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Case Report

Primary cutaneous anaplastic large cell lymphoma with lung metastasis

Nor Kamaruzaman Esa [a]*, Kahairi Abdullah[b], Shamim Rahman Abdul Rasheed[b]

[a] Department of Otolaryngology-Head & Neck Surgery, Faculty of Medicine, Universiti Sultan Zainal Abidin, Jalan Sultan Mahmud, 20400 Kuala Terengganu, Terengganu, Malaysia.
[b] Department of Otolaryngology-Head & Neck Surgery, Kulliyyah of Medicine, International Islamic University Malaysia, Jalan Hospital, 25100 Kuantan, Pahang, Malaysia.

* Corresponding author: liqa24@gmail.com

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Abstract Primary cutaneous anaplastic large cell lymphoma (PC-ALCL) is relatively a rare tumour of head and neck region. Without histopathological confirmation, it appears and behaved as like malignant epithelial tumor of the head and neck region as will be illustrated in the present case. Cutaneous involvement of the tumor with distant metastasis has made the prognosis unfavorable. This isolated case of head and neck PC-ALCL was highlighted as it has a tendency to behave aggressively and early diagnosis and treatment is crucial to avoid poor survival outcome.

Keywords: head and neck, lung metastasis, primary cutaneous anaplastic large cell lymphoma.

Introduction

Primary cutaneous anaplastic large cell lymphoma (PC-ALCL) is relatively uncommon in head and neck region as compared to malignant epithelial and salivary gland tumour. It is an anaplastic variation of T-cell lymphoma and currently, three types are recognized. They are anaplastic lymphoma kinase (ALK) positive ALCL, ALK negative ALCL and PC-ALCL (Stein *et al.,* 1985). PC-ALCL is predominantly a disorder of adults, with a median age at diagnosis of approximately 50 to 60 years. Most patients are older than 50 years of age.

Case report

A 58-year-old man presented with large ulcerative lesion at the left side of the neck for one month. It started as a small nodular swelling at the pre auricular region about seven month prior to the visit. According to the patient, an incision and drainage was done with an impression of an abscess. He denied of any procedures that have been done to get a sample for biopsy from the swelling before or during the incision and drainage. Following that, the lesion did not heal, instead it progressed to ulceration and eventually enlarged. There was no history of trauma, epistaxis, any voice change and dysphagia. Examination revealed a large ulcerative mass, which covered most of the left side of the neck area (Fig. 1). Left facial nerve function was intact. Cervical lymphadenopathy was not detected by palpation on both sides of the neck. Oral cavity, nasal and ear examination did not reveal any abnormality.

Tissue biopsy was taken under local anaesthesia from the centre and margin of the lesion. Computed tomography (CT) scans showed a large lobulated soft tissue density mass, which was measuring 12 cm x 6 cm in diameter and demonstrated contrast enhancement. Deeper tissue involvement was not apparent because there was an evidence of fat plane separating the lesion from the deeper tissues and it appeared as all the tissues were pushed medially (Fig. 2). There were

multiple nodular densities scattered throughout both lung fields which highly suggestive of lung metastasis (Fig. 3).

Under microscopic examination, the malignant cells were medium-to-large lymphoid cells having vesicular nuclei with irregular nuclear membrane, indented, kidney-shaped or multinucleated. Tumour cells strongly expressed CD30 (Fig 4) and CD45 RO. Cytokeratin was negative. The final diagnosis was primary cutaneous anaplastic large cell lymphoma (PC-ALCL) based on primary skin involvement with strong expression of CD30. While waiting for further medical treatment, the patient developed sepsis and breathing difficulty on sixth day of admission. His conditions continued to deteriorate and finally, he passed away.

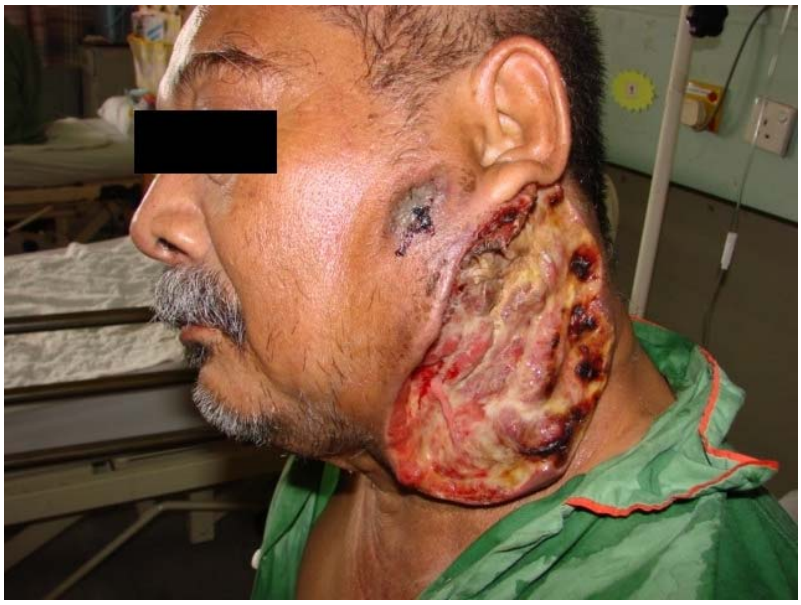

Fig. 1 Large ulcerative lesion over left side of the neck with some areas covered with necrotic tissues and suppuration.

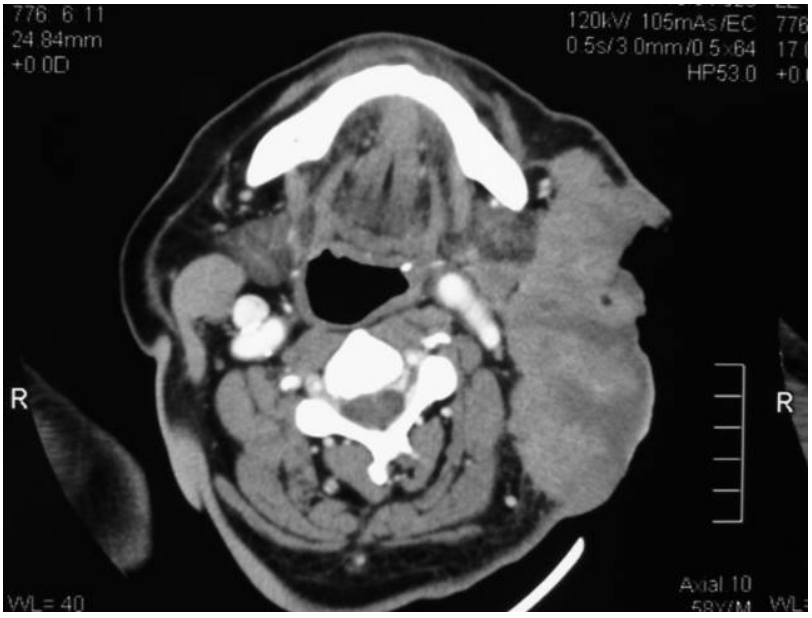

Fig. 2 An axial view of a CT scan of the neck revealed deeper extension of the mass.

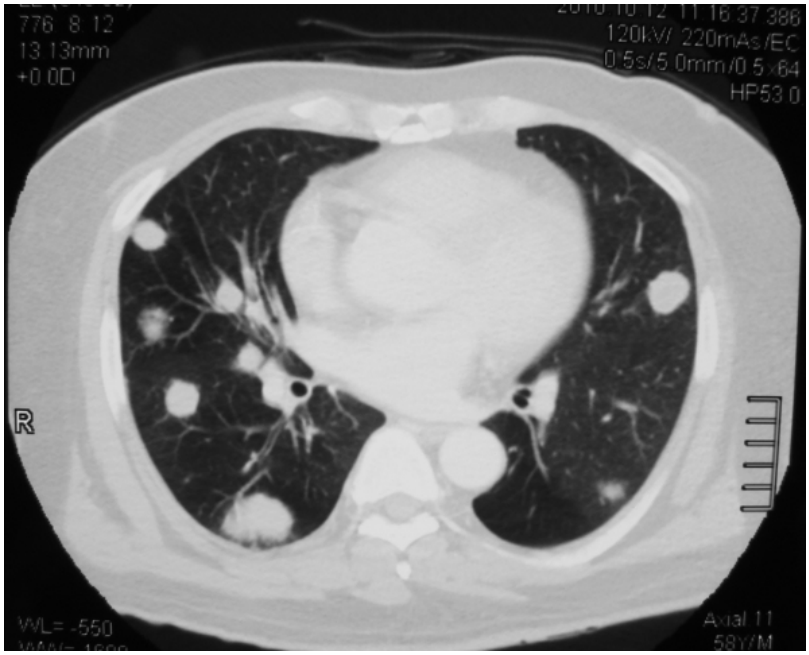

Fig. 3 An axial view of a CT scan of thorax showed evidence of multiple lung metastases on both lung fields.

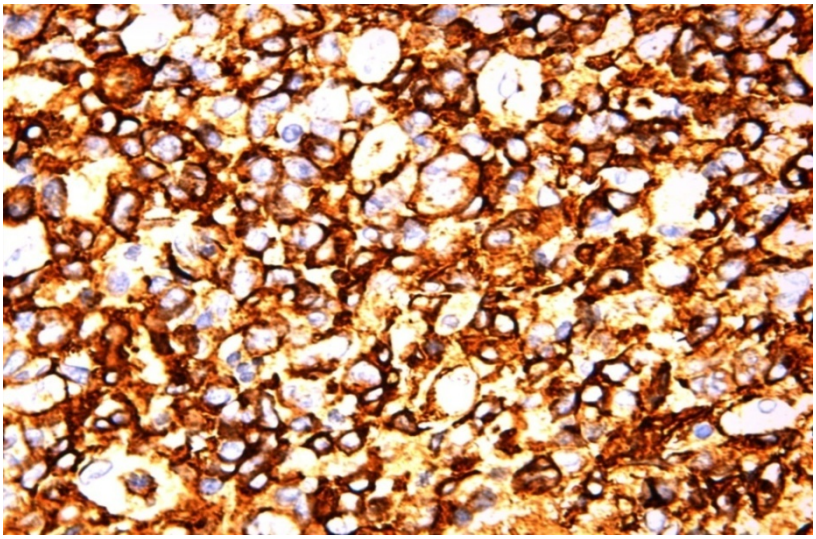

Fig. 4 Tumor cells strongly expressed CD 30 (H & E stain, original magnification x 600).

Discussion

ALCL was initially identified as unusual large cell lymphoma (Stein *et al*., 1985). It is a rare type of T-cell lymphoma and accounts about 1-2% of all cases of non-Hodgkin lymphoma (NHL). PC-ALCL is defined by skin-only involvement without systemic dissemination at presentation. It is the second most common skin T-cell lymphoma after mycosis fungoides (Bradford *et al.*, 2009).

It commonly presents as a solitary nodule that is often ulcerate and usually occur on the extremities, face, trunk and occurrence in the head and neck area has been reported as well. Extracutaneous involvement is observed in approximately 10 percent of cases and usually manifest as regional lymphadenopathy (Willemze *et al*., 2005).

By looking at its appearance and behaviour in the present case along with CT evidence of lung metastasis, squamous cell carcinoma, which was synonym in head and neck malignancy, should be ruled out first. Open biopsy is not recommended in suspected squamous cell carcinoma because the risk of seeding. In biopsy there is a possibility of dislodging and spreading neoplastic cells to surrounding site (Shyamala *et al.*, 2014) and was reported up to 20% risk of cancer cell dissemination (Kusukawa *et al*., 2000). Fine needle aspiration cytology (FNAC) is more preferred method of biopsy. Sensitivity and specificity of FNAC in the masses of the head and neck have been reported as 97 and 96% (Peters *et al.*, 1989). However in selected conditions that may need bigger sample, non-diagnostic FNAC or in lymphoma, open biopsy are justified to be performed. In the present case, histopathological examination had confirmed the diagnosis of ALCL and the case was referred to the haematologist for subsequent management. The ulceration of the lesion in the present case could have been contributed by the incision and drainage procedure which was done at the primary care setting and this most probably had expedited the growth of the tumour to such extent.

Lung involvement, is a rare extracutaneous manifestation in PC-ALCL (Rush *et al.,* 2000), of which nodal is more common. Not many cases were reported with regards to metastatic lesion in PC-ALCL. However in systemic ALCL the frequency of extranodular involvement is higher. Skin involvement about 21%, bone (17%), soft tissues (17%), bone marrow (11%), lung (11%), liver (8%) (Falini *et al*., 1999) and few cases of central nervous system involvement were also reported. Histologically, the tumour composed of large anaplastic cells which presented as a round, oval or irregular shaped nuclei and abundant cytoplasm. Classically, PC-ALCL has strong Golgi and membrane expression of CD 30 in almost every cell (Kinney *et al.*, 2011). As shown in the present case, it also showed a strong expression of CD 45 RO.

The standard treatment for PC-ALCL without extracutaneous involvement is surgical excision or radiation therapy. In the present case, if it was just based on CT appearance of the primary neck lesion whereby a fat plane was still visible, wide surgical excision with reasonable margin followed by vascularised free tissue transfer is possible. Nevertheless, multiple metastatic lung lesions as evidenced from the CT scan with TNM stage T1bN0M1 (Benner *et al*., 2009) have made the prognosis not favourable (Chadburn *et al.*, 1993). But in cases which resectable ALCL of the lung surgery was performed, they showed better survival rate (Rush *et al*., 2000).

In cases with systemic involvement, systemic chemotherapy with low-dose methotrexate or doxorubicin could be considered. Evaluation for prognosis in primary cutaneous of lymphoma was usually met with some difficulties. Primary cutaneous disease, spontaneous regression, absence of extracutaneous involvement, and younger age at onset (<60 years of age) has been suggested to be associated with better prognosis. Although PC-ALCL has a favourable prognosis with more than 90% 10-year disease-related survival (Bekkenk *et al.*, 2000), extensive involvement and

localization on the head and neck have been associated with a less favourable prognosis (Yu *et al*., 2008), as has been depicted in the present case. However, some study showed disease-specific survival (DSS) at 5 years was 95% for lesion in the head and neck region, while other patients with extensive limb involvement especially leg had poorer survival rate of 76% as compared without leg involvement at 96% (Benner *et al.*, 2009).

Conclusion

PC-ALCL is a rare head and neck tumour. It may behave aggressively and masquerading as malignant epithelial tumour. Open biopsy is contraindicated in an unknown head and neck lesions, especially with intact skin. Localization in the head and neck with lung metastasis is associated with poor prognosis. Nonetheless, early diagnosis and multidisciplinary approach for an early treatment are essential to improve survival.

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