

## CASE REPORT

# Rapidly lethal dermatomyositis associated with cutaneous lymphangitis carcinomatosa

Cristina Resende, Catarina Araújo, Maria Luz Duarte, Celeste Brito

Department of Dermatology and Venereology, Hospital de Braga, Braga, Portugal

**Correspondence to**

Dr Cristina Isabel Pinho Resende,  
cristinapresende@gmail.com

**SUMMARY**

A 70-year-old woman with a recent diagnosis of dermatomyositis (DM) presented to the dermatology department for study of a probably paraneoplastic syndrome. On examination, we observed discrete, indurated, reddish, painful plaques and nodules on her abdomen and both thighs. A cutaneous biopsy from an abdominal nodule, performed as part of the paraneoplastic workup, was suggestive of cutaneous lymphangitis carcinomatosa, secondary to unknown malignancy. An extensive investigation to locate the site of the primary tumour revealed no specific findings. A course of palliative chemotherapy with cisplatin and 5-fluorouracil was then given, but the patient's condition deteriorated and 6 months after her initial observation the patient died. We describe this case because, to our knowledge, the association between DM and cutaneous lymphangitis carcinomatosa has not been described yet in the literature and to highlight that, DM can be a rapidly lethal disease.

**BACKGROUND**

Dermatomyositis (DM) is an uncommon inflammatory myopathy of unknown aetiology, characterised by the association of progressive proximal muscle weakness, pathognomonic or specific cutaneous findings, and compatible electrophysiological and pathological findings.<sup>1 2</sup> The disease demonstrates a bimodal age distribution in juvenile and adult forms. In adults, it is often considered as a paraneoplastic syndrome.<sup>3</sup>

We present a case of rapidly lethal paraneoplastic DM, associated with cutaneous lymphangitis carcinomatosa, without a definitive diagnosis of the primary tumour.

**CASE PRESENTATION**

A previously healthy 70-year-old Caucasian woman was admitted to our department with heliotrope erythema, periorbital swelling, several proximal muscle weaknesses and painful nodules on her abdomen and thighs. All of these lesions appeared progressively over the previous 6 months. The patient presented with asthenia, anorexia and weight loss of 8 kg in 5 months. She also reported dysphagia, mainly at the initiation of the attempt and dysphonia. Her medical and family histories were unremarkable. She was not taking any medication.

A physical examination revealed discrete, indurated, reddish, painful plaques and nodules on her upper abdomen and both thighs (figures 1 and 2). The nodules were well circumscribed, firm and non-mobile, with diameters ranging from 6 mm to 5 cm. There were no other abnormalities on examination.



**Figure 1** Reddish, indurated plaques and nodules on her upper abdomen.

We also observed pink/violet macular hyperpigmentation and atrophy, with telangiectasias over the posterior shoulders, back, buttocks and a V-shaped area of the anterior neck and chest, suggestive of poikiloderma. Evaluation of muscle strength revealed a grade 4/5 weakness of the pectoral and pelvic girdle musculature. The laboratory tests revealed erythrocyte sedimentation rate 63 mm/h (normal range 1–20), creatine kinase 440 UI/L (normal range 26–192), lactate dehydrogenase 254 UI/L (normal range 84–246), normocytic and normochromic anaemia with haemoglobin 8.2 g/dL and platelet count  $29 \times 10^9/L$ . The serum levels of cancer antigen 125 (CA125), CA 19.9, CA 15.3, and carcinoembryonic antigen were 61.70 U/mL (normal <35), 7.50 (normal range <37), 33.50 U/mL (normal range <32.4) and 5 U/mL (normal range 0.0–5) ng/mL, respectively. Antinuclear antibodies, anti-Jo-1 antibody, antiribonucleoprotein antibody, and anti-Ro antibody were negative.

A deltoid muscle biopsy revealed perivascular and interfascicular inflammatory infiltrates with adjoining groups of muscle fibre degeneration/regeneration, suggestive of DM. Electromyography was suggestive of a myopathic pattern with multiple polyphasic low-amplitude potentials with sharp edges and short duration.

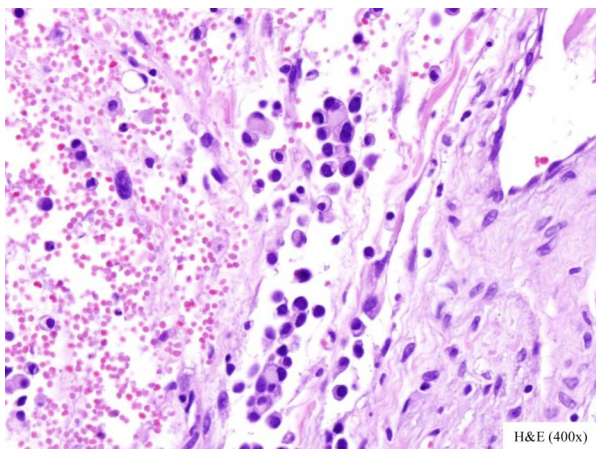
**To cite:** Resende C, Araújo C, Duarte ML, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-010376



**Figure 2** Reddish, indurated plaques and nodules on both her thighs.

Taken together, clinical, laboratory and pathological data suggested the diagnosis of a paraneoplastic DM, without a known malignancy.

The patient was initially treated with prednisone (60 mg orally daily) and the dose was gradually reduced over many weeks to months. She also received hydroxychloroquine (400 mg orally daily) and azathioprine (100 mg orally daily), with only partial response. The patient underwent extensive investigation to locate the site of the primary tumour, including chest x-ray, CT of the chest, abdomen and pelvis, oesophagogastroscopy, colonoscopy and mammography. This extensive workup revealed no specific findings. A cutaneous biopsy from an abdominal nodule, performed as part of the paraneoplastic workup, to exclude skin metastasis, demonstrated an infiltration of the dermal and hypodermal lymphatics by neoplastic cells (figure 3). Immunohistochemistry reports were positive for cytokeratin 7 (CK7; figure 4) and negative for CK20 and thyroid transcription factor-1 (TTF-1), or oestrogen receptors. Diagnosis of cutaneous



**Figure 3** Infiltration of the dermal and hypodermal lymphatics by neoplastic cells.

lymphangitis carcinomatosa, secondary to unknown malignancy was made.

#### TREATMENT

A course of palliative chemotherapy with cisplatin and 5-fluorouracil was given to the patient after the diagnosis of cutaneous lymphangitis carcinomatosa.

#### OUTCOME AND FOLLOW-UP

The patient's condition deteriorated and 6 months after her initial observation, the patient succumbed with Gram-negative sepsis, which failed to respond to treatment with broad-spectrum antibiotics. A postmortem examination was not performed.

#### DISCUSSION

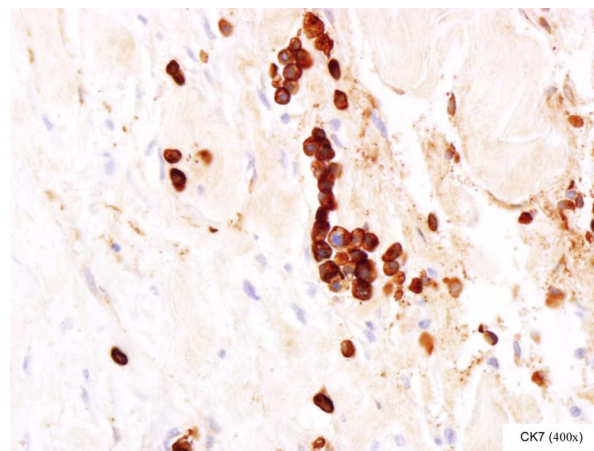
DM is related to an increased risk of cancer and is often considered as a paraneoplastic syndrome, although the strength of this relationship and its predisposing factors are not clearly defined.<sup>3-6</sup> This association is more frequently seen in adults with DM rather than in the paediatric population.<sup>7-8</sup> The reported frequency of an internal malignancy in adults above the age of 40 years with DM has been reported in the range 10–50%.<sup>6-9</sup>

The malignancies, most strongly associated with DM include the malignancy of gastrointestinal tract, pancreas, lung, breast, gynaecological cancers and non-Hodgkin's lymphoma.<sup>6-7-10</sup>

Cutaneous manifestations seen in paraneoplastic DM are similar to those of classic DM and include a heliotropic rash with periorbital oedema, nail fold telangiectasias, Gottron's papules on the finger joints and pink-violaceous poikiloderma overlying the chest, upper back, elbows and knees.<sup>8</sup> Autoantibodies are present in a high percentage of patients with DM without malignancy and the absence of autoantibodies may be predictive of an occult malignancy,<sup>8</sup> like in our patient.

The malignancy can occur before the onset of DM, concomitantly or afterwards.<sup>1-9</sup> In our patient, the malignancy appears a few months after the initial symptoms of DM, but this temporal relation is not clear in our case. In 5–10% of malignancies, the primary tumour is not known, despite full evaluation, and they first appear in one or more metastatic sites,<sup>6</sup> like in our patient.

Management of paraneoplastic DM focuses primarily on the treatment of DM and the underlying malignancy.<sup>8</sup> Paraneoplastic DM is generally more resistant to corticosteroids and cytotoxic therapies, compared with idiopathic DM.<sup>2-6</sup> Moreover, the immunosuppressive drugs that are used in the



**Figure 4** Tumour cells are positive for cytokeratin 7.

treatment of DM could theoretically exacerbate malignancy, but this is seldom seen in clinical practice.<sup>4 5 8</sup>

Cutaneous lymphangitis carcinomatosa, a type of cutaneous metastasis, was also diagnosed in our patient.

Cutaneous metastasis of cancer is rare.<sup>11–13</sup> The incidence of cutaneous metastasis in patients with metastatic cancer is approximately 5% and the skin represents the 12th site of metastasis for deep cancer.<sup>11–13</sup>

Cutaneous lymphangitis carcinomatosa is a rare presentation of skin metastasis, accounting for approximately 5% of all cutaneous metastasis and it results from an occlusion of the lymphatic channels of the dermis by neoplastic cells.<sup>11</sup>

The prognosis of cutaneous lymphangitis carcinomatosa is very poor with a mean survival of 3 months after diagnosis of cutaneous deposits.<sup>12 13</sup>

Our patient died 4 months after the diagnosis of cutaneous lymphangitis carcinomatosa. There are few published cases of cutaneous lymphangitis carcinomatosa and, to our knowledge, this is the first case of cutaneous lymphangitis carcinomatosa associated with paraneoplastic DM.<sup>11–13</sup>

Immunohistopathological stains showed a cytokine expression profile (CK7 positive and negative for CK20, TTF-1 or oestrogen receptors). Our findings in immunohistochemistry favour the hypothesis of a gastric carcinoma. However, the hypothesis of breast cancer with loss of oestrogen receptor cannot be excluded. These results make the hypothesis of pulmonary carcinoma unlikely.

The patient's condition deteriorated rapidly and the patient died without a diagnosis of the primary tumour.

We describe this case because the association between DM and cutaneous lymphangitis carcinomatosa is rare and to highlight that DM can be a rapidly lethal disease.

### Learning points

- ▶ The absence of autoantibodies in dermatomyositis (DM) may be predictive of an occult malignancy.
- ▶ Paraneoplastic DM can be a rapidly lethal disease.
- ▶ Cutaneous lymphangitis carcinomatosa is a rare presentation of skin metastasis, accounting for approximately 5% of all cutaneous metastasis.
- ▶ The prognosis of cutaneous lymphangitis carcinomatosa is very poor with a mean survival of 3 months after diagnosis of cutaneous deposits.

**Contributors** CIPR contributed to the literature research, data collection and article writing. CA contributed to the acquisition of data. MDLD contributed to the acquisition of data, interpretation of data and revision. CB contributed to the acquisition of data and revision.

**Competing interests** None.

**Patient consent** Obtained.

**Provenance and peer review** Not commissioned; externally peer reviewed.

### REFERENCES

- 1 Dourmishev LA, Popov JM, Rusinova D. Paraneoplastic dermatomyositis associated with testicular cancer: a case report and literature review. *Acta Dermatovenerol Alp Panonica Adriat* 2010;19:39–40.
- 2 Sabio JM, Vargas-Hitos JA, Jiménez-Alonso J. Paraneoplastic dermatomyositis associated with bladder cancer. *Lupus* 2006;15:619–20.
- 3 Tsoukalas N, Tolia M, Kostakis D, et al. Dermatomyositis as an early manifestation and a significant clinical precursor of lung cancer: report of a rare case and review of the current literature. *Int J Clin Exp Med* 2013;6:105–9.
- 4 Harrison BA, Heck SI, Hood AF. A fatal case of dermatomyositis with underlying metastatic esophageal adenocarcinoma. *Cutis* 2008;1:26–8.
- 5 Kim HI, Chung SH, Hwang JE, et al. Dermatomyositis associated with cancer of unknown primary site. *J Korean Med Sci* 2007;22:174–7.
- 6 Shah KR, Boland CR, Patel M, et al. Cutaneous manifestation of gastrointestinal disease. Part I. *J Am Acad Dermatol* 2013;69:189 e1–e21.
- 7 Narasimhaiah DA, Premkumar JA, Moses V, et al. Carcinoma of gall bladder presenting as dermatomyositis. *Ann Indian Acad Neurol* 2011;14:44–6.
- 8 Lalla SC, Aldridge RD, Tidman MJ. Carcinoma of the penis presenting with dermatomyositis. *Clin Exp Dermatol* 2001;26:556–8.
- 9 Langan SM, O'Briain S, Barnes L. Dermatomyositis associated with angiotropic lymphoma. *Clin Exp Dermatol* 2003;28:597–9.
- 10 Jouary T, Gracia C, Lalanne N, et al. Rapidly lethal dermatomyositis associated with metastatic melanoma. *J Eur Acad Dermatol Venereol* 2008;22:399–400.
- 11 Prat L, Chouaid C, Kettaneh A, et al. Cutaneous lymphangitis carcinomatosa in a patient with lung adenocarcinoma: case report and literature review. *Lung Cancer* 2013;79:91–32.
- 12 Keohane SG, Doherty VR, McLaren KM, et al. Cutaneous lymphangitis carcinomatosa: a rare primary manifestation of malignancy. *J R Soc Med* 1996;89:582–3.
- 13 Nakamura S, Hashimoto Y, Nishi K, et al. Cutaneous lymphangitis carcinomatosa metastasis of extra-ovarian primary peritoneal carcinoma. *Acta Derm Venereol* 2012;92:585–663.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact [consortiasales@bmjgroup.com](mailto:consortiasales@bmjgroup.com)

Visit [casereports.bmj.com](http://casereports.bmj.com) for more articles like this and to become a Fellow