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Enigmatic Presentation of Primary Cutaneous Lymphoma in a Boy

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Background: Primary cutaneous follicle center lymphoma (PCFCL) is a subtype of primary cutaneous B-cell lymphoma. It is exceedingly rare in the pediatric population, with less than two dozen cases documented in individuals younger than 20 years. The rarity of the disease, coupled with the scarcity of comprehensive clinical data, emphasizes the importance of a meticulous approach in the diagnostic process to avoid under-diagnosis or misdiagnosis.

Methods: We report a case of an eight-year-old boy who presented with a unique form of scalp lesion, ultimately diagnosed as PCFCL. A complete excision of the remaining defect was addressed by a rotational flap based on the superficial temporal artery.

Results: Adopting a multidisciplinary approach in managing PCFCL in the pediatric age group exemplifies the importance of collaborative care in addressing complex and rare conditions. The favorable outcome post surgical intervention reinforces the role of complete excision in treating localized PCFCL, aligning with current treatment guidelines for this patient demographic.

Conclusions: The management of pediatric PCFCL consists of complete surgical excision as the primary treatment modality, reserving radiotherapy for cases of relapse. This case adds to the scant literature on pediatric primary cutaneous B-cell lymphoma and highlights this rare entity's diagnostic challenges and clinical peculiarities. Ongoing research is essential to enhance our understanding, refine diagnostic criteria, and develop more effective treatment protocols for PCFCL.
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INTRODUCTION

Primary cutaneous lymphomas encompass a heterogeneous group of cutaneous neoplasms, manifesting primarily or secondarily as non-Hodgkin lymphomas. Among these, primary cutaneous B-cell lymphoma (PCBCL) is exceedingly rare in the pediatric and adolescent population, with only a handful of cases documented in the literature. These lymphomas represent approximately 20%–25% of all adult primary cutaneous lymphomas, underscoring their rarity in the pediatric demographic.[1]

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The incidence of PCBCL in children and adolescents is strikingly low, recorded at 0.12 per 1,000,000 person-years.[2] Within this category, primary cutaneous marginal zone lymphoma is the most prevalent subtype (77.1%), followed by primary cutaneous diffuse large B-cell lymphoma (12.5%) and primary cutaneous follicle center lymphoma (PCFCL; 10.4%).[2] PCFCL, originating from follicle center cells, is especially noteworthy for its infrequency, comprising less than 1% of all B-cell lymphoma cases.[3] Although it predominantly affects White men aged 50–60 years and constitutes the majority of PCBCLs, this subtype is exceptionally rare in children.[4,5]

This report delves into the case of an 8-year-old boy who presented with a unique form of scalp lesion, ultimately diagnosed as PCFCL. This case adds to the scant literature on pediatric PCBCL and highlights this rare entity's diagnostic challenges and clinical peculiarities.

CASE PRESENTATION

A healthy 8-year-old boy was referred from a local health center to our pediatric surgery clinic with a history of a slowly enlarging solitary nodule on the left parietal region

Disclosure statements are at the end of this article, following the correspondence information.

of the scalp. The nodule, first noticed 2 months prior, was associated with intermittent itchiness but no other symptoms. The family reported no episodes of fever, night sweats, or unexplained weight loss, which are often considered systemic symptoms in lymphoma cases.[6] On physical examination, the lesion was noted to be a smooth, firm, nonulcerated nodule measuring approximately 3×2 cm without evidence of regional lymphadenopathy (Fig. 1).

An excisional biopsy was performed, and histopathological examination revealed an unremarkable epidermis. However, the dermal layer was noted to have a dense nodular infiltrate composed of lymphoid follicles. These follicles were characterized by the absence of tingible-body macrophages and ill-defined to absent mantle zones extending to the resection margins. Immunohistochemical studies yielded positive results for paired box5, CD45 (Leukocyte Common Antigen), CD20, CD10, and B cell lymphoma 2 protein (Bcl)-6, indicative of atypical lymphoid follicles (Figs. 2–4). The samples were negative for CD5, CD3, and Bcl-2. CD21 and CD23 staining revealed focally disrupted follicular dendritic meshwork. The Ki-67 proliferative index was notably high (>30%). Immunoglobulin gene rearrangement studies confirmed clonality, supporting a diagnosis of lymphoma.[7]

Based on the clinical presentation and histopathological findings, the differential diagnosis included PCFCL, primary cutaneous marginal zone lymphoma, or primary cutaneous diffuse large B cell lymphoma (PCDLBCL). Comprehensive clinical, laboratory, and radiological investigations, including a positron emission tomography scan, did not reveal systemic involvement, thus favoring a diagnosis of PCFCL.[5]

Subsequently, the patient was referred to our plastic and reconstructive surgery clinic. The National Tumor Board committee advocated for reexcision with scalp reconstruction. Surgical re-excision was successfully performed, achieving 1.5-cm margins (Fig. 5). An intraoperative frozen section confirmed negative margins. The resultant 3.5-cm scalp defect was addressed with a rotational flap, based on the superficial temporal artery, ensuring tension-free closure (Figs. 6 and 7). The patient recovered well and was discharged the following morning. The patient was reviewed on a weekly basis for the first month and 5 months postoperatively, with no evidence of recurrence (Fig. 8). As the average follow-up period for current reports of pediatric primary cutaneous follicular cell lymphoma ranges between 2 and 5 years, we planned to follow up with our patient for 3 years.

Takeaways

Question: How can we accurately diagnose and treat the rare primary cutaneous follicle center lymphoma (PCFCL) in pediatric patients, given the limited clinical data?

Findings: Our case study of an 8-year-old boy demonstrates the effectiveness of a multidisciplinary approach, involving complete surgical excision and scalp reconstruction, in treating localized PCFCL. This method resulted in a favorable outcome and adheres to current treatment guidelines for pediatric patients.

Meaning: Successful treatment of pediatric PCFCL relies on precise diagnosis and a collaborative care approach, emphasizing the importance of surgical excision.

DISCUSSION

PCFCL is an exceedingly rare entity in the pediatric population, with less than two dozen cases documented in individuals younger than 20 years in the available literature[3] These lymphomas typically manifest in the skin without extracutaneous spread at diagnosis.[8]

The clinical presentation often includes solitary or multiple nodules, with a proclivity for the scalp, forehead, and trunk regions.[9] Condarco et al reported a noteworthy case involving an 8-year-old boy with a PCFCL on the frontal scalp, successfully managed through surgical excision, with no recurrence observed over a 4-year follow-up period.[10]

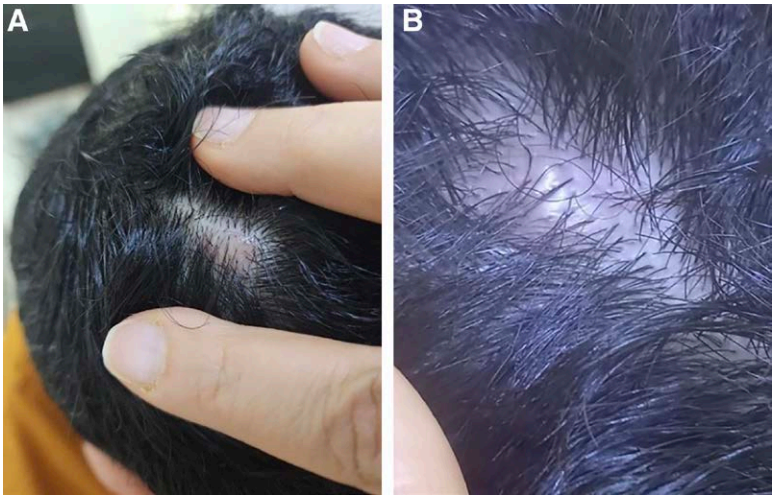

Fig. 1. Initial presentation of the scalp lesion as a nodular, firm mass about 3 × 2 cm. A, Long view. B, Close-up view.

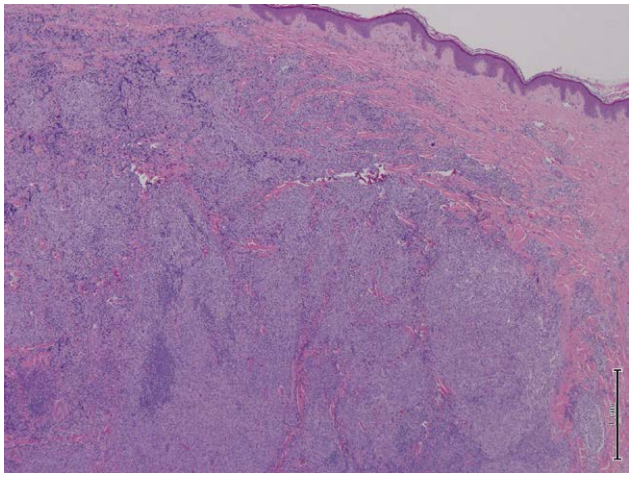

Fig. 2. Dense lymphocytic infiltrate in the dermis extends till the subcutaneous layer, with sparing of the epidermis in follicles, as demonstrated by H&E staining.

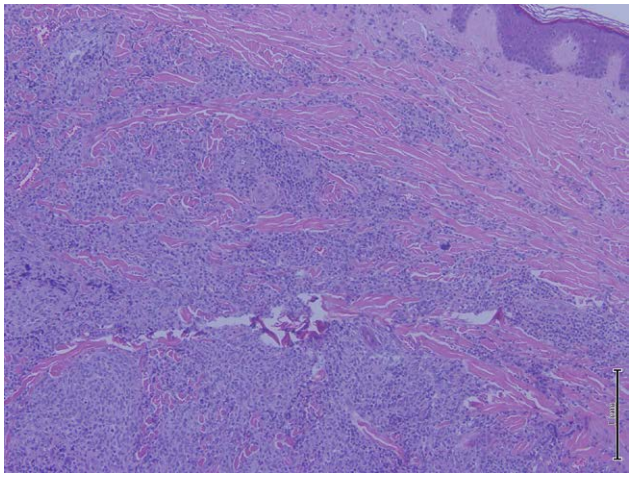

Fig. 3. Microscopic examination reveals abnormal lymphocytes of follicle center origin, resembling sheets stained by H&E.

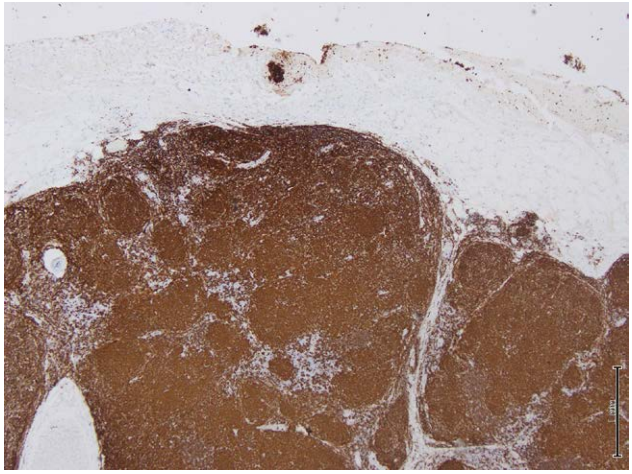

Fig. 4. This image highlights CD20 immunohistochemistry positivity in numerous neoplastic follicles.

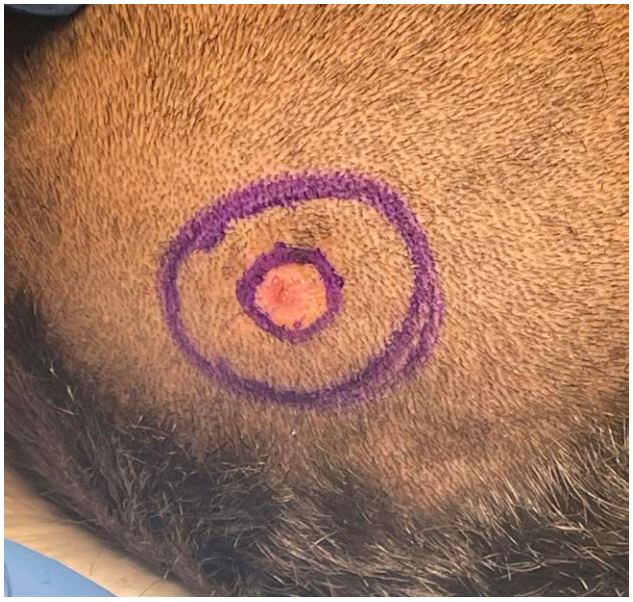

Fig. 5. Preoperative marking of the residual scar tissue from the previous excision over the left parietal region measuring 1.5 × 1 cm with additional 1.5-cm margins.

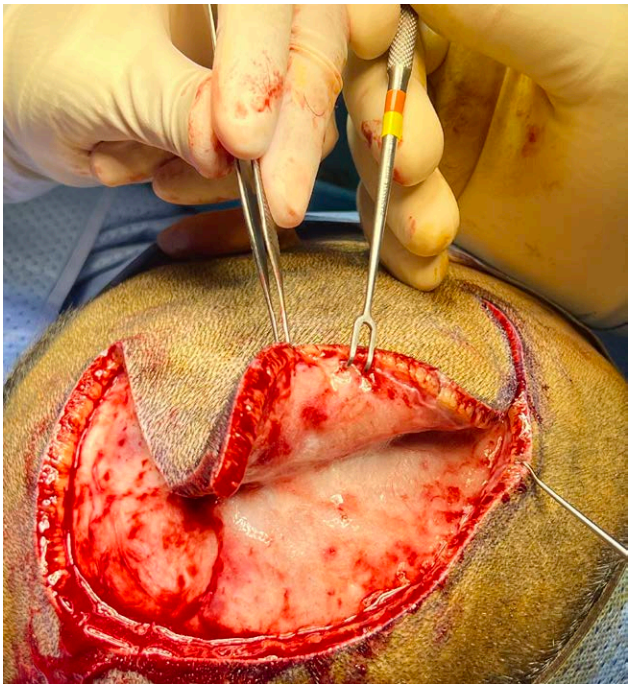

Fig. 6. Complete excision of the scalp lesion down to the periosteum with elevation of the advancement rotational flap within the subgaleal plane.

The pathogenesis of PCFCL remains an area of ongoing research. Chronic antigenic stimulation by viral agents, such as Epstein-Barr virus, human herpesvirus 8, and hepatitis B virus, has been postulated as a potential contributing factor.[11] Histopathologically, these tumors are characterized by dermal infiltration extending into

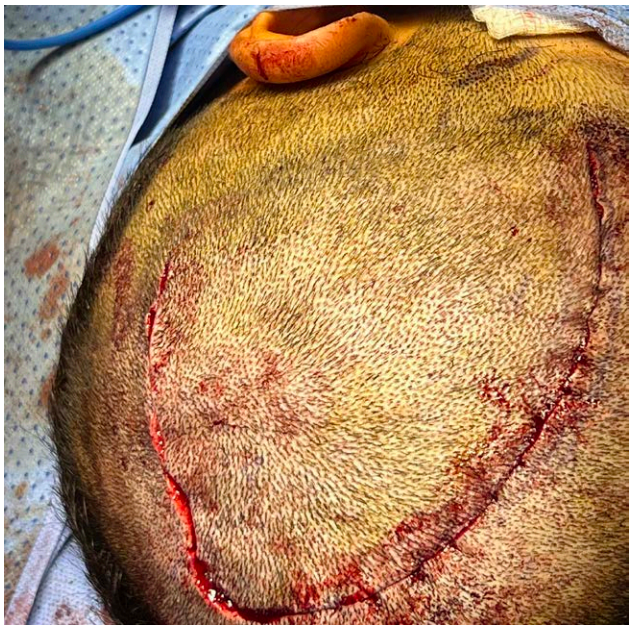

Fig. 7. Tension-free primary closure of scalp lesion in layers.

Fig. 8. Image depicting complete healing of the closure line 2 weeks postoperatively.

the subcutaneous fat. The neoplastic cells predominantly express B-cell markers, including CD20 and CD79a, and consistently BCL6, denoting their follicle center origin. Although the proliferation index, as indicated by Ki67 staining, is generally lower than in reactive follicles, some cases exhibit high proliferation indices.[5]Distinct from reactive follicular infiltrates, neoplastic follicles in PCFCL typically lack or have only a few tingible body macrophages.[1]

BCL2 expression, reported in a proportion of PCFCL cases,[12] is a critical discriminant in distinguishing PCFCL from systemic follicular lymphoma with secondary cutaneous involvement. In contrast to nodal follicular lymphoma, PCFCL tends to exhibit lower levels of expression of BCL2.[13]

To evaluate systemic involvement, comprehensive laboratory workups, including complete blood count, lactate dehydrogenase, comprehensive metabolic panel, and positron emission tomography/computed tomography, are essential per the National Comprehensive Cancer Network guidelines.[14]

The management of pediatric PCFCL, as per recent guidelines, emphasizes complete surgical excision as the primary treatment modality, reserving radiotherapy for cases of relapse due to its potential side effects, including alopecia and scarring of the scalp.[15] Although single-agent rituximab therapy has demonstrated efficacy in adult patients with extensive skin involvement, its application and effectiveness in the pediatric setting require further investigation.[16]

The prognosis for PCFCL is generally favorable, with a 5-year survival rate in adults exceeding 95%.[17] The prognosis in pediatric and adolescent patients is posited to be similar, although the limited number of cases necessitates cautious interpretation of these outcomes.[15]

The rarity of PCFCL in children necessitates a thorough and meticulous approach in the diagnostic process to prevent misdiagnosis. This case adds to the scant literature by highlighting unique clinical and diagnostic challenges faced in pediatric PCFCL.

Ongoing research is essential to enhance our understanding, refine diagnostic criteria, and develop more effective treatment protocols for PCFCL. It is imperative that we continue to accumulate clinical data to better understand, diagnose, and treat this rare lymphoma subtype.

CONCLUSIONS

This case underscores the complex nature of diagnosing and managing PCFCL in pediatric patients. Through a multidisciplinary approach and complete surgical excision, we achieved a favorable outcome, reinforcing the effectiveness of these strategies. Continued attention to such rare cases is crucial to improving outcomes and refining treatment approaches for future patients. This contributes to the growing compendium of knowledge on pediatric PCFCL, ultimately paving the way for improved patient outcomes and the development of targeted therapeutic strategies in the future.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

PATIENT CONSENT

Parents provided written consent for the use of patient's images.

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