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Ovarian Carcinoma Following Dermatomyositis Diagnosis: A Case Report And Review Of The Literature

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Abstract

Background: Dermatomyositis (DM) is a rare paraneoplasic neurological syndrome (PNS) that may precedes the development of malignancies. It´s an uncommon inflammatory myophaty with characteristic cutaneous manifestations. The association between DM and cancer has long been recognized but is becoming more clearly delineated. Several studies have documented an increased rate of malignant disease associated with DM over that observed in the general population. The incidence of ovarian cancer developing in the setting of DM was reported to be as high as 13.3 % in one series.

Case report: We report a case of a 50 years old postmenopausal woman who developed a progressive muscle weakness abd cutaneous rash that was finally diagnosed as DM. On CT abdomen exploratoin a complex right adnexal mass was described. CA -125 was elevated. Exploratory laparotomy found a nonadherent enlarged right ovary with no surface excrescences or peritoneal spread. Histologically was high-grade serous papillary adenocarcinoma.

Conclusion: Screening for tumours is important in patients with a previous diagnosis of DM or other PNS.

Keywords: Dermatomyositis, Ovarian cancer, Paraneoplasic neurological syndrome.

Introduction

DM is an inflammatory myopathy with well-defined pathological features including the microangiopathy. DM is frecuently associated with concomitant cancers, as many as 25% of cases are associated with an occult neoplasm [1]. This paraneoplastic syndrome may precede the development of malignancies such as lung, breast, or ovarian cancer by several months and even years [2]. It is a rare inflammatory myopathy with characteristic cutaneous manifestations. Criteria for this disease are progressive, proximal and symetrical weakness, and increased concentrationof muscle enzymes, an abnormal electromyogram, an abnormal muscle biopsy sample, and compatible cutaneous disease [3]. A rash that occurs in a shawl-like distribution is characteristic of this disease, as is proximal muscle weakness and elevation of serum creatine kinase levels. The cutaneous manifestations include heliotrope rash, cuticular changes, a photo distributed erythema, and a scaly alopecia. The pathogenesis of the skin lesions or dermatomyositis is not understood.

The association between DM and cancer has long been recognized but is becoming more clearly delineated. Several Scandinavian studies have documented an increased rate of malignatn disease associated with DM over the observed in the general population [4,5]. The incidence of ovarian cancer developing in the setting of DM was reported to be as high as 13.3 % in one series [2].

Screening for tumours is important in patients with a previous diagnosis of DM or other PNS as the early diagnosis of the tumour directly affects prognosis [6]. Because of the rare correlation between ovarian carcinoma and the DM, we present our case and review the literature-

Case

50 years-old white woman, who was 4 years postmenopausal was referred to the emergency department with muscle weakness in arms and legs. Furthermore, she had a pruritic skin rush for three months believed to be secondary tu sun exposure. Physical

examination found cutaneous lesions that were erythematous, slightly scaly, macular and papular, involving the neck, upper chest, front, back, arms and hands (Figure 1).

There was violaceous facial erythema with a heliotrope eyelid rash and deep violaceous color on the extensor surfaces of fer fingers, elbows and knees. She had loss of proximal muscle strength. The medical history included bipolar disorder, appendicectomy and carpal tunnel syndrome surgery. Mammogram was normal and her previous Papanicolaou smear and pelvic examination were normal 2 years before admission. Laboratory findings included creatinine phosphokinase of 3572 U/mL (normal 0-170), deshidrogenase lactate of 1006 (normal 230-460) and light elevation of others liver enzymes, aldolase of 25 U/Ml (normal 1.8), and a sedimentation rate of 67. Electromyelogram showed myophatic motor unit potentation and increased insertional activity consistent with inflammatory myophaty. CA 125 was 220.1 U/mL (normal 0-35), CA 15.3 was 48.67 (normal 0-25). CEA, alfafetoprotein and CA 19.9 were normal.

Following Good practice points given (6) her internist requested a CT-thorax/abdomen, US of the pelvic region and mammography. Trasvaginal ultrasound revealed a 4 cm complex right adnexal cyst with echogenic areas (Figure 2). On CT abdomen exploration a 4,4 x3,8 cm, complex right adnexal mass was describes withour ascytis or lymphadenopathy (Figure 3).

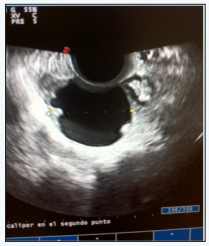

Figure 2

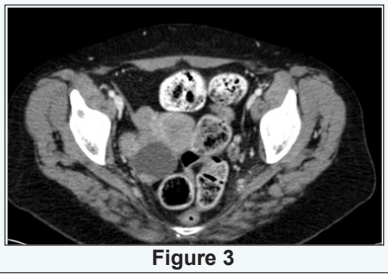

Figure 3

Exploratory laparotomy found a nonaherent enlarged right ovary with no surface excrescences or peritoneal spread. Her uterous, left ovary, anf fallopian tubes were normal. Intraoperative biopsy reveled adenocarcinoma. We did a hysterectomy and billateral salpingo-oophorectomy, omentectomy, and excision of lymph nodes from the common iliac and para-aortic regions. As well as peritoneal cytologi and apracolic biopsias were performed.

Grossly, the right ovarian neoplasm was a fluid-filled, cystic tumor measuring 6.5 cm with two solid protuberant masses of 3.5 and 2.5 cm respectively and papillary projections of 5 mm each one. The capsule was infiltrated. Definitive histopathologic examination accorging to the International Federation of Gynecology and Obstetrics (FIGO) revealed a serious papillary adenocarcinoma, stage IC ovarian carcinoma (Figure 4). The excised of para-aortic and common iliac lymph nodes showed no metastasis. Peritoneal cytology and paracolic biopsias were also negative.

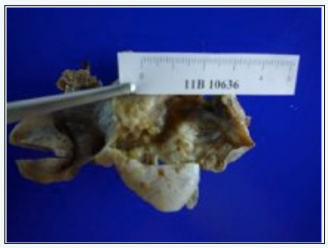

Figure 4

Postoperatively, she received five courses of chemotherapy with paclitaxel a carboplatin. Her skin condition and muscle weakness improved soon after the second course of chemotherapy, approximately 2 months later. The methylprednisolone dose was tapered until discontinued. Currently the patient is still alive for 2.5 years, without evidence of recurrent disease, since pelvic examination and transvaginal ultrasonography, as well as CT and biomarkers (Ca 125: 12 U/mL) are all normal.

Discussion

Despite evidence that ovarian cancer is the most common gynecologic malignancy in women with dermatomyositis, the association between the two is rarely appreciated by ginecologists [2,7-9]. To emphasize the significance of the coexisting disorders, an ovarian carcinoma presenting initially as dermatomyositis is described.

The first description of an existing relationship between malignant disease and myositis was done in 1916 [10]. Since then, several authors confirmed the association between malignancy and idiophatic inflammatory myopathies with a frequency ranging 6-40% [4,11-14]. However clinical outcomes of the studies are limited by referral bias, lack of controls and inclusion criteria for myositis.

Polymiositis and dermatomyositis are idiophatic inflammatory myophaties of undetermined etiology. Connective tissue diseases present with similar clinical symptoms and signs like the myopathies and are usually classified together. Diagnostic criteria por the PM and DM was first suggested by Bohan and Peter [3].

Two Scandinavian reports (Sweden & Finland) in a population-based study with PM and DM showed an overall incidence of associated malignancy 13% and 13.8% respectively [4,13]. The mean age of patients with myositis relationed with malignancy is 53 years, ranged from 40 to 66 years [8]. The

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association between dermatomyositis and malignant disease is better established than that of polymyositis. Usually the onset of dermatomyositis-polimyositis precedes evidence of carcinoma, ramking from 3 months to 6 years with a mean of 2 years [14], but it may also present simultaneous with this malignancy [2,4,15]. In contrast, the development of dermatomyositis after established diagnosis of ovarian cancer seems to be less common.

The present case report refers to patients with a serious papillary ovarian adenocarcinoma on a previously established diagnosis of DM. Our patient demostrates that dermatomyositis may develop in the setting of pre-existent ovarian cancer and that skin manifestations may precede clinically obvious, muscle symptoms by several months. Such patients may experience improvement in muscle strength with corticosteroid therapy, despite persistance of skin rash and progression of their underlying neoplasm.

An autoantibody was recently described in adult patients with dermatomyositis that seems to be associated with cancer in this population: the specific antibody transcriptional intermediary factor 1 gamma (TIF- gamma, initially described as antibody anti –p155/140) This antibody currently represents the best marker of neoplasia in dermatomyositis . The antibody positivity indicates high risk of neoplasia and forces the clinician to closely monitor the patient with exhaustive screening periodically, mainly during the first 3 years [16].

The reported frequency of malignancy in dermatomyositis varies from 6% to 60%, but large population-based cohort studies report a frequency of 20-25% [17]. Several cancer types show this association with dermatomyositis. The most common are ovarian, lung, pancreatic, stomach and colorectalcancers and lymphomas [18]. The risk for lymphoma was only raised the first year after diagnosis of dermatomyositis. For the others tumours, the risk is the highest within the first year of follow-up dropping substantially thereafter. The risk for ovarian, pancreatic and lung cancer remains above average even after 5 years [18]. At diagnosis, thorough examination is requested. In children, specific attention should be paid to splenomegaly or lymphadenopathy [19]. In adults, abnormalities should guide screening tactics, but lack of abnormalities does not imply no screening is needed. Although the risk rises with age, all adults' patients should be screened. Women should be screened by US of the pelvic region and CA-125 and mammography and by CT-thorax/abdomen. Men should be tested by CT- thorax/abdomen. Men under the age of 50 years should have a US of the testes. All patients over 50 years old (men and women) should have a colonoscopy). Screening is to be repeated annually for 3 years. Afterwards, screening is only performed if new symptoms or finding alerts to it [17-20]. Evidence regarding any additional value of FDG-PET is lacking.

In conclusion, the detection of ovarian cancer at an early stage is crucial for patient survival. The physician must be alert of the possibility of malignancy in patients with a previous diagnosis of myositis, especially of ovarian carcinoma in the female population. The current NCCN Clinical Practice Guidelines in Oncology recommend TV US, combined with cancer antigen 125 (CA-125) each 6 months in patients with genetic/familiar high risks for ovarian carcinoma [21].

However, screening of such carcinoma including pelvic examination, transvaginal ultrasonography and CA-125, is not always effecitve [22]. Because the risk for ovarian, pancreatic and lung cancer remains above Average even after 5 years [18], screening is to be repeated annually for 3 years. Afterwards, screening is only performed if new symptoms or finding alert to it [17, 20].

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