CASE REPORT

Non-healing pharyngeal ulcer in a middle-aged Caucasian man

Nwabundo Nwankwo, Raya Saba, Aibek E Mirrakhimov, Shesh Rao

Department of Internal Medicine, Saint Joseph Hospital, Chicago, Illinois, USA

Correspondence to
Dr Aibek E Mirrakhimov,
amirrakhimov1@gmail.com

SUMMARY

Extranodal natural killer NK/T-cell lymphoma is a rare malignancy. The disease is common in some Asian countries such as China, Japan as well as in South America. We present a case of extranodal NK/T-cell lymphoma which presented as a pharyngeal ulcer in a 35-year-old Caucasian man.

BACKGROUND

Extranodal natural killer NK/T-cell lymphoma is a rare malignancy. The disease is common in some Asian countries such as China, Japan as well as in South America. We present a case of extranodal NKT-cell lymphoma which presented as a pharyngeal ulcer in a 35-year-old Caucasian man.

CASE PRESENTATION

A 35-years-old Caucasian man presented to the hospital with a history of voice hoarseness for 2 years. His symptoms were previously investigated by his primary care physician and otolaryngologist. However, laryngoscopy was not performed at that time. The patient was told that his hoarseness is likely due to a viral upper respiratory infection, and he was advised to schedule an appointment in 1 month. However, the patient was not compliant and did not see his primary care physician in due time.

One month prior to encounter, the patient developed a non-healing painful ulcer on the posterior pharyngeal wall. Review of systems was positive for progressive fatigue, hoarseness of voice, throat pain and decreased appetite. On a physical examination, the patient was found to have an oral ulcer on the posterior pharyngeal wall measuring 1 cm in diameter. CT/positron emission tomography of the neck was performed, which showed right-sided palatine tonsil enlargement with increased uptake of F18 deoxyglucose (figure 1).

Laboratory work-up was significant for anaemia of chronic disease. The pharyngeal ulcer was biopsied (figure 2), and the diagnosis of extranodal NKT-cell lymphoma nasal type was made. The tumour was positive for CD2, CD56, Epstein-Barr virus (EBV) encoded small nuclear RNA (EBER) and a clonal rearrangement of γ receptor positive T lymphocytes. Bone marrow biopsy did not show the presence of malignant lymphocytes.

Radiation therapy and chemosensitisation with cisplatin followed by etoposide, ifosfamide and dexamethasone were started. The patient was discharged from the hospital and continued outpatient management with his oncologist.

Since the discharge, the patient was re-admitted several times to the hospital because of recurrent aspiration pneumonias. In November 2012, the patient was admitted to the hospital because of fever with chills, dyspnoea and productive cough. The patient had several episodes of desaturation and was transferred to the intensive care unit. CT of the neck with contrast showed subglottic narrowing of the trachea (figure 3). CT of the chest with contrast showed bilateral upper lobes infiltrates and right middle lobe infiltrate (figure 4). Bronchoscopy was performed which showed necrotic changes in tracheal cartilage, which was related to local irradiation and frequent intubations. Patient was started on intravenous levofloxacin, vancomycin and piperacillin–tazobactam.

DIFFERENTIAL DIAGNOSIS

Oral/pharyngeal ulcers

Squamous cell carcinoma, systemic lupus erythematosus, Behcet's disease, bullous pemphigoid, aphthous ulcers, herpes simplex infection coxsackievirus infection.

Voice hoarseness

Upper respiratory infection/laryngitis, polypoid corditis (in smokers because of irritation of true vocal cords), vocal cords polyp/nodule laryngeal cancer. Voice hoarseness lasting for at least 2 weeks should be investigated by otolaryngologist.

DISCUSSION

Extranodal NKT-cell lymphoma nasal type is a rare lymphocyte neoplasm of NK and/or T-lymphocyte lineage. This disease is more common in individuals of Asian descent. Mean age of onset ranges from 50 to 60 years.¹ The disease is strongly linked to EBV infection, and mutated cells contain EBER, which is a marker of EBV infection. The potential causative role of EBV infection is particularly intriguing since it is known that EBV infects lymphocytes through interaction with the CD21 receptor, which is normally presented on B lymphocytes.² The possible explanation for this is that NK and T lymphocytes may acquire CD21 from the B lymphocytes during cellular interaction, thus making them vulnerable for EBV infection.³

Clinically, the disease can manifest with the symptoms of upper airway involvement such as nasal stuffiness, oral ulcers and nasal bleeding. On rare occasions, the disease may manifest with the involvement of distant sites, such as testis and central nervous system.^{4 5} The disease portends a poor prognosis, and if not treated, the usual survival is typically several months.¹

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Figure 1 CT of the pharynx showing right palatine enlargement (arrow). Positron emission tomography CT scan showing increased uptake of F18 deoxyglucose.

Several aspects of our patient's demographics are notably different from that of a typical patient with NKT-cell lymphoma. First, the patient presented was of Caucasian descent, who generally have a low incidence of this malignancy for unclear reasons.¹ One explanation for that may be a greater prevalence of EBV infection in Asia and South America. Second, age of the presentation was around 33–35 years, which is much earlier than a mean age from 50 to 60 years at diagnosis.¹ However, this case reminds us that there is always a possibility of an exception in clinical medicine.

Therefore, it is necessary to keep a clinical suspicion and have a low threshold for evaluation of patients with chronic non-resolving upper airway and oral symptoms such as hoarseness and non-healing oral ulcers. The aetiology of non-resolving oral/pharyngeal ulcerations is broad and ranges from relatively benign causes such as aphthous ulcers and Lichen planus to malignancies. Individuals with hoarseness lasting for 2 weeks should be referred to otolaryngologist for a further evaluation. Biopsy of the lesion should be considered to establish a definitive diagnosis.

Figure 2 Large atypical lymphocytes with irregularly folded nuclei.

Figure 3 CT of the neck showing subglottic tracheal wall thickening and narrowing.

Learning points

- ▶ Extranodal natural killer T-cell lymphoma nasal type is a rare malignancy most commonly seen in Asia and South America.
- ▶ Disease is strongly linked to infection with Epstein-Barr virus.
- ▶ Chronic non-healing oral ulcers should be worked up with a biopsy to exclude a malignancy.

Figure 4 Bilateral pulmonary infiltrates.

Contributors Each author contributed equally to this manuscript.

Competing interests None.

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