

Extranodal Lymphoma of the face mimicking other pathology

J Scannell, J Combes, N Hyde

Citation

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Abstract

The literature was reviewed on a rare pathological condition that can mimic squamous cell carcinoma, infection and salivary gland pathology. We illustrate 3 cases of presentation of facial extranodal lymphoma which caused diagnostic difficulty. Diffuse large B cell lymphoma (DLBCL) worldwide, is the largest subtype of non-Hodgkin's lymphoma. DLBCL is characterised by relatively frequent extranodal presentation of stomach, CNS, bone testis and liver but rare in the orofacial region. The cure rate is 40-50% with CHOP (cyclophosphamide, adriamycin, vincristine, & prednisolone). Facial peripheral T-Cell lymphoma (PTCL) is a type of non-Hodgkin's lymphoma. The prognosis is poor with a 5 year overall survival of approximately 15-30%.

INTRODUCTION

Non Hodgkin's lymphoma can have extranodal presentation in approximately 25% of cases, unlike Hodgkin's lymphoma which rarely involves extranodal sites. Extranodal lymphoma in the head & neck region is extremely rare.

Diffuse large B cell lymphoma (Figure 1), worldwide is the largest subtype of non-Hodgkin's lymphoma characterised by relatively frequent extranodal presentation e.g. stomach, CNS, bone, testis and liver but rare in the oro-facial region. It has a bimodal age distribution affecting children under the age of 12 and adults above the age of 40 years. The three year disease survival rate for this type of lymphoma is 90% and 47% for stage 1 and 11 respectively. Combination chemotherapy and radiotherapy has a better complete response rate and relapse free survival rate than radiotherapy alone.

Facial peripheral T-Cell lymphoma (PTCL) is a type of non-Hodgkin's lymphoma, rarely encountered with limited literature. In 20 years there have only been nine reported nasal and paranasal cases in Scotland³, and eight buccal and gingivae submucosal cases in Japan². HTLV-1 induced adult T cell leukaemia/lymphoma primarily occurs in south west Japan.

PTCL (Figure 2) is characterised by infiltration or swelling mimicking inflammation or infection and is a recognised cause of midline granuloma syndrome³. Diagnosis of PTCL can be very difficult and excisional biopsy is strongly

recommended over core needle biopsies to allow immunohistochemistry and often molecular studies (including T-cell receptor gene rearrangements) to be performed. It has also been suggested that Positron Emission Tomography (PET) should be used in staging as well as Computed Tomography (CT) because CT alone has proven to be inadequate in assessing areas such as subcutaneous tissue, sinus and bowel.

Figure 1

Figure 1: Diffuse large B cell NHL: uniform, round nuclei with vesicular chromatin. Tumour cells are +ve for CD20

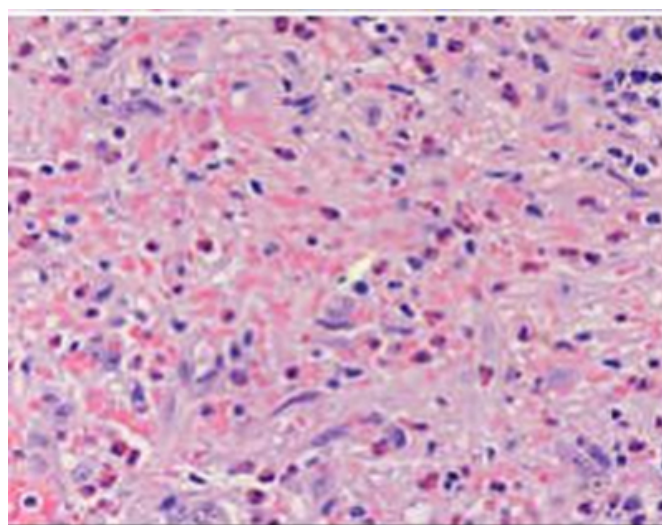
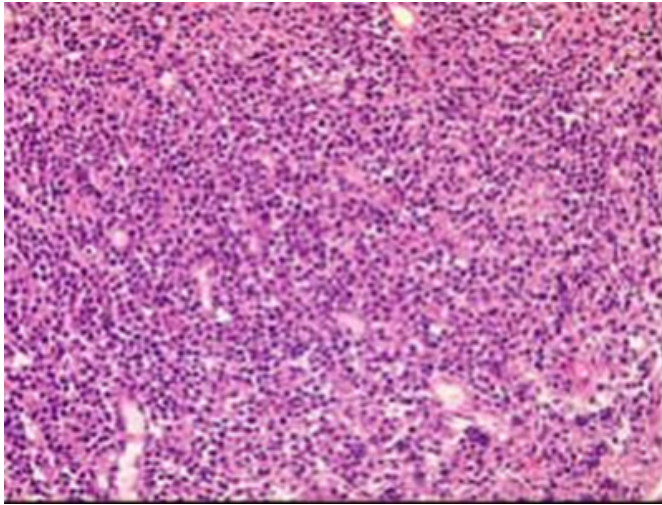


Figure 2

Figure 2: PTC: atypical lymphoid cells with scattered eosinophils & epithelioid cells in the background.



It has a poor prognosis with a 5 year overall survival rate of approximately 15-30%. Early local relapse is common and radiotherapy is disappointing. There is no widely accepted standard therapy⁴.

We report 3 cases of extranodal lymphoma of the face which caused diagnostic difficulty.

All cases were staged 1a disease and treated with radiotherapy and combination chemotherapy, CHOP (cyclophosphamide, adriamycin, vincristine, & prednisolone). All cases had a good response.

CASES

Case 1: 76 year old female of Chinese origin presented with a 4 month history of slowly enlarging painful left maxillary swelling (Figure 3). The patient was originally seen by her dentist who treated her with antibiotics. Orthopantomogram radiograph showed an anterior maxillary radiolucency consistent with an infected residual dental cyst.

Antibiotics were ineffective, so CT scan was carried out which revealed thickening of the maxillary sinus and a large right maxillary swelling with bony erosion (Figure 4). The intra-oral swelling became firmer and a new clinical diagnosis of salivary gland malignancy was made. However, intra-oral biopsy revealed a diffuse B cell lymphoma (DLBCL) according to the WHO classification⁵.

Staging CT showed pretracheal and retro-carinal nodes and enlargement of the thyroid gland with a focal lesion. The patient was treated with 3 cycles of CHOP which was complicated by an episode of neutropaenic sepsis but this led

to complete resolution of the facial swelling (Figure 5).

There was no evidence of recurrence at 8 months follow up.

Figure 3

Figure 3: clinical extraoral & intraoral appearance at presentation with left facial swelling



Figure 4

Figure 4: CT showing large anterior maxillary swelling with bony erosion

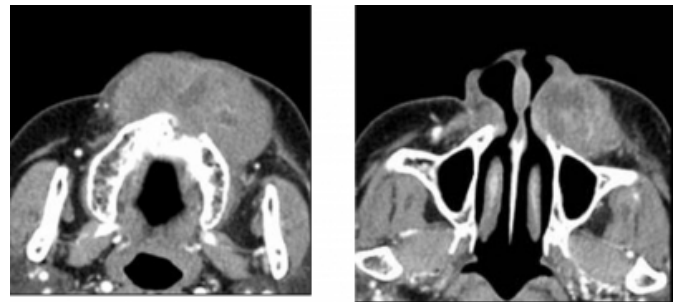


Figure 5

Figure 5: clinical appearance after treatment with 3 cycles of CHOP



Case 2: 66 year old male, retired engineer presented with a 5 week history of ulceration of right cheek and 1 stone weight loss; no other B symptoms were present. The patient was a lifelong heavy smoker with minimal alcohol intake and no significant past medical history. On examination he had a deep, indurated ulcer of the buccal mucosa, resembling Squamous cell carcinoma and a level 1 ipsilateral cervical lymph node. He was clinically staged as T3N1 SCC.

CT scan of facial bones revealed extensive soft tissue involvement of the right cheek and upper buccal mucosa extending to the mandible and tongue base. The patient was diagnosed with stage 1a diffuse large B cell non-Hodgkin's lymphoma of the right buccal mucosa and tongue base. The bone marrow aspirate was negative. He was treated with consolidation radical radiotherapy following CHOP. Both the intra-oral mass and cervical node regressed completely after 4 months of treatment. At 4 year follow up, the patient still remains disease free, at the time of writing this paper.

Case 3: A 57 year old female mental health worker presented with a 2 week history of lower lip ulcer (Figure 6). The patient was a heavy lifelong smoker but had no excessive sunlight exposure. A provisional clinical diagnosis of T1N0 squamous cell carcinoma (SCC) of the lower lip was made. However a biopsy revealed T-cell Lymphoma. The bone marrow aspirate was normal and no other lesions were seen on Staging CT scan. She was diagnosed with stage 1a T-cell lymphoma. Treatment consisted of 4 cycles of CHOP and 30 Gray of involved field radiotherapy. This led to immediate resolution. The patient remained disease free at 2 year follow up.

Figure 6

Figure 6: clinical appearance of lower lip ulcer at presentation



DISCUSSION

The cases have illustrated how the diagnosis of extranodal lymphoma in the head and neck region can be very difficult as they can mimic more common pathology such as infection and squamous cell carcinoma.

The literature has shown that diagnoses can often be delayed and difficult. Although PTCL is very uncommon, a higher ratio of T cell to B cell lymphoma occurs among extranodal head and neck non-Hodgkin's lymphoma in comparison to nodal non-Hodgkin's lymphoma. Extranodal PTCL in comparison to nodal PTCL in the head and neck region has a worse prognosis irrespective of clinical stage².

CONCLUSION

Extranodal non-Hodgkin's lymphoma are distinct from nodal non-Hodgkin's lymphoma in the head and neck region and consist of diffuse proliferation and relatively higher grade malignancy that may result in a poorer prognosis. It is important to be aware of extranodal lymphoma as a cause of facial swelling, oral malignant ulcer, or salivary neoplasm.

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Author Information

Jolie Scannell

Department of Oral & Maxillofacial surgery, St George's Hospital, London

Jamie Combes

Department of Oral & Maxillofacial surgery, St George's Hospital, London

Nick Hyde

Department of Oral & Maxillofacial surgery, St George's Hospital, London