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RESEARCH ARTICLE

PARANEOPLASTIC DERMATOMYOSITIS REVEALING OVARIAN CANCER: ABOUT A CASE

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Abstract

Dermatomyositis (DMs) is a systemic autoimmune disease affecting mainly muscle and skin. It may be associated with cancer in a paraneoplastic setting with a frequency ranging from 18 to 32% [1]. various types of cancer are described in the literature; however, the association with gynecological cancers, especially in women, is more frequent. the incidence of ovarian cancer in dermatomyositis in women is 13.3%; it is higher than in the general population (1%) [2]. We present the case of a patient with paraneoplastic dermatomyositis, who presented with a typical clinical symptomatology of paraneoplastic dermatomyositis, confirmed by other complementary examinations (creatine phosphokinase (CPK) + EMG (electromyogram) + skin biopsy). Etiological exploration by the ovarian specific tumor marker CA 125, and pelvic ultrasound completed by pelvic MRI oriented the diagnosis towards ovarian adenocarcinoma. Our results suggest that a complete gynecological workup including gynecological clinical examination, CA 125, CA 15-3 and pelvic ultrasound should be routinely performed in any woman over 40 years of age with DM.

Introduction:-

Dermatomyositis (DMs) is a rare connective tissue disorder that combines characteristic skin involvement and muscle involvement

Predominantly in the limbs. It is associated with underlying neoplasia occurring before, simultaneously, or after the diagnosis of cancer. The type of neoplasia associated with DMs varies by study and geographic region of the patient. Gynecological cancers, especially ovarian cancer, are the most common in women.

We report a case of a 68-year-old woman who was hospitalized in dermatology department for dermatomyositis and whose paraneoplastic workup revealed an ovarian adenocarcinoma.

Case presentation

The patient was 68 years old, with a history of arthrosis , and a heart rhythm disorder on a beta-blocker. She presented 21 days before her admission pruriginous erythematous skin lesions, associated with muscular weakness in a context of general asthenia. The general examination revealed an asthenic patient, tachycardia at 100 beats/min;

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polypneic at 32 cycles/min, and apyretic . the cutaneous examination find a poikilodermic erythema of the face and the neckline (**Figure 1**) , a Flagellate erythema of the back (**Figure 2**) , a erythema in the extensor surfaces of the 2 knees **(Figure 3)** , sign of the manicure, a erythema in band of the back of the hands and fingers. The skin signs were associated with a muscle deficit predominantly at the proximal level.

The investigations (biological, radiological, pathology) resulted in the diagnosis of ovarian adenocarcinoma associated with a paraneoplastic dermatomyositis which was initially suspected by clinical signs, then confirmed by the association of the increase in muscle enzymes (LDH at 1402 UI/l, CPK at 17923 IU/l, aldolase at 14,4U/l) ant the electromyogram (EMG) abnormality which showed a myogenic tracing. the skin biopsy came back in favor of a dermatomyositis.

The etiological work-up revealed an increase in tumour markers (CA 15-3 =70.4 ; CA 125=87.1), ultrasound revealed a suspicious ovarian mass, and a complementary pelvic MRI was performed, showing two bilateral latero uterine solid cystic formations of probable ovarian origin locally infiltrating classified ORADS 5 in relation to cystadénocarcinomes . The mammography completed by breast MRI reveals a lesion of the right breast classified as Birads 3 of the ACR **(Figure 4).**

The serology of hepatitis B was positive in our patient, so she was initially put on plaquenil for the skin involvement, Intravenous immunoglobulin (IVIG) was then administered for 2 days (1 g/kg/d) , followed by cortcotherapy 1mg/kg/d at day 15 of antiviral treatment with tenofovir.

The patient was transferred to gynecology; she had an exploratory laparotomy with bilateral adnexectomy; the anatomopathological study confirmed the diagnosis of ovarian adenocarcinoma. She completed tree courses of adjuvant chemotherapy, after her 3rd session she died of septic shock.

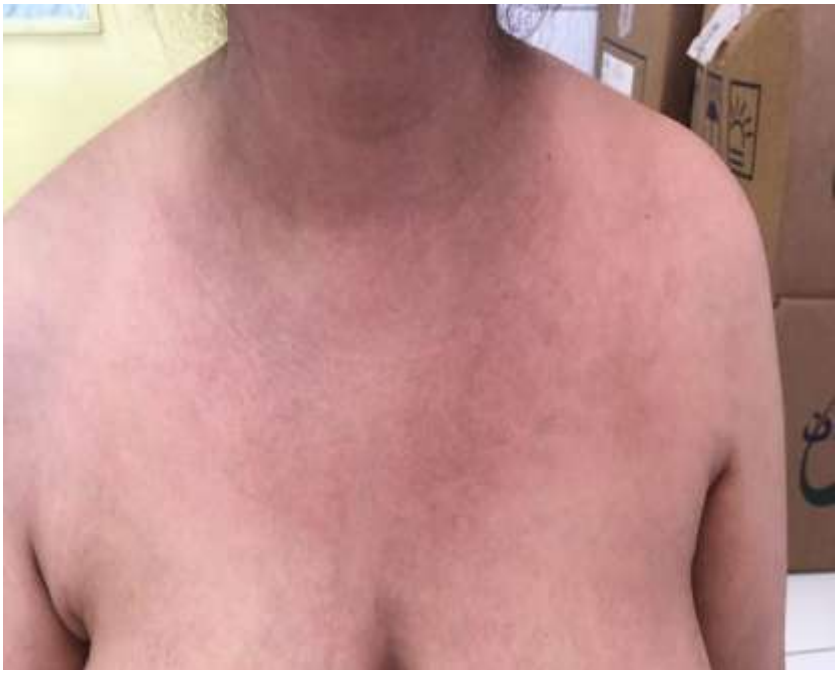

Figure 1:- Shawl Sign.

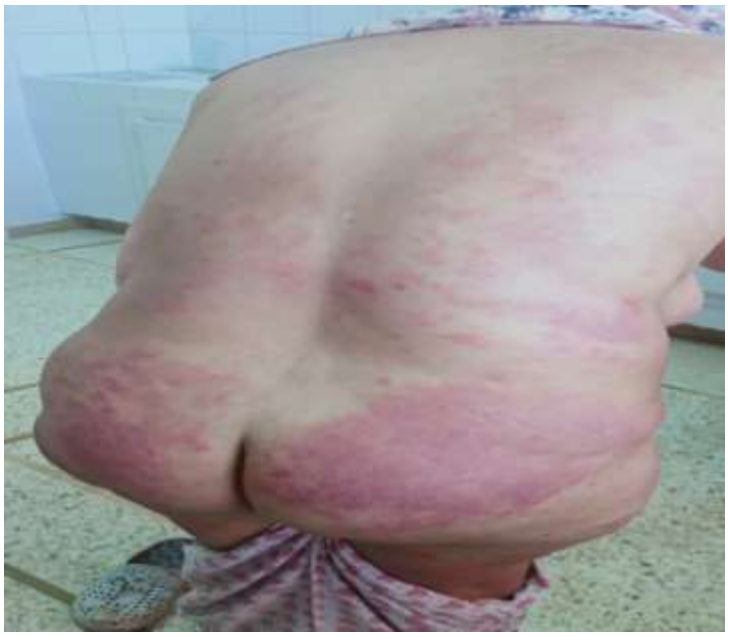

Figure 2:- Flagellate erythema of the back.

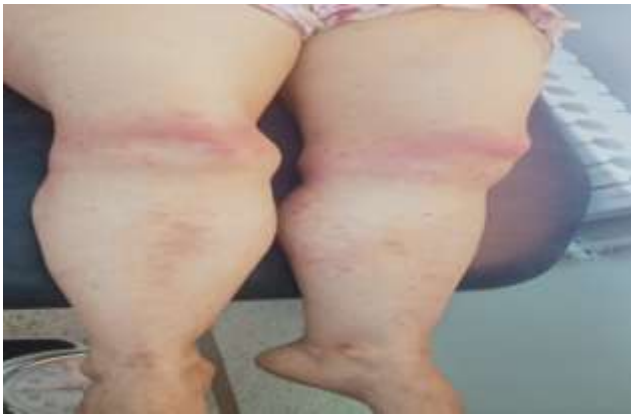

Figure 3:- Erythema of the knee extension faces.

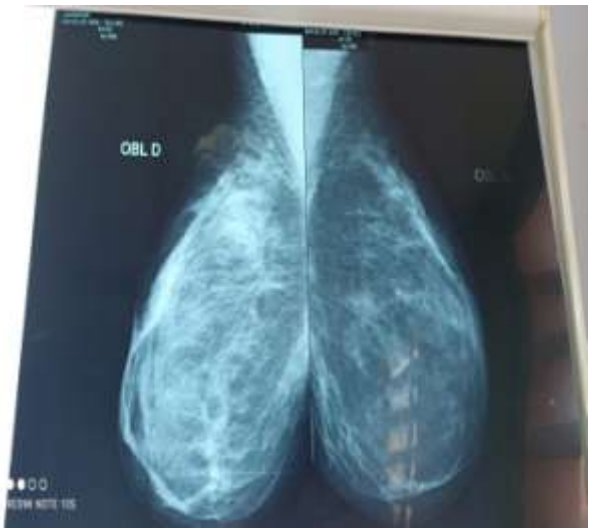

Figure 4:- Mammography (ACR 3 lesion of the right breast).

Discussion:-

Dermatomyositis is a rare disease, predominantly female with an estimated annual incidence between five and ten cases per million inhabitants and the annual prevalence between 06 and 07 cases per 100 000 people [3]. It is a connective tissue disease of unknown pathophysiology and etiology characterized by inflammatory involvement of the skin and striated muscles responsible for skin lesions and muscle weakness.

The most common malignancies associated with dermatomyositis are ovarian, breast and colon cancer, melanoma and non-Hodgkin's lymphoma [4]. Ovarian cancer was found in 8.3% of patients with dermatomyositis [4]. Cancer is most commonly diagnosed simultaneously or during the first year after the diagnosis of dermatomyositis although there continues to be an elevated risk of malignancy even after 5 years.

The pathogenesis is that of a microangiopathy, characterised by complement deposition in the microvasculature of the muscle and antibody mediated capillary destruction. Confirmation of the diagnosis requires tissue biopsy of the skin or muscle. Skin biopsy reveals an interface dermatitis on immunofluorescence, with vacuolar changes at the dermal-epidermal junction and a perivascular lymphocytic inflammatory infiltrate [5]. Muscle biopsy demonstrates a perivascular inflammatory infiltrate, capillary obliteration, capillary thrombosis, endothelial cell damage and perifasicular atrophy [6].

Two theories have been put forward: hormonal and immunoallergic. In the first hypothesis, it is the tumor that secretes hormonal polypeptides that are biologically active but inappropriate in terms of homeostasis. These polypeptides would be responsible for the different clinical syndromes of endocrine type. In the Immuno-allergic theory, the paraneoplastic syndrome would be the result of cross-reaction of antibodies produced against tumor antigens, with normal tissues having a similar structure .

The diagnosis of DM is based on five criteria according to Bohan and Peter [7]: progressive and symmetrical muscle weakness of the limb girdles and neck flexors; dermatological signs (periorbital heliotropic rash with edema, Grotton papules); muscle biopsy in favor of myositis; elevation of serum muscle enzymes showing muscle necrosis. Creatine kinase (CPK) is the most specific muscle enzyme, it provides information on objective suffering of the muscle tissue. They are increased in 75 to 85% of cases, however a normal level should not rule out the diagnosis; electromyographic profile in favor of muscle damage. The presence of three or four of these criteria, in addition to the rash, makes the diagnosis of DM, and the presence of two criteria associated with the rash is highly suggestive of DM.

The presence of 4 criteria allowed us to make the diagnosis in our case (skin signs; muscle weakness, increase in muscle enzymes; EMG abnormality in favor of muscle damage).

Malignancies are usually identified through a history, physical exam, basic labs and/or age-appropriate screening tests. In women, a transvaginal ultrasound and CA125 may be helpful to identify ovarian cancer. Although many studies have looked for traits that would enable us to predict when dermatomyositis has a higher risk of being associated with cancer, little has been achieved. Of all the parameters studied, it would appear that those most consistently associated with a higher risk of cancer in patients with adult dermatomyositis are male gender and more advanced age. One clinical trait that has repeatedly been related with paraneoplastic dermatomyositis in the literature is skin necrosis [**8**]. the only predictive factor of a paraneoplastic dermatomyositis in our patient was his advanced age of 68 years; necrotic lesions were absent.

By sex, the most common cancers associated with adult dermatomyositis in women are breast and ovary; in men are lung, colon and rectum. A targeted history must be taken from all patients and a complete physical examination performed. Blood tests (including prostate specific antigen in men and CA125 in women), urinalysis, and fecal occult blood should also be performed. In women, mammography and transvaginal ultrasound are also recommended.

Patients with dermatomyositis are advised to be screened for malignancy for the first 3 years after diagnosis of dermatomyositis, except for ovarian cancer, which can occur even 5 years after the diagnosis of dermatomyositis. Thus, patients are advised to have a Ca-125 test every 6 months; However, Ca-125 screening for ovarian cancer in dermatomyositis has a sensitivity of 50% [**9**].

The use of prednisolone at 1mg/kg up to a maximum of 80mg/day is recommended as first line treatment for dermatomyositis. This should be continued for 4-6 weeks with careful withdrawal. Muscle enzymes tend to normalize within 6 weeks; however, improvement in muscle strength tends to be delayed for several months [**10**]. . If there is not an adequate response or if treatment is to be prolonged, methotrexate may be beneficial at doses of 7.5 to 15 mg per week. in our case, the treatment was initially based on plaquenil for skin involvement; hepatitis B serology was positive with a positive viral load, which led to the choice of a course of immunoglobulin 1g/kg/d IV for 48 hours; prednisone was introduced after 2 weeks on tenofovir

The delay in diagnosis could be due to the insidious onset and slow progression of the dermatomyositis and the limitations of imaging studies. Ninety-four percent of patients were diagnosed as being at stage III or IV, which makes their prognosis extremely poor [1]. Mortality due to malignant ovarian tumors associated with dermatomyositis was 100%, and the mean survival time average since diagnosis was 11 months (range 0 to 28 months) [1]. the prognosis in our patient was poor, she died by her ovarian cancer at 4 months of the beginning of the cutaneous and muscular symptomatology of her dermatomyositis, which reminds the gravity of the paraneoplastic dermatomyositis in the old woman. the dermatologists and the gynecologists must always be vigilant in front of this diagnosis.

Conclusion:-

Dermatomyositis is associated with ovarian cancer in women over 45 years of age, its research by the tumor marker CA 125, by pelvic ultrasound should be systematic in any woman who consults for dermatomyositis.

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