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EDITED BY

Elena Massarotti,
Brigham and Women's Hospital and Harvard
Medical School, United States

REVIEWED BY

Naomi I. Maria,
Northwell Health, United States
Ozge Basaran,
Hacettepe University, Türkiye

*CORRESPONDENCE

Antonio Bustos-Merlo
✉ antoniobustosmerlo@gmail.com

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Case Report: Overlap syndrome of relapsing polychondritis and systemic lupus erythematosus observed in women: a comprehensive review and unique case presentation

Antonio Bustos-Merlo^{1*}, Nuria Navarrete-Navarrete¹,
Lourdes González-Navarrete² and María Martín-Armada¹

¹Autoimmune and Systemic Diseases Unit, Department of Internal Medicine, Hospital Universitario Virgen de las Nieves, Granada, Spain, ²Department of Internal Medicine, Hospital Universitario de Canarias, Tenerife, Spain

Relapsing polychondritis (RP) is an immune-related condition involving inflammation of cartilaginous structures and other body tissues, especially affecting the ears, nose, eyes, joints and respiratory tract. Approximately one-third of patients with RP have an identifiable form of systemic vasculitis, connective tissue disease, other autoimmune disease, or a malignant or premalignant condition. In this report, we present the case of a 50-year-old woman who met criteria for systemic lupus erythematosus (SLE) and was diagnosed with RP several months later. The main objective was to investigate cases of overlap syndrome involving RP and SLE. A literature review of reported cases with this coexistence was conducted, examining presentation, predominant symptoms, laboratory data, complications and systemic involvement. The review included a total of 12 cases of overlap syndrome of RP and SLE, analysing the epidemiology, temporal relationship, clinical features, diagnosis, treatment and evolution of the reported cases. Although this overlapping presence is rare (less than 1%), it emphasises the importance of maintaining a high index of suspicion for RP in patients with SLE who present with recurrent episodes of scleritis, audiovestibular involvement and chondritis.

KEYWORDS

relapsing polychondritis, systemic lupus erythematosus, chondritis, audiovestibular involvement, overlap syndrome

Introduction

Relapsing polychondritis (RP) is a rare inflammatory disease of unknown etiology and autoimmune basis. It is characterised by recurrent inflammatory lesions affecting cartilage, cardiovascular system and sensory organs. After its initial description by Jaksch-Wartenhorst in 1923 as polychondropathy (1), several authors have reported cases under different names, with Pearson's proposal, chronic atrophic polychondritis, being

Abbreviations

APS, antiphospholipid antibodies; ANA, antinuclear antibodies; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; RP, relapsing polychondritis; SLE, systemic lupus erythematosus; TLR, toll-like receptors; VEXAS, vacuoles, E-1 enzyme, X-linked, autoinflammatory, somatic.

the most widely accepted (2). The epidemiology of RP is unclear, with an estimated annual incidence between 0.71 and 2.0 per million people (3, 4). It affects both sexes equally, mainly seen in the fifth decade of life. Up to one third of patients with RP have a recognisable form of systemic vasculitis, connective tissue disease or other autoimmune disorder, or a malignant or pre-malignant autoimmune disease (5, 6). A high percentage of patients with a genetic disease described in 2020 called VEXAS syndrome (vacuoles, E-1 enzyme, X-linked, autoinflammatory, somatic) have an initial clinical diagnosis of RP (7). VEXAS syndrome is characterised by manifestations such as recurrent fever, skin and lung inflammation, auricular and nasal chondritis, vasculitis, deep vein thrombosis and arthralgias, together with progressive haematological abnormalities including macrocytic anaemia and thrombocytopenia.

In this report, we describe a rare case of overlap syndrome involving PR and systemic lupus erythematosus (SLE), accompanied by a review of the literature.

Clinical case

A 50-year-old Caucasian woman presented to the outpatient clinic for review after being diagnosed with SLE during a hospital admission four months earlier. Table 1 describes her clinical and laboratory features prior to the current admission, including those that scored as criteria for SLE classification. The patient had 19 ACR/EULAR points for SLE classification, with 10 being the minimum required in addition to the presence of antinuclear antibodies (ANAs). The patient was under treatment with prednisone (7.5 mg), hydroxychloroquine (100 mg) and acetylsalicylic acid (100 mg) daily. Despite treatment, the patient returned to experience daily fever up to 38°C, which she tolerated well. In addition, she had bilateral parotid swelling and auditory symptoms such as vertigo, hearing loss and tinnitus in both ears. He also experienced hemicranial headache, with partial improvement after the use of regular analgesics. Three months prior to the current visit, the patient sought relief from her headaches through multiple punctures of both pinnae at an acupuncture centre. The day after these procedures, she experienced marked swelling in the auricular cartilages, as seen in Figure 1.

The symptoms disappeared spontaneously two weeks after onset. On physical examination, the patient's vital signs were as follows: blood pressure 130/73 mmHg, heart rate 77 bpm and temperature 36.5°C. Ocular redness was observed left. Pupils were isochoric, normoreactive to light and accommodation, with preserved extrinsic eye movements. No alterations were detected in the cranial nerves. Sensibility and strength were preserved in all extremities. When gait was explored at, there was a tendency to fall to the left. The Romberg's test was negative. The parotid glands were enlarged and painful on palpation.

During the evaluation by the otorhinolaryngologist, a diagnosis of sudden sensorineural hearing loss in the left ear with a severe left vestibular deficit was made. Simultaneously, she was reviewed by the ophthalmologist, who diagnosed acute scleritis in the right

eye. Topical ocular glucocorticoid therapy was started, together with intravenous boluses of methylprednisolone (100 mg for 3 days) and immunoglobulins (20 mg daily for 5 days). Despite ocular improvement, the audiovestibular symptomatology showed little improvement. Therefore, it was decided to administer intratympanic dexamethasone. Subsequently, oral glucocorticoid treatment was maintained, associated with subcutaneous administration of methotrexate. The patient was left with severe auditory and vestibular sequelae. During follow-up in the following months, the patient presented recurrent episodes of polycondritis (affecting the auricular pavilions, the nose and the costosternal area) and recurrent scleritis. Colchicine was added to her usual treatment, the dose of methotrexate was increased to 15 mg weekly and adalimumab was added. These episodes resulted in a saddle-shaped nasal deformity. After 15 months of follow-up, the patient experienced progressive edematisation of the lower limbs with foamy urine. Methylprednisolone 250 mg boluses were administered for 3 days and a renal biopsy was performed, with a diagnosis of class V lupus nephropathy. Treatment with adalimumab, which had successfully controlled the episcleritis, was discontinued and remission induction with cyclophosphamide 500 mg fortnightly for 3 months and subsequent maintenance with mycophenolate mofetil was performed. Subsequently, tacrolimus was added, which was ineffective. Finally, indicated treatment with rituximab (1 g/15 days, 2 doses). Two months after starting rituximab, total protein excretion was 0.45 g per day. Currently, the patient undergoes regular check-ups and maintains a residual non-nephrotic range proteinuria of about 0.3 g per day.

Discussion

A comprehensive search of MEDLINE databases was conducted through PubMed and EMBASE until 1 February 2023. All articles describing cases of overlap syndrome of RP and SLE were considered for inclusion. Cases that did not meet the established criteria for the diagnosis of SLE or had incomplete information were excluded. No language restrictions were applied in the search. In addition, the references of the selected articles were screened to ensure the completeness of the search. In total, 20 references were included, covering 46 cases; however, clinical and analytical features were detailed in only 11 references, totalling 12 cases. The flow chart provides a visual representation of the complete search and the articles incorporated in the review (Figure 2).

A total of 12 cases were included and analyzed, all of whom were women. Their characteristics are presented in Tables 2–4 (8–18). The mean age of the published cases with this coexistence was 37.5 years with a median of 46.5 years (range 29–75). Although in our case, RP appeared after the diagnosis of SLE, we observed that it can appear simultaneously (15–17), later (10, 11, 16), with a latency period of up to 9 years (10) or prior to (12, 14). In terms of clinical manifestations, chondritis was present in all cases, with the most frequent affected site being the outer ear, either unilaterally or bilaterally. Ocular involvement

TABLE 1 Characteristics and laboratory data of our case patient.

		Previous admission	On admission	Fifteen months after admission
Medical history		Febrile syndrome accompanied by cutaneous lesions exacerbated by sunlight exposure ^a	Ocular redness, vertigo, hearing loss, and tinnitus in both ears	Progressive oedema in the lower extremities with foamy urine
Blood	Reference range, this hospital			
Haemoglobin (g/dl)	11–17	9.7	11.7	10.9
Haematocrit (%)	3.8–5.5	3.55	3.87	3.56
White-cell count (x10 × 3/μl)	3.5–10.5	2.29 ^a	5.15	3.32
Neutrophils (x10 × 3/μl)	1.5–7.7	1.63	3.57	2.11
Lymphocytes (x10 × 3/μl)	1.1–4.5	0.34 ^a	1.12	0.83
Platelet count (x10 × 3/μl)	120–450	63 ^a	177	266
Erythrocyte sedimentation rate (mm/h)	1–10	112	59	66
C-reactive protein (mg/L)	0–1.5	154	23.8	0.8
Glucose (mg/dl)	75–115	81	86	94
Creatinine (mg/dl)	0.51–0.95	0.55	0.69	0.48
Urea nitrogen (mg/dl)	18–37.7	22	34	24
Antinuclear antibodies		1/1280 ^a	U	U
Anti-dsDNA (U/ml)	0–20	23 ^a	3.5	45
Direct coombs test		Mono-specific C3D	Mono-specific C3D	Mono-specific C3D
Antiphospholipid antibodies		Anti-β2 glycoprotein and anticardiolipin +; lupus anticoagulant ^a	U	U
C3 (mg/dl)	90–180	65.4 ^a	86	100
C4 (mg/dl)	10–40	8.5 ^a	12.3	26
Proteinuria 24 h (g)	<0.5	0.4	0.5	0.3
Radiology				
Abdominal and renal ultrasound		Normal	Normal	Normal
Chest radiography		Normal	Normal	Normal
Cranial magnetic resonance		U	Normal	U
Histology				
Renal biopsy				Nephropathy class V

^aACR/EULAR classification criteria for systemic lupus erythematosus; U, unknown.

was present in 33.3%, in the form of conjunctivitis, scleritis or episcleritis (8, 12, 13, 17). Although we can see that the audiovestibular manifestations that presented in our patient are not so frequent; vestibular syndrome in 16.6% (11, 17) and sensorineural hypoacusis in 23.08% (11, 13, 17). The same applies to the involvement of the nasal cartilage, which was affected in 25% of cases (12, 16, 17). As it coincides with the series of patients with SLE, urinary alteration is seen in approximately half of the cases described. Cardiac involvement occurred in only 1 case (8). With regard to analytical parameters, which are not specified in most publications, elevated erythrocyte sedimentation rate (ESR) was found in five cases (8, 10–12, 15). Regarding the presence of published antibodies, are positive the double-stranded anti-dsDNA antibodies in five cases (8, 10, 13, 14, 18, 19). Regarding histopathology, renal biopsy was performed in four patients and skin biopsy in four other patients. The most frequently used treatment was oral corticosteroids together with hydroxychloroquine. There was only one case of rituximab administration for neurological manifestations (14). There was little information regarding patient follow-up, being unknown in most of them.

Complications included, as in our patient, saddle nose deformity in 16.6% (9, 12) and one case of respiratory distress syndrome due to tracheal stenosis and the need for urgent tracheostomy (12).

This review provides a comprehensive overview of the overlap of RP and SLE, highlighting the diversity of clinical manifestations and the need to consider this connection in clinical management. The differential diagnosis of RP should include granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis, Behçet's disease, Cogan's syndrome (interstitial keratitis and audiovestibular dysfunction) and leprosy. In the case of a male patient VEXAS, which may present with chondritis of the ear (usually unilateral) or of the nose, thrombopenia and macrocytic anaemia, should be noted. As with other immune-mediated diseases, coexistence with another immune-mediated disease, in this case SLE, is not uncommon. Interestingly, both our patient and all cases published in the literature are female. The reason for this is unknown, although we know that this prevalence of female gender is observed in most autoimmune and immune-mediated diseases in general. There are several hypotheses to explain the current gender bias. The human endosomal Toll-like receptors, TLR7 and TLR8, are important mediators of innate



FIGURE 1
Image of the patient, shows swelling of the left auricle.

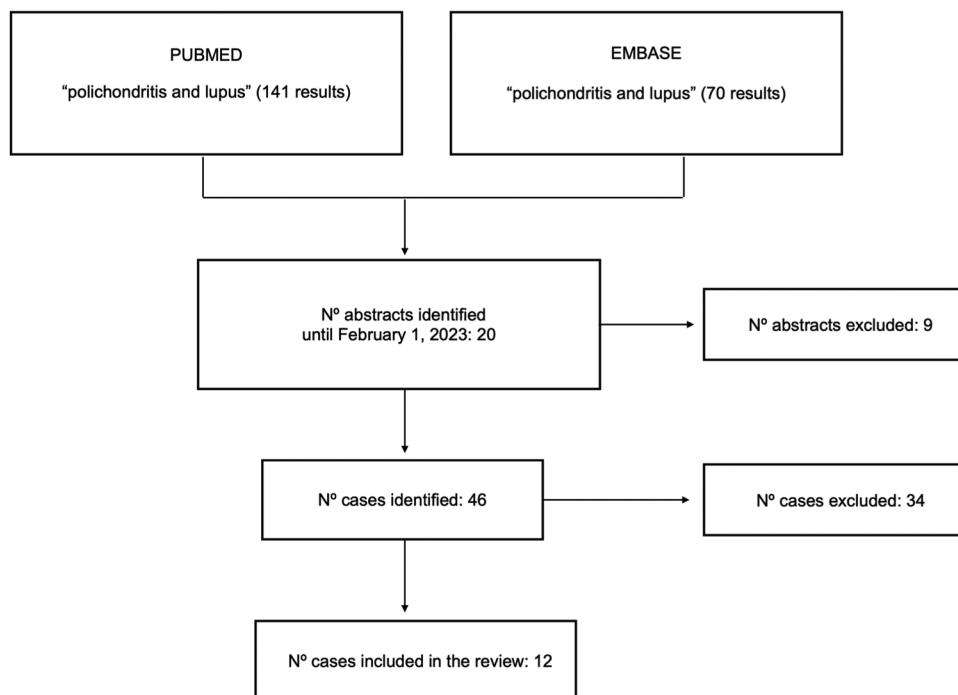


FIGURE 2
Flow chart of the selection of articles included in this review.

TABLE 2 Demographics and clinical findings of reported cases.

Characteristic		Median (range) or N (%)
Demographics	Females/Males	12 (100)/0
	Age, years	46.5 (26–75)
	White/Black/U	6 (50)/1(8)/5 (41)
Clinical findings	Chondritis	12 (100)
	Ocular inflammation	4 (33)
	ENT manifestations (hearing loss, vestibular syndrome, nasal discharge)	5 (42)
	Seronegative polyarthritis	9 (75)
	Cardiac involvement	1 (8)
	Neurologic involvement	2 (17)
	Haematologic involvement	6 (50)
	Kidney involvement	5 (42)
Temporal relationship debut SLE and RP		
Laboratory parameters	ANA	12 (100)
	Anti-dsDNA	5 (42)
Histology	Skin/cartilage biopsy	3 (25)
	Kidney biopsy	3 (25)
Treatment	Corticosteroids	10 (83)
	Other immunosuppressant	1 (8)
Complications	Saddle nose	2 (17)
	Airway stenosis	1 (8)

immunity and autoimmune pathogenesis. Their signalling promotes the earliest appearance of haematopoietic cells during development and thereafter influences the fate and function of primitive and effector immune cell types (20). TLR7 and TLR8 are encoded respectively by adjacent X-linked genes. Both evade X-chromosome inactivation in female immune cells. Cells harbouring foci of simultaneous TLR7 and TLR8 transcription are more frequent in females and males with Klinefelter syndrome than in euploid males, resulting in a sevenfold frequency difference (21). Active co-dependent transcription. In addition, it has been shown that there is a mechanism by which TLR7 and TLR8 direct monocyte fate and identify a specialised population of phagocytes responsible for inflammation and infection-associated anaemia and thrombocytopenia (22). Elevated levels of interferon gamma have also been corroborated in both murine models and human patients with SLE, systemic sclerosis and Sjögren's syndrome (23), indicating that interferon gamma may be another factor contributing to female bias in autoimmunity.

The clinical course of RP is unpredictable and variable between patients. In relation to ear involvement, unilateral or bilateral external ear inflammation is the most common presenting feature, with hearing impairment, tinnitus or vertigo being less common (24). The latter is a consequence of inner ear inflammation associated with destruction induced by Eustachian tube chondritis, endolymphatic hydrops or sensorineural hearing loss or vasculitis of the internal auditory artery or its cochlear branch. Hearing and/or vestibular involvement may occur acutely or insidiously at some point, and occurs in approximately one third of patients (25). Eye disease is a common problem,

affecting about 20% of patients at presentation and up to about 60% at some point during the course of the disease (24, 26, 27). Symptoms of nasal cartilage inflammation include congestion, crusting, rhinorrhoea, epistaxis, pain, redness and swelling of the bridge of the nose, and/or nasal pressure. Nasal chondritis is present in approximately 20% of patients at presentation and in 60% over the course of the disease (24). Cartilage destruction associated with sustained or recurrent episodes of inflammation may result in a characteristic saddle-shaped deformity of the nose, seen in about 10% of patients at presentation and in 25% during the course of the disease. Arthritis is seen at presentation in up to one-third of patients, and peripheral joint involvement eventually occurs in approximately 50%–70% (24, 28). There is no predilection for large or small joints. Cardiac involvement due to clinically significant aortic or mitral valve disease occurs in approximately 10% of patients (29). Renal involvement, as determined by biopsy or the presence of hematuria and/or proteinuria, remains unclear and is probably very rare, although there are series describing renal involvement in up to 23% of cases (30). Neurological manifestations are unusual in RP (31) and neurological involvement is presumed to be due to the presence of another associated disease, as it has rarely been documented in RP due to vasculitis phenomena (32). However, at that time there was no specific test to diagnose patients with antineutrophil cytoplasmic antibody-associated vasculitis. Laboratory blood tests are not used in the absence of a specific parameter for the diagnosis of RP. Elevations of ESR or C-reactive protein (CRP), and modest leukocytosis or thrombocytosis are associated with clinical and subclinical disease activity. The appearance of ANA and antiDNA positivity are not characteristic of RP and should raise suspicion of coexisting SLE (33). Antiphospholipid antibodies are associated with obstetric complications and thrombotic events, although they have also been associated with other symptoms, including chondritis. The association is not well known and studies are needed to establish it. The coexistence of RP and antiphospholipid syndrome (APS) is very uncommon and there is no convincing evidence regarding the association between them when APS occurs in RP. It seems to be more closely related to association with SLE (34). The cases discussed in this article did not present APS and the presence of APS is not detailed in the majority of the patients. Skin biopsy is reserved for situations where the patient does not have a multi-organ cartilaginous inflammatory syndrome or does not respond to glucocorticoids (5). Due to the rarity of RP, to date there are no standardised therapeutic guidelines on treatment, which means that therapy is tailored empirically according to disease activity and the nature of organ involvement (35). Glucocorticoids may decrease disease activity and reduce the frequency of recurrence (36, 37). Immunosuppressive agents such as azathioprine, methotrexate and cyclosporine may be given in non-responders or when a corticosteroid-sparing effect is needed (38–41). Although well-designed randomised clinical trials are lacking and the evidence for their use is mainly based on expert opinion, biologic agents are also showing a role in the therapeutic approach to these patients. However, data on rituximab

TABLE 3 Epidemiology and clinic of detailed reported cases.

Case no.	Ref.	Age, Sex	Race	Chondritis	Ocular inflammation	ENT manifestations (hypoacusis, vestibular syndrome, nasal discharge)	Seronegative polyarthritis	Cardiac involvement	Neurologic involvement	Haematologic involvement	Kidney involvement
1	8	38, F	W	Ears	Episcleritis	No	Large joints	Yes	No	Anemia	No
2	9	26, F	W	Yes	No	No	Yes	No	No	No	No
3	10	29, F	W	Ears	No	No	Yes	No	Headache, papilledema	Thrombocytopenia	Yes
4	10	34, F	W	Ears	No	No	No	No	No	No	No
5	11	55, F	U	Ears	No	Bilateral severe sensorineural hearing loss, tinnitus and vestibular syndrome	Large and small joints	U	No	No	Yes
6	12	62, F	W	Ears, nose, laryngeal and costal cartilage	Yes	Chronic sinusitis, hoarseness	Yes	No	No	No	No
7	13	75, F	W	Ears	Yes	Hearseness and right conductive hearing loss	Yes	No	No	No	No
8	14	32, F	U	Ears	No	No	Large and small joints	No	Ischemic stroke and multiple small infarcts	Anemia and thrombocytopenia	Yes
9	15	47, F	U	Ears	No	No	No	No	No	No	Yes
10	16	50, F	U	Arythenoid, nose and ears	No	Hearseness	No	U	U	Anemia	U
11	17	60, F	B	Ears and nose	Conjunctivitis	Audiovestibular dysfunction(left-sided sensorineural hearing deficit)	Yes	No	No	Anemia	Yes
12	18	46, F	U	Left ear	No	No	Yes	U	U	Thrombocytopenia and leukopenia	U

F, female; W, white; B, black; U, unknown; ENT, ear, nose and throat.

TABLE 4 Chronological relationship, complementary tests, treatment and evolution of the reported cases.

Case no.	Ref.	Temporal relationship debut SLE and PR	ANA	Anti-dsDNA	ESR (mm/h)	Skin/cartilage biopsy	Kidney biopsy	Treatment	Complications (saddle nose, airway stenosis)	Follow-up
1	8	U	Positive 1:800 speckled pattern	Positive	43	Yes	No	Oral prednisone 20 mg/daily	No	Lost
2	9	U	Positive	U	U	No	No	U	Saddle nose deformity	U
3	10	9 years before	Positive 1:800 rim pattern	Positive	Elevated	No	Class V: membranous nephritis	Corticosteroids for polycondritis	No	>10 years
4	10	U	Positive 1:128 homogenous pattern, positive LE cells	Negative	U	No	No	Topic treatment	No	U
5	11	4 years before	Yes	U	121	No	Subacute glomeronephritis	Oral prednisone	No	6 years
6	12	After	Yes Positive LE cells	U	90	Yes	No	Oral prednisone	Saddle nose deformity Respiratory distress (tracheostomy)	U
7	13	U	Positive 1:80	Positive	Normal	No	No	Oral prednisone	No	U
8	14	1 year later	Positive 1:1280	Positive	U	No	No	Oral prednisone (1 mg/kg), hydroxychloroquine, AAS, daily plasma (9 days) and rituximab (375 mg/m ² , 3 days)	No	Recovery from the neurological defects at 6 months
9	15	Simultaneous	Positive	Negative	Elevated	Yes	Class II: mesangial proliferative	Oral prednisone and hydroxychloroquine	No	U
10	16	Simultaneous	Positive	U	U	No	U	Oral prednisone 25 mg/day	No	U
11	17	Simultaneous	Positive	U	U	Yes	Yes	Oral prednisone	No	U
12	18	4 years before	Yes	Positive	U	No	U	Hydroxychloroquine and corticosteroids (disease outbreak)	U	U

ANA, antinuclear antibodies; anti-dsDNA, anti-dsDNA antibodies; APL, antiphospholipids antibodies; ESR, erythrocyte sedimentation rate; U, unknown.

administration are controversial and this agent is not recommended as a first-line biologic in the treatment of patients with RP (42).

Overlap syndrome between SLE and RP is extremely rare (<1%). Since the diagnosis of RP is based on clinical findings, it is crucial to maintain a high index of suspicion for RP in the presence of recurrent scleritis, audiovestibular involvement and chondritis in patients with SLE, especially those in the fourth-fifth decade of life. This case highlights the importance of early diagnosis and immediate therapeutic intervention, especially in situations of severe manifestations of RP, due to the sequelae damage that can result from severe and recurrent inflammation and deterioration in the patient's quality of life. The management of patients with 2 potentially aggressive diseases such as SLE and RP requires a treat-to-target strategy. The multidisciplinary approach and a very close follow-up will determine whether the treatment is adequate or whether dose adaptations, treatment changes or even non pharmacological interventions are required, e.g., assessment by a psychologist, control of anxiety or depression symptoms, indication of hearing aids, surgical interventions for aesthetic deformities or others. Undoubtedly, this work has some limitations, as it deals with a series of non-homogeneous cases. We have made an effort to compare them and to highlight the relevance of considering the diagnosis of relapsing polychondritis in patients with suggestive symptoms, despite the fact that SLE may also have similar chronic inflammatory signs. Specialists who treat patients with autoimmune diseases are well aware that no two patients are alike, and that the diseases can present in different ways. We also know that several of them can coexist in the same patient, a common finding in patients with immune-mediated diseases. The differentiating features of both diseases, SLE and RP, are what will lead to the suspicion of their coexistence. Only persistent study and updating of knowledge by specialists will allow suspicion, diagnosis and appropriate treatment of these patients. Under-diagnosis of one of the entities can have fatal consequences.

Patient perspective

A Story of Resilience: Living with Relapsing Polychondritis and Systemic Lupus Erythematosus.

The first symptoms of my illness started with tiredness, when it was seven o'clock in the evening I had to lie down and sleep, I didn't think anything of it, I put it down to work. The first episode of inflammation of the ears was with fever and a lot of pain for about 2 weeks, but since then I always had the feeling of having hard ears. After about a month, I began to have a severe headache every day, which was worse at night, as was the fever. As the days went by I had a very annoying noise in my left ear, and my eyes, alternately, became very red and very painful, I began to feel dizzy and had a very strange sensation when I walked. I went to my internal medicine specialist and was hospitalised, during which time I lost my hearing for about 8–10

days, but after treatment I recovered a little, which still allows me to hear with the help of hearing aids. For the first few months I needed a cane to walk as I had no balance. With the help of vestibular rehabilitation at home and in the hospital, as well as walking every day and doing yoga, I managed to improve partially. At first the episodes of red eyes were recurrent, with a lot of pain, accompanied by fever and headache quite often. Later I was diagnosed with lupus nephritis. The sun harms me a lot, it causes reddening of the skin and itching, I always wear sun protection.

Data availability statement

The original contributions presented in the study are included in the article/Supplementary Material, further inquiries can be directed to the corresponding author.

Ethics statement

Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

Author contributions

AB-M: Writing – review & editing, Writing – original draft. NN-N: Writing – review & editing. LG-N: Writing – original draft. MM-A: Writing – review & editing.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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