CASE REPORT

Breast cancer with dermatomyositis as initial presentation

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SUMMARY

Patients with breast cancer normally present with breast lump or abnormal mammogram. Dermatomyositis is rarely the first presentation. We present a case of a 63-year-old woman who had generalised dermatitis, progressive fatigue and muscle weakness. She was first diagnosed as dermatomyositis and subsequently breast cancer. Her rash and muscle weakness progressed drastically over a month. Tumescent mastectomy and axillary surgery was performed, which led to gradual regression of her dermatomyositis over six months. This case report emphasized in the benefit of early diagnosis and treatment of dermatomyositis and breast cancer. Pros and cons of tumescent mastectomy is discussed as well.

INTRODUCTION

Breast cancer is the most common cancer among women in Malaysia with the Age-Standardised Incidence Rate (ASR) of 39.3 per 100,000 populations.[1] In contrast, dermatomyositis (DMS) is an uncommon idiopathic inflammatory myopathy that primarily affects skeletal muscle and skin with well-characterized cutaneous findings. DMS is often associated with an increased risk of malignancy and may present as a paraneoplastic syndrome of an underlying malignancy. However, DM is rarely the initial presentation of breast cancer. Here, we describe a patient with breast cancer who initially presented with symptoms of DMS. Her DMS progressive deleteriously before treatment of her breast cancer and regressed following surgical management of the underlying malignancy.

CASE PRESENTATION

A 63-year-old woman with no known medical illness present with a month history of erythematous, scaly eruption over both metacarpophalangeal and interphangeal region of the dorsal aspect of the hand and extensor aspect of the elbows and knee. Flat erythematous lesions were also noticed over the chest and shoulders. Subsequently, she developed generalised malaise, joint pain and proximal myopathy of bilateral upper and lower limbs within a month. Laboratory tests revealed a normal ESR, C3 and C4 level. However, CK, white cell count, AST and ALT were elevated (Table I). In view of the rarity of DMS presenting in middle age, screening of malignancy was initiated. Meanwhile, oral prednisolone and azathioprine were started. Physical examination revealed a vague lump 2X2 cm in the outer lower quadrant of her left breast. Two months after onset of symptoms, she was still ambulant but generally weaker with worsening skin lesion. Ultrasound of breasts revealed a lobulated lesion of mixed echogenicity with increased vascularity in the outer lower quadrant of the left breast (fig 1). However, the mammogram showed neither a dominant mass nor a cluster of microcalcification in her breasts and there was no lymph nodes enlargement. Breast cancer was suspected and a core biopsy was offered for diagnostic purpose but she refused.

Ten weeks after the onset, her skin lesions and muscle weakness worsened and she was admitted to our hospital for intravenous hydrocortisone and immunoglobulin administration. The patient returned for core biopsy a month after discharged. At that time she was unable to walk and complained of dysphagia. A core biopsy from the left breast lump revealed an invasive carcinoma of no special type. Thorax, abdomen and pelvis computed tomography (CT) show neither sign of metastasis nor other occult malignancy. Surgical resection of the breast tumour was offered. However, the patient was still in denial state and she refused definitive surgical resection.

Fifteen weeks after onset, her general condition deteriorated further and she became bedbound. She required Ryles tube feeding due to a poor oral intake. Her general physical status deteriorated to ASA (American Society of Anaesthesiology) class IV and she was hospitalised. A tumescent mastectomy and axillary sentinel lymph nodes biopsy were performed. Histopathology showed that the tumour was completely resected with a clear margin and the lymph nodes were not involved. The tumour was a grade 3 (modified Bloom-Richardson system) invasive carcinoma of no special type with medullary features. 50% of the tumour cells were positive for estrogen receptors and occasionally positive for progesterone receptors. She was staged pT1N0M0.

Two weeks post-operatively, she developed an episode of DMS flare-up which subsided within a week. Her wound healed uneventfully and she was started with oral prednisolone, hydroxyurea and tamoxifen a month post-operatively. She regained muscle strength gradually over a five-months period and no skin lesions flare-up was observed. She was followed-up in both surgical and rheumatology clinic. However, she succumbed to death due to hospital pneumonia around one year after the surgery.

DISCUSSION

DMS is an idiopathic inflammatory myopathy, with an estimated prevalence rate of approximately 1 per 100000 in the general population.[2] The incidence peaks between the ages of 40 and 50, though it may affect individuals of any age. DMS is diagnosed and classified according to the criteria

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Table I: Laboratory values prior to the surgery, one months post-operatively and three months post-operatively

	Pre-op	2 months post-op	5 months post-op
White cell count (x10^9)	10.3	6.7	8.8
Creatine kinase (U/l)	6030	107	155
AST (U/l)	302	38	40
ALT (U/l)	121	23	35

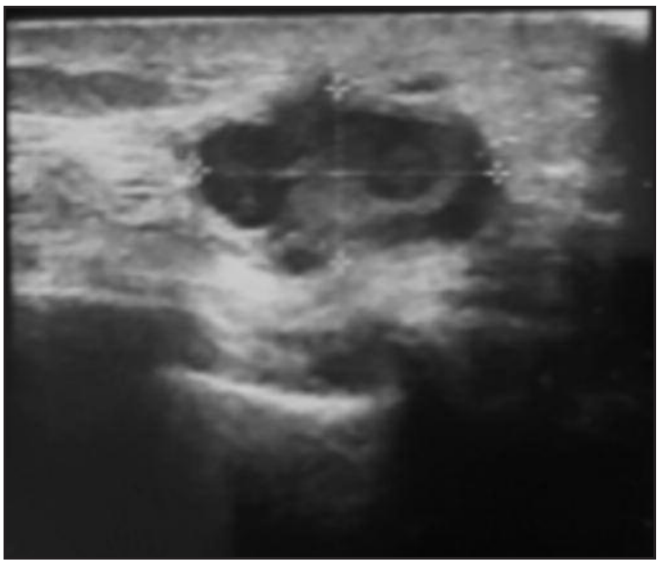

Fig. 1: Ultrasound image of the left breast lump.

set by Bohan and Peter in 1975.[2] Our patient, presented with a typical rash of DMS, symmetrical proximal muscle weakness and elevated in serum skeletal muscle enzymes, which fulfilled the criteria of probable DMS.

DMS is often associated with underlying malignancy especially in the elderly. Yang et al. reported that DMS is associated with an increased risk of malignancy in the ovary, breast, lungs, stomach, prostate, bladder, colorectal and cervical cancer in descending order.[3] The pathophysiology in which DMS is associated with malignancy remained unclear. Several studies hypothesized that the malignant cells induce the production of autoantibodies which cause DMS in the host.4 In general, DMS is associated with auto-immunity. A frequency of increased autoantibodies in individuals with malignancy seems to be higher than that in the general population with DMS.[4] This may explain the rapid progress of DMS in a patient with underlying malignancy.

The mainstay of therapy in patients with DMS is to control the cutaneous flare-up and to improve the muscle strength. Meanwhile, the treatment of breast cancer includes complete resection of a primary tumour with a negative margin to reduce the risk of local recurrence. Adjuvant treatment designed to treat micrometastatic disease. In our patient, the symptoms of DMS worsen drastically over 2-months despite aggressive immunosuppressive therapy. Her condition only improved after tumescent mastectomy with concurrent immunosuppressive therapy. These indicate the possibility of autoimmunity induced by the malignancy, in agreement with studies as aforementioned.

Tumescent mastectomy was opted as our patient had a high risk of complications if given general anaesthesia. Tumescent anaesthesia refers to a large amount of diluted local anaesthesia and epinephrine infiltration subcutaneously. This causes the surgical site to become swollen and tumescent, which favourably results in an extensive regional anaesthesia of the skin and subcutaneous tissue. Due to its vasoconstrictive property, epinephrine also results in a significantly less intra-operative bleeding and reduces the necessity for excessive cauterization. Tumescent anaesthesia was also shown to have the antibacterial effect that may reduce the development of post-operative wound infections.5 In addition, the expansion of skin, due to local infiltration analgesia, enhances skin closure that involved large defect. Tumescent mastectomy is not without complication: over-dosage may result in morbidity.[5] However, this can be prevented by administration of local anaesthesia within the recommended safe dosage.

CONCLUSION

In conclusion, malignancy should be taken into consideration when a patient in the older age group is newly diagnosed with DMS. Prompt diagnosis of malignancy and its removal may halt the progression of DMS and prevent the deterioration of the muscular function of the patient. Tumescent mastectomy is safe for patients who are at high-risk for a general anaesthesia.

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