Langerhans cell histiocytosis presenting as a primary penile lesion

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Langerhans cell histiocytosis (LCH) is a rare proliferative disorder of cells, with the phenotype of activated Langerhans cells. The diagnosis of LCH is often delayed or missed and its clinical course is highly variable, ranging from a self-healing solitary bone lesion to widely disseminated life-threatening disease. Langerhans cell histiocytosis affects mainly young children and features an accumulation of CD1a+ dendritic Langerhans cells in the bone, skin, and other organs. Few cases of Langerhans cell histiocytosis on the penis have been reported in the literature. The case is presented here of LCH on the penis, with a review of similar documented cases.

CASE REPORT

A 37 year-old man with an uneventful medical history presented with a solitary nodular lesion on the prepuce, 1.2 cm in maximum diameter, reddish with a smooth surface. His clinical examination was unremarkable and no similar lesions were noted elsewhere. Surgical resection of the nodule was performed and histological assessment revealed infiltration of neoplastic cells, some of which were giant cells with oval, indented nuclei, a small degree of nuclear atypia and eosinophilic cytoplasm. Eosinophils were not observed in the histology sample. Immunohistochemistry results were positive for vimentin, S-100 protein, CD1a and Langerin, while the neoplastic cells were negative for keratins AE1/AE3, SM-actin, HNF-35, Desmin, CD34 and CD99. The proportion of cell proliferation, Ki-67+, was almost 20%. These findings were consistent with the diagnosis of LCH. Investigation for metastases, including computed tomography (CT) of the thorax and abdomen, 99mTc bone scan, and bone marrow aspiration revealed no evidence of disease beyond the penis. The patient was not given any medical treatment but was monitored closely as an outpatient in the hematological department, as cutaneous LCH could represent the initial presentation of the multisystem disease of LCH.

COMMENT

LCH of the penis is very rare cutaneous lesion. Its etiology remains unknown and association with various stimuli, such as chemical or viral exposure, has not been proved. Review of the relevant literature revealed reports of seven additional cases of LCH presenting as a penile lesion, which were treated by surgical excision or topical steroid ointments, or chemotherapy in the case of multisystem involvement. The outcome in all cases was excellent and there are no reports of recurrence of the disease in the penis.

Cases of non-Langerhans cell histiocytosis have also been reported. Juvenile xanthogranuloma (JXG) is the commonest variant of non-Langerhans cell histiocytosis, and only four cases with a penile lesion have been reported. In addition, one case of intermediate cell histiocytosis presenting as a solitary lesion of penis has been reported.

The prognosis of solitary penile lesions of LCH is excellent, with no reports of recurrence of the disease. Nevertheless, careful assessment should be made for any systemic involvement and close monitoring is recommended.
ΠΕΡΙΛΗΨΗ

Ιστιοκυττάρωση από κύτταρα Langerhans σε δερματική αλλοίωση ακροποσθίας

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Η ιστιοκυττάρωση από κύτταρα Langerhans είναι σπάνια νόσος, συνήθως της παιδικής ηλικίας, που σχετίζεται με κλωνικό πολλαπλασιασμό των δενδριτικών κυττάρων CD1a+. Συχνές θέσεις προσβολής αποτελούν ο μυελός των οστών, οι πνεύμονες και τα οστά. Το κλινικό φάσμα της νόσου κυμαίνεται από την εμφάνιση μονήρων ασυμπτωτικών βλαβών έως και εκδηλώσεις πολυσυστηματικής νόσου. Για τη συμμετοχή των ανδρικών γεννητικών οργάνων στη νόσο υπάρχουν ελάχιστες αναφορές στη βιβλιογραφία. Περιγράφεται η σπανιότατη περίπτωση νεαρού άνδρα με ιστολογικά και ανοσοϊστοχημικά καταχωρημένη διάγνωση πρωτοπαθούς ιστιοκυττάρωσης Langerhans στην ακροποσθία. Ο πλήρης έλεγχος για σταδιοποίηση δεν αποκάλυψε παθολογική εστία νόσου. Από τη μελέτη της διεθνούς βιβλιογραφίας προκύπτει ότι η εντόπιση της ιστιοκυττάρωσης Langerhans στο πέος, όπως συμβαίνει στην παρούσα περίπτωση, είναι σπάνια. Η αιτιολογία είναι άγνωστη και δεν έχει αποδειχθεί η συσχέτιση αυτής της σπάνιας δερματικής αλλοίωσης με ποικίλα ερεθίσματα όπως χημικά ή ιογενή. Ως αντιμετώπιση αναφέρεται χειρουργική αφαί-