Case Report

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Burkitt lymphoma in a scalp region: a case report of it's recurrence in a 13-year-old child

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Introduction: Burkitt lymphoma (BL) is an aggressive non-Hodgkin lymphoma characterized by chromosome 8 MYC gene translocation. It manifests in three clinical types: immunodeficiency-related, sporadic (nonendemic), and endemic (African), each differing in epidemiology and clinical behavior. Treatment typically involves enrollment in clinical trials or intensive chemotherapy regimens like R-CODOX-M/IVAC. The authors present a case of recurrent BL following treatment.

Case report: A 13-year-old female presented with a gradually progressive swelling in the left parieto-occipital region. Examination revealed normal vital signs and a Glasgow coma scale, with seronegative findings on investigations. An excision of a subganglion soft tissue tumor was performed, revealing histopathological features suggestive of a small round blue cell tumor. After chemotherapy, the patient experienced a recurrence in the scalp region, diagnosed as BL.

Discussion: While scarce reports exist on BL in the scalp region, cases have been documented in various body locations. Treatment strategies, including chemotherapy and surgery, have shown promising results in managing the disease and improving symptoms.

Conclusion: The recurrence of BL is rare, highlighting the importance of vigilance in monitoring patients post-treatment. The authors report a case of recurrent BL in a 13-year-old female, emphasizing the need for continued research and surveillance in managing this aggressive malignancy.

Keywords: burkitt lymphoma, non-hodgkin lymphoma, recurrence

Introduction

Burkitt lymphoma (BL) is a very aggressive non-Hodgkin lymphoma of the B cell. It is defined by chromosome 8 MYC gene translocation and dysregulation. Three recognized clinical types of BL are: immunodeficiency related, sporadic (nonendemic), and endemic (African). These types differ in their epidemiology, clinical appearance, and genetic characteristics, although sharing the same histology and exhibiting comparable clinical behavior[1].

BL and Burkitt leukemia are considered different manifestations of the same disease in the WHO 5th edition (WHO5) of the classification of hematolymphoid tumor and the International Consensus Classification (ICC) of mature lymphoid neoplasms. Both classification systems also recognize three aggressive B cell lymphoma entities that resemble BL: 'Burkitt-like lymphoma with 11q aberration', 'High-grade B cell lymphoma with *MYC* and *BCL2* rearrangements', and 'High-grade B cell lymphoma, not otherwise specified'[2,3].

HIGHLIGHTS

- Genes and defects involve in Burkitt Lymphoma (BL).
- The burden of BL in the pediatric population.
- Recurrence of BL in the pediatric population.
- Prognosis of the disease.
- Management of BL in primary and recurrent cases.

Since the resource-poor countries with the highest apparent prevalence—such as equatorial Africa—are unable to gather the epidemiologic data required for exact diagnosis and case ascertainment, the precise global incidence of BL remains unknown. Cases of BL are typically classified into three different clinical types for epidemiologic and diagnostic purposes: endemic (African), sporadic (nonendemic), and immunodeficiency related. The geographic distribution of endemic and sporadic clinical forms of BL varies[4].

The standard of care has yet to be defined and our preferred treatment is enrollment in a clinical trial. However, for patients who are not candidates for such trials or for those who choose not to participate, we suggest intensive, short-duration combination chemotherapy with central nervous system prophylaxis. Our preferred regimen is R-CODOX-M/IVAC (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine, Methotrexate/ Rituximab, Ifosfamide, Etoposide, Cytarabine). CODOX-M/IVAC is highly toxic, primarily to the hematopoietic system, and most patients will have a prolonged hospital stay. Therapy should be initiated promptly and dose reduction should be avoided, if possible. For older or less fit patients who may not tolerate more aggressive regimens, we suggest infusional chemotherapy with dose-adjusted EPOCH plus

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rituximab. We recommend the addition of rituximab to combination chemotherapy. Although practice varies, we generally wait until the second cycle of chemotherapy to add rituximab in order to minimize tumor lysis[1]. Since, the recurrence rate of BL is very rare we could not find any data regarding any previous case reporting of the same. Thus, we hereby present the case reporting of recurrence of BL following treatment.

Case report

Here, we present the case of a 13-year-old-female with a complaint of swelling in the left side of parieto-occipital region for 2 months, nonpainful and gradually progressive. There is no h/o vomiting, trauma, cough, and SOB. There is no history of hypertension, diabetes, tuberculosis, and surgical intervention in the past. Normal bowel and bladder habits and is nonsmoker and does not consume alcohol. On examination, Glasgow coma scale: E4V5M6, pupil: B/L normal and reactive to light. Pulse: 82 beats/minute, BP: 110/70 mmHG, respiratory rate: 22 breaths per minute, temperature is afebrile, SPO_2: 98%. A respiratory examination is normal with no added sounds. The cardiovascular examination was normal.

On investigations

Seronegative, with all other investigations normal.

Treatment

Excision of a subganglion soft tissue tumor under general anesthesia. Operation theatre findings showed soft tissue of soft to firm inconsistency of size 4×3 cm in the left parieto-occipital region. The patient was admitted to the ward. All the preoperative investigations were done. At the time of discharge, the patient was hemodynamically stable and afebrile.

Advice at discharge

Daily dressing and suture removal after 7 days. Follow up in neurosurgery OPD after 10 days/SOS.

Histopathology

The excised tissue (as seen in Fig. 1) was sent for histopathological examination and following were the reports.

Sections examined reveals multiple bits of fibrocollagenous tissue showing diffuse proliferation of atypical blue looking cells arranged in loose sheets with fibrocollagenous tissue in between the sheets and also in the perivascular pattern (as seen in Fig. 2). Thin elongated vasculature some with hemangiopericytoma like appearance are observed in the sheets of these cells.

Individual cells are monotonous with round to oval nuclei, mild irregular nuclear membrane, inconspicuous nucleoli, coarse, and scant amount of eosinophilic cytoplasm. Some of the cells have clear cytoplasm. Interspersed atypical cells with vesicular chromatin are observed in between the dark looking cells. Occasional atypical mitotic figures and areas and hyalinization along with few foci of necrotic debris are seen. Some of the area shows thick walled blood vessels. Mild lymphocytic infiltration is observed is observed at the edge of some bits. The overall histological features (as seen in Fig. 2) are suggestive of small round blue cell tumor.

Figure 1. Excised part of the Burkitt lymphoma.

Course of the lymphoma after the surgical excision

After the first surgery, the patient underwent chemotherapy for 3 months. After 2 months of completion of his chemotherapy, the patient again presented with swelling in the scalp region (as seen in Fig. 3) which on histopathological examination diagnosed to be a BL.

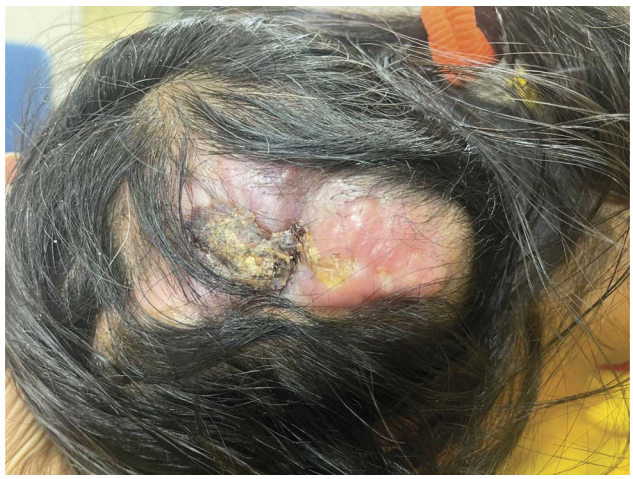

Figure 2. Histology showing the features of Burkitt lymphoma.

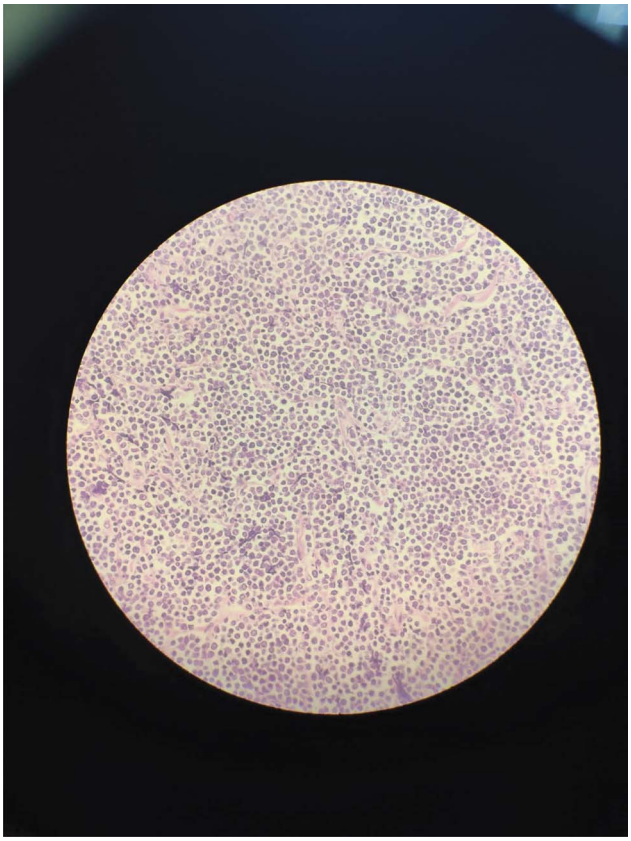

Figure 3. Current state of the scalp of the patient.

Discussion

We could not retrieve any case reporting on BL in the scalp region. However, there were many cases of BL in different regions of the body.

Giordano *et al*.[5] outlined the case of a 24-year-old Hispanic guy who presented with severe dysphagia that was becoming worse and was not getting better after receiving medical treatment in cycles. Upon closer examination, the mass revealed a sizable ulcerating mass involving the soft palate and right tonsillar area. During an incisional biopsy conducted under local anesthetic, BL was found to have inflammatory tissue, macrophages, and lymphoid cells scattered throughout, giving the impression of a starry sky. After the biopsy, HAART treatment was initiated to alleviate the symptoms.

Rector *et al*.[6] presented the case of a 70-year-old man who arrived in the emergency room with a subacute history of lower-extremity edema, nonspecific tiredness, and abdominal distension. In addition, he disclosed that he had recently had unintended weight loss of 20 pounds, constipation, and discomfort when eating. He denied having a temperature, chills, orthopnea, chest pain, nausea, or vomiting. Upon performing contrast-enhanced computed tomography (CT) of the abdomen and pelvis, a multinodular tumor was detected extending from the anterior pericardium to the right atrial and ventricular chambers. Under echocardiography and fluoroscopic guidance, the patient had a transluminal biopsy of the right ventricular tumor through the right jugular vein utilizing a 5.5F bioptome catheter. A high-grade B-cell lymphoma indicative of BL was observed. A bone marrow biopsy revealed no involvement of the marrow. The first round of chemotherapy included vincristine, doxorubicin, dexamethasone, and Rituximab while the patient was in the ICU and produced encouraging results. Hence, early chemotherapy showed promising results.

Bahashwan *et al*.[7] reported the case of a 65-year-old man who was diagnosed with benign prostatic hyperplasia and diabetes mellitus. The guy began experiencing erratic memory problems and a steady decline in awareness 4 days before to the presentation. The patient's vital signs were stable. The previously reported brain lesion underwent a mini-craniotomy and biopsy; the pathology report identified signs of encephalitis. A second CT scan revealed left supratentorial hydrocephalus along with a 3×1 cm increase in the extent of the left intraventricular lesion. Both an excisional biopsy and a craniotomy were performed on the left ventricular lesion. Histopathology revealed characteristics of a diffuse lymphocyte infiltration with a starry sky appearance. A ventriculoperitoneal shunt was inserted following surgery along with starting of corticosteroids. Chemotherapy was started with Rituximab, vincristine, and high-dose methotrexate. A brain CT scan on 10th day of treatment showed improvement.

Quimby *et al*.[8] highlighted the case of a 90-year-old Caucasian woman who had a big neck mass and a 6-week history of growing left-sided otalgia, stridor, and dysphonia. Examining revealed a hard 2×3 cm lump on the left side without any skin abnormalities overlaying it. Using flexible fiberoptic nasolaryngoscopy, a huge exophytic tumor completely blocked the airway was discovered. She was diagnosed with primary Hodgkin lymphoma in the left cervical node 8 years prior. After her presentation, she had a supraglottic tumor biopsy, laryngoscopy, and an emergency awake tracheostomy. Microscopic analysis revealed a starry sky with BL characteristics. Her palliative radiation treatment began with a dose of 3600 cGy divided into 18 fractions after she completed a 7-day course of high-dose prednisone (100 mg daily). She subsequently developed mucositis and was treated palliatively for symptom control. She had recurrent aspiration and developed bacteremia secondary to aspiration pneumonia for which she opted to forgo antibiotics.

Hosoda *et al*.[9] recounted a case of an 83-year-old man who had no symptoms about his abdomen but whose CT scan revealed an expansion of the periportal lymph node and a gallbladder tumor. The scan was done as part of a hypertension screening. The soluble interleukin 2 receptor and carcinoembryonic antigen levels were somewhat higher, according to the lab results. After undergoing periportal lymph node dissection, gallbladder bed excision, and bile duct resection, the patient's condition was finally determined to be Burkitt's lymphoma. After being sent to hemologists, he began taking vincristine, prednisone, doxorubicin, and cyclophosphamide at a lower dosage.

In our case, when the patient presented with the recurrent lesion over the scalp she was counseled with multidisciplinary approach including neurosurgeon, medical oncologist, radiologist, and pathologist. After the discussion the decision was made to initiate the chemotherapy by the medical oncologist.

Methods: All the work has been reported in line with the Surgical CAse REport (SCARE) 2023 Criteria[10].

Conclusion

As we conclude the above discussion the recurrence rate of BL is very low. Thus, here, we report a case of a 13-year-old female previously diagnosed to have BL. After surgical excision of the very tumor and undergoing chemotherapy, the patient again presented with the scalp swelling which on histopathology diagnosed to be the recurrence of the previous lesion.

Ethical approval

The ethics approval is not required for our case report.

Consent

Written informed consent was obtained from the patient's parents for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

A.B. and D.K.: research and writing; Dr A.D.: supervision and reviewing; Dr G.K.: reviewing.

Conflicts of interest disclosure

The author declares no conflicts of interest.

Research registration unique identifying number (UIN)

Not applicable.

Guarantor

Aashish Baniya.

Data availability statement

No data sheets were used to analyze the data as it is a case report.

Provenance and peer review

Externally peer-reviewed.

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