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Case Report Open Access

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Abstract

2YHUODS V\QGURPH LV D WHUP XVHG WR GHVFULEH WZR RU PRUH V\VWHPLF UKHXP HDFK GLVHDVH IXO¿OV LWV RZQ GLDJQRVWLF FULWHULD ,QÀDPPDWRU\ P\RSDWKLHV V risk of malignancy and interstitial lung disease. It may overlap with Systemic Sclerosis, Systemic Lupus Erythematosus, Mixed Connective Tissue Disease and less often Rheumatoid Arthritis.

Keywords: Amyopathic dermatomyositis; Rheumatoid arthritis; Overlap syndrome; Idiopathic in ammatory myopathy

Introduction

We presented a case of an atypical presentation of overlap syndrome with unremarkable labs ndings. is is a case of 30 y female who presented with violaceous eruption and edema on the upper eyelids (heliotrope rash). She visited emergency room few times and was diagnosed as an allergic skin reaction and treated with antihistamine medications without improvement of symptoms. Few months later she developed signi cant functional limitations marked by mechanic's hands, Gottron's papules, Gottron's sign, peryungal erythema with in ammatory arthritis and synovitis of the metacarpophalangeal joints, proximal interphalangeal joints, elbows and knees. Associated with fever episodes and alopecia without clinical evidence of muscle weakness. Laboratory data was signi cant for positive rheumatoid factor, anti-CCP antibodies, and normal CPK levels. Skin biopsy ndings were typical of dermatomyositis with super cial and deep perivascular lymphocytic in ltrate. Patient ful lled the criteria for both rheumatoid arthritis and amyopathic dermatomyositis. erapy with prednisone and methotrexate resulted in signi cant clinical improvement. Primary care physicians must be familiar with the clinical presentation of dermatomyositis for early, accurate diagnosis and appropriate clinical management. Prompt screening and evaluation for malignancy and interstitial lung disease are important to decrease morbidity and mortality.

Case Report

Overlap syndrome is a term used to describe two or more systemic rheumatic diseases identi ed in the same patient, in which each disease ful ls its own diagnostic criteria. In ammatory myopathies such as Dermatomyositis are associated with an increased risk of malignancy and interstitial lung disease. It may overlap with Systemic Sclerosis, Systemic Lupus Erythematosus, and Mixed Connective Tissue Disease and less o en Rheumatoid Arthritis. We presented a case of an atypical presentation of rheumatoid arthritis and amyopathic dermatomyositis in the same patient. e presence of both diseases simultaneously have been described in the literature only a handful of times, leading to the possibility of the existence of an overlap syndrome.

is is a case of 30-year-old Puerto Rican female, who had an unremarkable medical history and signi cant family history of mother with Systemic Lupus Erythematosus (SLE). She developed a violaceous erythema of the upper eyelids and periorbital edema (Figure 1).

Seven months later presented to the rheumatology clinic complaining of the former symptoms, marked functional disability, episodes of low grade fever, alopecia, arthralgia and bilateral in ammation of the hands. Physical examination of skin revealed edematous purple discoloration of periorbital tissue (heliotriope rash), periungual telangiectasias, erythematous papules on the extensor surface of the metacarpophalangeal (MCP) joints bilaterally (Gottron's papules), elbows, knees and ankles (Gottron's sign) and mechanic's hands was unable to make a st with both hands, swelling of the proximal interphalangeal joint (PIP) with some degree of sausage digits, synovial thickening of MCP, PIP, elbows and knees joints pain on exion of the knees without signs of muscle weakness.

Patient was initially treated with methotrexate (MTX) and Adalimumab for RA without improvement, and then changed to Etanercept. She requested a second opinion and Etanercept was discontinued. High dose prednisone was started with continuation of MTX, while new labs were available.

Laboratory results revealed (Table 1): WBC 4.4 g/dL, hemoglobin 8.9 g/dL, hematocrit 26.6%, blood urea nitrogen 4.0 g/dL, creatinine 0.4 g/dL, serum total protein 8.6 g/dL, sed rate 49, serum protein

Figure 1: Patient's face suggestive of Dermatomyositis heliotrope rash.

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electrophoresis M spike not observed, total globulin 5.3 g/dL. Le elbow skin biopsy showed super cial and deep perivascular lymphocytic in ammatory in ltrate with focal parakeratosis. e chest X- ray (Figure 2) revealed heart and pulmonary vasculature of normal appearance, essentially clear lungs. Hand x-ray (Figure 3) showed normal bone density and preserved articular spaces.

Test-Name	Results	Normal Range
CPK	168 U/L	30-170 U/L
ANTI-CCP	>250 units	0-19 units
ANA	Negative	<1:80 negative
RF	632.8 IU/ml	0.0-14.0 IU/ml
ANTI-JO-1	<0.20	<1.0 negative
ANTI-SSA	<0.20	<1.0 negative
ANTI-SSB	<0.20	<1.0 negative
ANTI-RNP	<0.20	<1.0 negative
ANTI-SM	<0.20	<1.0 negative
ANTI-DS-DNA	1 IU/ml	<5 IU/ml negative
AST (SGOT)	20	5-40 U/L
ALT (SGPT)	13	5-40 U/L
SED RATE	49	0-15 mm/h (Male)
		0-20 mm/h (Female)

Table 1: Laboratory results.

Figure 2: Patient's chest x-ray.

Figure 3: Patient's hand X-ray.

Azathioprine 100 mg PO daily was started and prednisone was tapper down. Also Hydroxychloroquine 200 mg PO BID was added. Pulmonary function tests (PFTs) and MDA5 were ordered. PFTs came back normal. Insurance company did not approve MDA5 antibody. Patient demonstrated clinical improvement (Figure 4), functional capacity and was able to return to her regular work.

Discussion

Overlap syndromes de ned as a patient who presents with multiple features of two are more well-de ned connective tissue disease (CTD) such as Systemic Lupus Erythematosus, Systemic Sclerosis Sjörgen's Syndrome, Polyarteritis Nodosa, and Rheumatoid Arthritis. In ammatory myopathies associated with overlap syndromes have been related to one or multiple connective tissue disorders with an incidence rate of 21% [1] More common in females than in males with a 9:1 ratio [1] about 11-40% of patients with dermatomyositis present as an overlap syndrome [2].

Dermatomyositis (DM) is an in ammatory myopathy disorder that includes cutaneous manifestations such as heliotrope rash: a red purple macular rash with periorbital edema, Gottron's papules and Gottron's sign: violaceous papules or macules involving the dorsal interphalangeal or metacarpophalangeal areas of the hands, elbow or knee joints that manifest in 70% of patients [3], symmetric proximal muscle weakness and elevated skeletal muscle enzymes rst described by Bohan and Peter's criteria in 1975 (Table 2) [4]. Also can present with periungal abnormalities, calcinosis cutis, psoriasiform changes in scalp, raynaud's phenomenon and mechanic's hands: hyperkeratotic, ssured skin on the lateral and palmar aspects of the ngers, seen in antisynthetase syndrome up to 30% of patients with DM [5]. e etiology remains unknown; few studies reported an association with histocompatibility antigens, autoimmunity and environmental agents [6]. e annual incidence of DM is approximately 1 per 100,000 in the general population [7]. Amyopathic dermatomyositis (ADM) also known as dermatomyositis sine myositis is a subtype of DM characterized by biopsy-con rmed hallmark skin rash of classic DM for 6 months without evidence of muscle weakness or elevated muscle enzymes as proposed by Sontheimer [8] is entity has been controversial because it does not exactly meets the criteria described by Bohan and Peter. e estimated annual incidence of this subtype was 0.2 per 100,000 persons in a study made in Olmstead County, Minnesota [4] Comprises 10 to 30% of DM cases [7].

Rheumatoid arthritis (RA) is another systemic autoimmune disease of unknown cause that presents with in ammatory polyarthritis that can lead to deformity through the laxity of ligaments and tendons

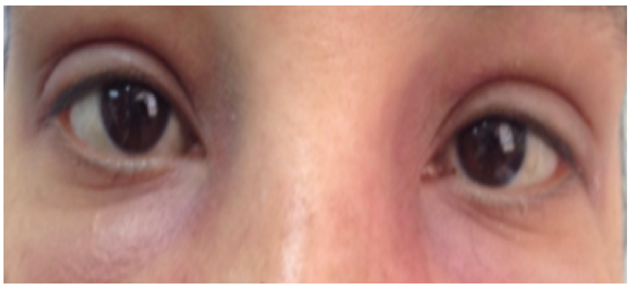

Figure 4: Patient's face while on treatment with Prednisone and Methotrexate.

Criteria	Description
A	3UR[LPDO DQG V\PPHWULFDO PXVFOH ZHDNQHVV RI WKH SHOYLF DQG VFRSXODU JLUG months, with or without dysphagia or involvement of respiratory muscles
B	Elevation of the serum levels of skeletal muscle enzymes: creatine kinase, aspartate aminotransferase, lactate dehydrogenase andaodolase
C	Electromyography characteristic of myopathy (short and small motor unites ibrillation, positive pointy waves, insertional irritability and UHSHWLWLYH KLJK IUHTXHQF\ ¿ULQJ
D	0XVFOH ELRSV\ VKRZLQJ QHFURVLV SKDJRF\WRVLV
E	Typical Cutaneous changes:
	1. Heliotrope rash with periorbital oedema and violaceoue erythema
	2. Gottron's sign: vasculitis in the elbow, metacarpophalangeal and proximal interphalangeal joints
Polymyositis	'H¿QH DOO RI $ '
	2. Probable - any three of A-D
	3 Possible - any two of A-D
Dermatomyositis	'H¿QH (SOXV DQG WKUHH RI$ '
	2. Probable - E plus and two of A-D
	3 Possible - E plus and one of A-D

Table 2: %RKDQ DQG 3HWHU FODVVL¿FDWLRQ IRU SRO\P\RVLWLV DQG GHUPDWRP

Joint istribution (0-5)	
1 large Joint	0
2-10 Large Joints	1
1-3 small Joints (Large joints not counted)	2
4-10 small Joints (Large joints not counted)	3
>10 Joints (at least one small Joint)	5
Serology (0-3)	
Negative RF and negative ACPA	0
Low Positive RF or Low positive ACPA	2
High Positive RF or high positive ACPA	3
SYMPTOM DURATION	
<6 Weeks	0
• :HHNV	1
ACUTE PHASE REACTANTS (0-1)	
Normal CRP and normal ESR	0
Abnormal CRP or abnormal ESR	1

Table 3: 2010 ACR/EULAR criteria for RA.

and destruction of bone and cartilage through the process of erosion. Classically afected joints include proximal interphalangeal and metacarpophalangeal joints of the hands and feet, and the wrist in a symmetric distribution. RA afects approximately 1% of the population worldwide, with a peak incident between ages 30 and 60 years. Women afected 2-3 times more ofen than men. For diagnosis, patient must have infammatory arthritis involving ≥3 joints, positive RF and/or anti-CCP, disease duration of >6 weeks, and elevated CRP or ESR which need a minimum of 6 out of 10 points for diagnosis (Table 3) [9].

Patient was found to have heliotrope rash, Gottron's papules, Gottron's sign, and arthralgias, periungal erythemas that are characteristic of Dermatomyositis. But, no proximal muscle weakness present, with normal CPK levels, and a biopsy that revealed superfcial and deep perivascular lymphocytic infammatory infltrate with focal parakeratosis that is consistent with DM. Based on the criteria proposed by Bohan and Peter and revised by Sontheimer this patient's clinical sign and symptoms are most consistent with amyopathic dermatomyositis due to the presence of cutaneous manifestations without elevation of muscle enzymes and muscle weakness. Also presented with swelling of the proximal interphalangeal joint (PIP) in both hands with some degree of sausage digits and synovitis, synovial thickening of MCP, PIP, elbows and knees joints, high positive titers for rheumatoid factor and anti-CCP antibodies, with elevated infammatory markers having 8 out of 10 points, fulflling the 2010 ACR/EULAR criteria for RA (Table 3) [9,10].

Conclusion

RA and ADM represent an unusual case of overlap syndrome. It is important to recognize Amyopathic DM as a subtype of DM as it represents an increased risk of interstitial lung disease and malignancy. Interstitial lung disease is an important complication of at least 10% of cases and may be associated with diaphragmatic and chest wall muscle weakness. Melanoma diferentiation-associated gene 5 (MDA5) antibodies detection is associated with rapidly progressive interstitial lung disease and poor prognosis. Early detection with this test will help reduce the risk of progressive pulmonary insufciency and death. Te incidence of cancer has been reported in the literature from 5 to 7-fold compared with the general population, particularly in the frst 2 years afer diagnosis Te most common types of malignancies are ovarian and gastric cancer, and lymphoma with only a few reported cases in the literature, this case report provides further insight into this rare presentation considerably high morbidity and mortality if not promptly recognized in a timely fashion.

Recommendations

1. Recommended PFTs, chest X-ray and CT scan if symptoms like dyspnea and persistent non-productive cough are associated.

2. Patients with MDA-5 antibody in DM have higher risk of ILD, including rapidly progressive presentation and high mortality.

3. Screening for internal organ malignancy.

4. Early Diagnosis and treatment of RA to prevent irreversible joint damage.

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