

Dermatomyositis and colorectal cancer: Case report and literature review

Mario Andrés Jaramillo-Santos,¹  Andrés Sánchez-Gil,²  Andrés Valencia-Uribe,^{3*}  Lázaro Arango-Molano.⁴ 

OPEN ACCESS

Citation:

Jaramillo-Santos MA, Andrés Sánchez-Gil A, Valencia-Uribe A, Arango-Molano L. Dermatomyositis and colorectal cancer: Case report and literature review. *Rev Colomb Gastroenterol.* 2021;36(Supl.1):91-97. <https://doi.org/10.22516/25007440.615>

¹ MD, Internist, gastroenterologist. Professor, Universidad de Caldas, Unión de Cirujanos S. A. S. Manizales, Colombia.

² MD, general surgeon, clinical gastroenterologist, gastrointestinal surgeon. Professor, Universidad de Caldas, Unión de Cirujanos S. A. S. Manizales, Colombia.

³ MD, general surgeon, clinical and surgical gastroenterology *fellow*, Universidad de Caldas, Unión de Cirujanos S. A. S. Manizales, Colombia.

⁴ MD, general surgeon, clinical gastroenterologist, gastrointestinal surgeon. Coordinator, clinical and surgical gastroenterology postgraduate program, Universidad de Caldas. Unión de Cirujanos S. A. S. Manizales, Colombia.

*Correspondence: Andrés Valencia-Uribe.
andres.valenciau@ucaldas.edu.co

Received: 10/07/20

Accepted: 22/02/21



Abstract

Patients with dermatomyositis are more likely to have an underlying malignancy, although the exact cause of this association is unknown. There are multiple possible anatomical sites, including ovaries, breasts, stomach, colorectum, blood, lungs, and prostate.

We present the case of a 58-year-old woman who during abnormal weight loss study showed severe muscle weakness and skin alterations, associated with finding of adenocarcinoma of the transverse colon.

Keywords

Dermatomyositis; Malignancy; Colorectal cancer; Paraneoplastic syndrome.

INTRODUCTION

Dermatomyositis is a widely studied collagen disease characterized by muscle weakness and associated with skin lesions. There is a higher frequency of malignancy in patients with dermatomyositis compared to the general population. Dermatomyositis frequently presents as a paraneoplastic syndrome, and the high rates of cancer detection following its diagnosis make the screening of cancer necessary taking into account the age group and clinical presentation of each patient (1-3). Additionally, dermatomyositis has been described as a paraneoplastic manifestation of gastrointestinal cancer.

The cancers most commonly associated with dermatomyositis are ovarian, pulmonary, breast, stomach, colorectal, pancreatic and non-Hodgkin's lymphoma (3-6).

CLINICAL CASE

This is the case of a 58-year-old woman without any relevant history of disease and a family history of colon cancer (her sister was diagnosed with it during the fifth decade of life) who experienced an abnormal weight loss of 6 kg during the last six months associated with scanty hematochezia and occasional diarrhea episodes. Three polyps were observed on a total colonoscopy: a 7 mm polyp in

the ascending colon (biopsy report: villous adenoma with low-grade dysplasia) (**Figure 1**), a 50 mm sessile polyp in the hepatic flexure (biopsy report: villous adenoma with low-grade dysplasia) (**Figure 2**) and a 30 mm polyp in the upper rectum (biopsy report: adenoma with low-grade dysplasia) (**Figure 3**); normal results in all 3 cell lines of the complete blood count test were reported. Endoscopic polypectomy of the rectal polyp (piecemeal resection) was performed using a hot loop, which was then classified as tubulovillous adenoma with low-grade dysplasia according to the histopathology report.

One month later, and provided that the endoscopic resection of the polyp located in the hepatic flexure was impos-

sible, the patient underwent a right hemicolectomy and a ileotransversostomy. The following findings were described in the histopathology report of the surgical specimens resected during these procedures: 2 tubular adenomas with focal high grade dysplasia without pedicle involvement, resection margins free of lesion and absence of lymph node involvement. Two months later, in addition to postsurgical changes, a follow-up total colonoscopy allowed identifying a 40 x 20 mm sessile polyp in the transverse colon immediately distal to the anastomosis (biopsy report: villous adenoma with low-grade dysplasia) (**Figure 4**); due to this finding, a new surgery consisting of colectomy extension, an omentectomy and a new ileocolic anastomosis was carried



Figure 1. 7 mm sessile polyp in the ascending colon.



Figure 2. 50 mm sessile polyp in the hepatic flexure of the colon.



Figure 3. 30 mm polyp in the upper rectum.



Figure 4. 40 x 20 mm sessile polyp in the transverse colon.

out (histopathology report: tubulovillous adenoma with high-grade dysplasia and *in situ* adenocarcinoma immediately distal to the previous anastomosis, with no signs of infiltration, free resection margins, and no lymph node or greater omentum involvement. Six months later, a 4 mm polyp in the upper rectum was observed in a new ileocolonoscopy, no other findings were reported; the polyp was resected using a cold loop (pathology report: tubulovillous adenoma with low-grade dysplasia) (Figure 5).

Four months before undergoing the first surgery, erythematous skin macules appeared on the patient's chest, which became more intense and larger and extended to the upper back, the lower back, the upper limbs and the face (Figures 6-11). Two months later she developed generalized muscle weakness experiencing falls while standing or walking and being unable to get up, together with edema and distal pain in the upper and lower limbs. Initial total serum creatine kinase (CK) levels were 2028 $\mu\text{mol/L}$, however after the colectomy extension, a progressive decrease to 1800 and 1028 $\mu\text{mol/L}$, together with symptomatology improvement, was evidenced. Chronic interface dermatitis with vacuolar changes compatible with dermatomyositis was reported in the skin biopsies of the right pectoral region and right arm. Based on these clinical, laboratory and histopathological findings and behaviors, it was concluded that the patient had dermatomyositis as a paraneoplastic manifestation of colon cancer. Tumor markers (Ca 19.9 and serum CEA) were normal, and no signs suggestive of

second malignancy were evidenced in both a CT scan of the chest a CT scan of the abdomen and pelvis. Currently, the patient is being monitored by the rheumatology and dermatology services; also, she is undergoing systemic steroid treatment and has reported the resolution of muscular symptoms and a significant improvement of skin lesions (Figure 12).

DISCUSSION

Dermatomyositis belongs to a heterogeneous group of connective tissue diseases known as idiopathic inflammatory myopathy (7). Its clinical manifestations include proximal musculoskeletal weakness and inflammation, as well as skin manifestations. An incidence of 5 to 10 cases per 1 million inhabitants, being more frequent in women (3:1 ratio), has been described in the in the United States. Its peak incidence occurs between 40 and 60 years of age (8). The association of dermatomyositis and malignancy was first established in 1916 in a patient with stomach cancer. Since then, multiple retrospective case series have reported this association in approximately 24% of cases (9).

The etiology of cancer-related dermatomyositis remains unknown, although several hypotheses have been proposed, such as the release of bioactive components that generate immune reactions in muscle fibers and the skin (10). It has been found that muscle fibers affected by autoimmune diseases overexpress specific autoantigens and that this



Figure 5. 4 mm polyp in the upper rectum.



Figure 6. Bilateral eyelid erythema and facial erythema (forehead, glabella and cheeks).



Figure 7. Erythematous plaques on the posterior cervical region, upper back and shoulders that constitute the classic “shawl sign” or capillary erythema sign.



Figure 8. Erythematous plaques extending to the lower back.



Figure 9. Violaceous erythematous plaques on the anterior triangle of the neck that extend to the chest.

overexpression is exhibited by tumor cells and regenerating myoblasts, which indicates that antigens expressed by both types of cells are similar (11). It has been proposed that there is a crossed immune response against regenerating

muscle cells, which leads to the development of dermatomyositis (12, 13).

In 1975, Boham & Peter (14) established the following diagnostic criteria for dermatomyositis: muscle weakness, high muscle enzymes, abnormal electromyography findings compatible with myopathy, compatible muscle or skin biopsy pathology report, and skin manifestations. Dermatomyositis diagnosis is made when the characteristic skin lesions are present and they are associated with 3 or more of the remaining criteria. In addition, when there is no muscle weakness, amyopathic dermatomyositis, which is also part of the same spectrum of paraneoplastic syndrome, is diagnosed (16, 17).

Leatham et al. (18), in a retrospective study conducted in 400 patients with dermatomyositis in the United States, reported that malignancy was found in 15.8% of cases, distributed as follows: breast cancer (24.5%), hematologic cancer (17%), colorectal cancer (9.4%) and prostate cancer (9.4%). Hill et al (4) found an association (in decreasing order of frequency) between the following types of cancer and dermatomyositis: ovarian, lung, stomach, colorectal, pancreatic and non-Hodgkin’s lymphoma. Sigurgeirsson



Figure 10. Violaceous erythematous plaques on the anterior aspect of the shoulder and arms.



Figure 11. Distal involvement of the upper limbs caused by violaceous erythematous plaques affecting the dorsum of the hands.

et al (19), in a case series conducted in 750 patients with dermatomyositis and polymyositis, reported that the most frequent cancers were colorectal and lung cancer. In Japan, Hatada et al (20) found that gastric cancer was the most frequent malignancy in patients with dermatomyositis (25.4%). In all case series, cancers of the digestive tract are among the most frequent malignancies in patients with this condition. In addition, several studies have reported that people with dermatomyositis have a higher risk of developing colorectal cancer (4, 19, 21-23).

There is ambiguity in the existing literature regarding the presence of clinical or laboratory and imaging predictors of malignancy in patients with dermatomyositis. In this sense, the presence of interstitial pneumonitis, elevated serum CK levels, skin necrosis (ulcers), poikiloderma, erythrocyte sedimentation rate (ESR) > 40 mm/h, elevated C-reactive protein (CRP) levels, hypoalbuminemia and age > 50 years have been proposed as clinical predictors (17, 24-29).

The treatment of patients with dermatomyositis and cancer is similar to that of cancer patients in general, pro-



Figure 12. Improvement of lesion on the cervical region, the upper back and the lower back.

vided that the cancer-dermatomyositis association does not imply any changes in the management of dermatomyositis. However, it has been reported that patients with dermatomyositis and malignancy tend to have a worse oncologic prognosis, since advanced stages are present at diagnosis and optimal resolution is not always possible (17). Successful surgical management usually leads to complete resolution of dermatomyositis symptoms (30, 31). Early diagnosis of malignancy in these patients is paramount to achieve a better prognosis.

Most of dermatomyositis with colorectal cancer cases reported have affected women (63 %), and adenocarcinoma is the most frequent histological finding in dermatomyositis (96.3 %). In 77.7% of patients, dermatomyositis manifested before colorectal cancer with elevated CK levels. Immediate improvement of symptoms after surgery occurs in half of patients, in the remaining, improvement occurs within the first months after undergoing the surgical procedure (31, 32, 33).

In the case reported here, dermatomyositis symptoms occurred a few months before the transverse colon carcinoma was found. Resolution of muscle symptoms and significant improvement of skin lesions took place several weeks after the lesion was surgically resected.

CONCLUSIONS

The exact cause of the association between dermatomyositis and the presence of malignancy is unknown. The presence of similar autoantigens in neoplastic, musculoskeletal and cutaneous cells has been suggested. Up to a quarter of dermatomyositis cases are associated, during the course of the disease, with the presence of cancer. At the time of diagnosis, patients with associated malignancy are already developing it, and generally the latter has a slow course of disease. When a patient is diagnosed with dermatomyositis, ruling out the presence of underlying malignancy, ideally in early stages, is of utmost importance to carry out interventions that positively contribute to the achievement of a better prognosis. Generally, after successful management (complete surgical resection) of the neoplastic lesion, dermatomyositis symptoms resolve; however, in case of tumor recurrence or metastatic cancer, they usually reappear.

The digestive tract is among the main sites of potential primary tumor origin, and within the digestive tract, stomach and colorectal cancer are the neoplasms most commonly associated with dermatomyositis. Finally, the evaluation of these patients must focus on their risk factors and associated clinical manifestations.

REFERENCES

1. Zantos D, Zhang Y, Felson D. The overall and temporal association of cancer with polymyositis and dermatomyositis. *J Rheumatol*. 1994;21(10):1855-9.
2. Barnes BE, Mawr B. Dermatomyositis and malignancy. A review of the literature. *Ann Intern Med*. 1976;84(1):68-76. <https://doi.org/10.7326/0003-4819-84-1-68>
3. Callen JP. Relationship of cancer to inflammatory muscle diseases. Dermatomyositis, polymyositis, and inclusion body myositis. *Rheum Dis Clin North Am*. 1994;20(4):943-53.
4. Hill CL, Zhang Y, Sigurgeirsson B, Pukkala E, Mellemkjaer L, Airio A, Evans SR, Felson DT. Frequency of specific cancer types in dermatomyositis and polymyositis: a population-based study. *Lancet*. 2001;357(9250):96-100. [https://doi.org/10.1016/S0140-6736\(00\)03540-6](https://doi.org/10.1016/S0140-6736(00)03540-6)
5. Callen JP. Relation between dermatomyositis and polymyositis and cancer. *Lancet*. 2001 Jan 13;357(9250):85-6. [https://doi.org/10.1016/S0140-6736\(00\)03535-2](https://doi.org/10.1016/S0140-6736(00)03535-2)
6. Maddison P. Cancer types in dermatomyositis and polymyositis. *Lancet*. 2001 May 5;357(9266):1443. [https://doi.org/10.1016/S0140-6736\(00\)04587-6](https://doi.org/10.1016/S0140-6736(00)04587-6)
7. Callen JP. Dermatomyositis. *Lancet*. 2000;355(9197):53-7. [https://doi.org/10.1016/S0140-6736\(99\)05157-0](https://doi.org/10.1016/S0140-6736(99)05157-0)
8. Kamiyama H, Niwa K, Ishiyama S, Takahashi M, Kojima Y, Goto M, Tomiki Y, Higashihara Y, Sakamoto K. Ascending Colon Cancer Associated with Dermatomyositis Which Was Cured after Colon Resection. *Case Rep Gastroenterol*. 2016;10(2):338-43. <https://doi.org/10.1159/000447289>
9. Zahr ZA, Baer AN. Malignancy in myositis. *Curr Rheumatol Rep*. 2011;13(3):208-15. <https://doi.org/10.1007/s11926-011-0169-7>
10. Wang J, Guo G, Chen G, Wu B, Lu L, Bao L. Meta-analysis of the association of dermatomyositis and polymyositis with cancer. *Br J Dermatol*. 2013;169(4):838-47. <https://doi.org/10.1111/bjd.12564>
11. Casciola-Rosen L, Nagaraju K, Plotz P, Wang K, Levine S, Gabrielson E, Corse A, Rosen A. Enhanced autoantigen expression in regenerating muscle cells in idiopathic inflammatory myopathy. *J Exp Med*. 2005;201(4):591-601. <https://doi.org/10.1084/jem.20041367>
12. Levine SM. Cancer and myositis: new insights into an old association. *Curr Opin Rheumatol*. 2006;18(6):620-4. <https://doi.org/10.1097/01.bor.0000245721.02512.77>
13. Zampieri S, Valente M, Adami N, Biral D, Ghirardello A, Rampudda ME, Vecchiato M, Sarzo G, Corbianco S, Kern H, Carraro U, Bassetto F, Merigliano S, Doria A. Polymyositis, dermatomyositis and malignancy: a further intriguing link. *Autoimmun Rev*. 2010;9(6):449-53. <https://doi.org/10.1016/j.autrev.2009.12.005>

14. Bohan A, Peter JB. Polymyositis and dermatomyositis (first of two parts). *N Engl J Med*. 1975;292(7):344-7. <https://doi.org/10.1056/NEJM197502132920706>
15. Dalakas MC, Hohlfield R. Polymyositis and dermatomyositis. *Lancet*. 2003;362(9388):971-82. [https://doi.org/10.1016/S0140-6736\(03\)14368-1](https://doi.org/10.1016/S0140-6736(03)14368-1)
16. Fung WK, Chan HL, Lam WM. Amyopathic dermatomyositis in Hong Kong -- association with nasopharyngeal carcinoma. *Int J Dermatol*. 1998;37(9):659-63. <https://doi.org/10.1046/j.1365-4362.1998.00453.x>
17. Wakata N, Kurihara T, Saito E, Kinoshita M. Polymyositis and dermatomyositis associated with malignancy: a 30-year retrospective study. *Int J Dermatol*. 2002;41(11):729-34. <https://doi.org/10.1046/j.1365-4362.2002.01648.x>
18. Leatham H, Schadt C, Chisolm S, Fretwell D, Chung L, Callen JP, Fiorentino D. Evidence supports blind screening for internal malignancy in dermatomyositis: Data from 2 large US dermatology cohorts. *Medicine (Baltimore)*. 2018;97(2):e9639. <https://doi.org/10.1097/MD.0000000000009639>
19. Sigurgeirsson B, Lindelöf B, Edhag O, Allander E. Risk of cancer in patients with dermatomyositis or polymyositis. A population-based study. *N Engl J Med*. 1992;326(6):363-7. <https://doi.org/10.1056/NEJM199202063260602>
20. Hatada T, Aoki I, Ikeda H, Tamura T, Okada K, Nakai T, Utsunomiya J. Dermatomyositis and malignancy: case report and review of the Japanese literature. *Tumori*. 1996;82(3):273-5.
21. Stockton D, Doherty VR, Brewster DH. Risk of cancer in patients with dermatomyositis or polymyositis, and follow-up implications: a Scottish population-based cohort study. *Br J Cancer*. 2001;85(1):41-5. <https://doi.org/10.1054/bjoc.2001.1699>
22. Chang SH, Park JK, Lee YJ, Yang JA, Lee EY, Song YW, Lee EB. Comparison of cancer incidence among patients with rheumatic disease: a retrospective cohort study. *Arthritis Res Ther*. 2014;16(4):428. <https://doi.org/10.1186/s13075-014-0428-x>
23. Yu KH, Kuo CF, Huang LH, Huang WK, See LC. Cancer Risk in Patients With Inflammatory Systemic Autoimmune Rheumatic Diseases: A Nationwide Population-Based Dynamic Cohort Study in Taiwan. *Medicine (Baltimore)*. 2016;95(18):e3540. <https://doi.org/10.1097/MD.0000000000003540>
24. Ohno S, Oshikawa K, Kitamura S, Saitoh K. Clinico-pathological analysis of interstitial pneumonia associated with collagen vascular disease in patients with lung cancer. *Nihon Kyobu Shikkan Gakkai Zasshi*. 1997;35(12):1324-9.
25. Lakhanpal S, Bunch TW, Ilstrup DM, Melton LJ 3rd. Polymyositis-dermatomyositis and malignant lesions: does an association exist? *Mayo Clin Proc*. 1986;61(8):645-53. [https://doi.org/10.1016/s0025-6196\(12\)62030-8](https://doi.org/10.1016/s0025-6196(12)62030-8)
26. Basset-Seguín N, Roujeau JC, Gherardi R, Guillaume JC, Revuz J, Touraine R. Prognostic factors and predictive signs of malignancy in adult dermatomyositis. A study of 32 cases. *Arch Dermatol*. 1990;126(5):633-7.
27. Rose AL, Walton JN. Polymyositis: a survey of 89 cases with particular reference to treatment and prognosis. *Brain*. 1966;89(4):747-68. <https://doi.org/10.1093/brain/89.4.747>
28. Dourmishev LA. Dermatomyositis associated with malignancy. 12 case reports. *Adv Exp Med Biol*. 1999;455:193-9. https://doi.org/10.1007/978-1-4615-4857-7_28
29. Marie I, Hatron PY, Levesque H, Hachulla E, Hellot MF, Michon-Pasturel U, Courtois H, Devulder B. Influence of age on characteristics of polymyositis and dermatomyositis in adults. *Medicine (Baltimore)*. 1999;78(3):139-47. <https://doi.org/10.1097/00005792-199905000-00001>
30. Nagano Y, Inoue Y, Shimura T, Fujikawa H, Okugawa Y, Hiro J, Toiyama Y, Tanaka K, Mohri Y, Kusunoki M. Exacerbation of Dermatomyositis with Recurrence of Rectal Cancer: A Case Report. *Case Rep Oncol*. 2015;8(3):482-6. <https://doi.org/10.1159/000439519>
31. Gkegkes ID, Minis EE, Iavazzo C. Dermatomyositis and colorectal cancer: a systematic review. *Ir J Med Sci*. 2018;187(3):615-620. <https://doi.org/10.1007/s11845-017-1716-7>
32. Dourmishev LA, Draganov PV. Paraneoplastic dermatological manifestation of gastrointestinal malignancies. *World J Gastroenterol*. 2009;15(35):4372-9. <https://doi.org/10.3748/wjg.15.4372>
33. Ouahbi H, Benhami M, Nouikh L, Acharfi N, Kelati A, Oualla K, Benbrahim Z, Elmrabet FZ, Arifi S, Mernissi F, Mellas N. Dermatomyosite et cancer rectal: à propos d'un cas avec revue de la littérature. *Pan Afr Med J*. 2019;33:122. <https://doi.org/10.11604/pamj.2019.33.122.14509>