International Journal of Research in Medical Sciences
Sanjeev CM et al. Int J Res Med Sci. 2025 Sep;13(9):3886-3889
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20252815

Case Report

Dermatomyositis associated with malignancy: a case highlighting paraneoplastic syndrome

Chavan Mayuresh Sanjeev*, Arun Krishnan, Jaykaran

Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India

Received: 08 August 2025
Revised: 23 August 2025
Accepted: 23 August 2025

***Correspondence:**
Dr. Chavan Mayuresh Sanjeev,
E-mail: mayureshchavan17@gmail.com

ABSTRACT

Dermatomyositis (DM) is a rare idiopathic inflammatory myopathy characterized by distinctive cutaneous manifestations and progressive, symmetric proximal muscle weakness. It affects both adults and children, with variable clinical presentations and disease severity. The pathogenesis involves autoimmune mechanisms, including complement-mediated microangiopathy, with a potential association with malignancies in adults**.** In this case study, we report a case of classical DM with incidence of malignancy. Timely diagnosis and administration of steroid led to better prognosis of the patient.

Keywords: Dermatomyositis, Malignancy, Paraneoplastic, Syndrome, Inflammatory

INTRODUCTION

Dermatomyositis (DM) is the rare form of autoimmune inflammatory myopathy characterized by progressive and symmetric proximal muscle weakness and the presence of typical skin manifestations. It occurs in adults and children but is clinically more important in adults because of its high paraneoplastic relationship with 15-25% of all cases having a paraneoplastic relationship. The pathogenesis is immune-mediated microangiopathy that may include skin and muscle; it is frequently associated with elevated levels of muscle enzymes, perifascicular atrophy on biopsy, and MRI or EMG manifestation of myositis. Hallmark signs (heliotrope rash, Gottron papules, and nailfold telangiectasia) help diagnosis though some cases may be atypical or even amyopathic.[1] New evidence, such as the one reported in the present study, highlights the potential of imaging and PET-CT toward the detection of concealed malignancies, especially GI and pancreatic cancer. Recognition of paraneoplastic DM is an emergency thus early treatment to avoid progression leading to other complications such as corticosteroid therapy and malignancy focused treatment is essential as this improves the prognosis.[2] As such, a multidisciplinary diagnostic-therapeutic approach is a necessity to achieve desirable outcomes in patients.

The analyzed case report brings to attention DM as a rare autoimmune inflammatory myopathy with a close connection to malignancy. Literature has reported that 15-25% of the cases of adult-onset DM are associated with cancer; common diseases associated include ovarian, lung, gastrointestinal cancer, pancreatic, and breast cancer.[3] Paraneoplastic DM also has acute onset with severe skin findings, muscle proximal weakness, and lack of response to immunosuppression is a predictive sign of occult malignancies. Among the diagnostic procedures, there are muscle enzyme assays, EMG, MRI, and biopsy, and PET-CT is useful in detecting malignancy. Corticosteroids are used as first-line treatment; they are enhanced by the use of immunosuppressants or IVIG in cases which fail to respond to initial treatment.[4] Cross-disciplinary screening and treatment methodologies have much better outcomes on the patients.

CASE REPORT

58 years old female patient who is a known case of cervical carcinoma presented to us with complains of difficulty in standing up from sitting position since 1 month.

Weakness of bilateral lower limb since 1 month which was insidious in onset progressive in nature. initially patient had difficulty in sitting on a bike, climbing stairs later which progressed to inability to standing up from chair. Proximal muscle weakness was more than the distal muscle weakness.

Weakness of bilateral upper limb with was insidious onset progressive more proximally than in distal muscles

Vital signs in the emergency department were normal. An examination revealed a chronically ill-appearing woman in no acute distress.

Diffuse non scarring alopecia was present.

Raised hyperpigmented papules were found with areas of depigmentation on face neck upper back hyperpigmented papules were seen on upper eyelids.

Hyper pigmented flat-topped papules and plaques were found on MCP an IP joints (Gottrons Papules) nail fold telangiectasia.

Proximal muscle tenderness was found.

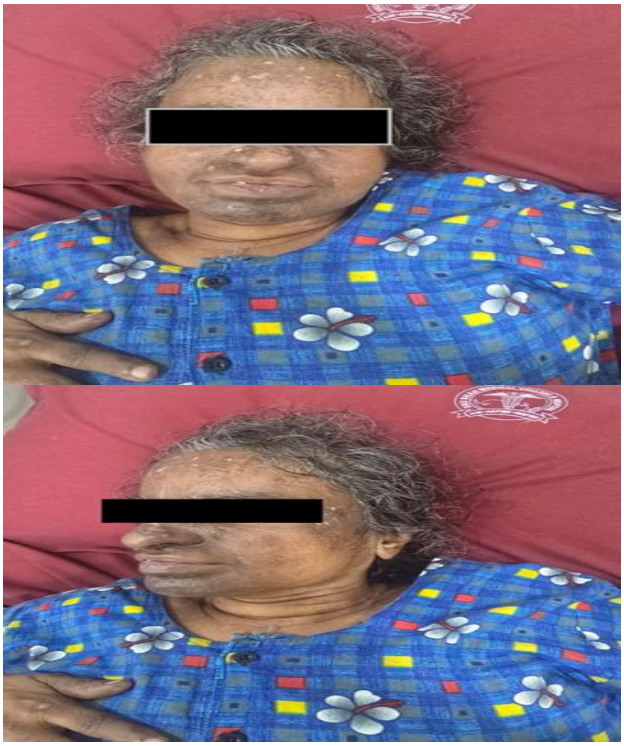

Figure 1: Clinical presentation of DM showing hyperpigmented facial rash and periorbital involvement in a 58-year-old female patient.

Nervous system examination

Higher mental function were found to be normal tone was normal.

Power was 2/5 in proximal muscles of all four limbs and 4/5 in distal muscles of all four limbs.

She had no ocular, facial or bulbar weakness or sensory deficits. Her deep tendon reflexes were trace at the biceps, brachioradialis and Achilles tendons. His patellar reflexes were 3+. The remainder of the exam was unremarkable.

All basic investigations were done. CBC RFT were found to be normal.

AST was 132.05 (normal<31), ALT was 109.40 (normal <34), Albumin was 3.30, Globulin was 2.41 CPK was 177, CPK MB was 390.9, ESR was 60, CPK total was 2884.5, LDH was 914, ferritin was 443.1 Urine routine showed ++ proteinuria.

ANA IIF was positive +++ speckled pattern ANA profile showed Mi 2 positive.

Skin Biopsy was done which showed focal vacuolar interface changes. Epidermal loss of rete ridges. Dermal pigment laden macrophages, increase collagen upto subcutis and the perivascular mononuclear cell inflammation.

EMG showed the myogenic pattern suggestive of the myositis.

MRI thigh showed Linear T2 /STIR hyperintensities involving all muscles of the thigh suggestive of myostis. Edematous changes in overlying subcutaneous plane as well predominantly along the anterolateral and posterior aspect of thigh.

Muscle biopsy was done which showed perifasicular atrophy altered muscle fibers with nuclear internalization. There is focal necrosis of muscle fibers with fragmentation and diffuse lymphohistiocytic inflammation (myositis). Features suggestive of myositis.

PET scan showed SSTR expressing enhancing lesion in head and uncinate process of pancreas consistent with neuroendocrine tumor. Diffuse tracer avidity in skeletal muscles possibly myositis.

Based on the above findings, a diagnosis of DM was made. Case was discussed with rheumatologist and neurologist was started on intravenous steroids and supportive measures.

Patient improved symptomatically, muscle tenderness improved within month. At present, she is orally administered 50 mg prednisolone therapy for maintenance therapy and still being followed up.

DISCUSSION

DM is an autoimmune inflammatory myopathy with hallmark cutaneous features and proximal muscle weakness. The paraneoplastic form of DM is of particular clinical significance, given its potential to serve as an early clue to an underlying malignancy.[5] Recognizing DM as a possible paraneoplastic syndrome is critical, especially in adult patients over the age of 40 or those presenting with atypical features or poor response to conventional therapy.

Numerous studies have established a strong association between DM and malignancies; with an estimated 15-25% of adult-onset DM cases linked to cancer. The most commonly associated malignancies include ovarian, lung, gastrointestinal (especially gastric and colorectal), pancreatic, and breast cancers, as well as non-Hodgkin lymphomas.[6] The risk appears to be highest within the first year of DM diagnosis but may persist for up to five years or longer.

Clinically, paraneoplastic DM (PNDM) may present identically to idiopathic DM, although certain features-such as abrupt onset, severe cutaneous findings with mild or absent myopathy (amyopathic DM), resistance to immunosuppression, and systemic symptoms like weight loss or fatigue-should raise suspicion for an underlying neoplasm.[7]

Our patient presented with classic cutaneous features of DM, such as heliotropo rash and Gottron's papules, along with progressive proximal muscle weakness. Creatine kinase (CK) and other muscle enzymes were elevated, supporting muscle involvement. Further confirmation was obtained via electromyography (EMG), muscle biopsy, or MRI findings.

In our case, the diagnosis of neuroendocrine tumor of pancreas shortly after the appearance of classic DM features, including proximal muscle weakness, Gottron papules strongly supports a paraneoplastic process. This clinical course is well-documented and highlights the importance of malignancy-directed therapy in managing paraneoplastic autoimmune syndromes. Immunosuppressive treatment alone is often insufficient or only partially effective in such cases. From a clinical standpoint, patients diagnosed with DM -particularly older adults or those with risk factors-should undergo thorough malignancy screening at baseline. Recommended evaluation includes comprehensive history and physical examination, age-appropriate cancer screenings (e.g., mammography, colonoscopy and Pap smear), cross-sectional imaging (CT chest/abdomen/pelvis or PET-CT), and targeted testing based on clinical context or serologic markers.

Treatment typically involves high-dose corticosteroids as first-line therapy, often combined with steroid- sparing agents such as methotrexate, azathioprine, or mycophenolate mofetil. In refractory or rapidly progressive cases, IVIG or rituximab may be used.[8] The patient in this report responded well to steroids, with marked improvement in muscle strength and resolution of cutaneous lesions.

This case reinforces the importance of a multidisciplinary approach to DM management, involving dermatology, rheumatology, neurology, and, when needed, oncology. Regular monitoring for disease activity and potential complications, including interstitial lung disease and calcinosis, is critical.

The case study showed typical DM signs and symptoms that include a heliotrope rash, Gottron papules, nailfold telangiectasias, and proximal muscle weakness, which were confirmed with biopsy, EMG, and MRI that showed evidence of perifascicular atrophy and diffuse inflammatory myositis.[9] High values of enzymes that include CK (2884.5) and LDH (914) confirmed the presence of active myopathy. PET-CT demonstrated a pancreatic neuroendocrine tumor which is the most plausible paraneoplastic cause. These data are in agreement with previous literature where 15-25 percent of adult DM patients developed malignancy most notably in the gastrointestinal and the pancreatic tract.[10] Clinical improvement was noted with steroid therapy, just like previous cases; early screening with imaging and multidisciplinary approach in improving prognosis is important

CONCLUSION

In conclusion, DM is a systemic autoimmune condition with a broad clinical spectrum. Prompt diagnosis and individualized treatment are essential for improving outcomes and minimizing long-term morbidity.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Gerami P, Schope JM, McDonald L, Walling HW, Sontheimer RD. A systematic review of adult- onset clinically amyopathic dermatomyositis (dermatomyositis sine myositis): a missing link within the spectrum of the idiopathic inflammatory myopathies. J Am Acad Dermatol. 2006;54(4):597-613.
2. Dalakas MC. Polymyositis, dermatomyositis and inclusion-body myositis. N Engl J Med. 1991;325(21):1487-98.
3. Petri MH, Satoh M, Martin-Marquez BT, Vargas-Ramírez R, Jara LJ, Saavedra MA, et al. Implications in the difference of anti-Mi-2 and -p155/140 autoantibody prevalence in two dermatomyositis cohorts from Mexico City and Guadalajara. Arthritis Res Ther. 2013;15(2):R48.
4. Salgueiro C, Poblete MJ, Robles-Silva C, Abarzúa Á,

Vera-Kellet C. Trichoscopic, oral, and periungual fold findings as activity and damage markers in dermatomyositis patients and their correlation with myositis antibodies. Arch Dermatol Res. 2023;315(6):1603-13.
5. Al Kanaan R, Nazer II, Al Nasser M. A Case of Juvenile Dermatomyositis Presenting with Inverse Gottron's Papules: A Case Report. Case Rep Dermatol. 2025;17(1):80-5.
6. Liluashvili S, Pataraia S, Kituashvili T. Dermoscopic features of connective tissue disease. Dermatol Rev. 2024;111(2):148-52.
7. Castillo RL, Femia AN. Covert clues: the non-hallmark cutaneous manifestations of dermatomyositis. Ann Translat Med. 2021;9(5):436.
8. McAtee CL, Lubega J, Underbrink K, Curry K, Msaouel P, Barrow M, et al. Association of rituximab use with adverse events in children, adolescents, and young adults. JAMA Network Open. 2021;4(2):e2036321.
9. Ross L, McKelvie P, Reardon K, Wong H, Wicks I, Day J. Muscle biopsy practices in the evaluation of neuromuscular disease: a systematic literature review. Neuropathol Appl Neurobiol. 2023;49(1):e12888.
10. Wu MJ, Liao WA, Lin PY, Sun YT. Muscle biopsy: a requirement for precision medicine in adult-onset myopathy. J Clin Med. 2022;11(6):1580.

Cite this article as: Sanjeev CM, Krishnan A, Jaykaran. Dermatomyositis associated with malignancy: a case highlighting paraneoplastic syndrome. Int J Res Med Sci 2025;13:3886-9.