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Primary cutaneous B cell lymphoma--leg type (NEW EORTC--WHO classification), with nasal sinuses involvement

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peritoneal contamination is likely to have contributed to a failure to achieve optimal cytoreduction in the case presented.

The role of adjuvant therapy in the management of ovarian squamous cell carcinomas is similarly unclear. Limited experience suggests that surgical management with close follow up alone would suffice for early stage 1A disease. Stages 1C and above have been targeted with a variety of adjuvant regimens with variable outcomes. Relatively poor disease control has been noted with the conventional regimens to treat epithelial ovarian cancers, i.e. cisplatin, vincristine, mitomycin C, and bleomycin (POMB). On the other hand remarkable responses have been achieved with paclitaxel based regimens in some cases.

Radiotherapy has also been used with the rationale of squamous cell carcinoma being a radiosensitive tumour; and has brought forth variable results for cancers within antecedent dermoids. In the latest case series and review of literature, albeit in cases of squamous cell cancers arising in mature cystic teratoma of the ovary, whole-pelvis radiation and concurrent weekly platinum-based chemotherapy following aggressive cytoreduction has been shown to be of benefit. It however remains uncertain whether patients with de novo primary squamous cell cancer of the ovary would benefit as much from similar adjuvant therapy. To conclude primary squamous cell cancers arising in the ovary is an extremely rare entity, and has not previously been described in patients of Southeast Asian heritage. In the absence of well designed trials, and based on the existing albeit sparse data, the primary management approach at present is surgical debulking, akin to the principles underlying the management of ovarian adenocarcinoma. Combination chemotherapy with newer drugs has shown some benefit and pelvic radiotherapy may have a role to play for local control. However, in the absence of quality data, except for the very early stages of presentation, a role for adjuvant therapy is at present unclear.

References

Case Report

Primary Cutaneous B Cell Lymphoma- Leg Type
(NEW EORTC - WHO Classification), with nasal sinuses involvement
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Abstract
Primary Cutaneous lymphomas of B cell origin are rare, there remains a controversy in truly classifying these lymphomas and an updated EORTC classification divides them on the basis of their distinct histopathological grounds rather than on the basis of their anatomic location as in WHO classification, while the new WHO- EORTC joint classification maintains some characteristics of both systems, We report an elderly gentleman who primarily had a typical Leg dominant Cutaneous lymphoma of B cell origin uniquely with involvement of nasal Sinuses , bearing the Immunohistochemical staining features of " Cutaneous lymphoma - Leg Type" befitting the new joint WHO-EORTC classification of Cutaneous B cell Lymphoma.

Introduction
Cutaneous lymphomas are uncommon skin tumours, with skin being either the primary site for the origin of the tumour or more often the secondary site. Approximately...
65% of the cutaneous lymphomas are T cell in origin and only 20-25% are thought to originate from the B cell, with majority of these being diffuse large B cell type. Most of the primary cutaneous lymphomas have an indolent behaviour.

Primary Cutaneous B-cell lymphoma (PCBCL) as an entity was introduced in 1990s to characterize a group of lymphoproliferative disorders characterized by clonal proliferation of B lymphocytes primarily involving the skin. According to the Revised European and American Classification of Lymphoid Neoplasms (REAL) / World Health Organization (WHO) classification the Non Hodgkin's lymphomas, lymphomas which have large cell morphology and diffuse growth pattern are designated as diffuse large B cell lymphoma (DLBCL). The European Organization for Research and Treatment of Cancer (EORTC) divided primary cutaneous B-cell lymphomas (PCBL) in four groups, one of them being The Primary cutaneous diffuse large B-cell lymphoma, 'leg type'; They are a heterogeneous group of lymphomas that primarily involve the skin but may have variable clinical, histopathologic, and immunologic phenotypes.

We are reporting a case of an elderly patient, with Primary Cutaneous Large B cell lymphoma (PCLBCL) rather unique in having primarily the involvement of both lower limbs along with simultaneous involvement of paranasal sinuses extending to involve the inferior orbital area.

Case Report

A 70 year old gentleman, presented in April 2007 with a history of painless skin nodules that developed over two months. Initially these nodules were small and on the shin of left leg but gradually they progressed to involve the right leg, right arm and left hand and right side of nose. The
skin covering these nodules first became black followed by development of blisters with serous discharge which finally became thick and crusted (Figure 1).

On examination there were bilateral blistering lesion at legs and on the dorsum of the right foot, crusted with mild serous discharge on bilateral shins, skin nodules on right side of nose, right and left forearm and swollen left middle finger. Rest of the systemic examination was unremarkable, his overall ECOG performance status was 2.

Laboratory investigations at the time of presentation were: Blood Urea Nitrogen: 19mg/dl, serum Creatinine: 0.9mg/dl, total Bilirubin: 1.0 mg/dl, Gamma Glutamine transferase : 11 IU/l, Alanine Aminotransferase: 9 IU/l, Alkaline Phosphatase: 78 IU/l, serum Albumin: 2.7 mg/dl, serum Calcium: 7.6 mg/dl, serum Lactic Dehydrogenase:1373 IU/l. Complete blood counts were Hb: 9.0 gm/dl, WBC: 7.1 x 10^9/l and Platelets: 262x 10^9/dl. Biopsy specimen of leg lesions gave the diagnosis of diffuse large B cell lymphoma, with the immunohistochemical stains being positive for LCA, CD20, CD79, Bcl-2, Bcl-6, MiB 1, MUM-1 and characteristically negative for CD3 (Figure 2), (Figure 3).

CT scans of paranasal sinuses, chest and abdomen showed an enhancing soft tissue mass involving nasal cavity, paranasal sinuses, nasopharynx on right side with orbital extension and abdominal lymphadenopathy.

Discussion

Primary B cell Lymphoma of the leg, which was identified as a distinct entity in the EORTC classification, has now been incorporated in the new WHO-EORTC classification of cutaneous B cell Lymphomas as 'Primary cutaneous large B cell lymphoma, Leg Type', ending the longstanding controversy of placement and categorization of the primary cutaneous B cell Lymphomas. The sub classification of PCBCL is now made on morphological features rather than anatomic site of presence and this is important to remember, as the type in question is apt to express Bcl-2, MUM-1, however and interestingly expression of the said immunophenotypic markers was not associated with worse prognosis, as opposed to other types of Cutaneous B cell lymphomas namely follicular Cutaneous B cell lymphoma (PCFCLs).  

There however is a growing consensus that the primary cutaneous DLBCL of leg is distinct in its behaviour as opposed to the DLBCL occurring at the other sites in that these patients are of older age group, more frequently females, have a short duration of skin lesion before diagnosis.

In a European multi centre study of 145 patients with primary cutaneous DLBCL, round-cell morphology, location on the leg, and multiple skin lesions at diagnosis were found to be independent adverse prognostic factors and these neoplasms are highly sensitive to radiotherapy and hence, it is the treatment of choice for localized disease at presentation or relapse. Combination chemotherapy with Cyclophosphomide, vincristine, Adriamycin and prednisone (CHOP), with or without addition of Rituximab, is preferred in patients with the tumour involving the leg, multiple skin lesions or with systemic involvement.

Our case is interesting in being a rare, histopathologically fitting the new WHO-EORTC classification of 'Primary Cutaneous diffuse large B-cell lymphoma, leg type' and with clinical involvement of Para nasal sinuses, as for primary Cutaneous lymphomas it's very unusual to involve other organ sites. The patient was managed in line of the recommended guidelines and he responded well to the treatment. This case reminds the practicing physicians of diversity with which lymphoma can present.

References