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DESCENDING NECROTIZING MEDIASTINITIS AS A FATAL COMPLICATION OF NK/T-CELL LYMPHOMA: A CASE REPORT

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Abstract

Introduction. Extranodal NK/T-cell lymphoma (ENKL) of the nasal type is a rare and aggressive tumor with a poor prognosis. The condition often presents with atypical symptoms, leading to delays in diagnosis and treatment. This lymphoma type is characterized by rapid progression, often involving the nasal passages, upper gastrointestinal tract, and other extranodal sites. Early detection and intervention are crucial, as the disease can lead to such severe complications as descending necrotizing mediastinitis, as observed in this case.
Aim. This report describes a fatal case of descending necrotizing mediastinitis due to ENKL and highlights the clinical challenges in its management.
Materials and methods. A 36-year-old male was diagnosed with ENKL after presenting with pharyngitis and laryngitis. Initial misdiagnosis and delayed histological findings prolonged the disease course over one year. Surgery, including a left-sided transcervical approach, was performed to drain the deep cervical phlegmon and mediastinum. Chest and neck CT scans were crucial in assessing disease extent and mediastinitis.
Results. Despite surgery and aggressive treatment, the patient's condition deteriorated. He developed purulent wound leakage, persistent fever, and respiratory distress, leading to fatal bleeding from the left external carotid artery on the 9th postoperative day.
Discussion. ENKL has an aggressive course and poor prognosis, often being misdiagnosed due to symptom overlap with other conditions like nasopharyngeal angiofibroma. In this case, delayed diagnosis and Epstein-Barr Virus (EBV) involvement contributed to rapid decline. Early biopsy and multi-specialist collaboration are essential for accurate diagnosis and treatment. Timely recognition and combined chemotherapy and radiotherapy are critical for improving outcomes.
Conclusions. This case illustrates descending necrotizing mediastinitis as a fatal complication of extranodal NK/T-cell lymphoma, nasal type. The rapid, necrotic spreading into cervical and mediastinal tissues emphasizes the importance of early biopsy, accurate histological diagnosis, and timely surgical drainage. Persistent upper airway lesions require thorough evaluation to avoid delayed diagnosis and life-threatening outcomes.

Keywords: **mediastinitis, transcervical approach, non-Hodgkin lymphoma, extranodal NK/T-cell, extranodal NK/T-cell lymphoma (ENKL), tracheoesophageal fistula**

INTRODUCTION

Extranodal NK/T-cell lymphoma of the nasal type (ENKL) is a rare, aggressive non-Hodgkin lymphoma that primarily affects the nasal cavity, paranasal sinuses, and upper gastrointestinal tract [5, 4]. Despite often appearing benign initially, ENKL is marked by rapid local tissue destruction, angiodestruction, and systemic toxicity [1, 6]. The disease is strongly associated with Epstein–Barr Virus (EBV), clonal T-cell receptor (TCR) gene rearrangements, and STAT3/STAT5B mutations, all of which correlate with poor prognosis [5, 7].

Diagnosis is challenging due to its rarity and nonspecific symptoms. Accurate identification requires histological and immunophenotypic analysis, with IHC markers such as CD2, CD3ε, CD5, CD56, TIA-1, and Ki-67. Risk stratification tools like PINK and PINK-E

help assess prognosis based on disease stage, EBV load, and clinical presentation [2].

ENKL can lead to rare but fatal complications, including descending necrotizing mediastinitis (DNM) – a polymicrobial infection spreading from deep neck spaces to the mediastinum [4].

We report a rare case of ENKL complicated by DNM, highlighting the diagnostic difficulties and emphasizing the importance of early multidisciplinary management to prevent fatal outcomes.

AIM

The study aims to present a case report of descending necrotizing mediastinitis as a fatal complication of NK/T-cell lymphoma.

MATERIALS AND METHODS

This study is based on a retrospective review of a clinical case involving a patient diagnosed with descending necrotizing mediastinitis (DNM) as a complication of extranodal NK/T-cell lymphoma. Clinical, surgical, and pathological data were obtained from hospital records and analyzed to assess the course of the disease, diagnostic challenges, and therapeutic interventions.

A 36-year-old man was admitted to the hospital in November 2020, with a suspected diagnosis of descending necrotizing mediastinitis. His medical history revealed progressive pharyngitis and laryngitis persisting for approximately one year following a common cold. A primary biopsy of the nasopharyngeal mass, performed in January 2020, initially yielded a diagnosis of juvenile nasopharyngeal angiofibroma.

Due to the development of stage III laryngostenosis, an inferior tracheostomy was performed in October 2020. During this procedure, a second laryngeal biopsy was obtained for further histopathological evaluation. Immunohistochemical (IHC) analysis of the laryngeal tissue in October 2020, revealed squamous and respiratory epithelium of the mucosal layer with necrotizing and ulcerative infiltrates, consistent with ENKL. Lymphoma cells exhibited diffuse expression of CD3 and granzyme B, with some cells showing positive CD56 and CD30 markers. The Ki-67 proliferation index was approximately 80%. Additionally, Epstein-Barr Virus (EBV) chromogenic *in situ* hybridization was detected [Fig. 1, 2].

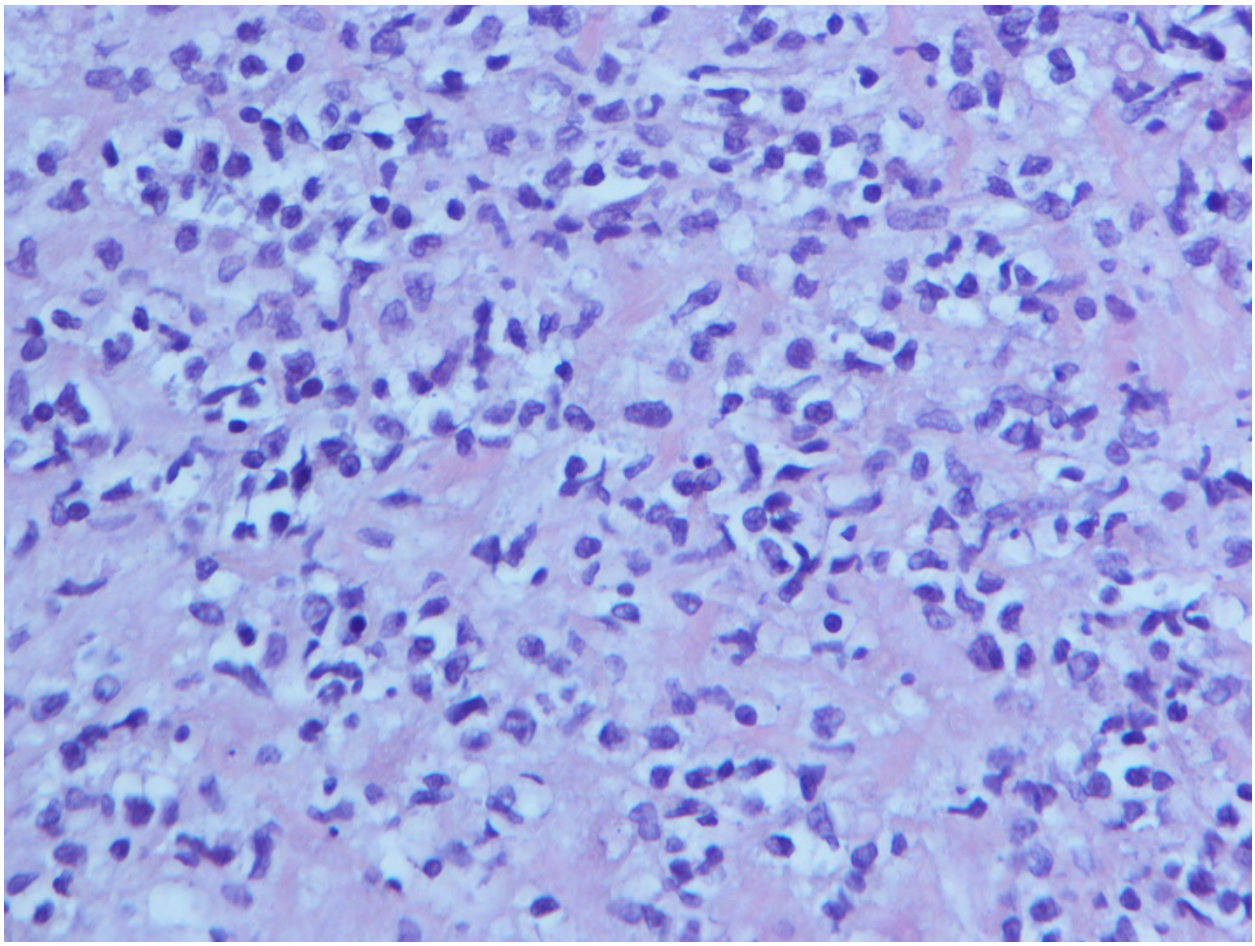

Figure 1. Histological picture of lymphoma cells exhibited diffuse expression of CD3 and granzyme B, along with a few cells showing positive CD56 and CD30 markers (H&E, ×40).

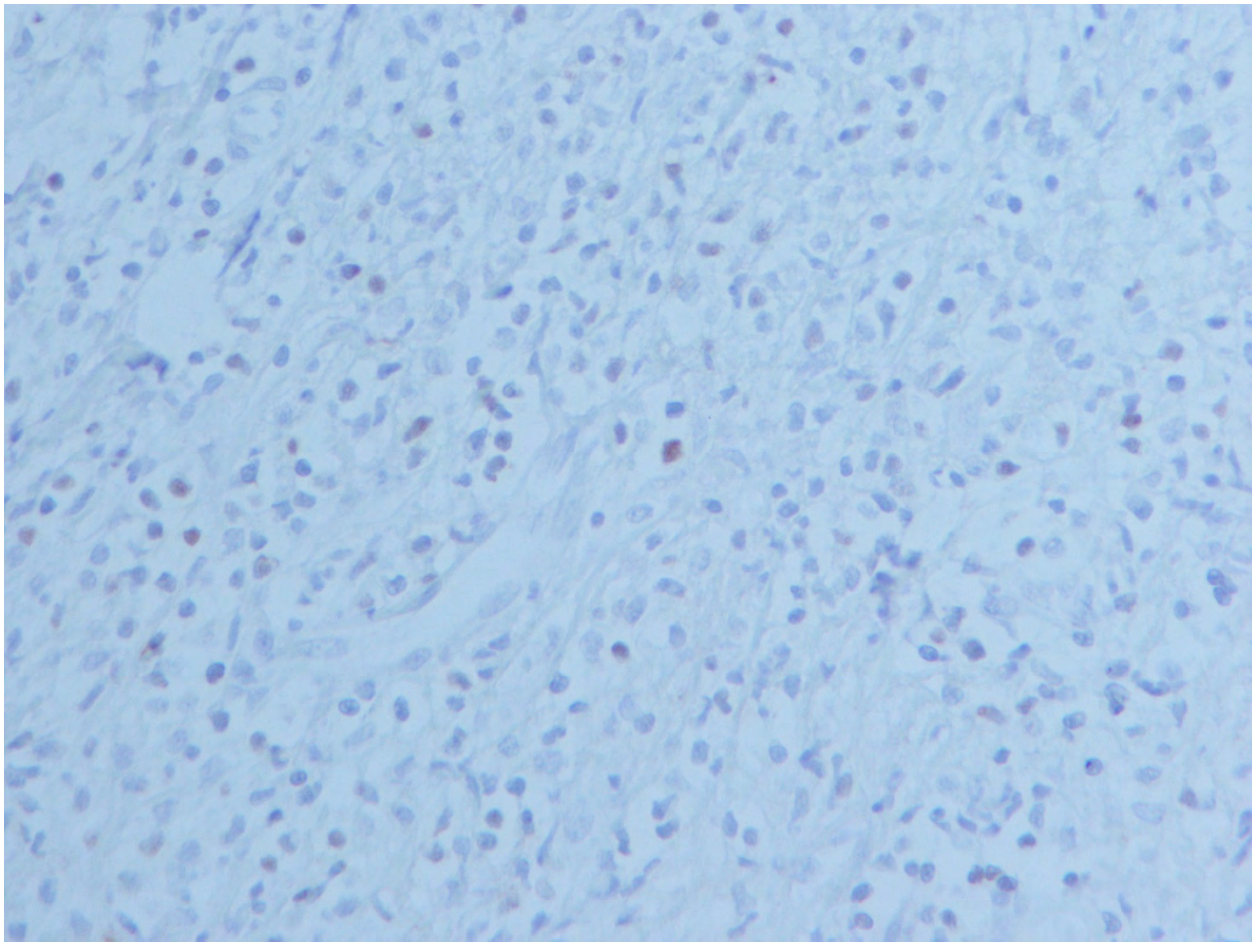

Figure 2. Histological picture of Epstein-Barr Virus (EBV) chromogenic *in situ* hybridization (H&E, ×40).

The patient was employed in an office setting and had no history of occupational exposure to hazardous substances or professional health risks.

On the 7th postoperative day after tracheostomy, the patient experienced bleeding from the superior thyroid artery, which was managed using a left-sided transcervical approach and vessel ligation.

RESULTS

The patient presented with severe general condition, including fever (38.5 °C), dyspnea, productive cough with purulent sputum, cyanosis, and neck swelling. On physical examination, a lower tracheostomy, nasogastric feeding tube, and a postoperative scar on the left side of the neck from a previous lateral cervicotomy were noted. The skin appeared pale; respiratory rate was 32 bpm; pulse was 105 bpm, rhythmic and weak. Auscultation revealed diminished breath sounds but no wheezing. A large amount of fetid purulent sputum in the tracheobronchial tree required regular bronchoscopic sanitation.

Chest and neck CT revealed heterogeneous infiltrates with air bubbles surrounding the pharynx, larynx, trachea, and esophagus, with luminal narrowing and parapharyngeal extension into the posterior superior mediastinum, consistent with Endo type I mediastinitis. A 4.2 × 3.8 × 3.6 mm cavitary lesion in the right middle lobe, resembling a lung abscess, was also identified. Subsolid ground-glass infiltrates were seen in both lungs. Free air was present in the neck's soft tissues, and pericardial effusion measuring 1.8 mm was noted. Laboratory findings showed signs of endogenous intoxication, including anemia (hemoglobin 106 g/L), leukocytosis (9.8 × 109/L) with a left shift (52% band neutrophils), hypoproteinemia (total protein 53 g/L), and proteinuria (0.066 g/L).

A preliminary diagnosis of deep cervical phlegmon and acute descending posterior superior mediastinitis was established. Emergency surgery was performed via a left-sided transcervical approach, with drainage of the cervical and mediastinal spaces. Intraoperative findings revealed extensive necrosis of the pharynx, larynx, cervical esophagus (which was resected and catheterized), and posterior tracheal wall with an esophagotracheal fistula [Fig. 3]. Purulent material with gas bubbles extended along the esophagus into the superior mediastinum. Osteomyelitis of the hyoid bone and purulent spondylitis of the C5–C6 vertebrae with bone sequestration were also evident.

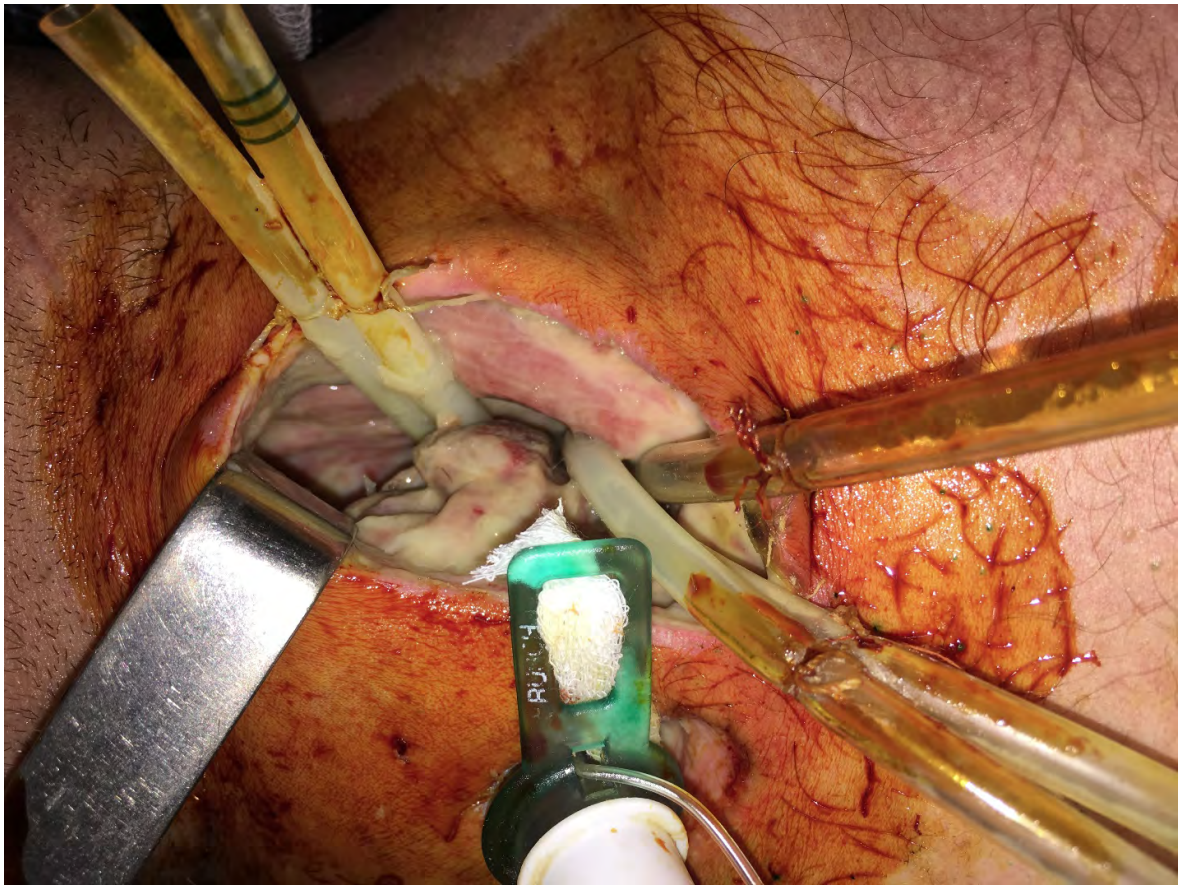

Figure 3. ***Status localis*** **of the cervical wound on the 6th postoperative day.**

A gastrostomy was performed for enteral nutrition (feeding). Multi-resistant *Pseudomonas aeruginosa* was cultured from the cervical wound. Exudative pleurisy developed on both sides, managed with pleural punctures and left-sided Bülau drainage. The patient required daily antiseptic wound irrigation, repeated necrectomies, and bronchoscopic sanitations to remove purulent secretions and maintain airway patency.

Despite comprehensive intensive and thoracic care, including broad-spectrum antibiotics (meropenem, linezolid, amikacin, metronidazole), analgesics, anticoagulants, beta-blockers, antipyretics, and albumin, the general condition gradually deteriorated. Purulent secretions from the cervical wound persisted, and esophagotracheal fistula contributed to ongoing pulmonary contamination. No significant reduction in sputum volume or fever was observed. The patient suffered daily from hyperthermia (36.5 °C-39.4 °C), severe neck and chest pain, dyspnea, and exhausting cough. On postoperative day 5, partial dehiscence occurred at the gastrostomy site with minor leakage of feed, requiring local repair under anesthesia.

On the 11th postoperative day profuse errosive bleeding occurred from the lower corner of the postoperative neck wound, temporarily controlled with tight gauze tamponade. During immediate surgical revision, the patient experienced cardiac arrest. Resuscitation efforts were unsuccessful, and biological death was declared.

The final postmortem diagnosis was: Extranodal NK/T-cell non-Hodgkin lymphoma, nasal type, with isolated involvement of the larynx and pharynx (Stage IVb, Group IV); phlegmonous-necrotizing pharyngitis and esophagitis with necrosis of the posterior and left lateral pharyngeal walls; total necrosis of the cervical esophagus; posterior tracheal wall necrosis with esophagotracheal fistula; deep neck phlegmon; superior mediastinitis; osteomyelitis of the hyoid bone; purulent C5–C6 vertebral spondylitis; sepsis; right middle lobe lung abscess; bilateral exudative pleurisy; exudative pericarditis; infectious-allergic myocarditis; Stage IIa heart failure; sinus tachycardia; hemoptysis; metaplastic anemia; massive hemorrhage from the cervicotomy wound (erosion of carotid artery); hemorrhagic shock; bedsores; tracheostomy; gastrostomy.

DISCUSSION

Currently, induction therapy is considered the optimal treatment strategy for extranodal NK/T-cell lymphoma (ENKL), nasal type, with combined chemotherapy regimens and radiotherapy playing key

roles in improving survival outcomes [1, 7]. However, diagnosis remains challenging, especially in early stages when the disease mimics benign or inflammatory conditions.

Previous rare reports have linked malignancies of the Pirogov-Waldeyer lymphoepithelial ring and various forms of non-Hodgkin lymphoma to deep neck phlegmon and descending necrotizing mediastinitis (DNM) [3]. While odontogenic infections are the most common cause of DNM, neoplastic infiltration and subsequent necrosis may provoke similar descending infections, particularly in immunocompromised or misdiagnosed patients.

DNM is a rare but extremely lethal complication, with reported mortality rates of 12%–24% [8]. Its outcomes are heavily influenced by early diagnosis, timely surgical intervention, and effective source control. In our case, the patient's prolonged disease course led to initially mistaken for nasopharyngeal angiofibroma and led to misdirected immunosuppressive treatment (glucocorticoids) and delayed accurate diagnosis (Stage IV ENKL).

The patient's fatal outcome was the result of multiple converging factors: late recognition of ENKL, positive EBV status (EBER-CISH), misleading histological findings, and delayed surgical management of infectious complications. The extensive local infiltration caused destructive changes in the pharynx, larynx, trachea, and esophagus, ultimately resulting in cervical necrosis, tracheoesophageal fistula, and mediastinitis. Due to the advanced stage and systemic deterioration (sepsis, anemia, cachexia), only palliative surgical interventions were possible, which could not improve survival.

Notably, one technical issue may have further worsened the course – primary closure of the cervicotomy wound without drainage after hemorrhage control. This may have contributed to abscess formation and DNM progression due to impaired wound drainage.

CONCLUSIONS

1. This case highlights descending necrotizing mediastinitis as a fatal complication of extranodal NK/T-cell lymphoma, nasal type – a rare but aggressive malignancy that often presents under the guise of benign ENT pathologies. The unique feature of this observation is the rapid destructive spreading of tumor-related necrosis into deep cervical and mediastinal spaces, causing multi-organ failure despite intensive multidisciplinary care.

2. ENKL should be considered in the differential diagnosis of persistent or atypical upper aerodigestive tract lesions. Delay in biopsy or histopathological misinterpretation can lead to catastrophic outcomes. The association with EBV and aggressive angiocentric tissue destruction further increases the risk of severe infectious complications such as DNM, especially when corticosteroids are misused.

3. Timely ENKL diagnosis requires early and repeated biopsy, combined with contrast-enhanced neck and chest CT and bronchoscopy. Multidisciplinary management is essential in complicated cases. Surgical management of necrosis must always include proper drainage, and postoperative wounds should be closed over a drain via a separate counterincision. Histological confirmation is mandatory for laryngeal or pharyngeal stenosis, with second-opinion pathology recommended when results are unclear.

Perspectives for further research. This case highlights the relevance of further research in Ukraine on early detection of extranodal NK/T-cell lymphoma and prevention of its surgical complications, including deep neck phlegmon and descending necrotizing mediastinitis. The results of this study are aimed at improving the quality of multidisciplinary care and surgical management of patients with rapidly progressive lymphomas.

FUNDING AND CONFLICT OF INTEREST

This research received no external funding. The authors declare no conflict of interest. All authors have given their consent for publication of this manuscript.

COMPLIANCE WITH ETHICAL REQUIREMENTS

This study was conducted in accordance with the principles of the Declaration of Helsinki and the recommendations established by the Committee on Bioethics at the National Pirogov Memorial Medical University, Vinnytsia, Ukraine, which approved the study protocol (Protocol No. 7, May 18, 2023).

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REFERENCES

1. Barber, B. R., Dziegielewski, P. T., Biron, V. L., Ma, A., & Seikaly, H. (2014). Factors associated with severe deep neck space infections: Targeting multiple fronts. Journal of Otolaryngology – Head & Neck Surgery, 43(1), 35. https://doi.org/10.1186/s40463-014-0035-5
2. Kim, S. J., Yoon, D. H., Jaccard, A., et al. (2016). A prognostic index for natural killer cell lymphoma after non-anthracycline-based treatment: A multicenter, retrospective analysis. The Lancet Oncology, 17(3), 389-400. https://doi.org/10.1016/S1470-2045(15)00533-1
3. Kimura, A., Miyamoto, S., & Yamashita, T. (2020). Clinical predictors of descending necrotizing mediastinitis after deep neck infections. The Laryngoscope, 130(11), E567–E572. https://doi.org/10.1002/lary.28406
4. Lungu, M., Telehuz, A., Voinescu, D. C., et al. (2021). NK/T-cell non-Hodgkin lymphoma: Case report and review of the literature. Experimental and Therapeutic Medicine, 21(1), 91. https://doi.org/10.3892/etm.2020.9523
5. Swerdlow, S. H., Harris, N. L., Jaffe, E. S., et al. (2017). WHO classification of tumours of haematopoietic and lymphoid tissues (Revised 4th ed.). IARC.
6. Tanaka, H., Mori, E., Akutsu, T., Saito, S., Tei, M., & Otori, N. (2023). Characteristics of extranodal NK/T-cell lymphoma, nasal type, compared with nasal diffuse large B-cell lymphoma. Clinical Medicine Insights: Oncology, 17, 11795549231156692. https://doi.org/10.1177/11795549231156692
7. Yoon, D. H., Kim, S. J., Jeong, S. H., et al. (2016). Phase II trial of concurrent chemoradiotherapy with L-asparaginase and MIDLE chemotherapy for newly diagnosed stage I/II extranodal NK/T-cell lymphoma, nasal type (CISL-1008). Oncotarget, 7(51), 85584-85591. https://doi.org/10.18632/oncotarget.11319
8. Yun, J. S., Lee, C. H., Na, K. J., Song, S. Y., Oh, S. G., & Jeong, I. S. (2023). Surgical experience with descending necrotizing mediastinitis: A retrospective analysis at a single center. Journal of Chest Surgery, 56(1), 35-41. https://doi.org/10.5090/jcs.22.110

Резюме

ГОСТРИЙ НИЗХІДНИЙ МЕДІАСТИНІТ ЯК ФАТАЛЬНЕ УСКЛАДНЕННЯ NK/T-КЛІТИННОЇ ЛІМФОМИ: КЛІНІЧНИЙ ВИПАДОК

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Вступ. Екстранодальна NK/T-клітинна лімфома (ENKL) назального типу – це рідкісна та агресивна пухлина з несприятливим прогнозом. Захворювання часто проявляється атиповими симптомами, що призводять до затримки діагностики та лікування. Цей тип лімфоми характеризується швидким прогресуванням, ураженням носових ходів, верхнього шлунково-кишкового тракту та інших екстранодальних ділянок. Раннє виявлення та інтервенція є надзвичайно важливими, оскільки хвороба може спричиняти важкі ускладнення, зокрема гнійний низхідний медіастиніт, який представлений у цьому випадку.
Мета. Описати летальний випадок гострого низхідного медіастиніту, спричиненого ENKL, та висвітлити клінічні труднощі в його лікуванні.
Матеріали та методи. Чоловік, 36 років, був діагностований з ENKL після звернення зі скаргами на фарингіт та ларингіт. Початкова помилкова діагностика та затримка гістологічних досліджень призвели до затяжного перебігу хвороби понад рік. Було виконано хірургічне втручання, зокрема лівосторонній трансцервікальний доступ для дренування глибокої флегмони шиї та медіастинуму. Комп'ютерна томографія грудної клітки та шиї виявила поширеність хвороби та діагностувала медіастиніт.
Результати. Попри хірургічне лікування та активний менеджмент хвороби, стан пацієнта погіршився. У нього прогресувала гнійна ранова інфекція, зберігалася лихоманка та дихальна недостатність, що призвело до летальної кровотечі з лівої зовнішньої сонної артерії на 9-ту добу після операції.
Дискусія. ENKL має агресивний перебіг і несприятливий прогноз, часто помилково діагностується через схожість симптомів з іншими захворюваннями, такими як носоглоткова ангіофіброма. У цьому випадку затримка діагностики та участь вірусу Епштейна-Барр (EBV) сприяли швидкому погіршенню стану пацієнта. Рання біопсія та співпраця фахівців різних напрямків є критично важливими для точної діагностики та лікування. Вчасне розпізнавання хвороби та комбінована хіміопроменева терапія є ключовими для покращення прогнозу.
Висновки. Клінічний випадок ілюструє гострий низхідний медіастиніт як фатальне ускладнення екстранодальної NK/T-клітинної лімфоми назального типу. Швидке поширення некрозу в клітковинні простори шиї та середостіння підкреслює важливість ранньої біопсії, точної гістологічної діагностики та своєчасного хірургічного розкриття та дренування. Важкі ураження верхніх дихальних шляхів потребують ретельної оцінки, щоб уникнути затримки у діагностиці та попередити розвиток небезпечних для життя ускладнень.

Ключові слова: **медіастиніт, трансцервікальний доступ, неходжкінська лімфома, екстранодальна NK/ T-клітинна лімфома, трахеоезофагеальна нориця**

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