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Grand Round

Dermatomyositis as a Paraneoplastic Syndrome in Carcinosarcoma of Uterine Origin

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ABSTRACT:
Paraneoplastic syndromes are a collection of disorders affecting an organ or tissue caused by cancer but occurring at a site distant from the primary or metastases. Dermatomyositis can occur in association with malignancy as a paraneoplastic phenomenon. We present a case of a patient presenting simultaneously with an advanced carcinosarcoma of the uterus and dermatomyositis. The diagnoses, pathophysiology and treatment of these two conditions are discussed and current published studies reviewed. Chandiramani, M. *et al*. (2006). *Clinical Oncology* 18, 641–648

Key words: Dermatomyositis, carcinosarcoma, malignant mixed Mullerian tumour, chemotherapy

Case Report

A 63-year-old women presented in March 2002 after a 6-week history of painless vaginal bleeding. She had completed her menopause 10 years previously and had no relevant past gynaecological or medical history. At the time of referral she reported an erythematous, scaly rash on her neck and upper thorax in a 'shawl-like distribution'. A similar rash was seen over the anterior aspects of her thighs. She complained of a short history of proximal muscle weakness affecting her legs and arms.

Clinically, she had a confluent, desquamating, scaly rash on her anterior and posterior chest wall, which extended up her neck (Fig. 1). Erythematous plaque-like lesions were present on the extensor surfaces of the thighs and elbows. Her hands and face were unremarkable. Muscle power was reduced to four-fifths in the proximal muscles in both arms and legs.

Cardiac, respiratory and abdominal examinations were all unremarkable. An examination under anaesthesia revealed a mobile, ante verted uterus, with a large tumour mass seen at the cervix, possibly arising from the uterine cavity or fundus. Blood parameters were all within normal range, apart from an elevated creatinine kinase (2897 iu/l) and alanine transaminase (172 iu/l). A rheumatological screen was negative.

The patient proceeded to exploratory laparotomy, where the tumour was found to be obliterating the pelvis and involving the bowel. Tumour nodules were felt on the liver and peritoneum. There were no obvious palpable lymph nodes. A total abdominal hysterectomy and a bilateral salpingo-oopherectomy were carried out, but residual tumour was left behind.

A histopathological examination confirmed the diagnosis of carcinosarcoma (malignant mixed Mullerian tumour). Macroscopically, the lower uterus was replaced by a pale and haemorrhagic mass extending through the wall of the uterus. The lower resection margin was disrupted with tumour. Microscopically, most of the tumour had the appearance of a largely necrotic, poorly differentiated endometrioid adenocarcinoma. The remainder of the tumour consisted of a malignant stromal component with pleomorphic stromal cells and epithelial cells with bizarre giant nuclei and multinucleated tumour cells. Occasional foci of squamous differentiation and some myxoid areas were seen (Fig. 2). The tumour extended into the endocervical stroma. Both fallopian tubes and ovaries had extensive metastatic deposits: peritoneal washings contained malignant cells.

Postoperatively, her myositis became so severe that she was unable to stand, her skin was deteriorating and her creatinine kinase did not improve. In view of her advanced disease, incomplete resection and aggressive dermatomyositis she was treated systemically with CAP chemotherapy. She received cyclophosphamide 600 mg/m^2, doxorubicin 50 mg/m^2 and cisplatin 50 mg/m^2 on a 21-day cycle.

Dermatology and rheumatology teams were involved in her care. Preoperatively her skin lesions had been treated with topical dermovate, as a rapid improvement was anticipated after radical surgery. Postoperatively, she received oral prednisone (40 mg) in addition to topical

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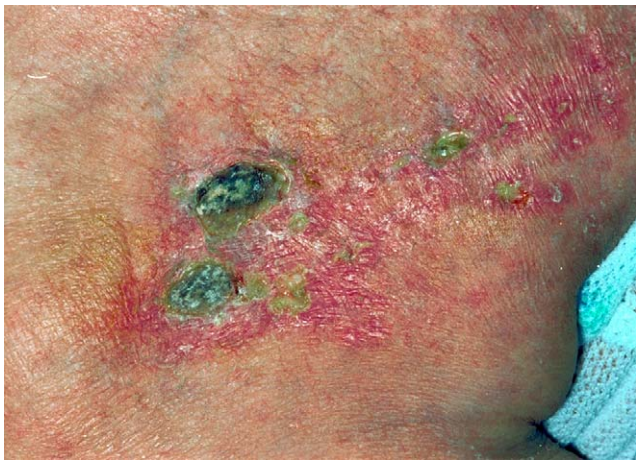

Fig. 1 — Dermatomyositis before chemotherapy.

steroids and emmolients. Baseline spirometry was carried out because of concern about respiratory muscle compromise. Four days after the start of oral steroids and her first cycle of chemotherapy, her myopathy deteriorated further. She was started on bolus intravenous methylprednisolone (500 mg daily for 3 days). Her muscle power and dermatology improved and her creatinine kinase fell to 832 iu/l. She was recommenced on oral prednisone (60 mg).

Ten days after her first cycle of chemotherapy she developed *Pseudomonas aeruginosa* septicaemia associated with grade 3 neutropenia. The source of the infection was from the chest wall dermatositis. She responded to intravenous antibiotics. As a result of her prolonged illness and hospital stay she lost one-sixth of her total body weight (11 kg lost). She had no difficulty swallowing, but her oral intake was only meeting 50% of her needs and nasogastric feeding was commenced with the patient's agreement. By the time of her second cycle of chemotherapy, the power in her legs was graded 4 out of 5 and skin was improving. Her serum creatinine kinase level was normal at 59 iu/l. She was receiving intensive physiotherapy, including hydrotherapy. Her prednisone dose was reduced to 50 mg and she continued to reduce this by 5 mg per week. She was receiving oral bisphosphanates, calcium and vitamin D prophylactically for steroid-induced osteoporosis.

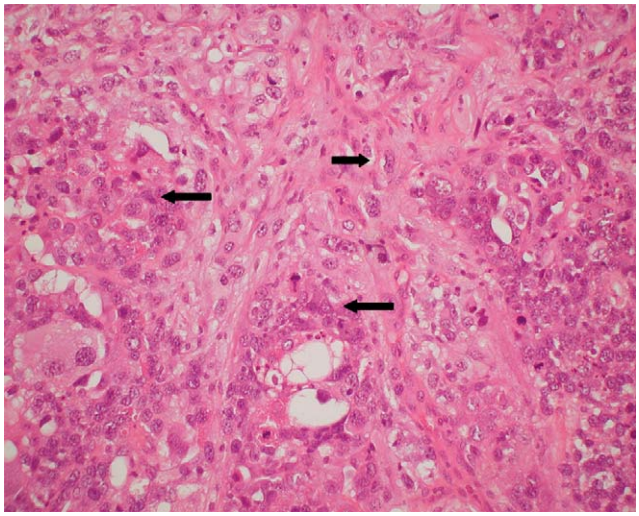

Fig. 2 — Carcinosarcoma featuring malignant glands formed by highly atypical epithelium supported (arrows) by a sarcomatous stroma (×200 original magnification).

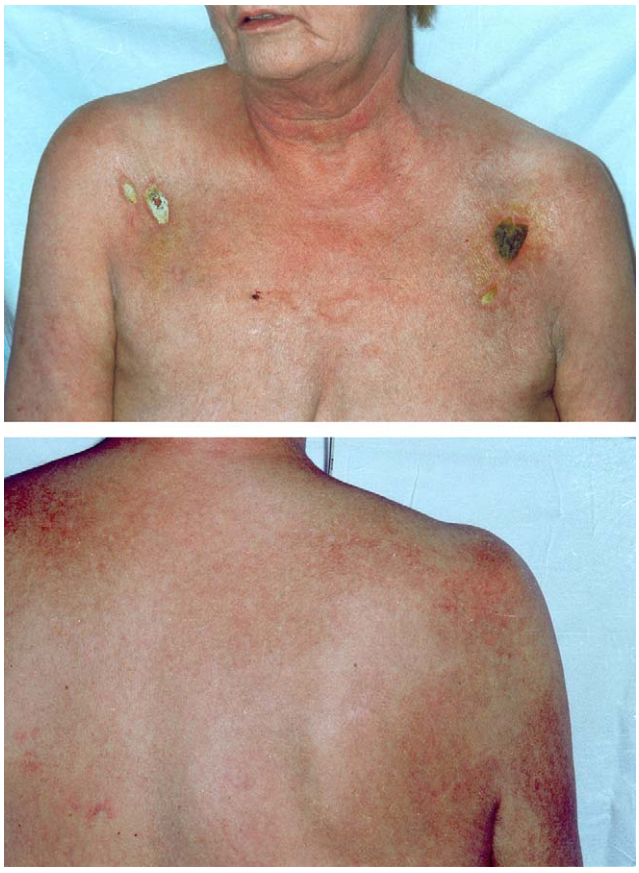

Fig. 3 — Improvement in dermatomyositis after three cycles of cisplatin, doxorubicin and cyclophosphamide.

She was discharged to her local cottage hospital for continued rehabilitation after her third cycle of chemotherapy (Fig. 3). She had been an inpatient for 12 weeks. She tolerated her chemotherapy without further significant toxicity and was able to walk into the clinic by her fourth cycle of chemotherapy. When she completed chemotherapy she was on a maintenance dose of 5 mg of prednisone. Post-chemotherapy computed tomography showed no evidence of residual disease. She entered a period of surveillance.

One year after her diagnosis her steroids were stopped completely. She still had some residual weakness in her arms, but otherwise had made a complete recovery. It is now 4 years since her diagnosis. She was recently reviewed in the combined gynae-oncology clinic where she has no evidence of recurrence, clinically or radiologically (Fig. 4), of either her dermatomyositis or uterine carcinosarcoma.