Dermatomyositis in the course of primary peritoneal cancer

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Summary
Dermatomyositis (DM) is a rare paraneoplastic syndrome. The case of a 47-year-old woman who was hospitalized in a neurological clinic due to non-specific neurological and dermatological symptoms is described. The results of laboratory tests (including CA 125 > 2,000 U/ml), histopathology and imaging, as well as the lack of response to treatment with steroids and methotrexate were indicative of DM in the course of malignancy. Diagnosis of primary peritoneal cancer was established on the basis of reconnaissance laparoscopy. Due to the significant weakening of muscle strength, weakening of cough reflex and swallowing disorders on the second day after surgery, the patient underwent a percutaneous tracheostomy. As a result of causative treatment with cytostatics (carboplatin with paclitaxel), paraneoplastic symptoms receded. The patient is currently in remission.

Key words: Paraneoplastic syndromes; Dermatomyositis; Peritoneal cancer.

Introduction
Paraneoplastic syndromes are a group of pathological, heterogeneous symptoms occurring in about 8% of people affected by malignancy. They are found at every stage of its development: in the preclinical stage, at the time of diagnosis and treatment [1].

Dermatomyositis (DM) represents a rare disease syndrome occurring at a frequency of 1:1,000,000 people/year [2]. It is an acquired idiopathic myositis with concomitant dermatitis [3]. It mainly affects women in their fifties and sixties. In 15-25% of cases, dermatomyositis is accompanied by a cancerous process and in these patients, breast cancer accounts for 20% of cases, and ovarian cancer for a further 20%. Furthermore, it may occur with cancers of the lung, stomach, intestines, pancreas, nasal cavity, throat, prostate, and bladder as well as non-Hodgkin’s lymphomas. Single cases of thyroid cancer with accompanying dermatomyositis have also been described [4-7].

Case Report
The 47-year-old patient was admitted to the neurology department due to pain and a progressive decrease in upper and lower limb muscle strength, which resulted in difficulties moving, as well as issues with swallowing, particularly during consumption of solid foods. In addition, the patient’s face was affected by erythema which had been developing for several years. The neurological examination revealed weakening of the proximal upper and lower limbs, proximal paresis of the upper and lower limbs 2/3 according to the Lovett scale, without the presence of pathological symptoms. Laboratory tests revealed increased ESR, increased activity of ALAT-139 U/l and AspAT-536 U/l, CPK 18222 U/l enzymes, and presence of antinuclear and cytoplasmatic antibodies both of granular type at a concentration of 1:320. The indirect immunofluorescence (IF) methods p-ANCA and cANCA detected no anti-mitochondrial antibodies or antigens from the group of soluble nuclear antigens (RNP, Sm, Ro/SS-A, La/SS-B, PM-Scl, Scl-70, Jo-1, Ku, Rib-P) nor against antigens neutrophil cytoplasm. The ANA3 profile was negative (no muscle-specific antibodies were found).

Myogenic damage was found during the electromyographic examination of the biceps muscle of the right arm, additionally a high resting activity in the right shoulder muscle was noted. Electroneurography (ENG) revealed small features of axonal damage of left sagittal motor fibers, secondary to changes at root level. Other adduction parameters in the motor and sensory fibers of the examined nerves were within the normal range.

A skeletal muscle fragment was taken for histopathological examination, which revealed the presence of a minimal inflammatory infiltration within the perimisium with a few CD4 lymphocytes, and granulocytes among inflammatory cells. The severity of the changes is described as insignificant. The picture described may correspond to dermatomyositis.

High-resolution chest imaging was performed (only atelectatic and inflammatory changes present in HRCT, abdominal cavity, and pelvic cavity (ascites with tumor in the peritoneum). The radiological image suggested the presence of a neoplastic process emerging from the right ovary. In a laboratory study, the CA-125 antigen level was 2,215 U/ml.

Dermatomyositis in the course of ovarian cancer was suspected. Therefore, the patient was transferred to the Department of Oncological Gynecology for further diagnosis. For the purpose of verifying the clinical diagnosis, a laparoscopy was performed. Malignant dissemination of cancer on the peritoneum, diaphragm, and network was found. Fragments of the network and the peritoneum were taken as representative. Based on histopathological examination, primary peritoneal cancer was diagnosed. After the diagnostic procedure due to a significant decrease in muscle strength, weakness of the cough reflex and swallowing disorders, the patient was admitted, as planned, to the intensive care unit where on the second day of hospitalisation a tracheostomy was performed using the percutaneous Ciaglia method without any further...
complications. On the third day, the patient was disconnected from the respirator; the patient breathed spontaneously through the tracheostomy tube with simultaneous therapy with passive oxygen. The patient was then transferred to the Department of Oncological Gynecology, where, after obtaining the result of histopathological examination (primary peritoneal cancer), chemotherapy with Paclitaxel + Carboplatin was initiated. Due to persistent difficulties in swallowing, a percutaneous endoscopic gastrostomy (PEG) was implanted to ensure an adequate supply of nutrients in a way that was as close to physiological as possible. During subsequent stays in the Oncological Gynecology Department, cycles of chemotherapy were continued. A gradual improvement in the clinical condition was observed: the return of a cough reflex, normal swallowing, and increased muscle strength. This enabled the removal of the tracheostomy tube and intensive rehabilitation. The patient is currently in a good general condition and undergoing cytostatic treatment.

Discussion

The occurrence of DM may precede the appearance or accompany cancer. Generally, it is manifested by a symmetrical weakening of the proximal muscles: the shoulder girdle, the iliocostal part of the abdominal muscle, the quadriceps, crural, buttocks and shoulder girdle. This weakens the respiratory muscles leading to respiratory failure. Approximately 30% of patients develop muscle weakness in the throat, esophagus, and larynx, causing dysphonia and dysphagia. Tenderness and muscle pain affect 20-50% of patients.

In DM, skin changes can precede myositis or occur alone. The pathognomic symptom is erythema, a violet-coloured erythema resembling glasses around the eyes (in 30-60% of patients). Erythema can also be located around the neck, line, neck, shoulders (shawl sign), lateral surface of the thighs, and hips (holster sign) [8, 9]. In 30-40% of cases there are symptoms of intermittent lung disease manifesting in a dry cough and increasing dyspnea. Other symptoms may be due to the involvement of the gastrointestinal tract (gastroesophageal reflux), peripheral joints (pain), heart (tachycardia or bradycardia), or the organ of vision (mynagmus, blurred vision) [1, 10].

Additional research highlights an increase in the markers of muscle damage (CK, AST, ALT, LDH, aldolase, myoglobin), an increase in ESR and CRP, the appearance of PM / DM-associated antibodies (antibodies against aminoacyl-tRNA synthetases, anti-SP, amin Mi-2), and companion antibodies (anti-Ro, anti-La)[2].

Electromyographic features of primary muscle damage include fibrillation potentials at rest. Histopathological examination usually has inflammatory infiltration mainly from CD4 T cells and activated B lymphocytes. Therefore, the patient met the criteria of dermatomyositis, which includes [2]: 1) Symmetrical, increasing weakness of the shoulder girdle and hip rim. 2) Typical histological results for muscle inflammation. 3) Increased serum CK or aldolase activity. 4) Electromyographic features of primary muscle damage. 5) Typical skin changes.

In the case described, the skin and muscle syndrome, and the lack of response to treatment with high doses of steroids and methotrexate led to the suspicion of a neoplastic disease. Full diagnostics were implemented (laboratory, histopathological examination of muscle fragment, electromyographic, and electroneurographic). At the same time, possible sites of cancer were investigated. On the basis of the imaging diagnostics (the presence of implants on the peritoneum and changes in the appendix), the presence of ovarian cancer was suspected.

The patient was transferred to the Department of Oncological Gynecology, where, after establishing a diagnosis of peritoneal cancer based on histopathological examination, causative treatment was initiated. The planned application of invasive methods of maintaining airway patency in the postoperative period and ensuring the possibility of full enteral nutrition by PEG deserves attention. This enabled the immediate implementation of causative treatment and early, intensive rehabilitation, leading to a significant improvement in the clinical condition and withdrawal of clinical exposures of the skin and muscle syndrome. The patient is in partial remission and undergoing further chemotherapy.

References


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