Austin Therapeutics

Open Access

Austin Publishing Group

Case Presentation

Hyperviscosity Syndrome due to Hashimoto's Disease: A Case Report

Nagata T[1]*, Sekine S[1], Hayashi S[2] and Imura J[2]

[1]Department of Surgery and Science, Graduate School of Medicine and Pharmaceutical Sciences for Research, University of Toyama, Japan

[2]Department of Pathology, Graduate School of Research into Medicine and Pharmaceutical Sciences, University of Toyama, Japan

***Corresponding author:** Takuya Nagata, Department of Surgery and Science, Graduate School of Medicine and Pharmaceutical Sciences for Research, University of Toyama, 2630 Sugitani, Toyama, 930-0194, Japan

Received: June 13, 2016; **Accepted:** August 16, 2016; **Published:** August 23, 2016

Abstract

Hyperviscosity Syndrome (HVS) sometimes associated with the collagen disease, chronic infections, chronic liver disease, and malignant tumors, such as multiple myeloma, macroglobulinemia, and leukemia, and the reports of HVS due to a thyroid disease is rare. Here we report two cases of HVS presumably caused by hypergammaglobulinemia associated with Hashimoto's disease. Both patients presented with significant thyroid enlargement, suggesting an association with hypergammaglobulinemia. They were treated with steroid pulse therapy and, after HVS were relieved, could undergo subtotal thyroidectomy, but one of them died of postoperative acute myocardial infarction. The thyromegaly associated with Hashimoto's disease is not a rare condition, but should be treated with caution because hypergammaglobulinemia or HVS may fail to be identified, which put patients at high risk.

Keywords: Hyper viscosity syndrome; Hashimoto's disease; Thyromegaly; Steroid

Abbreviations

HVS: Hyper Viscosity Syndrome; CT: Computed Tomography

Case Presentation

Case 1

A 61-year-old woman presented with a chief complaint of thyroid gland swelling and a feeling of dyspnea. She had noticed a thyroid mass but left it untreated for the previous 5 years. With recent difficulty breathing when bending her head down, she visited our department for a diagnostic evaluation and treatment. Both lobes of the thyroid were significantly enlarged (Figure 1a and 1b). Blood sampling was difficult as her blood was viscous. A hematological examination showed polyclonal hypergammaglobulinemia and increased coagulability, and she was diagnosed with HVS based on blood viscosity testing [1]. There was no clear evidence of autoimmune disease or malignant tumor.

To relieve HVS, plasma exchange was initiated with a catheter placed through the right internal jugular vein, but thrombosis caused complete occlusion of the internal jugular vein, resulting in perivasculitis (Figure 1c). For the treatment of hypergammaglobulinemia presumably responsible for HVS, steroid pulse therapy was administered, which led to a decrease in gamma globulin levels and the viscosity of blood (Figure 2). After vasculitis was relieved, the blood-flow around thrombus in right jugular vein was partly recovered (Figure 1d), and the patient underwent total thyroidectomy. The weight of resected thyroid was 690 g (Figure 3a), and the histological examination showed the existence of Hashimoto's thyroiditis with infiltration of many plasma cells containing a lot of IgG (Figure 3b). After the operation, gamma globulin levels decreased, and HVS was relieved until now.

Case 2

A 83-year-old man presented with a chief complaint of thyroid enlargement and dyspnea on exertion. He had had a thyroid mass detected and been followed by a local physician for the previous 10 years. He was on treatment for diabetes and atrial fibrillation. With recent difficulty breathing when climbing stairs, he was referred to our department for a complete diagnostic evaluation and treatment. Both lobes of the thyroid were significantly enlarged; the right lobe, in particular, was so swollen as to extend into the mediastinum

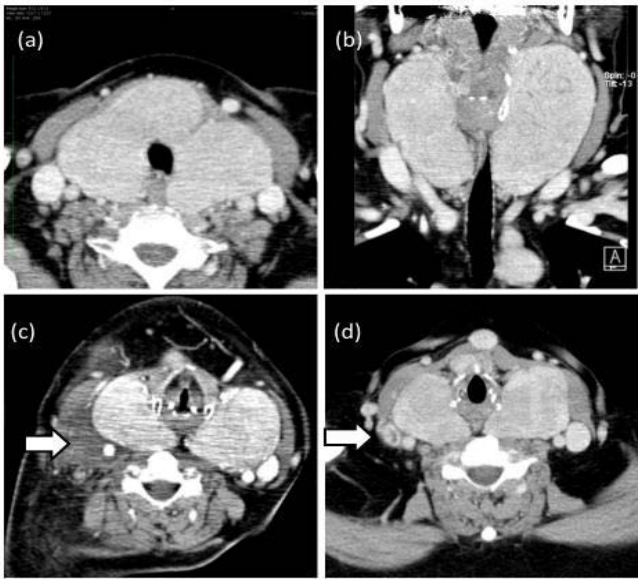

Figure 1: Computed Tomography (CT) of the thyroid in case1. (a) Horizontal and (b) vertical dislocation of pre treatment phase was revealed the enlargement in both sides of thyroid tumor. (c) After plasmapheresis phases that the occlusion of right internal jugular vein (solid white arrow) and perivasculitis was shown. (d) Three weeks later, perivasculitis was disappeared and the blood flow around the rt. internal jugular vein was partially recovered (solid white arrow).

Citation: Nagata T, Sekine S, Hayashi S and Imura J. Hyperviscosity Syndrome due to Hashimoto's Disease: A Case Report. Austin Therapeutics. 2016; 3(2): 1028.

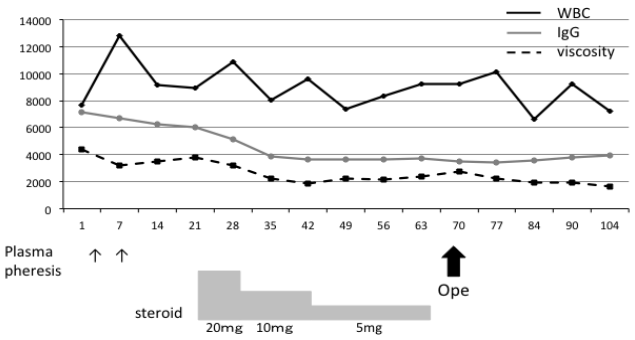

Figure 2: The progress of HVS after treatments. WBC (/mm^3), IgG (mg/dL) and viscosity (x10^{3}cp) were shown. Small arrow: plasmapheresis, grey box: steroid pulse treatment and large arrow: operation (total thyroidectomy) were indicated.

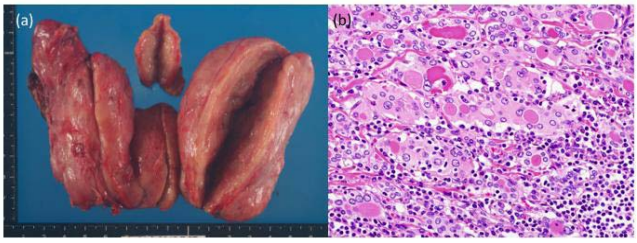

Figure 3: (a) Resected thyroid, volume: 11x14cm, weight: 690g. (b) Histological examination of thyroid tumor (x400).

(Figure 4a & 4b). A hematological examination showed an elevated IgG level of 2857 mg/dL and a high viscosity of 3.520 cp. A needle biopsy of the thyroid showed evidence of chronic thyroiditis, suggesting HVS caused by IgG hypergammaglobulinemia associated with Hashimoto's disease. Steroid pulse therapy resulted in a decrease in gamma globulin levels, and the patient underwent total thyroidectomy. After the operation, he experienced bronchitis and hoarseness, for which symptomatic therapy was continued. He also remained on heparin for atrial fibrillation. On postoperative day 9, he was found collapsed on the bed after breakfast. He was in cardiac arrest, treated with cardiopulmonary resuscitation unsuccessfully, and died. A postoperative hematological examination and autopsy imaging (Ai) led to the diagnosis that his death was caused by acute myocardial infarction.

Discussion

Hyperviscosity Syndrome (HVS) was first reported by Waldenstrom, et al. as a B-cell lymphoproliferative disorder in 1944 [2]. HVS often accompanies collagen disorder, chronic infection, chronic liver disease, and malignant tumors such as multiple myeloma, macroglobulinemia and leukemia [3,4]; reports of HVS resulting from thyroid disease are rare. It is characterized by increased blood viscosity that causes local circulatory disorder, resulting in symptoms such as visual impairment, loss of consciousness and bleeding tendency. The most common factor responsible for increased blood viscosity is IgM. IgM is a pentameric immunoglobulin with a molecular weight of 900,000, tends to bind to other proteins, and is associated with increased relative serum viscosity at a concentration of 3 g/dL or more [5]. Plasmapheresis is reported to be effective in

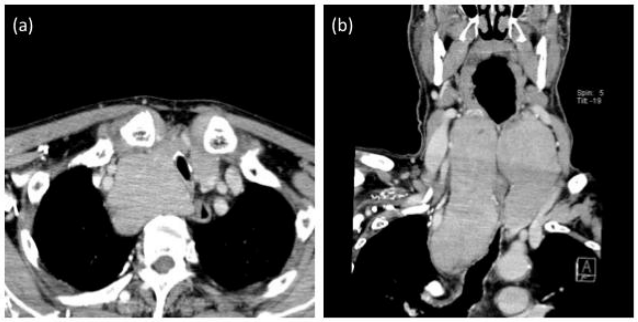

Figure 4: CT of the thyroid in case 2. (a) Horizontal and (b) vertical dislocation was shown.

the treatment of IgM hypergammaglobulinemia [6]. In contrast, IgG is a monomer with a molecular weight of 150,000, and thought to be less likely to cause HVS. However, HVS due to blood IgG forming polymers has been reported in patients with multiple myeloma [7,8].

Our patients were free of HVS-related symptoms, and exhibited significantly high levels of IgG and only slightly higher-than-normal levels of IgM at the time of admission. In Case 1, perivasculitis resulting from thrombosis occurred immediately after plasma exchange for the treatment of HVS. Although heparin was administered as a continuous intravenous infusion, this patient was at high risk for thrombosis, and plasma exchange should have been performed through a peripheral vein without catheter placement. Two sessions of plasma exchange caused only minimal changes in viscosity, failing to achieve the desired effect. In contrast, steroid pulse therapy was highly effective in reducing IgG levels and blood viscosity, making safe thyroidectomy possible. She has remained free of recurrent hypergammaglobulinemia and HVS for two years since the operation. In Case 2, only steroid pulse therapy was administered preoperatively, without plasma exchange. After IgG levels and blood viscosity decreased, thyroidectomy was performed. Despite thorough anticoagulant therapy, he developed acute myocardial infarction after the operation, indicating that HVS should be managed with great caution.

Conclusion

This report presents a first case that steroid pulse therapy was effective for HVS caused by hypergammaglobulinemia resulting from Hashimoto's disease.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Acknowledgement

We acknowledged to Dr. Kazuhiro Tsukada died at 5/ Mar/ 2016, for giving us a lot of helpful suggestions about this work, and Dr. Takashi Hori, for performing core needle biopsy of thyroid tumor. This work was partly supported by JSPS KAKENHI Grant Number 15K10181.

References

1. Trope GE, Lowe GD, McArdle BM, Douglas JT, Forbes CD, Prentice CM, et al. Abnormal blood viscosity and haemostasis in long-standing retinal vein occlusion. Br J Ophthalmol. 1983; 67: 137-142.

2. Waldenström J. Incipient myelomatosis or "essential" hyperglobulinemia with fibrinogenopenia: a new syndrome? Acta Med Scand. 1944; 117: 216-222.

3. Stone MJ, Bogen SA. Evidence-based focused review of management of hyperviscosity syndrome. Blood. 2012; 119: 2205-2208.

4. Chen LY, Wong PC, Noda S, Collins DR, Sreenivasan GM, Coupland RC. Polyclonal hyperviscosity syndrome in IgG4-related disease and associated conditions. Clin Case Rep. 2015; 3: 217-226.

5. Merchionne F, Procaccio P, Dammacco F. Waldenström's macroglobulinemia. An overview of its clinical, biochemical, immunological and therapeutic features and our series of 121 patients collected in a single center. Critical Reviews in Oncology/Hematology. 2011; 80: 87-99.

6. Vos JM, Minnema MC, Wijermans PW, Croockewit S, Chamuleau ME, Pals ST, et al. HOVON Multiple Myeloma Working Party; HOVON Lymphoma Working Party. Guideline for diagnosis and treatment of Waldenström's macroglobulinaemia. Neth J Med. 2013; 71: 54-62.

7. MacKenzie MR, Fudenberg HH, O'Reilly RA. The hyperviscosity syndrome. I. In IgG myeloma. The role of protein concentration and molecular shape. J Clin Invest. 1970; 49: 15-20.

8. Ray PK, Besa E, Idiculla A, Rhoads JE Jr, Bassett JG, Cooper DR. Efficient removal of abnormal immunoglobulin G from plasma of a multiple myeloma patient. Description of a new method for treatment of the hyperviscosity syndrome. Cancer. 1980; 45: 2633-2638.

Austin Therapeutics - Volume 3 Issue 2 - 2016
ISSN: 2472-3673 | www.austinpublishinggroup.com

Citation: Nagata T, Sekine S, Hayashi S and Imura J. Hyperviscosity Syndrome due to Hashimoto's Disease: A Case Report. Austin Therapeutics. 2016; 3(2): 1028.