East African Medical Journal Vol. 91 No. 2 February 2014
CUTANEOUS INVOLVEMENT OF HODGKIN LYMPHOMA IN A CHILD – CASE REPORT
L. A. Renner, MBChB, FRCPCH, Department of Child Health, University of Ghana Medical School, N. Salifu, MBChB, MWACP, Department of Child Health, L. Edusei, MBChB, FWACP, Department of Pathology, Korle Bu Teaching Hospital, C. Segbefia, MBChB, MWACP, Department of Child Health, I. Ekem, MBChB, FWACP, Department of Haematology, M. Lartey, MBChB, FWACP, Department of Medicine, University of Ghana Medical School, P. O. Box 4236, Accra, Ghana

Request for reprints to: L. A. Renner, Department of Child Health, University of Ghana Medical School, P.O. Box 4236, Accra, Ghana.

CUTANEOUS INVOLVEMENT OF HODGKIN LYMPHOMA IN A CHILD – CASE REPORT

L. A. RENNER, N. SALIFU, L. EDUSEI, C. SEGBEFIA, I. EKEM and M. LARTEY

SUMMARY

Specific cutaneous involvement in Hodgkin Lymphoma is rare and has not been reported in the younger paediatric age group. We report a case of a ten year old girl who presented with specific cutaneous involvement, confirmed using immunohistochemical stains. Treatment with combination chemotherapy resulted in rapid disappearance of the lesions and contrary to the generally poor prognosis associated with most other such reported cases in adults, she has clinically remained disease free two and a half years post treatment. Obtaining an accurate pathological diagnosis is essential to ensure appropriate treatment even in resource limited settings as illustrated by this case.

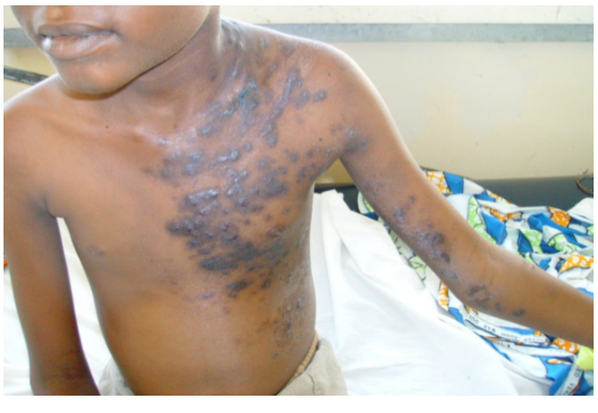
INTRODUCTION

Specific cutaneous involvement in Hodgkin Lymphoma is rare and may be due to retrograde lymphatic spread, direct extension from lymph nodes or haematogenous dissemination (1-4). Skin involvement is an indication of stage IV disease and is generally associated with a poor prognosis (1,3,5,6). Its incidence has decreased and this is likely due to improved treatment of Hodgkin Lymphoma (1,7). There have been no reports of cutaneous Hodgkin Lymphoma in the younger paediatric population. We report a case of a ten year old girl who presented with specific cutaneous involvement of Hodgkin Lymphoma, confirmed by immunohistochemical stains, who responded well to chemotherapy and has remained disease free two and a half years post treatment completion.

CASE REPORT

The patient was referred to Korle Bu Teaching hospital, in Accra, Ghana, at ten years of age with a six month history of fever, weight loss and a two month history of left sided neck swelling and skin rash. She had received two months anti-tuberculosis treatment at a District Hospital for possible extra-pulmonary tuberculosis. She appeared cachectic, weight 26.5Kg, febrile, pale and had generalised non-tender discrete firm cervical, axillary and inguinal lymphadenopathy. There were hyperpigmented firm to hard papules, nodules and plaques with areas of scaling of the left side of the neck, left haemithorax, inner aspect of left arm and left upper quadrant of abdomen (Fig.1). The lesions were pruritic. She also had a swelling of the medial canthus of the left eye, hyperpigmented lips and violaceous plaques on her hard palate. There was haepatomegaly measuring 4cm below the right subcostal margin. The spleen was not palpable. The differential diagnosis included Kaposi's sarcoma, lymphoma and extrapulmonary tuberculosis.

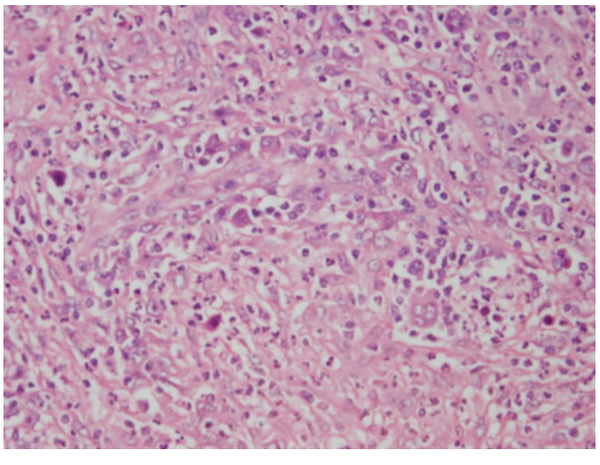
Figure 1
Skin lesions



Investigations : These were limited due to resource constraints. HIV serology was negative. Full blood count revealed Haemoglobin 9.9g/dl, white blood count 22.1 x 10^9/l (neutrophils 17.6 x 10^9/l),

platelets 92 x 10^9/l. Electrolytes, urea, creatinine, uric acid were normal. Bone marrow aspirate was not taken prior to starting chemotherapy due to logistic reasons but a specimen taken at week four prior to the second cycle of chemotherapy showed no evidence of malignant infiltration. Her chest Xray was normal. Abdomino-pelvic ultrasound scan revealed mild haepatomegaly and retroperitoneal lymphadenopathy. No CT/MRI scans were done due to lack of funds and PET scans are not available locally. Skin biopsies showed normal to slightly hyperplastic epidermis overlying aggregates and diffuse infiltration of lymphocytes, histiocytes, neutrophils and eosinophils. There were mononuclear, binucleated and multilobed Reed-Sternberg cells irregularly distributed. Occasional lacunar cells were seen and stromal fibrosis was evident. Lymphnode biopsy showed complete effacement of architecture by malignant lymphoma and presence of diffuse and nodular fibrosis. In the background of polymorphous infiltrate including neutrophils, eosinophils, lymphocytes and plasma cells were multinucleated, mononuclear lacunar Reed Sternberg cells (Figure 2).

Figure 2
Lymph node biopsy (H&E x40)



A tentative diagnosis of cutaneous involvement of Nodular Sclerosing Hodgkin Lymphoma was made. Specimens sent to South Africa and immunoperoxidase stains done showed: MUM 1 – definite nuclear staining in sub-population of tumour cells. ALK 1 – negative. CD45 – negative in skin and lymph node biopsy. EMA – difficult to interpret. CD30 – strong membrane and/or paranuclear staining observed in tumour cells. CD15 – no definite positive staining tumour cells. CD20 – negative in tumour cells. CD3 – negative in tumour cells. The definitive diagnosis was Hodgkin Lymphoma with cutaneous involvement.

She received eight cycles of treatment consisting of Chlorambucil, Vinblastine, Procarbazine, Prednisolone alternating with Adriamycin, Bleomycin, Vincristine and Dacarbazine (UKCCSG Hodgkins 2000 Protocol). The skin lesions cleared completely by the fouth cycle. Follow up investigations including ultrasound scans and Chest Xrays have remained normal and she is clinically disease free three years after first presentation.

DISCUSSION

Hodgkin lymphoma, when it presents as systemic disease and in advanced stages can infiltrate multiple lymph node sites, spleen and bone marrow. Non-specific cutaneous manifestations of the disease are not uncommon and some authors have put the prevalence between 13-50% (4,7). These lesions include; pruritis and associated prurigo, acquired ichthyosis, Addison-like hyperpigmentation, herpes zoster and alopecia (7).

Specific cutaneous disease, which is rare, was first described by Grotz in 1906 (5) and in reports occurred in 0.5 – 7.5% of Hodgkin lymphoma (1,3,5). It is thought that the incidence has declined with the advent of multimodal therapy and stem cell transplant for Hodgkin lymphoma (1,7). On literature review, it has never been reported in a child younger than 19 years of age (8,9).

The characteristic lesions are usually painless erythematous papules and nodules that frequently ulcerate. Three mechanisms have been proposed as the possible mode of spread; haematogenous dissemination, lymphatic spread or retrograde direct extension from affected lymph nodes to the skin (1,2,7,10). It is believed that retrograde direct extension is the most common mode of spread as the lesions commonly appear over areas drained by tumour-positive lymph nodes (4,7). This appears to be the mechanism of spread with this patient who had axillary and cervical nodes associated with the lesions on the chest and arm. Involvement of the trunk is considered to be the most frequent site (1,5).

The majority of reported cases occurred in patients with Nodular Sclerosing Type of Hodgkin Lymphoma (1, 10) as in this case. The results of immune-histo-chemistry using peroxidase technique were in keeping with findings reported in literature with CD30 positivity, CD45 negativity and CD3 negativity. CD15 negativity may be observed in cutaneous infiltrates (10).

This patient has remained disease free two and a half years post treatment completion contrary to reports of poor outcomes associated with most cases in the adult population (1,3,5). There have been some reported cases of good response to treatment (11).

This case emphasises the importance of precise pathological diagnoses of lesions, even in resource limited settings, to ensure appropriate treatment.

ACKNOWLEDGEMENTS

Dr Dorothy Lewis and Dr A. Gildenhuys, Lancet laboratories, South Africa for their expertise in making a definite diagnosis.

REFERENCES

1. Smith J. L, Butler J. J, Skin Involvement in Hodgkin's Disease. *Cancer* 1980; **45**: 354-361
2. Benninghoff D. L, Medina A, Alexander L. L, Camiel M. R. The mode of spread of Hodgkin's Disease to the Skin. *Cancer* 1970; **26**: 1135-1140
3. Tassies D, Sierra J, Montserrat E, Marti R, *et al*. Specific Cutaneous Involvement in Hodgkin's Disease. *Hem. Onc*. 1992; **10**: 75-79
4. Hayes T. G, Rabin V. R, Rosen T, Zubler M. A, Hodgkin's Disease presenting in the skin: case report and review of the literature. *J Am Acad Dermatol.* 1990; **22**: 944-947
5. White R. M, Patterson J. W. Cutaneous Involvement in Hodgkin's Disease. Cancer 1985; **55**: 1136-1145
6. Rubins J. Cutaneous Hodgkin's Disease. *Cancer* 1978; **42**: 1219-1221
7. Introcaso C. E, Kantor J, Porter D. L, Junkins-Hopkins J.M. Cutaneous Hodgkin's Disease. *J. Am. Acad. Dermatol.* 2008; **58**: 295-298
8. Fink-Puches R, Chott A, Ardigó M, Simonitsch I *et a*l. The Spectrum of Cutaneous Lymphomas in Patients Less than 20 Years of Age. *Pediatr Dermatol.* 2004; **21**: 525–533
9. Erkiliç S, Erbagci Z, Koçer NE, Bayazit YA, Camci C. Cutaneous involvement in Hodgkin's lymphoma: report of two cases. *J Dermatol.* 2004;**31**:330-334.
10. Cerroni L, Beham-Schmid C, Kerl H. Cutaneous Hodgkin's Disease: an immunohistochemical analysis. *J Cutan Pathol.* 1995; **22**: 229-235
11. Hsia C, Howsan-Jan K, Rizkalla K. Hodgkin Lymphoma with cutaneous involvement. *Dermatology Online Journal*. 2009; **15**: 5