

Sinus Histiocytosis with Massive Lymphadenopathy and Subglottic Stenosis: A Case Report

Chalermchai Chintrakarn, MD*,
Mana Rochanawuthanon, MD**

* Department of Otorhinolaryngology, Faculty of Medicine, Ramathibodi Hospital, Mahidol University

** Department of Pathology, Faculty of Medicine, Ramathibodi Hospital, Mahidol University

An 18 year-old woman presented with massive cervical lymphadenopathy. She also had bilateral parotid gland enlargement, and later upper airway obstruction from subglottic mass compression. Surgical excision of lymph nodes and bilateral total parotidectomy with facial nerve preservation were selected.

Direct laryngoscopy and CO₂ laser excision were used to remove the subglottic mass. Airway was secured by tracheostomy and after that it was removed. Histologic finding was Sinus histiocytosis with massive lymphadenopathy (SHML) that involved the subglottic. SHML is very rare, probably less than 1000 cases reported in the literature and this is the 6th case in the literature that involved the subglottic. There are no standard treatment modalities. The treatment is comprised of surgery, steroid, chemotherapy and local control. In the present case the authors chose surgery, steroid and local control of disease in the subglottic by using CO₂ laser excision. Now, the disease is under control.

Keywords: Sinus histiocytosis, Subglottic stenosis, Laser surgery

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Sinus histiocytosis with massive lymphadenopathy (SHML) was diagnosed by Rosai and Dorfman in 1969^(1,2). As a tribute to these authors, SHML was then called a Rosai-Dorfman disease. The disease is thought to be a disorder of the mononuclear phagocyte and immunoregulatory effector (M-PIRE) system⁽³⁾. SHML is diagnosed by large histiocyte-like cells, Rosai-Dorfman (RD) cells.

The disease may occur at the extranodal site even without any clinical involvement of lymph nodes. It is characterized by painless cervical lymphadenopathy, fever, leukocytosis, increased erythrocyte sedimentation rate, and hypergammaglobulinemia⁽²⁾. The disease has been found in patients from 7 months to 67 years old. However, most of cases occur before age 20 years⁽⁴⁾.

Twenty-eight percent of patients have extranodal disease⁽⁵⁾. Mostly in the upper respiratory tract⁽⁶⁾, eyelids, orbits⁽⁷⁾ and skin⁽⁸⁾. The most common

site of upper respiratory tract disease is the nasal cavity. The tongue, oral mucosa, pharynx, tonsils and paranasal sinuses are also involved. The upper respiratory tract symptoms are nasal obstruction, rhinitis, epistaxis and dyspnea⁽⁹⁾. In the present case, the patient had subglottic involvement which is the 6th case in the literature^(4,10-13).

Histologically, the involved lymph nodes show marked dilation of the subcapsular and medullary sinuses. These are filled with histiocytes which often contain phagocytized lymphocytes. These histiocytes have a benign appearance but do show rare mitoses. The histologic differential diagnosis of SHML comprises a variety of diseases, including malignant lymphomas of histiocytic type, histiocytosis⁽²⁾ and histiocytic medullary reticulosis⁽⁵⁾. The main histologic distinctions of this disorder are the relatively unremarkable appearance of the histiocytes and phagocytized lymphocytes.

Case Report

An 18 year-old woman complained of a left neck mass for four months.

Correspondence to : Chintrakarn C, Department of Otorhinolaryngology, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok 10400, Thailand.

4 months PTA: She began to have left preauricular swelling. After that she had submental and left neck masses. She was seen in a rural hospital and had cervical lymph node excisional biopsy. Histologic result was benign sinus histiocytosis of lymph node. AFB was not found. She did not receive any specific treatment.

2 months PTA: She had bilateral preauricular masses, submental and bilateral cervical masses that grew very rapidly albeit with antibiotic treatment. Then she decided to go to Ramathibodi Hospital.

ENT physical examination revealed

- Left preauricular lymph node, 2 cm in diameter
- Left submandibular lymph node, 5 cm in diameter
- Left jugulodigastric lymph node, 3 cm in diameter
- Right submandibular lymph node, 3 cm in diameter
- Left pretracheal lymph node, 1 cm in diameter

Fine needle aspiration biopsy was performed and the result was abnormal lymph node possibly sinus histiocytosis.

She was scheduled for left submandibular lymph node excision on September 15, 2000. Histologic result was sinus histiocytosis with massive lymphadenopathy.

1 month after that, the other lymph node did not subside so she was scheduled for right submandibular lymph node excision on October 18, 2000. Histologic result was the same.

1 month after that (November 30, 2000), the cervical lymph nodes not only persisted but also grew in all cervical lymph nodes area. She began to have an airway problem.

Ultrasound of the abdomen was negative

CT scan of the head and neck revealed:

There were masses at the nasopharynx, supraglottic, para-glottic, subglottic and trachea with 50% obstruction and bilateral true vocal cord paralysis. Multiple lymph nodes at both retropharyngeal, deep cervical, both paratracheal and left supraclavicular area were also found.

She was scheduled for tracheostomy, direct laryngoscopy with laser, bilateral functional neck dissection, bilateral submandibular gland excision and left preauricular node excision on January 11, 2001.

Operative finding:

- Multiple cervical lymph nodes ranging from 1 cm to 5 cm in diameter in level I-V, bilaterally.

- Well encapsulated lymph nodes.
- Two left preauricular nodes 1.5 cm in diameter.
- Subglottic masses at anterior and posterior walls.
- Subglottic stenosis with Cotton's grade 2 and 1 cm in length.
- CO₂ laser 10 watt, repeated pulse 0.5 second was selected to vaporize the subglottic masses.

Two months after the operation, the tracheostomy tube was closed and then the tube was removed. Unfortunately, the bilateral parotid glands were enlarged. Prednisolone was started at dose 30 mg per day and then tapered off in one month. November 15, 2001 bilateral parotid gland was further enlarged along with the right jugulodigastric node.

She was scheduled for bilateral total parotidectomy and lymph node excision on December 18, 2001.

Operative finding:

- Masses in both superficial lobes of parotid glands, 4 cm in diameter
- Right jugulodigastric node, 3.5 cm in diameter.

Two months after surgery, no facial palsy and no mass had occurred. She had no airway problem.

She has followed up at 3 monthly intervals until December 2003. The disease is under control. She can breath normally and no mass has occurred.

Histological findings

The specimens submitted for pathological examinations revealed 2 to 2.5 cm lymph nodes admixed with salivary glands. There were some lymph nodes which were within the salivary glands and showing the compression effect (Fig. 1) The lymph nodes showed good encapsulations and lobular yellow-tan cut surfaces. Histologically, they showed a slight effacement of the lymphoid architectures, as characterized by small to absence of the germinal centers, irregular paracortical areas and remarkable sinusoidal dilations. Within the sinusoids, there were numerous histiocytes which showed medium sized nuclei and prominent nucleoli. The cytoplasm were abundantly amphophilic with distinct but thin plasma membranes. Small lymphocytes and some red blood cells were present within the cytoplasm of the histiocytes (emperipolesis). Within the paracortical area

there were smaller numbers of small lymphocytes and prominent plasma cells, but not eosinophils (Fig. 2). All these findings were the characteristic features of sinus histiocytosis with massive lymphadenopathy⁽¹⁾.

Discussion

SHML is a rare idiopathic histiocytic proliferative disorder occurring in all age groups but is most common in children and young adults. The ratio between male and female is 58:42. The etiology is not well understood. It is a very rare but established disease.

Sinus histiocytosis with massive lymphadenopathy

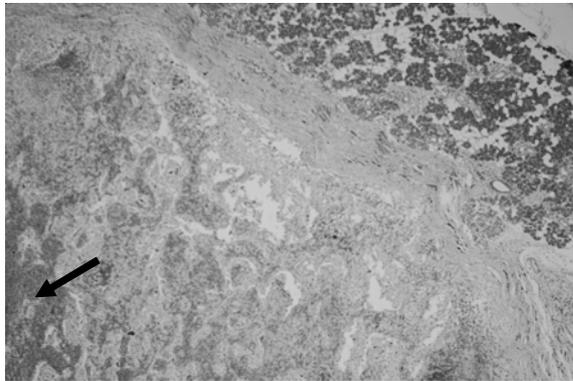


Fig. 1 Low power picture of the lymph node showing markedly dilated sinusoids, small germinal centers (arrow), thick fibrous capsule and compressed salivary gland (H & E x 20)

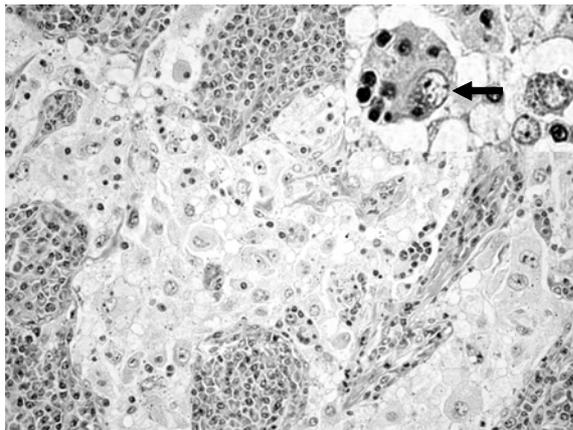


Fig. 2 The dilated sinusoids contained numerous histiocytes with ample amounts of pale cytoplasm and thin plasma membranes. Plasma cells infiltrated within the lymphoid parenchyma. In set showed emperipolesis of small lymphocytes and red blood cells in the circulating histiocytes (arrow)

denopathy should be considered in the differential diagnosis of cervical lymphadenopathy especially if extranodal abnormalities are found. The natural history of the disease is variable. Most patients are asymptomatic and the lymph nodes resolve spontaneously in a few months to several years. This woman is the 6th case of SHML that involved the subglottic.

Treatment modality comprises of antibiotics, corticosteroid, antituberculosis, radiotherapy, chemo-

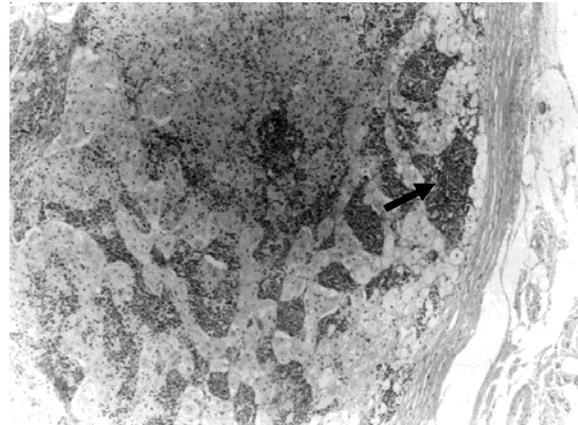


Fig. 3 Low power field of the lymph node of the subglottic area reveals markedly dilated sinusoids which are filled up by histiocytes to produce paler areas in this picture. Between the sinusoids darker areas of lymphocytes and plasma cells are present. (arrow) The capsule of the lymph node is thick and fibrotic (H&E x 40)

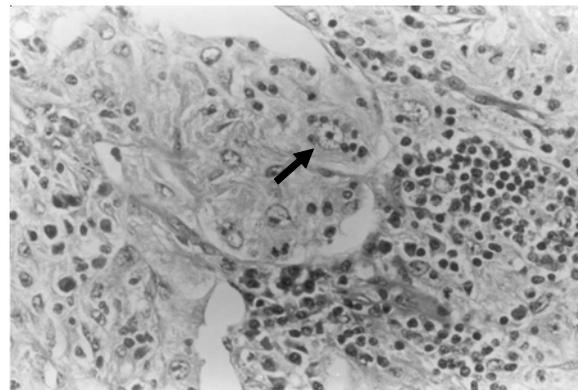


Fig. 4 Higher magnification of the lymph node in Fig. 3 reveals clusters of histiocytes in the sinusoids. They show abundant cytoplasm and medium sized nuclei. The nuclei are vesicular and containing prominent nucleoli (arrow). Some of the histiocytes contain lymphocytes, and plasma cells. Between the sinusoids are clusters of small lymphocytes and plasma cells (H&E x 200)

therapy and surgery⁽⁶⁾. Combination of prednisolone with chlorambucil has shown some benefit⁽⁴⁾. In general, the most selected treatment is surgery, steroid and local control of disease^(4,10-13).

In the present case, the authors choose surgical excision of the lymph nodes along with bilateral total parotidectomy with facial nerve preservation. Direct laryngoscopy and CO₂ laser excision were selected to remove the subglottic mass. Airway was secured by tracheostomy and after that it was removed. Now, the disease is controlled.

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รายงานผู้ป่วยโรคไซนัสฮิสติโอไซโตสิสที่มادت้วยต่อมน้ำเหลืองขนาดใหญ่ร่วมกับมีภาวะอุดตัน ไตสายเสียง

เฉลิมชัย ชินตระการ, มานะ โจรนวุฒนนท์

หญิงไทยใสด อายุ 18 ปี มาด้วยต่อมน้ำเหลืองที่คอโตขนาดใหญ่ทั้ง 2 ข้าง ร่วมกับมีต่อมน้ำลาย parotid โตขึ้นทั้ง 2 ข้าง ต่อมาเริ่มมีอาการหอบเหนื่อยแบบภาวะอุดกั้นทางเดินหายใจส่วนบน ตรวจร่างกายพบมีก้อนที่หลอดลม บริเวณไตสายเสียง ผู้ป่วยรายนี้ได้รับการรักษาโดยการผ่าตัดเอาต่อมน้ำเหลืองที่คอออก ร่วมกับการผ่าตัดต่อมน้ำลาย parotid ออกทั้ง 2 ข้าง โดยเก็บเส้นประสาทสมองคู่ที่ 7 ได้ทั้ง 2 ข้าง ส่วนเนื้องอก ที่ไตสายเสียง ได้รับการรักษาโดย Direct laryngoscopy และใช้คาร์บอนไดออกไซด์เลเซอร์ ผู้ป่วยได้รับการเจาะคอในช่วงแรก ต่อมาได้ถอดออกเมื่อผู้ป่วย หายดีแล้ว ผลการตรวจทางพยาธิวิทยาพบเป็น Sinus histiocytosis with massive lymphadenopathy (SHML) ซึ่งถือว่าเป็นโรคที่พบได้ยากมาก โดยพบน้อยกว่า 1000 รายใน world literature นอกจากนี้ ผู้ป่วยรายนี้ยังเป็นรายแรก ในประเทศไทย ที่พบที่บริเวณหลอดลมไตสายเสียง และเป็นรายที่ 6 ใน world literature ที่พบในบริเวณหลอดลม ไตสายเสียง การรักษาโรคนี้ในปัจจุบันยังไม่มีมาตรฐาน โดยทั่วไปจะใช้วิธีการผ่าตัด, ให้น้ำ steroid, ให้น้ำเคมีบำบัด, การฉายรังสี และควบคุมอาการเฉพาะที่ ในรายนี้ ได้ใช้วิธีผ่าตัด, ให้น้ำ steroid, และควบคุมอาการเฉพาะที่บริเวณ ไตสายเสียงโดยใช้ CO₂ laser ขณะนี้ผู้ป่วยสบายดี ไม่มีอาการผิดปกติ
