

Case Report

Unusual presentation of paediatric anaplastic large cell lymphoma

M Irfan¹, M M Yusri², A B M Farveen³

Abstract:

Anaplastic large cell lymphoma (ALCL) is a rare disease. Among childhood non-Hodgkin lymphoma, it constitutes less than 20% of the incidence of all cases. The clinical presentation though is known to be much diversified, most of the patients will present with an enlarged palpable cervical lymphadenopathy. Other reported features include fever of unknown origin, nonspecific pain, cough, shortness of breath, fatigue and malaise. We report a case of ALK-positive ALCL in a patient who presented with submandibular abscess. After defaulted treatment, the mass became fungating externally with everted edge that mimic squamous cell carcinoma.

Keywords: ALCL, pediatric, clinical presentation

Case Summary

A 10-year-old Malay girl presented with history of right submandibular swelling of 3 months duration. It began with a small painless mass which progressively increased with time. Initially she was afebrile but subsequently developed recurrent fever with increasing intensity of pain. Examination revealed a fluctuant mass at right submandibular region. The diagnosis of submandibular abscess was made. Incision and drainage

was planned. However, she and the guardian refused the planned treatment and requested for at own risk discharge.

One month prior to second hospital admission, the swelling break into the skin. The surface became ulcerated. She sought treatment from a general practitioner and every other day dressing was applied. This problem had caused her absence from school for one month.

^{1,2}Department of Otorhinolaryngology-Head & Neck surgery, ³Department of Pathology, School of Medical Sciences, Universiti Sains Malaysia

Corresponds to:

Dr. Irfan Mohamad, Department of Otorhinolaryngology-Head & Neck Surgery
School of Medical Sciences, Universiti Sains Malaysia, 16150 Kota Bharu
Kelantan, Malaysia. Email : irfan@kb.usm.my

She was again brought to our attention after a referral by the treating family doctor. At that she was found to be with tachypnoea and tachycardia. Clinically she was anaemic and lethargic. Our assessment revealed her to be in hypovolaemic condition. Her haemoglobin level was 5 g/dl and the heart rate consistently above 120 beats/minute. Specific examination of the neck revealed a fungating right sided neck mass with rolled edges. The mass measured about 4x4 cm in diameter with ulceration and was covered by fibrinopurulent exudates (Fig.1). Contact bleeding was present.

Stabilization of the general condition was achieved after packed cell transfusion and intravenous hydration. She was planned to under go urgent computed tomography (CT) scan and examination under anaesthesia with incisional biopsy.

CT scan revealed multiple cervical lymphadenopathies involving all levels (Fig. 2). Axillary, mediastinal, paraortic and inguinal lymph nodes were enlarged. Bilateral massive pleural effusions were noted as well as collapsed consolidation of the lung lobe. Massive hepatomegaly was also present.

Examination under anaesthesia was performed. A nasopharyngeal mass with ulcerated surface was noted and biopsied. Incisional biopsy of the fungating neck mass was also performed. Histopathological examination concluded the mass to be an anaplastic large cell lymphoma, ALK-positive (Fig. 3). Referral to hematological team was made and chemotherapy was planned for. Unfortunately, the patient died later on because of sepsis.

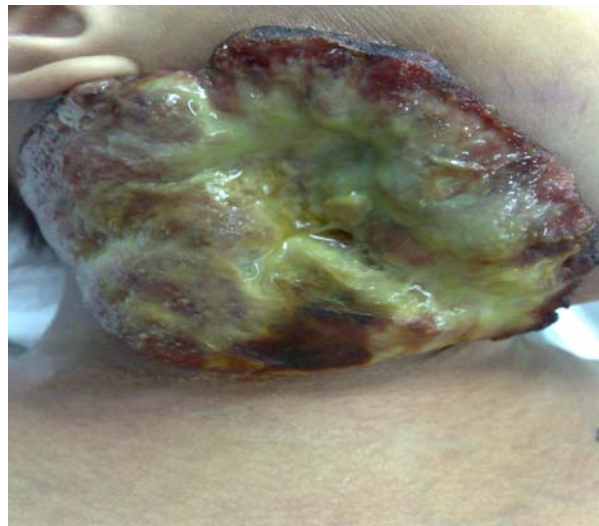


Figure.1: The mass with everted edge and ulcerated surface.

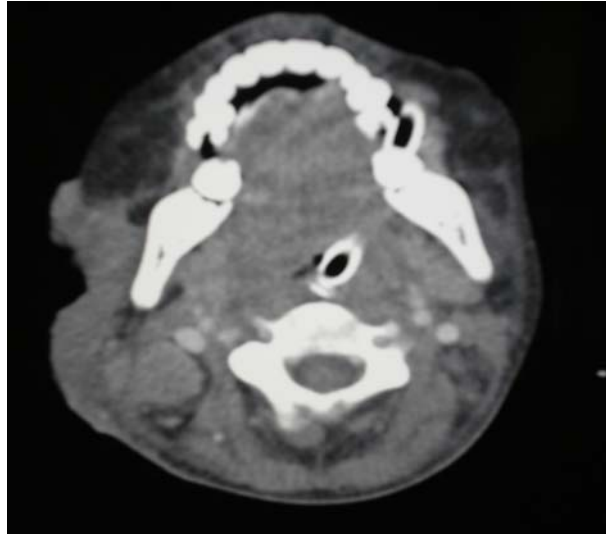


Figure. 2: Fungating mass burst out to the skin with underlying multiple cervical lymphadenopathies.

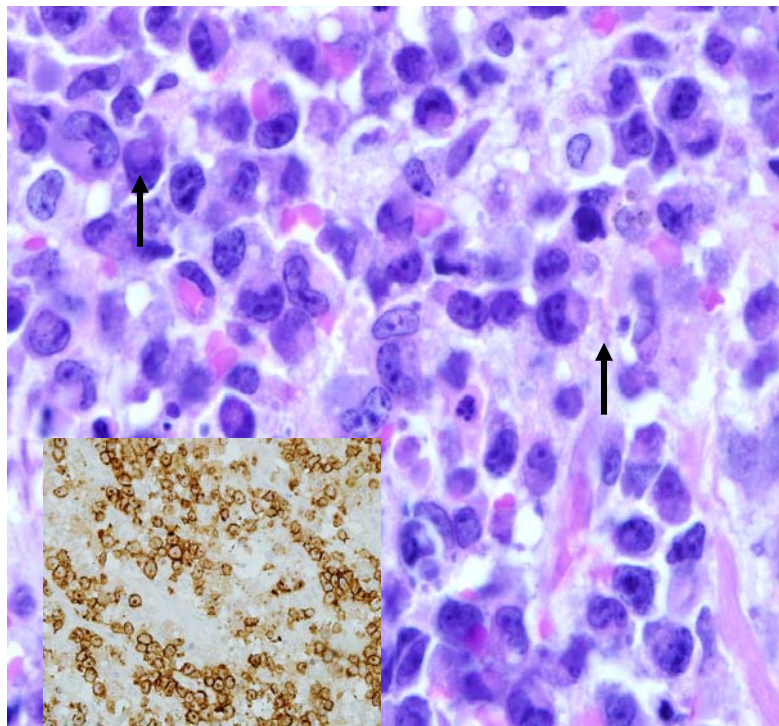


Figure. 3: Histopathological examination confirmed the diagnosis of ALK-positive ALCL. Some of the characteristic cells are seen, including *hallmark* cells (arrows), with reniform nuclei. Inset: positive ALK protein expression.

Discussion

Anaplastic large-cell lymphoma (ALCL) is a rare disease, accounting for less than 5% of all cases of non-Hodgkin's lymphoma¹. In childhood, ALCL constitutes 10-15% of non-Hodgkin lymphoma² ALCL is found to show heterogeneity in its clinical presentation³.

Although an enlarged and easily accessible peripheral lymph node is considered a classic diagnosis, a fluctuant neck lymphadenopathy which is diagnostic hallmark of an abscess is a rare presentation of ALCL. There was possibility in our patient that the diseased node became infected and lead to an abscess formation. However, early diagnosis could not be made because of the refusal of planned treatment in the first place and the patient defaulted from follow up for a few months.

Among 23 cases of ALCL studied by Mosunjac et al in 2008, 5 of them presented with unusual systemic features. Apart from fever of unknown origin, there were nonspecific pain, cough, and shortness of breath, asthma-

like symptoms, fatigue, malaise, night sweats, waxing and waning skin rash with recurrent epistaxis⁴.

She came to our attention again with an ulcerative lesion with everted margins. This was another rare presentation of a lymphoma. Clinically it looked like a metastatic squamous cell carcinoma in the neck with skin involvement. As the mass already breached the skin, there was no role of needle aspiration as compared to the superiority of incision biopsy.

Histologically, ALK-positive ALCL shows a broad morphologic spectrum. As in this case, the morphologic findings were the usual ones. Histopathological examination revealed a variable proportion of cells with eccentric, horseshoe- or kidney-shaped nuclei (hallmark cell) as well as doughnut cells and multinucleated ones. Brisk mitotic figures and apoptotic bodies were observed. Immunohistochemically, these cells are positive for CD3, CD30 and ALK protein, thus confirmed the diagnosis of ALK- positive ALCL.

References

1. Kadin ME, Moris SW. The t (2:5) in human lymphomas. *Leuk Lymphoma* 1998; **29**(3-4): 249-256.
2. Burkhardt T, Zimmermann M, Oshlies I et al. The impact of age and gender on biology, clinical features and outcome of non-Hodgkin lymphoma in childhood and adolescence. *Br J Hematol* 2005; **131**(1): 39-49.
3. Fotopoulos TN, Shenefelt PD, Messina J. Anaplastic large cell lymphoma mimicking noduloulcerative tertiary syphilis. *Cutis* 1997; **60**(4): 211-4.
4. Mosunjac MB, Sundstrom JB, Mosunjac MI. Unusual presentation of anaplastic large cell lymphoma with clinical course mimicking fever of unknown origin and sepsis: autopsy study of five cases. *Croat Med J* 2008; **49**(5): 660-8.