Polymyositis in association with polycythaemia vera

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ABSTRACT

Inflammatory myopathies are reported to be associated with various malignancies. This association is more commonly observed in cases of dermatomyosis and less frequently in polymyositis. Malignancies commonly reported in association with inflammatory myopathies include non-Hodgkin's lymphoma and ovarian, lung and gastric carcinomas, as well as nasopharyngeal malignancies. Polycythaemia vera as a cause of polymyositis is unknown. We report a case of polycythaemia vera in a female patient who developed polymyositis three years later.

Keywords: malignancy, polycythaemia vera, polymyositis

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INTRODUCTION

Inflammatory myopathies, such as polymyositis (PM) and dermatomyositis (DM), are characterised by idiopathic immune-mediated attacks on skeletal muscles, which result in muscle weakness. Various malignancies have been reported in association with PM and DM, but reports of polycythaemia vera (PV) developing in PM are relatively unknown.

CASE REPORT

A 47-year-old Muslim woman presented with hypertension, redness of both hands and feet and a burning sensation all over the body, especially in the legs. She claimed that these symptoms had been present for the past three years. During that time, she also experienced vertigo, dizziness and tinnitus, as well as recurrent epigastric pain, anorexia, palpitation and increased perspiration. Throughout this period, the patient underwent recurrent phlebotomies (300 ml of blood) from the peripheral veins. Three months before presentation, the patient had developed one episode of melaena, whereas two months ago, she had experienced a sudden weakness in the proximal muscles of both her upper and lower limbs; the condition involved both muscle pain and tenderness. The patient also developed nasal intonation of voice and nasal regurgitation during food intake. She did not have a history of smoking or stay at high altitude, and had no previous respiratory or cardiac ailments. No significant family history was present. Five years before presentation, the patient had undergone an appendicectomy and a cholecystectomy. A hysterectomy with bilateral salpingo-oophorectomy was performed three years ago, and histopathology had revealed adenomyosis.

Physical examination revealed bilateral conjunctival suffusion, palmar erythema, pedal oedema, blood pressure 150/110 mmHg and regular pulse at 105/min. No lymphadenopathy was found. The patient was found to have huge splenohepatomegaly. Breast examination was normal, but neurological examination revealed a limb-girdle pattern of weakness, absent gag reflex and decreased palatal movement. Planter was bilaterally flexor, and deep tendon jerks were reduced but preserved. Respiratory and cardiovascular system examinations revealed normal functioning, and no evidence of nasopharyngeal malignancy was found

Blood picture revealed haemoglobin 24.7 gm/dL, haematocrit 55.5%, mean corpuscular volume 89 fL, mean corpuscular haemoglobin (MCH) 28.9 pg/cell, MCH concentration 32.6 gm/dL and erythrocyte sedimentation rate 2 mm/hour. Total leucocyte count was 20,100/cu mm, with neutrophil 88%, lymphocyte 8%, monocyte 1%, basophil 2% and eosinophil 1%. Platelet count was 5.5×10^9/L. Bone marrow biopsy revealed hypercellular marrow with trilineage hyperplasia. Megakaryocytes were increased in number and varied in size; many were unusually large, with hyperlobulated nuclei. Bone marrow iron store was decreased. Reticulin fibres were slightly increased. Pulse oximetry revealed an arterial O_2 saturation of 98%. The patient's blood sugar, urea, creatinine, Na^+ and K^+ levels were all normal. Serum uric acid level was 15.7 mg/dL. Serum erythropoietin level was 12 U/L (normal range 4–27 U/L). JAK2 V617F mutation was detected.

Serum creatine phosphokinase was raised at 2,319 U/L (normal range for females 39–238 U/L). Cerebrospinal fluid study was normal. Electromyography and nerve conduction velocity were suggestive of inflammatory myopathy. Muscle biopsy revealed

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inflammatory myopathy with endomyseal infiltrate of inflammatory cells. In addition, ultrasonography and computerised tomogtaphy imaging of the patient's abdomen revealed splenohepatomegaly. No space-occupying lesion was found in the liver and kidneys. Her uterus, overies and gall bladder could not be examined as they were removed during previous surgeries. Upper gastrointestinal endoscopy revealed antral gastritis, bile in stomach and superficial ulceration in the first part of the duodenum. Rapid urease test was negative. Magnetic resonance imaging of the brain revealed generalised cerebral and cerebellar atrophic changes with focal ischaemic leukoaraiosis but no acute infarction or space-occupying lesions. The brainstem was normal. PM in association with PV was diagnosed.

DISCUSSION

Malignancies are more frequently associated with DM than with PM.[1] Currently, malignancies of the lung, breast, ovary, stomach and colon, nasopharyngeal carcinoma and non-Hodgkin's lymphoma are reported to be associated with both PM and DM.[2] Studies from Asian populations have shown a higher association of nasopharyngeal carcinomas with PM/DM.[3-5] Harris et al reported the only male PM patient to date who was subsequently diagnosed with PV.[6] However, in our case, a female patient first developed symptoms of PV followed by features of PM three years later.

Work-up for detection of malignancy in patients with PM or DM should be individualised according to the patient's age, gender and clinical features. Extensive undirected searches often result in very low yields.[7] Screening for malignancies should include a physical examination, routine blood tests, chest radiography, mammography and gynaecologic examination for women. Imaging of the chest, abdomen and pelvis, as well as serum tumour markers (CA125 and CA19-9) may augment the discovery of underlying malignancy.[8-10]

Inflammatory myopathies may precede or follow the diagnosis of malignancy. The removal of malignancy may result in an improvement of PM or DM, thus supporting its paraneoplastic nature. Conventional immunosuppressive treatment may not be helpful in malignancy-associated PM/DM. There is a need to enforce routine screenings for tumours if the symptoms of PM/DM do not respond to conventional immunosuppressive treatment. PM may be a rare paraneoplastic feature of PV, but large case series are required in order to establish a strong association between the two.

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