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Dermatomyositis Revealing Nasopharyngeal Carcinoma

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Abstract

Dermatomyositis (DM) may be an opening event of neoplasia. Nasopharyngeal carcinomas (NPCs) were rarely associated with this phenomenon. We report the case of a 42 year old patient with lilac table eyelid erythema lasting for six months with the notion of earache and recent epistaxis. The diagnostic inquiry concluded at a stage T2 N1 M0 NPC associated with DM. After 6 courses of chemo and radiotherapy skin symptoms disappeared completely with a clear regression of the tumor.

Key words: Dermatomyositis, Nasopharyngeal carcinoma.

Background

Dermatomyositis DM may be idiopathic, but is also commonly related to malignancy. Some epidemiological studies showed that 32% of DMs were associated with cancers. Most of them due to ovary, lung, pancreas, breast or gastrointestinal tract neoplasias [1]. The first observation of nasopharyngeal carcinoma NPC associated with DM was reported in 1969 [2]. The global estimation of DM and NPC association was at 0.086% [3].

Observation

A 42-year-old male patient, presented with swelling redness around eyes evolving for the last 6 months and lilac erythema of the peri-orbital region and the cheeks (Figure 1). He had symmetrical proximal muscle weakness and myalgias for the last 6 months. In addition, he complained of intermittent nasal stuffiness for the last 3 months along with episodes of epistaxis and this symptomatology was associated with rebel right earache. A clinical diagnosis of dermatomyositis was made. The nasal and ear symptoms were evaluated in consultation with the ENT department. The ENT examination objectified a secretory otitis media. Biological investigations showed high creatine phosphokinase (CPK) and lactate dehydrogenase (LDH) levels (420 and 410, respectively; normal highest values were at 260 and 268 respectively). CT scan of the nasopharynx showed a homogenous enhancing soft tissue mass in the nasopharynx, thickening of the posterior wall of the upper nasopharynx right lateralized reaching the retro pharyngeal space and filling the fat of the anterior para pharyngeal space with crossing the basilar fascia. There was no bone destruction. The histopathological examination showed an appearance characteristic of undifferentiated carcinoma. On the basis of histomorphology, a diagnosis of undifferentiated non-keratinizing nasopharyngeal carcinoma was retained. The tumor was ranked T2 N1 Mx and treated with chemoradiotherapy. After completing six cycles of chemoradiotherapy, the nasopharyngeal mass significantly reduced the patient's skin symptoms disappeared. The patient has now been off all treatment for 6 months with no evidence of either nasopharyngeal malignancy or dermatomyositis relapse.

Figure 1: Lilac erythema of the peri-orbital region and the cheeks.

Discussion and comments

Most practitioners should be aware of DM and neoplasia association. Even uncommon nasopharyngeal cancer should be suspected in specific guidance signs. Our observation has indeed highlighted a rare association. It focuses on the parallelism between the evolutions of both diseases.

Commonly the activity of DM reflects that of the malignancy. Authors have observed that, after remission following therapy for

NPC, the symptoms of DM disappeared or improved. The relapse of DM was reported to be correlated to local regional recurrence or presence of metastases. When the patient enters a period of cancer remission, their DM activity can be used to monitor for early relapse [4]. There is, however, a case where DM does not mirror the treatment and response of the malignancy [5]. This was noted in our present patient. The prognosis of NPC with DM has not been shown to be different from NPC in general, despite immunosuppressive therapies. In Hu et al.'s case-control study of 90 patients, actuarial survival at 5 and 10 years was 50% and 34.5% respectively for the group of NPC patients with DM versus 57% and 55% respectively for the group of NPC patients without DM [3].

Conclusion

NPC with paraneoplastic DM is a rare but non-specific entity. The course of DM may follow that of the NPC, but may also evolve independently of the cancer.

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