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Histiocytosis X of the vulva

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Introduction

Langerhans cell histiocytosis is a rare disorder characterized by proliferation of Langerhans cells. It usually presents as a multiple lesions involving multisystem or occasionally as a solitary lesion involving a single organ or location. We are presenting a case of Langerhans'cell histiocytosis presenting as erythematous, pruritic and ulcerated lesion involving the right labia majora and clitoris. It was treated by surgical excision of the lesion by simple vulvectomy. In published literature there are only eight reported cases treated by surgery.

Case Report

A 41 years old nulliparous, married woman was seen in gynecological outpatient clinics complaining of intense pruritis of the vulva and blood stained discharge for the last two years. She was of Asian origin and belonged to the lower socioeconomic class.There was no significant medical or gynaecological history. General and systemic examination was unremarkable. Pelvic examination revealed an ulcerating, blood stained, erythematous lesion involving the clitoris and the right labia minora. Vagina, cervix, uterus and the adenexae were normal. No lympadenopathy was noted. Pap smear, ultrasound pelvis and the whole abdomen was reported to be normal. Vulval biopsy (from several areas) under local anaesthesia was performed with the provisional diagnosis of carcinoma of the vulva. The histpathologic evaluation done on haematoxylin and eosin stained slide showed skin with stratified squamous epithelium and focal ulceration. There was a dense inflammatory infiltrate composed mainly of histiocytes and eosinophils mixed with polymorphonuclear neutrophils, lymphocytes and plasma cells extending into the deeper dermis. There were also groups of Langerhan cells identified in the deeper dermis. There cells showed characteristics Kidney shaped nuclei and nuclear groves. Immunohistochemistry performed using DAKO envision method (DAKO CA USA), showed reactivity for S-100 and CD-68 in the histiocytic cell population. Histological and immunohistochemical findings were characteristic of Langerhan cells histiocytosis.
Chest radiotherapy, serum biochemistry was normal. Water deprivation test followed by vasopressin infusion test; to rule out the co-existence of diabetes insipidis was also performed and found to be normal. A diagnosis of "pure" genital Langerhan cell histiocytosis (LCH) was made.
A simple vulvectomy was performed. Post-operative course was unremarkable and she was discharged home on the fifth post-operative day. Follow-up six weeks later showed the patient to be symptom free.

Discussion

Langerhans cell histiocytosis (LCH) affecting the vulva alone, is rare. At present, there are 51 published cases of LCH affecting the genital area. Of these, only 8 had "pure" Langerhan Cell Histiocytosis. In these cases there was no subsequent systemic spread of the disease[1].
Langerhans cell histiocytosis (LCH) is a rare clonal disorder that consists of single or multiple mass lesions composed of cells with an abnormal langerhans cell population. Despite extensive search for evidence of consistent cytogenetic abnormalities, gene re-arrangements or viral genomes, it's etiology, remains unknown. The biologic behaviour of LCH ranges from spontaneous remission to lethal dissemination. Such behaviour cannot be predicted on the basis of histologic features. The presence and degree of organ dysfunction, together with the patient's age at diagnosis, remain the most reliable indicators of prognosis[2].
Four distinct patient groups, segregated on the basis of initial presentation and subsequent anatomic extent of the disease have been categorized: (a) "Pure" genital LCH (b) Genital LCH with subsequent multiorgan involvement: (c) Oral and cutaneous LCH with subsequent genital and multiorgan involvement: (d) Diabetes insipidus with subsequent genital and multiorgan disease[3].
The treatment of genital LCH is not well defmed and is highly individualized. Therapy has included surgery, radiation, topical corticosteroids, topical nitrogen mustard, and systemic chemotherapy and combination therapy; mixed results have been obtained with treatment modalities. Although no modality has been shown to yield a superior outcome, complete surgical excision is advocated as initial therapy[4].

References

1.Solano T, Espama A, Sola J, et a!.. Langerhan's cell histiocytosis on the vulva. Gynecol.Oncol .,2000; 78 251-54.
2.Herzog KM, Tubbs RR. Langerhans cell histocytosis. Adv. Anat. Pathol., 1 998;5: 347-58.
3.Axiotis CA, Menno MJ. Duray PH. Langerhans cell histocytosis of female genital tract. Cancer, 1991;67:1650-60,
4.Lin KD. Lin JD, Hsu HH, et al. Endocrinological aspects of Langerhans cell histiocytosis complicated with diabetes insipidus. J. Endocrinol. lnves., 1998;21: 428-33.