**International Journal of Clinical and Experimental Medical Sciences**
2018; 4(1): 14-17
http://www.sciencepublishinggroup.com/j/ijcems
doi: 10.11648/j.ijcems.20180401.13
ISSN: 2469-8024 (Print); ISSN: 2469-8032 (Online)

# A Rare Case of POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal Gammopathy, and Skin Changes) Syndrome with the Isolated Adrenal Tuberculosis

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**To cite this article:**
Li Tian, Xuewen Xiao, Yang Cao, Jingbo Li, Shourong Shen, Xiaoyan Wang, Anliu Tang, Xiayu Li, Fen Liu, Feiyan Ai. A Rare Case of POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal Gammopathy, and Skin Changes) Syndrome with the Isolated Adrenal Tuberculosis. *International Journal of Clinical and Experimental Medical Sciences*. Vol. 4, No. 1, 2018, pp. 14-17.
doi: 10.11648/j.ijcems.20180401.13

**Received**: January 17, 2018; **Accepted**: February 2, 2018; **Published**: February 23, 2018

**Abstract:** POEMS syndrome is a rare paraneoplastic syndrome characterized by polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes. The diagnosis of this disease is often challenging due to its rarity and its multisystemic involvement. Isolated adrenal tuberculosis is also a rare disease which is also difficult to diagnose. Therefore it is incredibly challenging to diagnose a patient with POEMS syndrome when he has isolated adrenal tuberculosis. In this report, we describe a 43-year-old man suffering from POEMS syndrome with right adrenal tuberculosis. His symptoms were very complicated and initially we misdiagnosed him with endocrinal disturbances and the adverse effects of antituberculosis drugs. It was not until we accumulated enough evidences of POEMS syndrome that we could make the final diagnosis. To the best of our knowledge, this is the first report of POEMS syndrome with isolated adrenal tuberculosis.

**Keywords:** POEMS Syndrome, The Isolated Adrenal Tuberculosis, Diagnosis

## 1. Introduction

POEMS syndrome is a rare paraneoplastic syndrome whose acronym represents some of the defining features of this disease, including polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes. The other important features involve papilledema, extravascular volume overload, sclerotic bone lesions, thrombocytosis, elevated vascular endothelial growth factor (VEGF), and abnormal pulmonary function [1]. The syndrome is a manifestation of an underlying plasma cell disorder and its pathogenesis currently remains unclear [2]. The acronym POEMS was first coined by Bardwick et al. and is also known as osteosclerotic myeloma, Takatsuki syndrome, and Crow-Fukase syndrome [3-6]. Its prevalence is ~0.3 per 100,000 which was calculated by a nationwide Japanese survey in 2003[7]. Currently, the therapy of POEMS is mainly classified into two major parts: the first part is targeting the plasma cell clone, which includes radiation, chemotherapy, autologous peripheral blood stem cell transplantation (ASCT); the second part is targeting the rest of this syndrome, such as the elevated VEGF [8].

Isolated adrenal tuberculosis is also a rare disease. Its incidence is about 1 per 1000 [9]. It can lead to endocrinal disturbances such as Addison's disease. The computed tomography (CT) scanning, MRI (Magnetic Resonance Imaging) and histopathological examination of the suspected adrenal lesions are of great help in diagnosis [10-11].

## 2. Case Report

A 43-year-old man was admitted to our hospital on Feburary,

12th, 2016 due to the weakness of his lower extremity for four months and abdominal distension for a month. Prior to admission to our hospital, the patient had been diagnosed with right adrenal tuberculosis and underwent right adrenalectomy in local hospital on October, 19th, 2015. After the surgery, the patient was taking antituberculosis drugs and he had gradually felt the weakness in the lower extremity over the course of a few days. As a result, the doctors in the local hospital had adjusted the antituberculosis drugs' regimen several times but had failed to adequately treat his lower extremity weakness. About one month earlier before admission to our hospital, he had developed abdominal distension gradually without any obvious causes. The patient complained of having lost approximately 10 kilograms during the past four months. In addition, he also had the past history of contacting the patient cavernous pulmonary tuberculosis one year ago.

Physical examination demonstrated a thin man with hyperpigmentation in skin and mucosa. The liver could be palpated at about 4cm below the right costal margin and the spleen could be palpated at about 2cm below the left costal. Moreover, his abdomen was distended and there was moderate pitting edema of his lower extremities. The examination of lymph nodes showed there were enlarged lymph nodes in the cervical region, axillary fossa, and groin, which were ductile, not painful, and could be removed easily. Neurological examination revealed the 4/5 muscle strength of the lower extremities, while the muscular tension is normal.

Computed tomography (CT) of abdomen showed hepatosplenomegaly (Figure 1A). Ultrasound of gastroscopy revealed the enlargement of lymph nodes in abdominal cavity, the swelling of the left adrenal gland whose size is about 4.2*1.0cm, and the echo of the left adrenal gland is homogeneous (Figure 1B). The bone imaging suggested no metabolic abnormalities in all of bones.

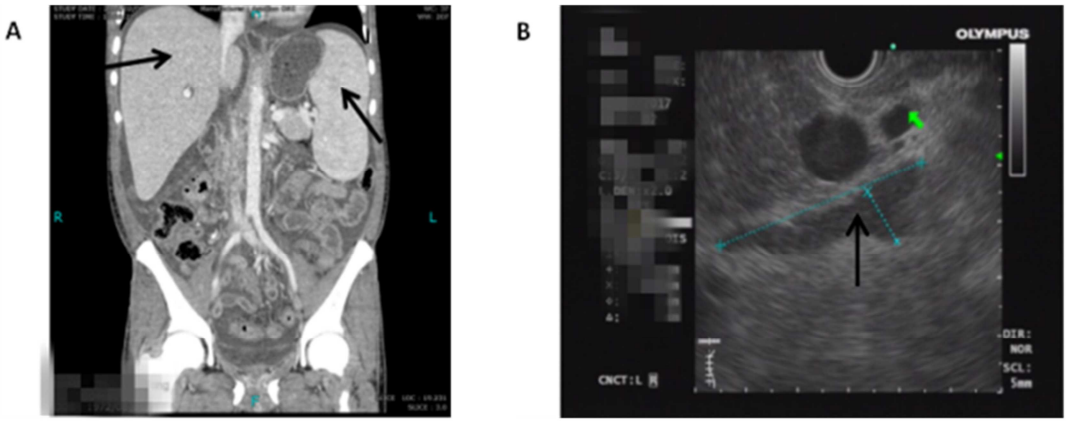


***Figure 1.*** *A. CT scan of the abdomen showing hepatosplenomegaly (black arrows). B. Ultrasound of gastroscopy showing the echo of the left adrenal gland is homogeneous (black arrow).*

Laboratory results (Table 1) showed anemia and proteinuria. T-spot was positive. The hormonal function tests revealed hypogonadism and hypothyroism, with Testosterone was 170.2ng/dL and TSH was 11.11μIU/mL. And the rhythm of cortisol and ACTH was disturbed. Full automatic immunofixation electrophoresis revealed IgA λ type of M protein was positive while λ type of M protein was suspiciously positive. Serum VEGF levels were in a normal range.

***Table 1.*** *Laboratory results of the patient.*

| Variable | Value(normal range) | |
|---|---|---|
| Hemoglobin, g/L | 102 | 130-175 |
| neutrophile granulocyte's percent, % | 75.4 | 40-75 |
| lymphocyte's percent, % | 16.4 | 20-40 |
| 24-hour urinary protein quantity, mg/24h | 2066 | 0-150 |
| Albumin, g/L | 30.4 | 40-55 |
| Globulin, g/L | 20.9 | 25-32 |
| Urea, mmol/L | 7.42 | 1.84-7.14 |
| Creatinine, μmol/L | 117 | 45-104 |
| Uric acid, μmol/L | 447 | 155-425 |
| Erythrocyte sedimentation rate, mm/hr | 49 | 0-15 |
| T-spot | Positive | negative |
| Anti-nuclear antibody | 1:160 | |
| High-sensitivity c-reactive protein, mg/L | 19.6 | 0-10 |
| FT3, pmol/L | 2.83 | 3.1-6.8 |
| Free thyroxine, pmol/L | 7.21 | 10.3-22.65 |
| TSH (oid-stimulating hormone) μIU/mL | 11.11 | 0.27-4.2 |
| Cortisol(0 o'clock), μg/dL | 14.17 | 2.3-11.9 |
| Cortisol(8 o'clock), μg/dL | 13.95 | 6.2-19.4 |
| Cortisol(16 o'clock), μg/dL | 14.26 | 2.3-11.9 |
| ACTH(0 o'clock), pg/mL | 32.92 | 7.2-63.3 |
| ACTH(8 o'clock), pg/mL | 116.6 | 7.2-63.3 |
| ACTH(16 o'clock), pg/mL | 6.3 | 7.2-63.3 |
| Estradiol, pmol/L | 146.1 | 28.01-156.3 |
| Testosterone, ng/dL | 170.2 | 249-836 |
| LH(luteinizing hormone), mIU/ml | 22.38 | 1.7-8.6 |
| Prolactin, ng/mL | 26.28 | 4.04-15.2 |
| IgAλtype of M protein | Positive | negative |
| λtype of M protein | suspiciously positive | negative |
| VEGF | 132.25pg/ml | ＜160pg/ml |

The examination of bone marrow showed bone marrow cells proliferated actively while plasma cells were in a normal range. And we reexamined the specimen of the right adrenal gland and reconfirmed the existence of the right adrenal tuberculosis (Figure 2A and 2B). Besides, the biopsy of an enlarged left cervical lymph node suggested the patient suffered from Castleman disease (Figure 2C and 2D).

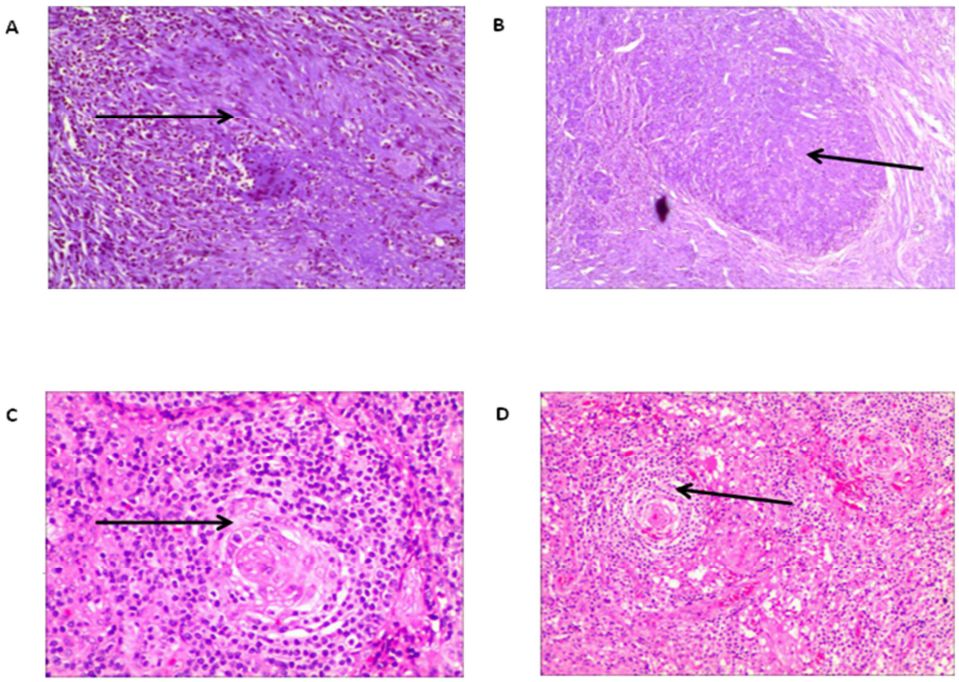


***Figure 2.*** *A. B. Granulomatous inflammation and giant cells of right adrenal tuberculosis (black arrows) (acid-fast stain; magnification, A, x40 and B, x10). C. D. Castleman disease in left cervical lymph node (black arrows) (hematoxylin and eosin stain; magnification, C, x40 and D, x10).*

As the evidences of POEMS syndrome accumulated, the diagnosis of POEMS syndrome was made based on polyneuropathy, monoclonal plasma cell disorder with IgA lambda, Castleman disease, organomegaly (hepatomegaly and splenomegaly), extravascular volume overload, endocrinopathy, skin changes, weight loss according to the criteria for the diagnosis of POEMS syndrome.

The patient was treated with glucocorticosteroid therapy and antituberculosis drugs. After these treatments, the abdominal distension and the weakness improved slightly. Physical examination showed the shrinkage of the liver and spleen. The patient was discharged with outpatient follow-up. At the recent follow-up examination, in September 2016, the patient's weakness and abdominal distension relieved greatly.

## 3. Discussion

POEMS syndrome is a rare disease associated with multisystem lesions, such as polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes. The diagnostic criteria of POEMS syndrome involve mandatory major criteria, other major criteria, and minor criteria. To be explicit, mandatory major criteria include polyneuropathy, typically demyelinating, and monoclonal plasma cell-proliferative disorder, almost always λ. Other major criteria include Castleman disease, sclerotic bone lesions, and VEGF elevation. Minor criteria include organomegaly (splenomegaly, hepatomegaly, or lymphadenopathy), extravascular volume overload (edema, pleural effusion, or ascites), endocrinopathy (adrenal, thyroid, pituitary, gonadal, parathyroid, and pancreatic), skin changes (hyperpigmentation, hypertrichosis, glomeruloid hemangiomata, plethora, acrocyanosis, flushing, and white nails), papilledema, thrombocytosis/polycythemia clubbing. Only both of the mandatory major criteria, one of the three other major criteria, and one of the six minor criteria are present can a patient be diagnosed with POEMS syndrome [12]. Our patient fulfilled two mandatory criteria, one of the three other major criteria, and four of the six minor criteria, which shown in Table 2.

***Table 2.*** *Clinical manifestations in our patient.*

| Criteria | Evidence |
|---|---|
| Polyneuropathy | Progressive weakness in the lower limbs |
| monoclonal plasma cell-proliferative disorder | IgA λ type of M protein (+) |
| Castleman disease | Left cervical lymph node confirmed its existence |
| Organomegaly | Hepatomegal, splenomegaly |
| Extravascular volume overload | Pericardical effusion, pleural effusion, or ascites |
| Endocrinopathy | Hypogonadism, hypothyroism, and the disturbance of corticsol |
| Skin changes | Hyperpigmentation |

However, at first, diagnosis of POEMS syndrome in this patient was highly complicated by the patient's history of a right adrenalectomy due to the right adrenal tuberculosis. Initially, we diagnosed the patient with endocrinal disturbances and the adverse effects of antituberculosis drugs. As the evidences of POEMS syndrome gradually been collected, we finally diagnosed the patient with POEMS syndrome.

To the best of our knowledge, this is the first report of POEMS syndrome with the isolated adrenal tuberculosis. Given the rarity of POEMS syndrome and its diverse

symptoms [13], it is extraordinarily difficult to make the right diagnosis especially when the patient just suffered from isolated adrenal tuberculosis and had the surgery of adrenalectomy. This case report may enlighten some doctors to take POEMS syndrome into consideration facing a complicated case with some of the specific symptoms of POEMS syndrome.

In conclusion, we report the first case report of POEMS syndrome with isolated adrenal tuberculosis. We hope this case report may remind some doctors of this rare disease facing an intractable case, especially the patient have or had the isolated adrenal tuberculosis. Moreover, this case report may give the scientists some enlightenments since our patient suffered from POEMS syndrome just after the right adrenalectomy.

## Acknowledgements

This work was supported by the National Nature Science Foundation of China (No.81670504). The author thanks the patient who was described in this report agrees to publish his information in public.

## Disclosure of Conflict of Interest

None.

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