

## DERMATOMYOSITIS ASSOCIATED WITH FALLOPIAN TUBE ADENOCARCINOMA IN A FILIPINO

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### ABSTRACT

Dermatomyositis is a type of inflammatory myopathy, which may be associated with malignancy, particularly in the elderly. This case report is that of a 73 year old female who presented with classical manifestations of dermatomyositis, accompanied by marked weight loss. More thorough investigation including tumor markers led to the diagnosis of fallopian tube adenocarcinoma.

**Keywords:** Malignancy, Dermatomyositis (DM), Polymyositis (PM)

### INTRODUCTION

Dermatomyositis (DM) belongs to the group of idiopathic inflammatory myopathies (IIM). These are systemic autoimmune diseases characterized by progressive, symmetrical weakness of the proximal muscles and in the case of DM, by cutaneous lesions. Researchers and clinicians have been interested for decades in the association of malignant diseases with IIM, mainly with DM1. In 1916, Stertz was described a patient with biopsy-proven DM and stomach adenocarcinoma<sup>2</sup>. Later, several studies examined the relationship between DM/PM and malignant diseases, and an increased incidence of malignancy in myositis patients, particularly in DM, was reported in most studies.<sup>3,4</sup>

We report an elderly female with the classical features of dermatomyositis, where a more intensive work up led to the diagnosis of fallopian tube adenocarcinoma – a rare form of gynecologic malignancy.

### Case Report

A 73 year old female was admitted because of rashes and generalized weakness.

The patient had reportedly felt well until 2 weeks prior when she developed erythematous rashes at the periorbital area, chest and upper extremities. A week later, she noted weakness of all extremities described as difficulty in dressing up, combing her hair, taking a bath or climbing stairs. She also had abrupt weight loss.

The patient is widowed, a pharmacist managing a private pharmacy. She was diagnosed with hypertension and diabetes mellitus 2 years ago maintained on losartan 50mg/ day, felodipine 5mg/ day and metformin 500mg thrice daily. She has had 8 successful pregnancies, and had natural menopause at age 52. She does not smoke cigarettes nor drink alcoholic beverages. Her mother died at an early age of unknown cause and her father had history of hypertension. She has one sibling who has no known illness.

Physical examination disclosed a fully conscious, conversant lady with stable vital signs. There were erythematous rashes at the periorbital area more pronounced at the upper lids (“heliotrope”), upper chest (“shawl” and “V” sign), Gottron’s papules on the knuckles of the hands, and scattered lesions on the peri-ungual areas, palms and soles. Palpebral conjunctiva was pale, sclera anicteric; there were no abnormal lesions nor masses in the oral cavity and breasts, and no cervical, axillary nor inguinal lymphadenopathies. Examination of the heart and lungs were unremarkable. There were no palpable abdominal masses nor hepatosplenomegaly, and rectal exam yielded brown stools. Muscle strength was 3/5 on all proximal muscle groups, and 5/5 on distal extremities. Peripheral pulses and deep tendon reflexes were normal.

Hemoglobin was 107g/L, WBC 9.2 x10<sup>9</sup>/L, aspartate aminotransferase (AST) 75 U/L, alanine aminotransferase (ALT) 41.8 U/L, total creatine kinase (CK) 1057 U/L (NV 26-192), CK-MM 1004 U/L (NV 19-167), LDH 354 U/L (NV 100-190). Among

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the tumor markers, CA-125 [ovarian] was 492.40 U/L (NV 0-35), carcinoembryonic antigen (CEA) 39 ng/ml (NV 0-5), alpha-feto protein (AFP) 2.57 (NV 0-7). Antinuclear antibody (ANA) was negative, serum complement (C3) was 0.82 g/L (NV 0.9-1.8) and HBsAg non-reactive. Serum calcium 1.09 mmol/L, alkaline phosphatase 60.5 U/L (NV 35-104), creatinine 0.79 mg/dl and glycosylated hemoglobin (HbA1c) 7.17%. Urine protein was +1, red blood cells (rbc) 0-2/hpf and pus cells 4-8/hpf. Upper gastrointestinal endoscopy showed chronic atrophic gastritis and acute gastric mucosal erosions at the antrum; colonoscopy was normal. Ultrasound of the whole abdomen showed normal sized liver with diffuse parenchymal changes and cholelithiasis; pancreas, spleen and kidneys were normal. Abdominal CT scan showed cholelithiasis, pancreatic tail cyst and a mid pelvic mass suggestive of an omental new growth. The transvaginal ultrasound showed small uterus with thin endometrium and a solid mass measuring 1.14 x 1.02 x 1.08cm.

The patient underwent pelvic laparotomy, followed by hysterectomy with bilateral salpingo-oophorectomy and omentectomy; fresh frozen section biopsy of the parametrial mass disclosed adenocarcinoma. The final surgical pathology result reported subtotal hysterectomy with bilateral salpingo-oophorectomy with bilateral lymph node dissection and infra-coli omentectomy. Final histopath disclosed poorly differentiated papillary serous carcinoma involving the right fallopian tube with extensive blood vessel invasion into the left parametrium, right ovary, corpus uteri. There was a predominant serosal infiltration into the outer myometrium and omentum, and peritoneal fluid cytology was positive for malignant cells consistent with adenocarcinoma.

The patient was given methylprednisolone 48 mg/day later shifted to prednisone 50mg/day. There was improvement of the muscle strength with fading of the rashes over the next few days and was discharged with instructions to taper prednisone and follow up for chemotherapy.

## DISCUSSION

The patients presented with classical features of proximal muscle weakness, elevated muscle enzymes and the characteristic heliotrope rash, V-sign and shawl sign of dermatomyositis as described by Bohan and Peter<sup>5</sup>. The first report of the frequency of malignancy in DM/PM using the criteria of Bohan and Peter was an analysis of the myositis patients seen at

UCLA medical center between 1956 and 19716 and within this study group of 153 patients, 13 (8.5%) had an associated malignancy. The percentage of adults with DM and malignancy was 11.8% (8 of 60 [13.3%] for DM and 5 of 50 [10%] for PM. DM patients have a greater risk of malignancy than PM patients, taken as a whole, it appears that roughly 25% of patients with DM have had or will develop an internal malignancy.<sup>7</sup> The frequency of malignancy was 28.8% and 36% in a 21 year and another 30 year retrospective study.<sup>1,8</sup> The link between malignancy and inflammatory myopathy relates to the expression of common autoantigens between cancer tissue and muscle tissue in some patients with dermatomyositis.<sup>9</sup>

Cancer can be diagnosed before, simultaneously with, or after the diagnosis of dermatomyositis. C.T. scan of the whole abdomen and transvaginal ultrasound in our patient showed a mid pelvic solid mass. The positive result yield of blind malignancy search was only 13% (11 of 87), but reached 28% (5 of 18) for blind abdominal-pelvic and thoracic computed tomographic scans.<sup>10</sup>

Tumors of the ovary and stomach are more frequently observed than in the general population. Adenocarcinomas of the cervix, lung, ovaries, pancreas, bladder, and stomach account for approximately 70 % of malignancies associated with inflammatory myopathies.<sup>11</sup> The tumor site varied, the uterus, lung, and stomach were affected slightly more often, but the difference was not significant according to one retrospective study.<sup>8</sup> Breast and gastrointestinal tumors were the most frequent, followed by the occurrence of lung tumors in another study<sup>1</sup> The first reported case of DM and concurrent fallopian tube carcinoma in the United States was a 62-year-old woman.<sup>11</sup> There was no other case report cited.

Primary tumors of fallopian tube cancer is a rare gynaecological malignancy.<sup>12</sup> Both benign and malignant forms account for 0.1-1.8% of all gynaecological cancers.<sup>13</sup> Overall incidence recorded is one study was 0.41 per 100,000 women.<sup>14</sup>

Early discovery of malignancy is critical in cases of DM.<sup>8</sup> In the patients who had DM without cancer, the survival rate was 73.6% at 6 years, but in the patients who had DM with cancer it was 10% at 5 years;<sup>4</sup> comparing it to 1-year survival rate of 56% in the latest study.<sup>1</sup> The most frequent cause of death in these patients was pneumonia (50%) and respiratory insufficiency respectively.<sup>1,8</sup>

## CONCLUSION

This case illustrates the association of malignancy specifically fallopian tube adenocarcinoma in a patient with dermatomyositis. It is crucial to recognize this association particularly in the elderly, as the overall prognosis and response to treatment are mutually interdependent.

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