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Original research article

Nasopharyngeal carcinoma in dermatomyositis patients: A 10-year retrospective review in Hospital Selayang, Malaysia

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ABSTRACT

Aim: The objective of our review is to investigate the association between dermatomyositis patients and nasopharyngeal carcinoma (NPC) together with the clinical presentation of the patients and their management in otorhinolaryngology.

Background: NPC is a malignant disease with good prognosis on early diagnosis. However, the relationship between the dermatomyositis and NPC and its management is not well defined.

Materials and methods: A 10-year retrospective review of case records of 21 dermatomyositis patients seen in Otorhinolaryngology Department of Hospital Selayang from January 2000 to November 2010.

Results: These patients ranged from 19 to 74 years old and a total of 8 (38%) out of 21 adults with dermatomyositis were detected to have malignancy. Five out of 8 patients had NPC (62.5%). The mean age of patients with NPC and dermatomyositis was 48 years. NPC is diagnosed in 4 out of 5 patients (80%) in the first year of diagnosis of dermatomyositis. The clinical findings of the examination of nasopharynx ranged from hyperemia to exophytic nasopharyngeal mass. Histologically, it is only related to NPC of WHO types II and III.

Conclusions: There is a strong relationship between dermatomyositis and malignancy, especially NPC. Clinicians should have a high index of suspicion for malignancy in all

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dermatomyositis patients. Rigid nasoendoscopies and biopsies, serum Epstein–Barr viral capsid IgA antibody and imaging studies are helpful in detecting NPC in dermatomyositis patients.

1. Background

Dermatomyositis is defined as an idiopathic inflammatory myopathy with clinically distinctive cutaneous manifestations.[1] However, when the symptoms are associated with malignancies, the condition is known as paraneoplastic syndrome which represents the clinical manifestation of the remote and indirect effects produced by tumor metabolites or other products.[2]

Dermatomyositis is highly associated with various types of malignancies, namely carcinoma of nasopharynx, breast, lung, colorectal, uterus and non-Hodgkin lymphoma.[3] 10–47% of dermatomyositis patients have an underlying malignancy.[4–6] Out of these, nasopharyngeal carcinoma (NPC) had been reported as the most common type of cancer related to dermatomyositis in Asia[3,4,7] where NPC is found in more than 40% of the total cases of malignancies in dermatomyositis patients.[3,4,7,8] The onset of NPC in dermatomyositis patients is usually within the first 5 years from the diagnosis of dermatomyositis.[5]

2. Aim

The objective of our review is to investigate the association between dermatomyositis patients and nasopharyngeal carcinoma (NPC) together with the clinical presentation of the patients and their management in otorhinolaryngology.

3. Materials and methods

In this 10-year retrospective study, the case records of 21 patients who were diagnosed with dermatomyositis from January 2000 until November 2010 were reviewed using the hospital's electronic medical records. All of the patients were confirmed to have dermatomyositis by physicians based on the clinical features and investigations including muscle biopsy.

The criteria for diagnosis of dermatomyositis were based on Tanimoto's classification and diagnostic criteria for polymyositis and dermatomyositis.[9] Skin lesions include heliotrope rash, Gottron's sign and erythema or purpura on extensor surfaces of the extremity joints.[9] The items included in the criteria were proximal muscle weakness, muscle grasping and spontaneous pain, non-destructive arthritis or arthralgia, elevated creatinine kinase or adolase, presence of systemic inflammatory signs, myogenic changes on electromyography, positive anti Jo-1 antibody and pathologic findings compatible with inflammatory myositis.[9] Dermatomyositis were diagnosed if patients had at least one out of three skin lesions and four out of eight other criteria items.[9]

Patients diagnosed to have dermatomyositis were referred to the otorhinolaryngology department for head and neck examination and diagnostic work up to detect head and neck malignancy.

The diagnostic work out includes thorough history taking, general examination of the ear, nose and throat followed by flexible nasopharyngolaryngoscopy and biopsy of the post nasal space of any suspicious lesion supplemented by fine needle aspiration of any cervical lymph node.

All patients who had no detectable head and neck malignancy were reviewed again by the otorhinolaryngologist particularly if the work up for any malignancy was negative and the dermatomyositis symptoms frequently relapsed, or for those who showed no improvement despite being on immunosuppressive treatment.

4. Results

A total of 21 dermatomyositis patients were reviewed from January 2001 until November 2010. There were 10 male (48%) patients and 11 female (52%) patients. The youngest patient was 19 years old and the oldest patient was 74 years old. The mean age of presentation was 43.8 years old. Eight of the dermatomyositis patients were below 40 years old (38%) and the remaining 13 were above 40 years old (62%).

Eight out of 21 patients (38%) were detected to have malignancies (breast, nasopharynx and lymphoma) and 5 out of the 8 (62.5%) had NPC. Among those who were diagnosed to have NPC, the male to female ratio was 4:1.

The mean age of the NPC patients with dermatomyositis was 55.2 years (range 39–74 years). The maximum age incidence was in the fifth decade. In our center, 8 dermatomyositis patients were below 40 years old (3 males and 5 females) and only 1 (12.5%) was related to malignancy (NPC).

Meanwhile, 7 out of 13 patients (54%) aged 40 years and above had a malignancy. There were 7 males and 6 females in this group. Out of these 7 who had a malignancy, 4 patients (31%) had NPC (1 female and 3 males), 2 patients had breast carcinoma and 1 had lymphoma (Fig. 1).

There were a total of 5 patients with nasopharyngeal carcinoma. Three were detected concurrently with the diagnosis of dermatomyositis while the other 2 patients were diagnosed during subsequent follow ups which ranged from 2 months to 38 months. Thus, the diagnosis of NPC was made in as many as 80% of the patients within the first year of the diagnosis of dermatomyositis (3 concurrently and 1 after 2 months during follow up). The fifth patient was diagnosed 4 years from onset of dermatomyositis. One of them presented with epistaxis and otitis media and another with a submandibular mass. It is