ORIGINAL ARTICLE

A Review of Dermatomyositis Cases at Hospital Besar Kuala Lumpur 1989 – 1993

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Summary

Eight cases of dermatomyositis were seen in the Skin Department, Hospital Besar Kuala Lumpur between 1989 and 1993. There was one case of juvenile dermatomyositis. There was a majority of Chinese patients (87.5%). There were three patients (37.5%) with underlying malignancies, two of which (66.7%) were nasopharyngeal carcinomas; all were under the age of fifty years. It is important to screen all adult patients for underlying malignancies, even those in the younger age group.

Key Words: Dermatomyositis, Malignancy

Introduction

Dermatomyositis is an uncommon inflammatory myopathy of unknown cause associated with a characteristic skin rash. Bohan and Peter[1] defined the five diagnostic criteria of dermatomyositis as i) a typical skin rash of dermatomyositis; ii) proximal symmetrical muscle weakness; iii) elevation of serum muscle enyzymes; iv) electromyographic features of myopathy; and v) muscle biopsy evidence of an inflammatory myopathy. The same authors have also suggested that a definite diagnosis of dermatomyositis be made if 3-4 criteria are present plus rash; a probable diagnosis if 2 criteria plus rash are present; and a possible diagnosis if 1 criterion plus rash are present. In the adult, dermatomyositis may be associated with an underlying malignancy. This paper reports 8 cases of dermatomyositis seen at the Skin Department, Hospital Besar Kuala Lumpur, between January 1989 and December 1993.

Materials and Methods

Case records from the Skin Department, Hospital Besar Kuala Lumpur, were examined from January 1989 until December 1993 for patients who fulfilled at least 2 of the Bohan and Peter's criteria plus skin rash. This paper reports eight such patients (Table I). Patients were screened for underlying malignancy: this consisted of review by an ENT surgeon to exclude nasopharyngeal carcinoma and review by a gynaecologist of female patients in addition to routine full blood counts, serum creatinine, urea, electrolytes, liver function tests, serum immunoglobulins, urinalysis, chest X-ray, and physical examination which included a rectal and vaginal (in females) examination. Of the two patients who were not screened, one had childhood dermatomyositis and the other adult patient was lost to follow up before screening could be performed. All the patients had skin biopsies performed in the Skin Department. All patients had blood taken for serum muscle enzymes, namely creatinine kinase and lactate dehydrogenase. Electromyography was requested for all patients but only 5 patients had EMGs because of technical problems. Only two patients consented to having muscle biopsies performed.

Results

The majority of the patients (62.5%) were between 41-60 years of age. One case (12.5%) of juvenile dermatomyositis was seen.

Table I
Clinical Findings in 8 Patients with Dermatomyositis

	Age	Sex	Race	Presentation	Evidence of Weakness	Malignancy	Remarks
Case 1	35	F	C	Heliotrope rash Gottron's papules Periorbital edema Proximal myopathy	raised muscle enzymes EMG characteristic	NPC	DXT steroids defaulted after 2y
Case 2	54	F	C	Photosensitive rash on face, arms Periorbital edema Proximal myopathy	raised muscle enzymes	nil	steroids well after 3y
Case 3	78	F	C	Photosensitive rash on face, arms, back Proximal myopathy	raised muscle enzymes EMG normal	nil	steroids defaulted after 3m
Case 4	44	M	C	Heliotrope rash Gottron's papules Periorbital edema Proximal myopathy	raised muscle enzymes muscle biopsy normal	NPC	DXT steroids well after 10y
Case 5	4	F	C	Heliotrope rash Gottron's papules Proximal myopathy	raised muscle enzymes EMG characteristic muscle biopsy characteristic	nil	well calcinosis steroids
Case 6	47	M	C	Photosensitive rash on face, arms Periorbital edema Gottron's papules Proximal myopathy Dysphagia	raised muscle enzymes EMG characteristic	occult 1	steroids azathioprine died after 3y metastases to cervical nodes, liver
Case 7	45	F	C	Photosensitive rash on face, arms Ragged cuticles Alopecia Proximal myopathy	raised muscle enzymes	nil	defaulted after 1m
Case 8	60	F	My	Heliotrope rash Photosensitive rash on face, arms, and back Ulcers on face Proximal myopathy Dysphagia Dysphonia	raised muscle enzymes EMG characteristic	nil	steroids azathioprine died after 2m pneumonia

Key: M=male, F=female, C=Chinese, My=Malay, EMG=electromyography, y=years, m=months NPC=nasopharyngeal carcinoma, DXT=radiotherapy

Seventy five per cent of the patients were female. The majority of patients (87.5%) were Chinese, with one Malay patient (12.5%). Twenty-five per cent of the patients attending the Skin Department are Chinese, 50% are Malays and 25% are Indians (personal communication from Dr. T. Ganesapillai, Head of Skin Department).

All the patients presented with either the typical heliotrope rash (37.5%), a photosensitive rash (50%), or both (12.5%). Four (50%) of the patients had Gottron's papules (violaceous papules over the knuckles), four (50%) had periorbital edema, and one (12.5%) patient had alopecia and ragged cuticles. The case of childhood dermatomyositis developed subcutaneous calcinosis around her knees. All the patients presented with proximal myopathy, which was confirmed in all cases by elevation in serum muscle enzymes (CK and LDH). Four patients had characteristic EMG findings, and one had a normal EMG. Two patients had muscle biopsies, one of which showed changes consistent with inflammatory myositis. Two (25%) of the patients presented with dysphagia in addition to the proximal myopathy.

Skin biopsies were performed on all the patients. Seven showed changes consistent with the diagnosis of dermatomyositis, namely hyperkeratosis, atrophy and flattening of the epidermis associated with vacuolar degeneration of the basal layer, dermal edema and a perivascular lymphohistiocytic infiltrate. One was reported as "nonspecific dermatitis". Immunofluorescent examination of skin biopsy specimens revealed granular deposits of IgG, IgA, IgM, and C3 in various combinations along the dermoepidermal junction; these findings are nonspecific.

Three of the patients (37.5%) had an underlying malignancy. All were under fifty years of age. Two of the malignancies (66.7%) were nasopharyngeal carcinomas, and the third patient had an occult primary with metastases to cervical lymph nodes and liver. In one patient the dermatomyositis preceded the diagnosis of NPC by a month, and the NPC was in fact found on routine screening for malignancy. In the other patient, the NPC preceded the dermatomyositis by two months. Both cases of NPC were confirmed by biopsy. Dermatomyositis preceded the diagnosis of malignancy by three years in the patient who had an occult primary with metastases.

Two patients died during follow-up and both were adults; one died from pneumonia, and the other succumbed to his malignancy, which was only revealed when he presented with PR bleeding and metastases to the cervical lymph nodes and liver 3 years after the diagnosis of dermatomyositis was made. He had previously defaulted from follow-up. The two patients with NPC responded to radiotherapy and systemic steroids; one was lost to follow-up 2 years after successful treatment, and the other remains well 10 years after the initial diagnosis was made. The remaining patients continued to attend follow-up, with the exception of two who defaulted (Cases 3 and 7).

All the patients with the exception of two responded to systemic steroids, with improvement in the cutaneous lesions and proximal myopathy. The two who required azathioprine in addition to the steroids eventually died. Cases 2, 4 and 5 continue to attend followup at the skin department and are on low dose maintenance oral steroids.

Discussion

Our patients in this study have confirmed similar findings in other studies, in particular those reported from the Far East. There is a female preponderance[2] and the majority of the patients were Chinese[3]. However, the number of patients in our study was too few to be representative.

Over half of the patients (62.5%) presented with a photosensitive rash. This finding underlines the importance of having dermatomyositis as a differential diagnosis whenever a patient with photosensitivity presents for investigation.

One patient had juvenile dermatomyositis which is thought to be a distinct entity from the adult type[4]. The prognosis is good except for contractures and calcinosis which can be disabling.

Three (37.5%) of the patients had malignancies, which is in keeping with other series which have reported the incidence of malignancy in patients with

dermatomyositis as varying from less than 10% to over 50%[5,6]. Although the association with malignancy is still controversial, most studies suggest that there may be an increased frequency in malignancy in patients with dermatomyositis[5,6,7,8,9]. Two thirds of the malignancies in our study were due to nasopharyngeal carcinoma, an association which has been reported in other Asian series[3,10,11]. Nasopharyngeal carcinoma should be searched for thoroughly and excluded in all adult patients with dermatomyositis in our part of the world, especially if they are Chinese.

It has been reported that underlying malignancy is common only in patients presenting in the fifth and sixth decades[12,13]. In our study, the malignancies occurred in patients who were relatively young, i.e.<50 years of age. One of the patients with NPC was 35 years of age. This underlines the importance of investigating for malignancy in all adult patients with dermatomyositis, regardless of age[3].

Extensive investigations in the absence of abnormal clinical findings are controversial because in the majority of cases malignancy is discovered on the basis of clinical signs, symptoms, or routine screening tests[14,15]. However, there are anecdotal reports of nondirected tumour searches resulting in the discovery of an underlying malignancy[16]. In fact one of our patients, Case 4, had NPC diagnosed on the basis of a post nasal space biopsy performed routinely as part of the workup following the diagnosis of dermatomyositis made in the preceding month. Suggested routine investigations for the screening for malignancy[15,17] are 1) complete medical history, systems review, and physical examination including rectal, vaginal (in females), and breast (in females) examination; 2) complete blood count, ESR, routine serum electrolytes, urea, creatinine, liver function tests, serum protein electrophoresis, fecal occult blood testing, urinalysis; 3) chest X-ray; 4) mammography (in females); 5) review by a gynaecologist and pelvic ultrasound in females[15,18]; and 6) review by an ENT surgeon, post nasal X-ray and biopsy[3]. In addition, any abnormality in these screening tests should be investigated further, and more extensive investigations should be done in patients who have suffered from a previous malignancy, in those who respond poorly to treatment, or who present with an unexplained relapse of the dermatomyositis[15].

In our study, the interval between dermatomyositis and malignancy varied between one month and three years. In one study, the longest interval was eight years[19]. In the majority of patients the malignancy and dermatomyositis occur within a short period of time of one another[12]. The cancer can precede, occur concurrently with, or follow the dermatomyositis[9].

Prognostic factors for adverse outcome have been reported as the presence of malignancy, advanced age, severity of the myositis, the presence of dysphagia, resistance to treatment, fever, hyperleucocytosis, a high ESR, extensive cutaneous lesions on the trunk, severity of initial clinical presentation, and long delays in starting treatment[20,21]. Juvenile dermatomyositis has a good prognosis[4]. The two patients who died in our study had a number of poor prognostic factors: one had a malignancy; both had dysphagia in addition to the proximal myopathy; both had difficulty walking due to the severity of the myopathy on initial presentation; both had extensive cutaneous lesions on the face, trunk and limbs; and both required azathioprine in addition to systemic steroids.

Systemic steroids are the mainstay of therapy, regardless of the absence of controlled data[2]. Our patients noted improvement in cutaneous lesions, muscle power and muscle enzymes on steroids, and those that needed immunosuppressives in addition did poorly.

Conclusion

Eight patients with dermatomyositis were seen in the Skin Department, Hospital Besar Kuala Lumpur between 1989 and 1993. Based on our results, the screening for underlying malignancy in all adult patients is recommended, even the younger adult age group. Nasopharyngeal carcinoma in particular should be thoroughly searched for, given the high incidence of this malignancy in the Chinese population in our part of the world. Some would even advocate repeated ENT examination as part of longterm follow-up[3]; indeed, all the patients should be followed up longterm and assessed periodically for underlying malignancy.

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