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Respiratory Diseases

Paraneoplastic Dermatomyositis Associated with Small-Cell Lung Carcinoma

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Abstract | **Case Report**

Dermatomyositis (DM) is frequently associated with neoplastic disease, particularly in elderly individuals. All histological types and locations of cancer can be involved. The association between DM and bronchogenic carcinoma has been demonstrated in several studies. We report a case of dermatomyositis revealing a small-cell bronchogenic carcinoma.

Keywords: Dermatomyositis, Bronchogenic Carcinoma, Paraneoplastic Syndrome.

INTRODUCTION

Dermatomyositis is a rare disease characterized by muscle inflammation and skin rashes. When it occurs in association with cancer, it is referred to as paraneoplastic dermatomyositis. This phenomenon is particularly associated with several types of cancer, including lung cancer.

CAS CLINIQUE

A 58-year-old male presented with progressive muscle weakness initially affecting the lower limbs and later spreading to the upper limbs. He was a chronic smoker with a 30 pack-year history, without any other significant medical or family history of cancer. The history revealed a 10-month progression of pruritic erythematous lesions on the neckline and face, with photosensitivity. The patient also reported exertional dyspnea evolving over one month, along with a general decline in health.

Physical examination revealed a V-shaped erythema on the chest, facial erythroedema, and linear erythematous lesions on the extensor surfaces of the forearms (Figure 1).

The hands showed erythematous scaly papules on the proximal interphalangeal and metacarpophalangeal joints consistent with Gottron's papules (Figure 2). The right lower limb also showed erythematous-squamous lesions.

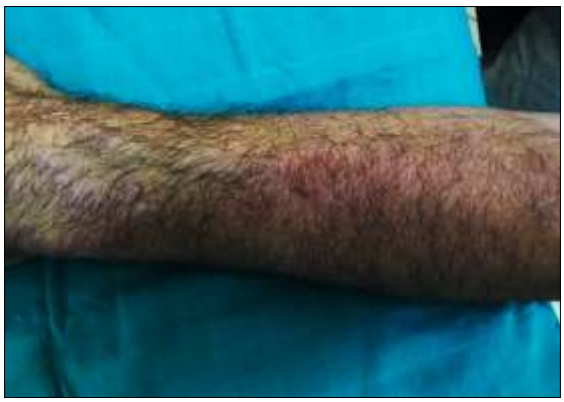

Figure 1: Linear erythematous lesions on the extensor surfaces of the forearms

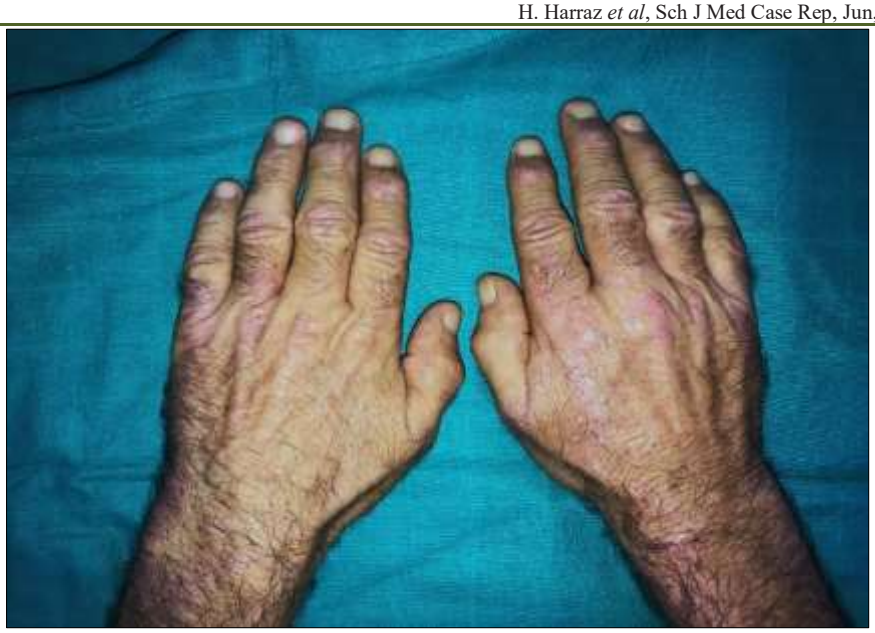

Figure 2: Gottron's papules on the extensor surfaces of the hands

Neurological examination revealed assisted standing and walking. The patient could hold the Barré position but not the Mingazzini. Muscle strength was 4/5 for distal muscles and 3/5 for proximal muscles. The "coat hanger" and "stool" signs were positive.

Laboratory tests showed elevated CPK (195 U/L) and LDH (253U/L) levels. CBC, AST/ALT, albumin, and CRP were normal. Electroneuromyography suggested a myogenic syndrome. Muscle biopsy revealed inflammatory foci in the inter-fascicular septa and endomysium, consistent with necrotizing inflammatory myopathy. Autoimmune screening for myositis-specific antibodies (anti-Mi-1, anti-Mi-2, anti-Jo, anti-PL, anti-SCL) was negative.

Thoracic CT revealed a right mediastino-pulmonary mass centered on the carina and the right main bronchus, measuring 80 mm in its largest axis (Figure 3). Flexible bronchoscopy showed a budding tumor completely obstructing the right upper lobar bronchus and another tumor bud in the middle lobe (Figure 4). Bronchial biopsies were consistent with a small-cell neuroendocrine carcinoma.

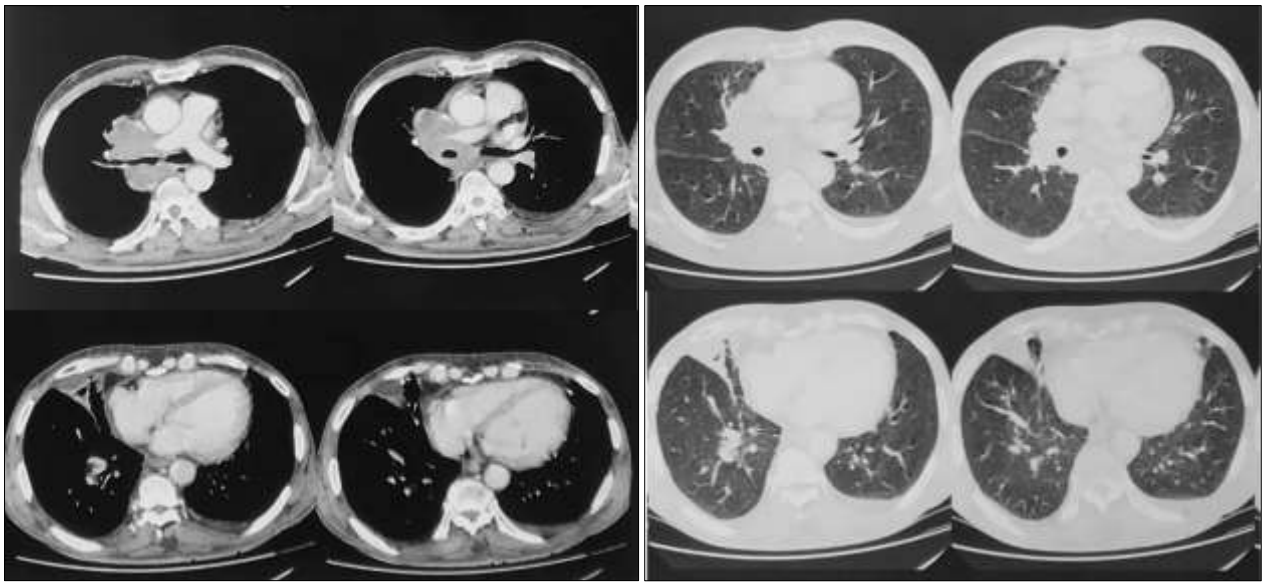

Figure 3: Thoracic CT showing a right mediastino-pulmonary mass centered on the carina and right main bronchus (80 mm)

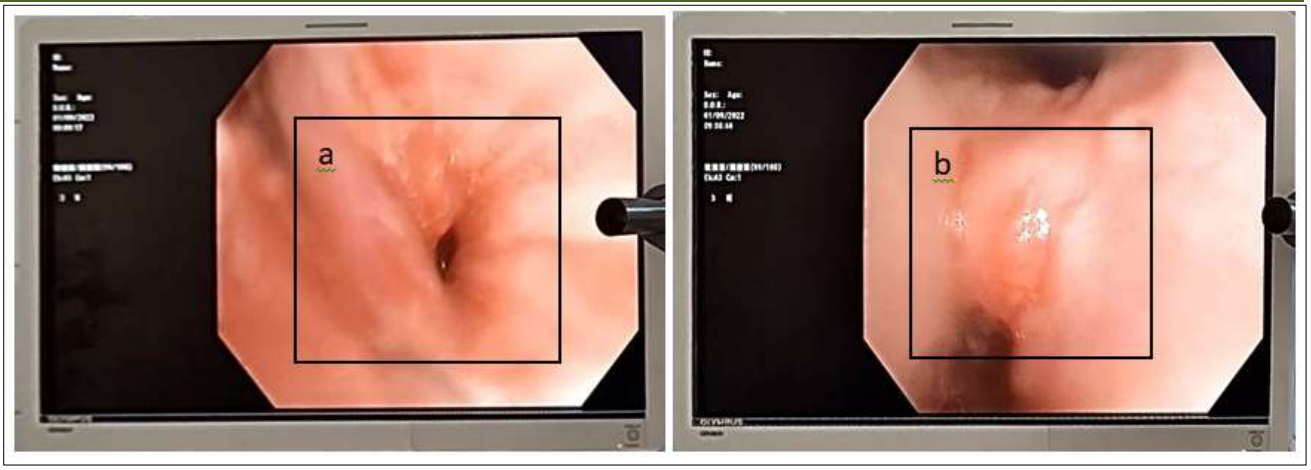

Figure 4: Bronchoscopy: (a) Tumor completely obstructing the right upper lobar bronchus; (b) Tumor bud in the middle lobe

The patient received intravenous methylprednisolone (2g/day) for 3 days, followed by oral corticosteroids at 1.5 mg/kg/day (80 mg) combined with hydroxychloroquine. Topical emollient and corticosteroid creams were prescribed for skin lesions. Muscle and skin signs did not improve with treatment, though there was a decrease in CPK and LDH levels.

Extension workup, including brain and thoraco-abdominopelvic CT and bone scintigraphy, revealed osteocondensing lesions in the right iliac wing and femoral head, and a metastatic occipital nodule on brain CT. This confirmed a small-cell neuroendocrine carcinoma, staged as T4N3M1b (Stage IVA).

The multidisciplinary oncothoracic team concluded a diagnosis of paraneoplastic dermatomyositis revealing metastatic small-cell neuroendocrine carcinoma. The therapeutic decision was palliative chemotherapy with Cisplatin and Etoposide.

After the first cycle of chemotherapy, muscle strength improved, though cutaneous signs persisted. Unfortunately, the patient died before receiving the second cycle due to swallowing difficulties.

DISCUSSION

Dermatomyositis (DM) is a rare connective tissue disease that belongs to the group of inflammatory myopathies. Its diagnosis is based on a combination of clinical, enzymatic, electromyographic, and histological findings, with characteristic cutaneous involvement. The annual incidence is 1 per 100,000 inhabitants [1], with a sex ratio of two women for every man [2]. It can occur at any age but shows a peak incidence between 5 and 14 years of age, and a second peak during the fifth or sixth decade of life [2].

In elderly patients, dermatomyositis is often associated with an underlying malignancy. The risk of cancer is increased by approximately 3 to 7 times in patients with dermatomyositis, with a prevalence of neoplasms reaching 8.6% in individuals recently diagnosed with DM [3].

Paraneoplastic dermatomyositis may appear before, simultaneously with, or after the diagnosis of cancer, and it can be associated with all histological types and anatomical sites of cancer observed in the general population [4]. It represents a poor prognostic factor for cancer.

The mechanism of paraneoplastic dermatomyositis/polymyositis remains poorly understood. According to several studies, it results from dysregulation of both humoral and cellular immunity. Two main theories have been proposed: a hormonal theory and an immunoallergic theory.

According to the hormonal theory, the tumor secretes active hormonal polypeptides responsible for the various clinical syndromes. The immunoallergic mechanism involves a cross-reaction of antibodies produced against tumor antigens with normal tissues that share similar structures. In DM, there is an initial injury to the capillary endothelium of the endomysium, leading to complement activation with immune complex deposition in the capillary bed. This results in endothelial cell edema, vacuolization, and capillary necrosis. Perivascular inflammation, ischemia, and destruction of muscle fibers are also present in dermatomyositis/polymyositis.

The histological type of bronchogenic carcinoma most strongly associated with paraneoplastic dermatomyositis is small-cell carcinoma (29%), followed by squamous cell carcinoma (21%) and adenocarcinoma (8%) [5].

Cutaneous manifestations are characteristic of dermatomyositis and may precede myositis by several months. They may present as photosensitive erythroedema, predominantly affecting exposed areas

such as the face, anterior neck, shoulders, and extensor surfaces of the limbs, or as a heliotrope rash, which is a violaceous periorbital erythema. Gottron's papules are erythematous or violaceous plaques that appear over the dorsal aspect of the interphalangeal and metacarpophalangeal joints, and less commonly over the elbows and knees. These lesions are considered pathognomonic of dermatomyositis. A painful periungual erythema, known as the manicure sign, may also be observed on palpation.

Motor symptoms are also present, typically as a bilateral, symmetric, non-selective muscle weakness. The weakness is myogenic in nature and predominantly affects proximal muscles, such as those of the shoulder and pelvic girdles, as well as the cervical muscles.

Other possible manifestations include general symptoms (fever, malaise, and weight loss), pulmonary involvement (interstitial lung disease or respiratory muscle weakness), musculoskeletal (arthralgia or synovitis), gastrointestinal (malabsorption), cardiac (atrioventricular tachycardia or myocarditis) and vascular symptoms (Raynaud's phenomenon).

The clinical manifestations of DM do not differ between patients with or without associated neoplasia. The following clinical features may be associated with the presence of malignant tumors in patients with DM: the presence of systemic symptoms, the absence of Raynaud's phenomenon, and the rapid onset of myositis. Elevation of at least one muscle enzyme (CK, AST, LDH, aldolase) is observed in 70 to 90% of patients. ESR and CRP are normal or moderately increased. Autoantibodies associated with dermatomyositis must be investigated as they provide insight into the origin of the dermatomyositis. Anti-TIF1-γ antibodies are associated with an increased risk of malignancy and are often found in patients with paraneoplastic dermatomyositis [6]. Anti-Mi-2, anti-SRP, and anti-P155/P140 antibodies may also be present in the context of neoplasia. Muscle biopsy is not systematic. It is performed on proximal and/or weakened muscles (deltoid or quadriceps). Possible lesions include predominantly perimysial and perivascular inflammation composed of B and T lymphocytes, CD4+ cells, as well as complement deposition in capillaries and capillary vasodilation. Perifascicular necrosis and atrophy, and endomysial and perimysial fibrosis may also be present.

The positive diagnosis is based on the Bohan and Peter criteria, which are as follows:

- Progressive and symmetrical muscle weakness of the girdles and neck flexors.
- Dermatological signs (periorbital heliotrope rash with edema, Gottron's papules).
- Muscle biopsy in favor of myositis.
- Elevation of serum muscle enzymes indicating muscle necrosis.
- Electromyographic findings consistent with muscle involvement.

In the presence of three or four criteria, along with the skin eruption, the diagnosis of DM is confirmed. If only two criteria are associated with the skin eruption, the diagnosis of DM is very likely. The goal of treatment is to induce remission of muscular, cutaneous, and possible other manifestations, as well as to treat the underlying cancer. Treatment must be adjusted according to disease evolution and involves several therapeutic approaches. Corticosteroid therapy is prescribed at 1 mg/kg/day initially, followed by progressive tapering. Methotrexate is indicated in cases of steroid resistance or dependence. Intravenous immunoglobulins are administered in cases of swallowing disorders. Synthetic antimalarials (Plaquenil®) are used to improve cutaneous involvement. Muscle physiotherapy is indicated to improve motor deficit. Management remains primarily etiological and depends on the effectiveness of the causal cancer treatment. This relationship is confirmed by the recurrence of muscle weakness observed in cases of cancer relapse. The 3-year survival rate of patients with DM is 57%, whereas the 2-year survival does not exceed 25% [7]. The following factors, when present during the first year after diagnosis, are indicators of poor prognosis: the DM-neoplasia association, advanced age, lack of improvement in muscle strength during the first month of treatment, and deterioration of general health status [8].

CONCLUSION

Paraneoplastic dermatomyositis may reveal various cancers, particularly bronchogenic carcinoma. The search for occult neoplasms should be systematic in patients presenting with DM in order to allow for earlier diagnosis.

Bronchogenic carcinoma is more frequent in patients with dermatomyositis and should be systematically investigated, especially in the presence of other risk factors (age, toxic habits).

REFERENCES

1. Mastaglia FL Phillips, B. A. Rheum Dis Clin North Am. 2002 Nov;28(4):723-41.
2. Isak V and Jorizzo, J. L J. Dermatol. Treat. 2017;29:450–459.
3. Zahr ZA and Baer, A. N. Curr Rheumatol Rep.2011;13:208–215.
4. Santacroce L. Drugs & Diseases. 2017 Updated December 06 2018.
5. Zahr ZA and Baer AN. Curr Rheumatol Rep. 2011;13:208–215.
6. Wen C.E. Semin. Oncol. 2016;43:366–369.
7. Basset-Seguin et N, Roujeau J. C, Gherardi R. Arch Dermatol. 1990;126:633–637.
8. Sparsa A et Liozon E, Herrmann F, Ly K, Lebrun V, Soria P, LoustaudRatti V. Arch Dermatol. 2002;138:885–890.