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Penile skin lesions and urinary tract obstruction: A rare presentation of Langerhans cell histiocytosis

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ABSTRACT

Langerhans cell histiocytosis (LCH) can manifest in any organ or system, but the occurrence of cutaneous lesions on the penis, causing urethral stenosis, is particularly uncommon. The diagnosis primarily relies on typical clinical manifestations and pathological examination. Treatment involves the excision of local lesions combined with chemotherapy, with a generally favorable prognosis.

A 3-year-old male patient experienced voiding difficulties after circumcision, revealing penile skin lesions upon examination. Postoperative pathological analysis and immunohistochemistry confirmed the diagnosis of LCH.

Langerhans cell histiocytosis can present as cutaneous lesions on the penis, leading to symptoms of urinary tract obstruction.

1. Introduction

Langerhans cell histiocytosis (LCH) is a disease characterized by abnormal proliferation of Langerhans cells, which can spread to various body organs. The clinical manifestations of LCH are mainly based on the site of Langerhans cell proliferation. LCH can occur at any age, but it primarily affects patients under 15 years of age. The peak incidence is between 1 and 4 years of age, and approximately 2% of LCH is present during the neonatal period.[1] The clinical presentation of LCH is atypical. It varies according to the location and extent of involvement. LCH is less common in single-system lesions and more common in multisystem disease. Cutaneous involvement is typically representative of multisystem disease. Isolated cutaneous disease accounts for only 2% of total cases.[2] Furthermore, isolated genital cutaneous defect is a rare occurrence. This case report presents a case of LCH with penile skin lesions as the primary manifestation.

2. Case presentation

A 3-year-old boy was admitted to our institute with symptoms of progressively worsening urinary obstruction, five months after circumcision in another hospital. During the physical examination, meatus stenosis and scarring hyperplasia were observed in a sub-coronary location, alongside a significant granulomas lesion at the junction of the penis and scrotum (Fig. 1A). The patient was initially considered with balanitis xerotica obliterans (BXO) and subsequently underwent urethrotomy, granulomas lesion excision, and buccal mucosa grafting for urethral defects in a single operation. The diagnosis of LCH was confirmed postoperatively by pathology and immunopathology (Fig. 1B), characterized by the infiltration of eosinophils, T-lymphocytes, neutrophils, and Langerhans cells for the granulomas lesion, and expression of the histiocytic markers CD1a, S100 (Fig. 1C & D). Afterward, the head CT, chest X-ray, and abdominal ultrasound were performed which were negative, eliminating the possibility of any concurrent systemic disease.

The oncology team oversaw the administration of chemotherapy treatment for the child. The postoperative chemotherapy plan encompassed two phases: intensive therapy and maintenance therapy.[3] The intensive regimen involved the intravenous infusion of vinblastine at a dosage of 6 mg/m^2 once a week for 6 weeks, in conjunction with daily oral consumption of prednisone at a dosage of 40 mg/m^2 for 4 weeks. This intensive treatment was repeated for two cycles, spanning almost 3 months. Subsequently, the maintenance regimen consisted of the intravenous infusion of vinblastine 6 mg/m^2 every 3 weeks, accompanied by oral administration of prednisone at 40 mg/m^2 twice daily for 5 days, once every 3 weeks. This maintenance protocol was sustained for 9

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months. Follow-up assessments were systematically conducted at 3 months, 6 months, 1 year, and 2 years post-treatment to monitor the child's progress.

One year after completing chemotherapy, the patient underwent urethroplasty. Over the subsequent five years of follow-up, the child's condition remains satisfactory, showing no signs of recurrence, normal urine stream during urination, and overall health is good.

3. Discussion

Langerhans cell histiocytosis (LCH) is a rare disease characterized by the accumulation and proliferation of Langerhans cells in various tissues, with its etiology remaining unknown. The clinical manifestations of LCH are diverse and can affect almost all organs. Illustrating the rarity of LCH presentations, we highlight a unique instance involving lesions on the penile skin. This phenomenon has been documented in only five related case reports in individuals under the age of 18 years.[4]

Diagnosing penile skin lesions can be challenging, as they may be confused with balanitis xerotica obliterans (BXO), a chronic inflammatory skin condition primarily affecting the external genitalia of men. Recognized as the male genital variant of Lichen Sclerosis (LS), BXO typically manifests in school-aged children, presenting with porcelain-white atrophic patches or plaques on the foreskin and glans penis.[5] During the initial presentation of urinary obstruction, pediatric urologists may find it challenging to consider LCH as a potential diagnosis, often leaning toward the more common diagnosis of BXO.

The therapy options for Langerhans cell hyperplasia in children depend on the extent of the disease and the location of the disease. There are significant differences in the treatment of single and multisystem LCH. Single-organ skin involvement can progress to multisystem involvement or heal spontaneously.[2] However, no standardized initial treatment protocol exists when the skin is the only affected organ. Treatment modalities encompass observation, topical corticosteroids, surgical excision of isolated lesions, and chemotherapy. In the previous 5 reports of LCH involving the penile skin in children, surgical excision and systemic treatment with corticosteroids and chemotherapy with vinblastine were administered, and only one case received topical corticosteroids.[4] After treatment, lesions subsided and symptoms were remitted.

In the presented case, LCH progressed rapidly, manifesting symptoms of urinary obstruction caused by urethral stricture. Following symptom relief through surgical intervention, consultation with oncologists led to the decision of chemotherapy. And the chemotherapy regimen is designed for 12 months to prevent recurrence. The post-operative chemotherapy targeting LCH revealed no impact on the survival of the prepositioned oral mucosal (Fig. 1E) for next-stage urethral reconstruction (Fig. 1F). Subsequently, the five-year follow-up revealed no recurrence of LCH, indicating the efficacy of the chemotherapy regimen in preventing disease relapse.

4. Conclusion

A lesion of Langerhans Cell Histiocytosis (LCH) in the penile skin resulting in urinary obstruction symptoms is exceptionally rare. Diagnosis of LCH primarily relies on pathological assessment. The presented case demonstrates the successful management of LCH with urological involvement using a combination of surgical and chemotherapeutic modalities, leading to favorable long-term outcomes.

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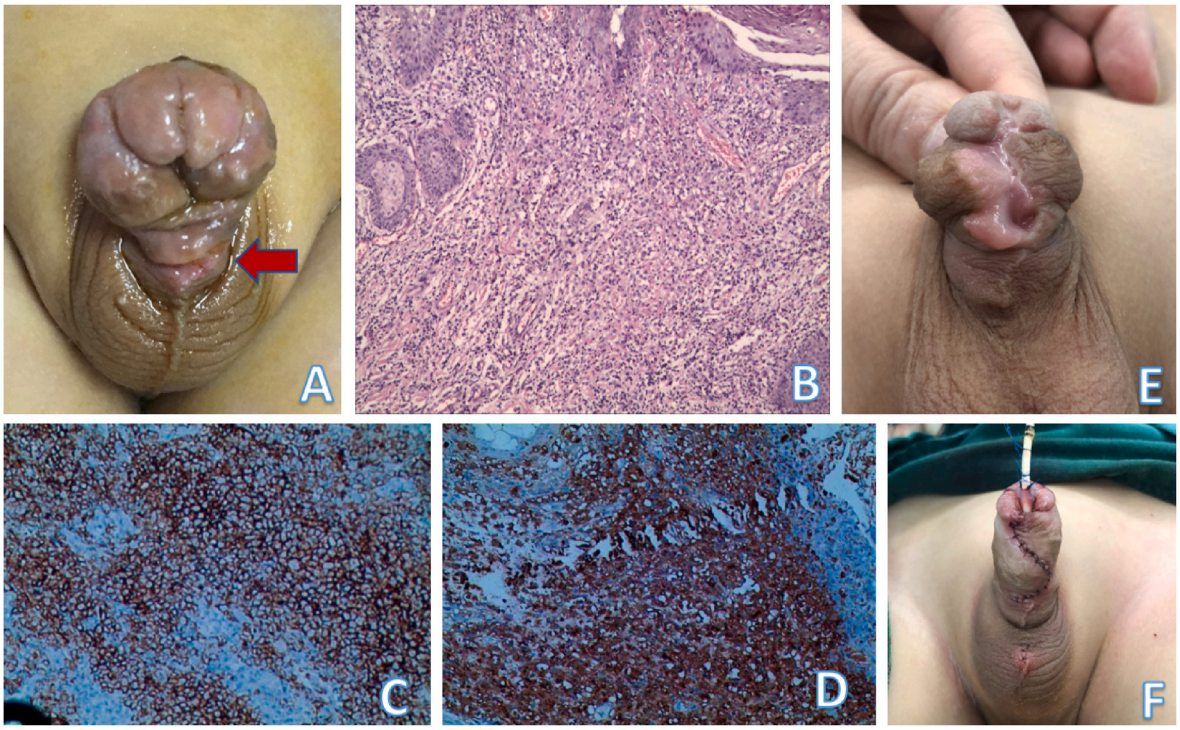

Fig. 1. A: The appearance of penile skin lesion.
Fig. 1B: H&E stain section from the penile skin lesion (200 ×). A large proliferation of histiocytes in the dermis with eosinophilic and lymphocytic infiltration.
Fig. 1C: Immunohistochemical staining of pathological sections with antibodies CD1a (+).
Fig. 1D: Immunohistochemical staining of pathological sections with antibodies S100 (+).
Fig. 1E: Post-chemotherapy, buccal mucosal grafts are taken for urethralplasty.
Fig. 1F: The post-operative appearance of urethral reconstruction.

CRediT authorship contribution statement

Yanxue Jiang: Writing – original draft. **Ting Zhang:** Writing – review & editing, Conceptualization. **Jun Lu:** Methodology. **Yun Zhou:** Supervision.

Declaration of competing interest

The authors have nothing to disclose.

References

1. Fraitag S, Emile JF. Cutaneous histiocytoses in children. *Histopathology*. 2022;80(1):196–215. https://doi.org/10.1111/his.14569.
2. Krooks J, Minkov M, Weatherall AG. Langerhans cell histiocytosis in children: history, classification, pathobiology, clinical manifestations, and prognosis. *J Am Acad Dermatol*. 2018;78(6):1035–1044. https://doi.org/10.1016/j.jaad.2017.05.059.
3. Haupt R, Minkov M, Astigarraga I, et al. Langerhans cell histiocytosis (LCH): guidelines for diagnosis, clinical work-up, and treatment for patients till the age of 18 years. *Pediatr Blood Cancer*. 2013;60(2):175–184. https://doi.org/10.1002/pbc.24367.
4. Thomas C, Mesnard C, Petrot D, Fraitag S, Legroux J, Barbarot S. Uncommon preputial localization of Langerhans cell histiocytosis. *Pediatr Dermatol*. 2021;38(2):500–501. https://doi.org/10.1111/pde.14468.
5. Lansdale N, Arthur F, Corbett HJ. Circumcision versus preputioplasty for balanitis xerotica obliterans: a randomised controlled feasibility trial. *BJU Int*. 2021;128(6):759–765. https://doi.org/10.1111/bju.15508.