Metastatic non-seminomatous germ cell tumour and dermatomyositis

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Summary: Twenty to thirty percent of cases of dermatomyositis in adults are associated with malignancy. We report a case in which dermatomyositis heralded the development of a testicular germ cell tumour. Germ cell testicular tumours are treatable and our case emphasizes the importance of measuring blood levels of beta-human chorionic gonadotrophin and alpha-fetoprotein and examination of the testes in young males with indications of possible malignancy.

Introduction

In adults with dermatomyositis, an underlying malignancy is found in 20–30%.^{1,2} We report the first case to our knowledge of dermatomyositis heralding a testicular germ cell tumour.

Case report

In June 1987 a 30 year old Caucasian man developed an erythematous rash over his face (especially peri-orbitally), shoulders, elbows, upper chest and back 13 hours after routine inoculation for polio, tetanus, typhoid and cholera. He then noticed that the skin overlying the metacarpophalangeal joints became purple, with painful ulcers on his finger tips and intermittent dysphagia with occasional regurgitation of food through his nose. His rash worsened over the next month, he lost weight and he developed pain, stiffness and weakness of his shoulders. He had difficulty raising his arms above his head and sitting up due to proximal muscle weakness. Investigations revealed creatine kinase 1,171 IU/l (upper limit of normal 50 IU/l), myopathic pattern on electromyogram (EMG) and abnormalities consistent with the diagnosis of dermatomyositis on skin and muscle biopsy. His autoantibody screen was negative and he had a normal chest X-ray. A barium swallow demonstrated pharyngeal weakness. Oral prednisolone 60 mg and azathioprine 125 mg daily led to partial improvement in his rash and weakness. However, the dysphagia persisted and he lost further weight and so cyclophosphamide 75 mg daily was added.

Apart from the skin rash his symptoms became tolerable, but over the next 4 months he was admitted on 4 occasions with acute non-specific abdominal pain. On each occasion the pain spontaneously resolved and chest X-ray and abdominal examination, including testicular examination, were normal. On the final occasion in December 1987, the chest X-ray revealed multiple round opacities ranging from a few millimetres to almost 2 cm in diameter with hilar and mediastinal lymphadenopathy. At this time a small mass was palpable in the left testis and he was referred to us for investigation. Computed tomographic (CT) scan confirmed widespread mediastinal lymphadenopathy and opacities in both lung fields but also revealed enlarged para-aortic and retrocaval nodes and multiple liver deposits. Malignant cells, possibly of germ cell origin, were found in bronchoalveolar lavage fluid. Serum beta-human chorionic gonadotrophin and alpha-fetoprotein levels were raised markedly at 611 µg/l (normal less than 10) and 45 IU/l (normal less than 4). The diagnosis of metastatic testicular germ cell tumour with dermatomyositis was made and later confirmed by orchidectomy. He was treated with 4 courses of intensive chemotherapy using bleomycin, vincristine and cis platinum. His tumour markers became normal and CT scan showed only one 1 cm node in the para-aortic region. His symptoms and signs from dermatomyositis resolved and muscle enzymes returned to within normal limits. He did not receive bleomycin with the fourth course of chemotherapy in view of lower zone chest X-ray shadowing and the development of a restrictive pattern of lung function attributable to bleomycin. Unfortunately his dermatomyositis recurred in March 1988 and investigations revealed relapse of his germ cell tumour. The tumour did not respond

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to high dose carboplatin and high dose methotrexate causing death in July 1988.

Discussion

Dermatomyositis is a disorder mainly of skin, muscle and blood vessels in which characteristic erythematous and oedematous changes are associated with muscle weakness and inflammation.³ The cause is unknown but appears unrelated to infection, vaccination, immunisation or medication. The tumours associated with dermatomyositis occur most commonly in the lung, breast, female genital tract, stomach or colon.¹ Dermatomyositis has also been associated with reticulosis including Hodgkin's disease.⁴

This is the first case of dermatomyositis heralding a testicular germ-cell tumour. Review of the literature has revealed only two other case reports of dermatomyositis and germ-cell tumour. Tsuchiya *et al.*⁵ reported the case of a female patient with a mediastinal germ-cell tumour and lung

secondaries at the time of presentation with a three month history of dermatomyositis. Her skin rash improved during chemotherapy for her tumour but oral prednisolone was required to control her myositis. Three years later she was weaned off prednisolone without recurrence of her dermatomyositis. Fife *et al.*⁶ reported a patient in whom dermatomyositis, unresponsive to prednisolone, developed after successful treatment for metastatic testicular germ-cell tumour.

This case emphasizes the need to measure levels of beta-human chorionic gonadotrophin and alpha-fetoprotein and to examine the testes in young men with indications of possible malignancy, including dermatomyositis. The early diagnosis of non-seminomatous germ-cell testicular tumours is important as there is effective chemotherapy. However, the survival rate is related to tumour volume and tumour marker levels⁷ and unfortunately the majority who present with metastatic disease have lung involvement at the start of chemotherapy.⁷ Treatment of our patient's tumour led to rapid resolution of the symptoms and signs of dermatomyositis.

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