



AUTOIMMUNE CONNECTIVE TISSUE DISEASES

DERMATOMYOSITIS AS PARANEOPLASTIC SYNDROME IN PATIENTS WITH UTERINE TUMOR

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BACKGROUND: Dermatomyositis (DM) is an idiopathic inflammatory myopathy, more common in women (2: 1) with a peak incidence between 40 and 50 years. It's clinically characterized by proximal and symmetrical muscular weakness and cutaneous lesions, with Gottron's papules, heliotrope, shawl or "V" sign, erythema and periungual capillary ectasias, and, more rarely, vesiculobullous formations. The frequency of cancer is markedly increased in dermatomyositis, especially in elderly patients. Up to 32% of the patients have an overlying disease.

OBSERVATION: 51 years old, female, with diffuse erythema, violaceous and slight scaling on the face and upper portion of the thorax, desquamative erythematous-violet papules in proximal metacarpophalangeal and interphalangeal joints, oral ulcers and proximal muscular weakness, starting 10 months ago. Three-month vaginal discharge with bloody appearance associated with septal cystic mass, pelvic cavity, posterior to the bladder, midline and focal endometrial thickening and hematometrial. Increased muscle enzymes (aldolase and creatine phosphokinase). Anatomopathological examination of the endometrium: cohesive grouping of atypical cells with bulky, hyperchromatic nuclei and eosinophilic cytoplasm. Anatomopathological examination of the skin: psoriasiform acanthosis, basal layer degeneration foci, thickening of the basement membrane and papillary dermis with melanophages. Initiated prednisone 1mg / kg / day, with important improvement of muscular weakness, but with maintenance of the skin lesions.

KEY MESSAGES: Malignancy in patients with dermatomyositis is 5-7 times more frequent in relation to general population, behaving as paraneoplastic syndrome. The adenocarcinomas of the cervix, lung, ovaries, pancreas, bladder and stomach correspond to approximately 70% of the cancers associated with inflammatory myopathies. Some possible risk factors for association with neoplasias are: leukocytoclastic vasculitis, presence of anti-p155 / p140 or anti-p1557 autoantibody, excessive elevation of muscle enzymes, male sex, DM with vesiculobullous manifestation, advanced age at the time of the diagnosis of DM.

