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Case Report

One Case of POMES Syndrome with "Limb Pain" As the Main Presentation

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Abstract

Objective: To summarize the clinical manifestations, imaging features and gene mutation sites of a patient with pomes syndrome with "limb pain" as the main manifestation, so as to improve the understanding of this rare disease.

Medical History: The patient was a 34 year old male with limb pain and fatigue for more than 20 days.

Symptoms and Signs: The tongue is in the middle, the uvula is in the middle, the bilateral pharyngeal reflex disappears, the neck is soft, the muscle strength of both upper limbs is grade 5, the proximal muscle strength of both lower limbs is grade 4, and the distal muscle strength is grade 5. The muscle tension of the extremities is normal, the tendon reflex of the extremities disappears, and no obvious abnormalities are found in the examination of deep and shallow sensation and ataxia.

The diagnostic method bone marrow puncture showed thrombocytosis. The pathological results showed that the hematopoietic cells of the three lines were proliferative, the oligoclonal bands in CSF were positive, and the oligoclonal bands in serum were positive. Serum immunofixation electrophoresis (March 30, 2018): IgG λ Abnormal monoclonal bands were found in the lane, and the type of monoclonal immunoglobulin was IgG- λ Type. Blood analysis suggested thrombocytosis (738 × 10°several L).

There is no specific therapeutic drug, but hormone and hydroxyurea are effective.

The clinical outcome was improved and the disease was gradually alleviated.

The clinical conclusion is that poems syndrome is a rare clinical syndrome with various forms. Early intervention is effective for the prognosis of patients.

Keywords: POMES syndrome; thrombocytosis; monoclonal plasma cells

Introduction

Also known as Crow-Fukase syndrome, is a systemic lesion related to plasmaccytosis, mainly manifested as: polyneuropathy (polynel Jropath, P), organomegaly (Organmegaly, 0), endocrinopathy (Erldocfinopathy, E), M protein (M-protein, P) and skin lesions (Skin-change, S), etc. In 1980, Bardwic abbreviated the English prefix for the clinical features of the syndrome. The cause and principle of the disease are not very clear, and may be related to tumors, metabolic disorders, or abnormal immune response. One case of POMES syndrome with "limb pain" is mainly reported, aiming to improve the awareness of the disease, and reduce misdiagnosis and missed diagnosis.

Clinical Data

General Information

The 34-year-old male was admitted to hospital with "limb pain and weakness for over 20 days". Patients more than 20 days ago no obvious cause of lower limb pain, knee upward, upper limbs pain is not obvious, gradually appear lower limbs fatigue, leg difficulties, mainly proximal, affect work, and facial, lower limbs swelling, then to our hospital, check blood analysis suggests thrombocytosis (73810 L), nuclear antibodies, immunoglobulin, complement tests, 14 days ago the patient to the provincial hospital, review platelets still elevated (81310 L), bone marrow puncture shows thrombocytosis, suggest to exclude reactive thrombocytosis considering myelocymoplastic tumor. Electromyography suggested multiple symmetric peripheral nerve damage, the patient was recommended to take oral "hydroxyurea and aspirin" treatment, the patient had oral traditional Chinese medicine treatment, the swelling symptoms were reduced, the pain was relieved, still felt limb fatigue, walking was "duck step", for further clear diagnosis, we came to our hospital. Since the onset of the disease, the body temperature was high, obvious in the afternoon and evening, with the highest 38°C, and a recent weight loss of about 5kg. Night sweats" for 3 months, upper respiratory tract infection history of 2 months, has been cured. Denial history of hypertension, denied blood history, denied history of hepatitis tuberculosis. A history of trauma and allergy was denied. Denied cigarette smoking and alcohol consumption history.

Check Up; Inspect

Physical examination: Heart, lung and abdominal physical

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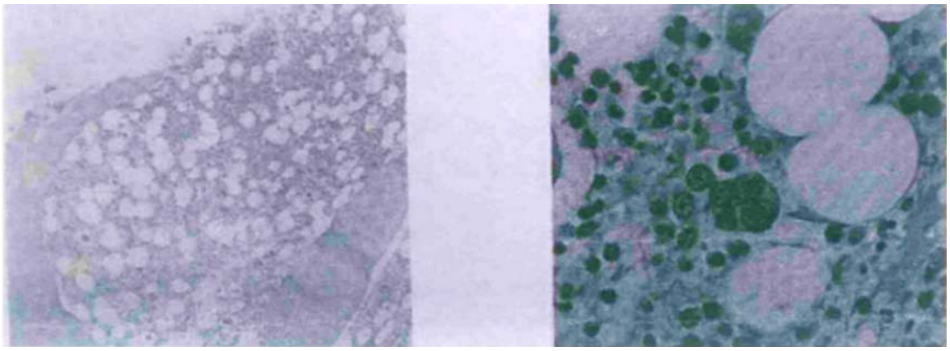

Figure 1: The Patient's first bone marrow puncture (3.15) showed a pathological hyperplasia of triple-lineage hematopoietic cells, with no significant increase in primitive cells and abnormal lymphocytes.

examination (-), superficial lymph nodes did not touch the enlargement. Ditting oedema in both ankles. Nervous system physical examination: mind, spirit, fluent speech, binocular ball movement flexible, no obvious nystagmus, bilateral nasolabial groove slightly symmetrical, extended tongue centered, udra, bilateral pharyngeal reflex disappear, the neck is soft, double upper limb muscle level 5, lower limb proximal muscle level 4, distal muscle level 5, four limb muscle tension is normal, limb tendon reflex disappear, deep feeling and mutual aid exercise examination no obvious abnormalities. Bilateral pathological signs (-), meningeal stimulation signs (-).

Auxiliary examination: prehospital immunoglobulin + complement, C opritin, anti-O, rheumatoid factor, antonuclear antibody, and anti-cyclic citrulline polypeptide antibody showed no abnormalities. Bone marrow puncture indicated thrombocytosis, and the pathological results (Figure 1) showed the hyperplasia of tertiary hematopoietic cells, with no significant increase in primitive cells and abnormal lymphocytes. The EMG suggests multiple symmetric peripheral nerve (root) damage. Cervical disc MR: C4 / 5 disc herniation with disc degeneration, cervical osteosis. Abdominal color ultrasound: indicating cholecystitis, no liver and spleen and other organomegaly. arteriovenous color ultrasound of both lower limbs: no abnormality.

After admission, perfect tumor markers, tuberculosis infection T cell spots, virus screening and A work were all normal. Routine blood platelets and leukocytes were increased for multiple tests (Table 1). Lumbar puncture was performed, pressure 220mmH, O, CSF cytology suggested color colorless transparent, leukocytes 010 several L, protein quantification 101.4mg/dL (one), sugar quantification 4.84 mmol/L (1), lymphocytes 0%, monocytes 0%, and neutrophils 0%. Prolactin 22.3ng/mL (all). CSF oligoclonal band was positive and serum oligoclonal band was positive. Chest CT: a small amount of exudative lesion in the lower lobe of both lungs. Cranial MR: bilateral frontal lobe abnormal signal, considering small ischemic areas. Review EMG: proximal nerve root damage (demyelination). Serum protein electrophoresis (2018.3.30): The M protein band was found. Serum immunoimmobilized electrophoresis (2018.3.30): abnormal monoclonal bands were found in IgG and swimming lane, and the monoclonal immunoglobulin type was IgG-type. On day 5 of admission, the patient had retroaural nodules and multiple lymph nodes. Ophthalmic consultation suggested bioptic papilledema. Orthotopic radiographs of the lumbar spine and pelvis indicate

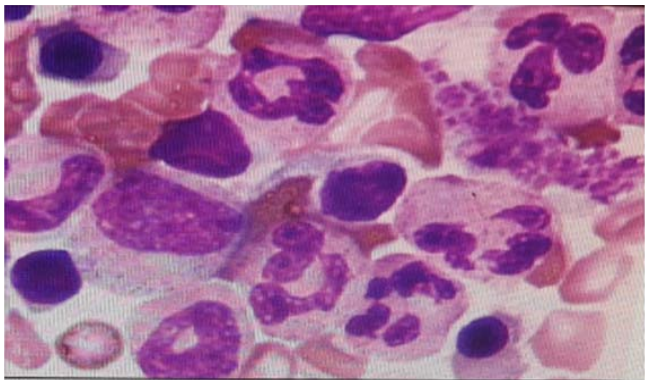

Figure 2: Patient's second bone marrow (4.20) puncture smear: hyperplasia III/VII, G=0.700, E=0.220, The Whole slice of megakaryocytes was found in 123, with a plasma-cell ratio of 0.5%.

bone hyperplasia. Review of abdominal color ultrasound: a small amount of abdominal fluid accumulation. Thyroid color ultrasound was not abnormal. On April 16, the patient was transferred to bone marrow puncture (Figure 2), showing hyperplasia III / VII, G=0.700, E=0.220,123, with the plasma-cell ratio of 0.5%. The ECT results All were negative, with neutrophil alkaline phosphatase 128 (1).

Diagnosis and differential diagnosis Location diagnosis: limb pain, fatigue is located in: peripheral nerve or cervical pulp. Multiple blood tests of conventional blood platelets, white blood cells are increased, bone marrow puncture shows thrombocytosis, qualitative and other comprehensive judgment of the patient pomes is more likely.

Pomes often need to identify with the following 1, multiple myeloma, the first symptom of the disease is bone pain, rather than multiple peripheral neuropathy, its onset age than POEMS syndrome, liver and splenomegaly is common, but surrounding lymph node enlargement than the latter, faster blood sink, blood calcium and M protein, urine-protein frequency is significantly higher than POEMS syndrome, X-line generally shows osteoporosis, multiple osteolytic destruction, bone sclerosis is rare. Combined renal damage is more common, with amyloidosis, less endocrine changes than POEMS syndrome, and significantly higher bone marrow puncture plasma cells.2. Chronic Green-Barre syndrome, mainly manifested as multiple peripheral neuropathy and the increase of cerebrospinal fluid

Table 1: The Patient tested higher blood routine platelets several times.

	2018.03.10	2018.03.14	2018.03.19	2018.03.29
White blood cell	8.45X10^9/L	8.84X10^9/L	11.4X10^9/L	10.97X10^9/L
Platelet	738X10^9/L	813X10^9/L	843X10^9/L	864X10^9/L
	2018.04.20	2018.04.24	2018.04.28	
White blood cell	18.5X10^9/L	13.1X10^9/L	13.23X10^9/L	
Platelet	425X10^9/L	329X10^9/L	358X10^9/L	

protein, generally shows no skin damage and endocrine dysfunction, no skeletal damage and M protein, plasma cell infiltration, etc.3. Connective tissue disease can also cause multisystem damage, but its muscle damage is mostly myogenic, serum protein electrophoresis does not appear M protein, and autoantibodies can be measured in the blood to identify. However, there are reports of scleroderma or systemic lupus erythematosus coexisting with POEMS syndrome.

Treatment After admission, "hydroxyurea and aspirin" were treated. The patient was treated with 1g of methylprednisolone sodium succinate, gradually reduced, and was treated with prednisone 30mg orally 13 days later. He was transferred to the hematology department for hospitalization and continued treatment with oral prednisone tablets and hydroxyurea.

Treatment results, follow-up and return After the application of hormones, the weakness and pain were gradually improved than before, gradually could walk by themselves, and the swelling was not obvious. The patient improved and was discharged from hospital, and his symptoms gradually improved after discharge. After 2 months of follow-up, the patient stopped his medication by himself and basically returned to normal life.

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Discussion

POEMS syndrome is a clinically rare clonal plasmacytosis, which was formally proposed by Bardwick in 1980, and its main clinical manifestations are multiple peripheral neuropathy (Polyneuopathy), organoma (Organomelaly), endocrine lesions (Endocrinopathy), M protein (Monoclonal gammopathy), and skin changes (Skin changes), each named as [1]. POEMS syndrome has the characteristics of low incidence, high misdiagnosis rate, high disability rate, the etiology and pathogenesis is not fully clear, may be related to monoclonal plasmachyperplasia, EB virus and human herpes virus-8 infection, 14g32 translocation and chromosome 13q14 deletion, proinflammatory cytokines and Vascular Endothelial Growth Factor (VEGF) excessive [2]. There is no unified diagnostic standard for POEMS syndrome at home and abroad. At present, the diagnostic criteria proposed by Dispenzieri et al were in 2003: (1) two main criteria: (1) polyneuropathy and (2) monoclonal plasma cell dysplasia (M protein).(2) 7 secondary criteria: (1) bone sclerosing lesions: (2) Casileman disease: 3) organomegaly (liver, spleen, lymph nodes enlargement) (4) edema (edema, pleural effusion or ascites) (5) endocrine disorders (adrenal, thyroid, pituitary, gonads, parathyroid, pancreatic dysfunction): (6) skin changes (pigmentation, hairy, hemangioma, etc.) (7) optic disc edema. POEMS syndrome can be diagnosed if two main criteria are met, plus more than one of seven secondary criteria. This patient is the first for peripheral nerve demyelination and axonal damage, detected M protein (type), but then review M protein turned negative, at the same time patients with prolactin higher than normal, skin pigmentation and pale nails, abdominal effusion, swollen lymph nodes, lower limb edema, papillary edema, in addition, patients also have recent weight loss, thrombocytosis such as [3]. At present, POEMS syndrome is more considered. However, no plasma cell hyperplasia was found in the two bone marrow punctures, and the laboratory results of M protein Yin conversion could not support this diagnosis. Some literature points out that the detection rate of M protein is not 100%, but if the bone marrow puncture and/or M protein detection are combined, the positive rate reaches 100%, so the negative M protein cannot rule out the diagnosis of [4] in this disease. When the M protein test is negative and this disease is highly suspected, the multisite bone marrow puncture and biopsy and the suspicious lesion tissue biopsy should be performed for immunohistochemistry to find the evidence of monoclonal plasma cell hyperplasia, [5]. Differential diagnosis: 1. Chronic Inflammatory Demyelinating Polyneuropathy (CIDP): often accompanied by cranial nerve damage, generally no skin damage and endocrine dysfunction, no skeletal damage and M protein, plasma cell infiltration, etc. Usually without VECF elevation, even if elevated, also <500g /L, the POEMS syndrome can reach 1500-2500ng.

Conflict of Interest

Conflicts of Interest Statement All authors declare that no conflict of interest exists.

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