Case Report
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A case of plantar localization of juvenile xanthogranuloma and review of the literature

Plantar yerleşimli bir jüvenil ksantogranülom olgusu ve literatürdeki olgular

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Abstract

Juvenile xanthogranuloma (JXG) is the most common type of non-Langerhans cell histiocytosis. The most common sites for development are the head and neck, and peripheral involvement is rare. Here, we present a 19-month-old patient who had a plantar lesion that did not clinically look to be JXG but received a histopathological diagnosis and review of the relevant literature.
Keywords: Juvenile xanthogranuloma, plantar, histiocytosis

Öz

Jüvenil ksantogranülom (JKG), Non-Langerhans hücreli histiyositozların en sık görülen tipidir. Baş ve boyun en sık lokalize olduğu bölgeler olup periferik tutulum daha azdır. Burada ayak tabanı yerleşimli, klinik olarak JKG düşündürmeyen ancak histopatolojik inceleme sonucunda tanısı konulan on dokuz aylık bir hasta sunulmuş ve literatürde yayınlanmış plantar yerleşimli olguların değerlendirilmesi yapılmıştır.
Anahtar Kelimeler: Jüvenil ksantogranülom, plantar, histiyositoz

Introduction

Juvenile xanthogranuloma (JXG) is the most common form of non-Langerhans cell histiocytosis. Seventy-five percent of cases arise in early years of life. The disease is commonly limited to skin and self healed. Based on the number and size of the lesions, JXGs can be classified as small nodular and big nodular forms, also it is defined as subcutaneous, clustered, papillated, plaque like, keratotic and giant, according to clinical appearance[1]. Here an atypical case of JXG is reported with similiar cases in the literature.

Case Report

A 19-month-old male patient was examined in outpatient clinic with a complaint of plantar solitary nodule which has been persisted for 1 year. Nine months before administering our clinic, 5 fluorouracil-salicylic acid combination solution was applied to the lesion for 1 month with a diagnosis of plantar wart. On physical examination there was approximately 1 cm soft, centrally ulcerated, hemorrhagic nodule with keratotic collar like rims on right foot base second metatharsophalangeal joint (Figure 1). Lesion was excised with prediagnosis of pyogenic granuloma, pilomatricoma, fibrosarcoma, melanoma.

Histopathologic examination revealed a dense histiocytic infiltration spreading through deep dermis. Most of the histiocytes were small, round and oval shaped, and had eosinophilic and foamy cytoplasms. In focal areas, Touton

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type giant cells were detected (Figure 2a). By the immunohistochemistry performed, the histiocytes were positive for CD68 (Figure 2b), but negative for S-100 (Figure 2c). According to the histopathological and immunohistochemical findings, the diagnosis of JXG was established.

Discussion

JXG is characterized as sharply demarcated, 0.5-2 cm pink-red colored papule or nodule which turns yellowish by time. Lesions are commonly located on head and neck area, followed by trunk, upper extremities and lower extremities[2]. Atypical areas like lip, tongue, eye lid, penis, nail bed, scrotum are reported in the literature with only 5 plantar JXG cases in our search on PubMed data base[3-7]. Our patient's features are summarized with the other literature cases in Table 1. Ages of the cases of plantar JXG's were between 4 months and 38 years. Only one case was female, four cases were male. Male patients had solitary and female patient had multiple lesions. The common feature of the cases was about their atypical clinical appearence that they were excised with prediagnosis like adnexial tumor, pyogenic granuloma, melanoma. Another remarkable feature in some cases was hyperkeratotic rim around the lesion. Mervak and friends defined that this sign can be helpful for diagnosis[7].

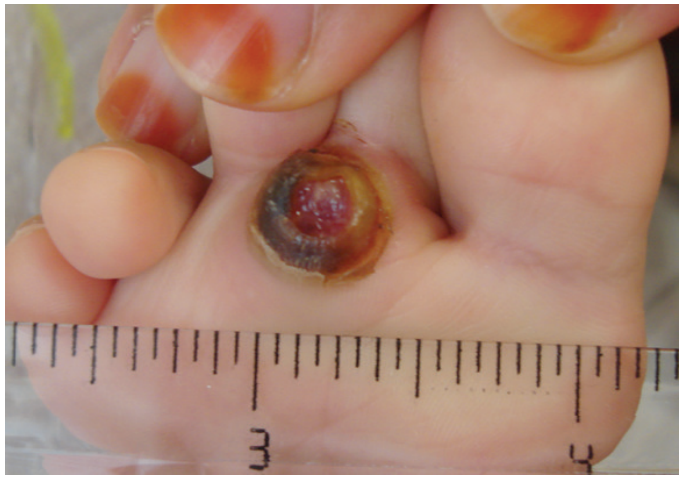

Figure 1. A 0.4 inch (~1 cm) hemorrhagic nodule on the sole with collarette-like hyperkeratotic rim

For diagnosing atypical cases there is a certain need for histopathological examination and it is important to perform immunohistochemical staining especially for excluding JXG from other histiocytosis. Invading histiocytes in JXG are negative for Langerhans cell markers CD1a and S100 and positive for CD68.

Extracutaneous involvement of JXG is rare. The most affected organs are eye, lung and liver in order of frequency. Heart, spleen, kidney, gonad, bone, central nervous system can be less affected. Risk of systemic involvement raise in patients who have multiple skin lesions[8]. Eye involvement commonly appears in the first two years of life. The most common features of ocular involvement is hyphema and glaucoma that may cause blindness[9]. The widest study about ocular involvement risk in cutaneous JXG is designed by Chang[10]. In this

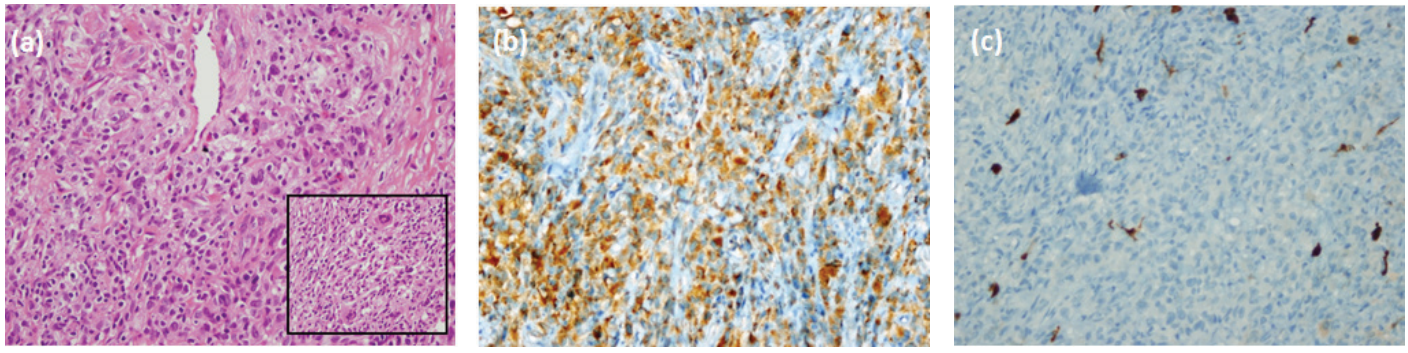

Figure 2. (a) Foamy cells, intermingled with sparse lymphocytes and eosinophils. Note the Touton type giant cell in the inset (H&E x100). (b) Histiocytes staining positive with CD68 (x200). (c) There were only a few cells staining for S100 (x200)

Table 1. Clinical features of plantar localized juvenile xanthogranuloma cases

Case / reference	Sex	Age	Number of lesions	Clinical features	Prediagnosis
1/3	M	14 months	Solitary	1 cm, centrally depressed dark brown nodule with hyperkeratotic rims	Adnexial tumor
2/4	M	38 years	Solitary	0.7 cm erythematous eroded nodule	Poroma
3/5	M	37 years	Solitary	1.2 cm red nodule surrounded with serous exsudate	Pyogenic granuloma
4/6	F	4 months	Multiple	1-3 mm plane red-brown polygonal papules	NA
5/7	M	11 months	Solitary	8 mm nodule with hyperkeratotic rims	Traumatized nevus, foreign body reaction, infection, melanoma, fibroma
Present case	m	7 months	Solitary	1 cm hemorrhagic centrally ulcerated nodule with hyperkeratotic rims	Pyogenic granuloma, pilomatricoma, fibrosarcoma, melanoma

F: Female, M: Male, NA: Not applicable

study ocular involvement risk is found 0.3%. Also JXG is defined to be associted with neurofibromatosis type 1 and juvenile myelomonocytic leukemia[11].

Our patient has no detected visceral involvement and hematologic abnormality after laboratory evaluation, ophthalmologic examination and abdominal ultrasonography. During 3 year of follow up period after excision there has been no recurrence.

We would like to share this case for unusual localization and clinical presentation of JXG with similiar cases in the literature, point the possibility of JXG in plantar localized papules especially with keratotic rims, and to emphasize the role of pathologic evaluation for the right diagnosis.

Ethics

Informed Consent: Consent was obtained from patients family.
Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Esra Saraç, Kıvılcım Karadeniz Cerit, Concept: Esra Saraç, Ayşe Deniz Yücelten, Design: Esra Saraç, Ayşe Deniz Yücelten, Data Collection or Processing: Esra Saraç, Analysis or Interpretation: Cuyan Demirkesen, Esra Saraç, Literature Search: Esra Saraç, Writing: Esra Saraç.

Conflict of Interest: No conflict of interest was declared by the authors.

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