	No typical episode(s)	0
	Joint(s) other than wrist, knee or 1st MTPJ	5
	Wrist	8
	Knee	9
D	Related metabolic diseases	
	None	0
	Present	6
E	Synovial fluid crystal analysis from a symptomatic joint	
	CPP crystals absent on ≥ 2 occasions	–7
	CPP crystals absent on 1 occasion	–1
	Not performed	0
F	OA of hand/wrist on imaging (defined as present if the Kellgren and Lawrence score is ≥ 2)	
	None of the following findings or no wrist/hand imaging performed	0
	Bilateral radio-carpal joints	2
	≥ 2 of the following: STTJ OA without 1st CMCJ OA; 2nd MCPJ OA; 3rd MCPJ OA	7
G	Imaging evidence of CPPD in symptomatic peripheral joint(s)	
	None on US, CT, or DECT (and absent on CR or CR not performed)	–4
	None on CR (and US, CT, DECT not performed)	0
	Present on either CR, US, CT, or DECT	16
H	Number of peripheral joints with evidence of CPPD on any imaging modality regardless of symptoms	
	None	0
	1	16
	2–3	23
	≥ 4	25

CMCJ – carpometacarpal joint, CPPD – calcium pyrophosphate disease, CR – conventional radiography, CT – computed tomography, DECT – dual-energy computed tomography, MCPJ – metacarpophalangeal joint, MTPJ – metatarsophalangeal joint, OA – osteoarthritis, STTJ – scaphotrapeziotrapezoid joint, US – ultrasound.

We look forward to future prospective studies applying the recent EULAR/ACR criteria, in order to better classify patients with polymyalgic manifestations and CPPD.

Likewise, further studies are required to confirm the possibility that PMR/CPPD may be a non-vasculitic pattern of disease.

Disclosures

Ciro Manzo and Paolo Falsetti participated equally in preparing this study.

Conflict of interest: The authors declare no conflict of interest.

Funding: There was no external funding regarding the work described in the article.
Ethics approval: Not applicable.
Data availability: The data that support the findings of this study are available on request from the corresponding author (C.M.).

References

1. Sharma A, Mohammad AJ, Turesson C. Incidence and prevalence of giant cell arteritis and polymyalgia rheumatica: A systematic literature review. Semin Arthritis Rheum 2020; 50: 1040–1048, DOI: 10.1016/j.semarthrit.2020.07.005.
2. Cimmino MA, Zaccaria A. Epidemiology of polymyalgia rheumatica. Clin Exp Rheumatol 2000; 18 (4 Suppl 20): S9–S11.
3. Manzo C. Incidence and Prevalence of Polymyalgia Rheumatica (PMR): The Importance of the Epidemiological Context. The Italian Case. Med Sci (Basel) 2019; 7: 92, DOI: 10.3390/medsci7090092.
4. Espígol-Frigolé G, Dejaco C, Mackie SL, et al. Polymyalgia rheumatica. Lancet 2023; 402: 1459–1472, DOI: 10.1016/S0140-6736(23)01310-7.
5. Colombo MG, Wetzel AJ, Haumann H, et al. Polymyalgia rheumatica. Dtsch Arztebl Int 2022; 119: 411–417, DOI: 10.3238/arztebl.m2022.0218.
6. Dasgupta B, Cimmino MA, Kremers HM, et al. 2012 provisional classification criteria for polymyalgia rheumatica: A European League Against Rheumatism/American College of Rheumatology collaborative initiative. Arthritis Rheum 2012; 64: 943–954, DOI: 10.1002/art.34356.
7. Manzo C, Camellino D. Polymyalgia rheumatica: diagnostic and therapeutic issues of an apparently straightforward disease. Recenti Progress Med 2017; 108: 221–231, DOI: 10.1701/2695.27559 [Article in Italian].
8. Mukhtyar CB, Twohig H, Hodgson H. Getting it right for polymyalgia rheumatica. Rheumatology (Oxford) 2022; 61: 4221–4222, DOI: 10.1093/rheumatology/keac187.
9. Ceccato F, Uña C, Regidor M, et al. Conditions mimicking polymyalgia rheumatica. Reumatol Clin 2011; 7: 156–160, DOI: 10.1016/j.reuma.2010.09.001.
10. Pascart T, Filippou G, Lioté F, et al. Calcium pyrophosphate deposition disease. Lancet Rheumatol 2024: S2665-9913(24)00122-X, DOI: 10.1016/S2665-9913(24)00122-X.
11. Bouvet JP, le Parc JM, Michalski B, et al. Acute neck pain due to calcifications surrounding the odontoid process: the crowned dens syndrome. Arthritis Rheum 1985; 28: 1417–1420.
12. Richette P, Bardin T, Doherty M. An update on the epidemiology of calcium pyrophosphate dydrate cristal deposition disease. Rheumatology 2009; 48: 711715, DOI: 10.1093/rheumatology/kep081.
13. Dieppe PA, Alexander GJM, Jones HE, et al. Pyrophosphate arthropathy: a clinical and radiological study of 105 cases. Ann Rheum Dis 1982; 41: 371–376, DOI: 10.1136/ard.41.4.371.
14. Aouba A, Vuillemin-Bodaghi V, Mutschler C, De Bandt M. Crowned dens syndrome misdiagnosed as polymyalgia rheumatica, giant cell arteritis, meningitis or spondylitis: an analysis of eight cases. Rheumatology (Oxford) 2004; 43: 1508–1512, DOI: 10.1093/rheumatology/keh370.
15. Pego-Reigosa JM, Rodriguez-Rodriguez M, Hurtado-Hernandez Z, et al. Calcium pyrophosphate deposition disease mimicking polymyalgia rheumatica: a prospective follow up study of predictive factors for this condition in patients presenting with polymyalgia symptoms. Arthritis Rheum 2005; 53: 931–938, DOI: 10.1002/art.21585.
16. Salaffi F, De Angelis R, Grassi W; MArche Pain Prevalence; INvestigation Group (MAPPING) study. Prevalence of musculoskeletal conditions in an Italian population sample: results of a regional community-based study. I. The MAPPING study. Clin Exp Rheumatol 2005; 23: 819–828.
17. Yanai H, Yoshida H, Tada N. Clinical, radiological, and biochemical characteristics in patients with diseases mimicking polymyalgia rheumatica. Clin Interv Aging 2009; 4: 391–395, DOI: 10.2147/cia.s7489.
18. Siau K, Lee M, Laversuch CJ. Acute pseudogout of the neck – the crowned dens syndrome: 2 case reports and review of the literature. Rheumatol Int 2011; 31: 85–88, DOI: 10.1007/s00296-009-1145-7.
19. Falsetti P, Acciai C, Volpe A, Lenzi L. Ultrasonography in early assessment of elderly patients with polymyalgic symptoms: a role in predicting diagnostic outcome? Scand J Rheumatol 2011; 40: 57–63, DOI: 10.3109/03009742.2010.486766.
20. Oka A, Okazaki K, Takeno A, et al. Crowned Dens Syndrome: Report of Three Cases and a Review of the Literature. J Emerg Med 2015; 49: e9–e13, DOI: 10.1016/j.jemermed.2015.02.005.
21. Manzo C, Natale M, Traini E. Diagnosis of polymyalgia rheumatica in primary health care: favoring and confounding factors – a cohort study. Reumatologia 2018; 56: 131–139, DOI: 10.5114/reum.2018.76900.
22. Ottaviani S, Goossens J, Demaria L, et al. Ultrasound shoulder assessment of calcium pyrophosphate disease with suspected polymyalgia rheumatica. Clin Exp Rheumatol 2020; 38: 1170–1175.
23. Conticini E, Falsetti P, d'Alessandro M, et al. Clinical, laboratory and ultrasonographic findings at baseline predict long-term outcome of polymyalgia rheumatica: a multicentric retrospective study: Polymyalgia rheumatica predicted by ultrasonographic findings polymyalgia rheumatica outcome predicted early by ultrasound. Intern Emerg Med 2023; 18: 1929–1939, DOI: 10.1007/s11739-023-03373-x.
24. Guggino G, Ferrante A, Macaluso F, et al. Pathogenesis of polymyalgia rheumatica. Reumatismo 2018; 70: 10–17, DOI: 10.4081/reumatismo.2018.1048.
25. Manzo C, Milchert M, Natale M, Brzosko M. Polymyalgia rheumatica with normal inflammatory indices at the time of diagnosis: can we just move a step forward? Reumatologia 2020; 58: 184–186, DOI: 10.5114/reum.2020.96549.
26. Sirotti S, Scanu A, Pascart T, et al. Calcium Pyrophosphate Crystal Formation and Deposition: Where Do we Stand and What Does the Future hold? Curr Rheumatol Rep 2024; 26: 354–365, DOI: 10.1007/s11926-024-01161-w.
27. Manzo C, Nune A, Castagna A. Why would immuno- and endocrino-senescence, age-related changes in the gut microbiota, and susceptibility to infection favour polymyalgia rheumatica over seronegative elderly-onset rheumatoid arthritis? Clin Exp Rheumatol 2023; 41 Suppl 135: 25–26, DOI: 10.55563/clinexprheumatol/hb7van.

28. Naim T, Kham D, Ali M, Fanciullo J. Crowned Dens Syndrome: A Challenging Diagnosis in Older Adults Presenting With Acute Neck Pain. J Gen Fam Med 2023; 24: 171–177, DOI: 10.1002/jgf2.616.
29. Shams S, Khan B, Jeffries A. Crowned dens syndrome – case of crystal deposition in cervical spine. Oxf Med Case Reports 2022; 2022: omab135, DOI: 10.1093/omcr/omab135.
30. Isono H, Haruka Kuno H, Hozumi T. Crowned dens syndrome: A case series of 72 patients at eight teaching hospitals in Japan. Cureus 2023; 15: e37101, DOI: 10.7759/cureus.37101.
31. Filippou G, Miguel-Perez M, Bong D, et al. The ultrasonographic ''pseudodouble contour'' sign in calcium pyrophosphate deposition disease: an anatomic explanation and how to distinguish it from gout. Arthritis Rheumatol 2023; 75: 639–640, DOI: 10.1002/art.42397.
32. Tedeschi SK, Becce F, Pascart T, et al. Imaging features of calcium pyrophosphate deposition disease: consensus definitions from an international multidisciplinary working group. Arthritis Care Res (Hoboken) 2023; 75: 825–834, DOI: 10.1002/acr.24898.
33. Rosenthal AK. Basic calcium phosphate crystal-associated musculoskeletal syndromes: an update. Curr Opin Rheumatol 2018; 30: 168–172, DOI: 10.1097/BOR.0000000000000477.
34. Sirotti S, Terslev L, Filippucci E, et al. OMERACT Ultrasound working group-CPPD subgroup. Development and validation of an OMERACT ultrasound scoring system for the extent of calcium pyrophosphate crystal deposition at the joint level and patient level. Lancet Rheumatol 2023; 5: e474–e482, DOI: 10.1016/S2665-9913(23)00136-4.
35. Mandl P, D'Agostino MA, Navarro-Compán V, et al. 2023 EULAR recommendations on imaging in diagnosis and management of crystal-induced arthropathies in clinical practice. Ann Rheum Dis 2024; 83: 752–759, DOI: 10.1136/ard-2023-224771.
36. McCarty D. Pseudogout. In: Hollander JL (ed.). Arthritis and Allied Conditions: a Textbook of Rheumatology. 7th ed. Lea & Febiger, Philadelphia 1966; 947–964.
37. Abhishek A, Tedeschi SK, Pascart C, et al. The 2023 ACR/EULAR Classification Criteria for Calcium Pyrophosphate Deposition Disease. Arthritis Rheumatol 2023; 75: 1703–1713, DOI: 10.1002/art.42619.
38. Doherty M, Abhishek A. Calcium pyrophosphate crystal associated arthropathy. In: Hochberg MC, Silman AJ, Smolen JS, et al. (eds.). Rheumatology. Elsevier, Philadelphia 2011; 1882.
39. Voulgari PV, Venetsanopoulou AI, Drosos AA. Recent advances in the therapeutic management of calcium pyrophosphate deposition disease. Front Med (Lausanne) 2024; 11: 1327715, DOI: 10.3389/fmed.2024.1327715.