Descending necrotizing mediastinitis complicating hyperimmunoglobulin E syndrome

Yusuke Takanashi¹, Takamitsu Hayakawa¹, Hiroshi Neyanai¹ & Kazuhito Funai²

¹Department of Thoracic Surgery, Fujieda Municipal General Hospital, Fujieda, Japan.
²Department of Surgery 1, Hamamatsu University School of Medicine, Hamamatsu, Japan.

Keywords
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Correspondence
Yusuke Takanashi, Department of Thoracic Surgery, Fujieda Municipal General Hospital, 4-1-11 Surugadai, Fujieda, Shizuoka, Japan.
E-mail: nashimed@yahoo.co.jp

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Abstract
Descending necrotizing mediastinitis (DNM) is a potentially lethal disease that originates from a deep neck infection (DNI); it is often associated with an immunocompromised state. Hyperimmunoglobulin E syndrome (HIES) is an extremely rare complex immune deficiency characterized by recurrent abscesses of staphylococcal etiology. A rare case of a 34-year-old woman wherein HIES putatively promoted the development of DNI into DNM is described. She presented with cervical purulent lymphadenitis and retropharyngeal abscess. Despite immediate cervical drainage and use of broad-spectrum antibiotics, she developed DNI and subsequently DNM. Mediastinal drainage with thoracotomy and subsequent deep neck drainage were performed. Bacterial culture of the abscess isolated methicillin-resistant Staphylococcus aureus (MRSA). Although a postoperative recurrent deep neck abscess required a second surgery, we succeeded in conservative remission of recurrent mediastinal abscess with long-term use of anti-MRSA drugs. Sufficient drainage under thoracotomy and robust administration of postoperative antibiotics resulted in successful management of HIES-associated DNM.

Introduction
Descending necrotizing mediastinitis (DNM) is a rare, potentially lethal disease originating from a deep neck infection (DNI) and is often associated with an immunocompromised state [1]. Hyperimmunoglobulin E syndrome (HIES) is a complex immune deficiency characterized by elevated immunoglobulin E levels in serum and recurrent abscesses of staphylococcal etiology [2]. Although HIES is known to be complicated with skin abscesses [2], cases of HIES associated with DNM have not been previously reported. A rare case of a young woman wherein HIES putatively promoted the development of DNI into DNM is described.

Case Report
A 34-year-old woman presented to our hospital because of fever and right cervical pain for 3 days. She had a medical history of HIES with associated recurrent skin abscesses. On arrival, physical examination revealed a body temperature of 40.4°C and swelling and redness of the right parotid area and right pharyngeal wall. Laboratory studies revealed elevated white blood cells (24,000/μL), C-reactive protein (28.2 mg/dl; normal value < 0.2 mg/dl), and immunoglobulin E (7900 IU/mL; normal value < 170 IU/mL). Cervical-enhanced computed tomography (CT) disclosed right cervical purulent lymphadenitis (Fig. 1A: arrow) and a retropharyngeal abscess (Fig. 1B: arrow). She was admitted to the otolaryngology service for right cervical drainage by paracentesis. Despite the immediate administration of meropenem (Nippon Chemiphar, Tokyo, Japan) (2 g/day), the abscess rapidly progressed, and 5 days after admission, she required a tracheotomy for laryngeal edema. The antibiotics were changed to vancomycin (Kohayashi Kako, Awara, Japan) (2 g/day) because of her medical history of HIES and its association with recurrent abscesses of staphylococcal etiology. This achieved temporary remission of the inflammatory reaction on laboratory studies and achoresis of the abscess; however, it was followed by sudden recrudescence 28 days after admission. Enhanced CT revealed bilateral deep neck abscesses (Fig. 1C: arrowheads) that extended down the right upper mediastinum along the carotid sheath to the subcarinal level, forming a right-
sided mediastinal abscess (Fig. 1C, D: arrows), and the patient was referred to our department of thoracic surgery with a diagnosis of DNM (Type IIB). Emergency mediastinal drainage with right transaxillary thoracotomy and subsequent bilateral deep neck drainage were performed. Intraoperative findings showed a white, nebulous collection of pus along the mediastinal pleura extending from below the subcarinal level of the posterior mediastinum to the right superior paratracheal lymph node of the superior mediastinum (Fig. 1E). The mediastinal pleura and retrovisceral space (Fig. 1F) were widely opened for the placement of chest tubes for postoperative drainage. Bacterial culture of the deep neck abscess isolated methicillin-resistant *Staphylococcus aureus* after the surgery, and antibiotics were changed to linezolid (Phizer, Manhattan, New York, USA) (1.2 g/day). Complete elimination of the abscesses was confirmed on CT immediately after the surgery. However, during the postoperative course, the recurrence of deep neck and mediastinal abscesses was detected on CT (Fig. 2A, B: arrows); the former required a second surgery for drainage, and the latter was observed using CT at intervals of 3 to 4 days and treated with continuous linezolid. Complete remission of the abscesses and negative conversion of inflammatory reaction were confirmed 65 days after the initial surgery (Fig. 2C, D).

**Discussion**

This case is a rare presentation of DNM wherein HIES is suspected to play a role as an exacerbating factor for infection.

Descending necrotizing mediastinitis is a potentially lethal disease originating from the downward spread of DNI. The most common etiology of DNM is attributed to DNI secondary to odontogenic, pharyngeal, and cervicofacial infections. Although DNM is often associated with an immunocompromised state with a high incidence of 38% reported by Ridder et al. [1], to the best of our knowledge, cases of HIES-associated DNM have not been previously reported.

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*Figure 1.* Cervical-enhanced computed tomography (CT) on admission to hospital revealed the following: (A) right cervical purulent lymphadenitis (arrow) and (B) retropharyngeal abscess (arrow). Twenty-eight days after admission, enhanced coronal (C) and sagittal (D) CT images showed bilateral deep neck abscesses (arrowheads) that extended down the right upper mediastinum along the carotid sheath to the subcarinal level, forming a right-sided mediastinal abscess (arrows). Intraoperative findings showed a white, nebulous collection of pus along the mediastinal pleura (E). The mediastinal pleura and the retrovisceral space were widely opened (F).
HIES is an extremely rare complex immune deficiency with a low incidence of <1:1000 000 [2,3] and is characterized by elevated immunoglobulin E in serum and recurrent skin abscesses of staphylococcal etiology [2]. The etiology of HIES is explained by a transcription factor mutation of the signal transducer and activator of transcription 3 that results in impairment of T helper lymphocytes and signaling of various cytokines required for immune response [2,3]. In the current case, cervical purulent lymphadenitis resulted in DNI, which subsequently spreads downward along the carotid sheath to form mediastinal abscesses; HIES was presumed to have promoted the development of DNI into DNM.

Optimum strategies for the surgical management of DNM remain controversial, and inadequate mediastinal drainage is the major cause of mortality in DNM. Endo et al. [4] classified DNM according to abscess distribution and established guidelines for surgical management: Type I is localized above the carina and can be adequately drained by transcervical mediastinal drainage; Type IIA extends to the lower anterior mediastinum and may require subxiphoidal drainage in addition to transcervical approach; and Type IIB affects the anterior and posterior mediastinum and demands aggressive mediastinal drainage via thoracotomy. Endo’s criteria have been widely applied for the surgical management of DNM; however, in recent years, some authors advocate more aggressive approaches with thoracotomy regardless of the degree of abscess extension [5]. In the present case, preoperative CT showed the mediastinal abscess had encompassed the subcarinal level of the lower posterior mediastinum (Type IIB), and thoracotomy for the provision of satisfactory mediastinal drainage was selected without hesitation. We believe the underlying HIES facilitated the recurrent abscess formation during the postoperative course and complicated infection control. Although the recurrent deep neck abscess required a second surgery, we believe that the combination of aggressive drainage with thoracotomy and robust administration of postoperative antibiotics resulted in the successful treatment of mediastinal abscess without second surgery even though in the presence of HIES.

Disclosure Statements
No conflict of interest declared.
Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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References
1. Ridder GJ, Maier W, Kinner S, et al. 2010. Descending necrotizing mediastinitis: contemporary trends in etiology, diagnosis, management, and outcome. Ann. Surg. 251:528–534.
2. Freeman AF, Holland SM. 2008. The hyper-IgE syndromes. Immunol. Allergy Clin. North Am. 28:277–291.
3. Jończyk-Potocznia K, Szczawińska-Popłonyk A, Warzywoda M, et al. 2012. Hyper Ig E syndrome (job syndrome, HIES) -

Figure 2. The recurrence of deep neck (A, arrow) and mediastinal abscess (B, arrow) was detected during the postoperative course, which was resolved by the long-term use of anti-methicillin-resistant Staphylococcus aureus drugs and deep neck drainage (C, D).
radiological images of pulmonary complications on the basis of
three cases. Pol. J. Radiol. 77:69–72.
4. Endo S, Murayama F, Hasegawa T, et al. 1999. Guideline of sur-
gical management based on diffusion of descending necrotizing
mediastinitis. Jpn. J. Thorac. Cardiovasc. Surg. 47:14–19.
5. D'Cunha J, James M, Antonoff MB, et al. 2013. Descending nec-
rotizing mediastinitis: a modified algorithmic approach to define
a new standard of care. Surg. Infect. (Larchmt.) 14:525–531.