CASE REPORT

Imaging findings of childhood B-cell lymphoblastic lymphoma in the mental region: a case report

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Lymphoblastic lymphoma (LBL) is an uncommon neoplasm that accounts for about 5% of all non-Hodgkin's lymphomas. Approximately 90% of LBLs are of immature T-cell lineage, with B-cell lymphoblastic lymphoma (B-LBL) accounting for only about 10%. Owing to infrequency, the clinicopathological features and the imaging findings of B-LBL are not well defined. LBLs frequently affect extranodal sites. The most common site is the skin, followed by the bone, and the head and neck region is rarely involved. We report herein a case of childhood B-LBL occurring in the mental region, with emphasis on the findings of several imaging studies, including contrast-enhanced CT, MRI and fluorodeoxyglucose-positron emission tomography/CT.

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Introduction

Lymphoblastic lymphoma (LBL) is an uncommon neoplasm that accounts for about 5% of all non-Hodgkin's lymphomas (NHLs).¹ Histopathologically, LBL comprises immature lymphocytes derived from the B-cell lineage [B-cell lymphoblastic lymphoma (B-LBL)] or T-cell lineage (T-cell lymphoblastic lymphoma). B-LBL is the less common type, accounting for only 10% of all LBLs.² Clinically, LBL frequently affects extranodal sites. The site most commonly affected is the skin, followed by the bone.² The head and neck region is rarely involved. In particular, B-LBL involving the head and neck is extremely rare, and only seven cases have been reported since 2007.^{3–8} Radiologic imaging studies have characterized B-LBL as showing lytic or sclerotic changes that mimic benign or malignant primary bone lesions.² However, few studies have evaluated image findings from B-LBL in detail. We report herein a case of childhood B-LBL occurring in the

mental region, with emphasis on the findings of several imaging studies.

Case report

A 9-year-old female visited a private dental clinic with a chief complaint of swelling in the right side of the mandible and mobility of the first deciduous molar of the right mandible. Under a clinical diagnosis of inflammatory odontogenic process, the tooth was extracted and antibiotics were prescribed. Furthermore, the socket was periodically irrigated with iodine solution for 2 months. However, the swelling remained. Given this clinical history, she was referred to our hospital for further investigation and treatment.

On the first visit to our hospital, clinical examinations revealed facial asymmetry with an elastically hard, painless mass in the right mental region, measuring 32 × 22 mm ([Figure 1](#)). Intraorally, a socket 12 mm in depth was present at the site of the right mandibular first

Figure 1 A clinical examination is revealing facial asymmetry with reddening and swelling of the right mental region.

deciduous molar. Before histopathological examinations, imaging studies were performed, including panoramic radiography, CT, MRI and fluorine-18 fluorodeoxyglucose (FDG)-positron emission tomography (PET)/CT.

Panoramic imaging revealed a well-defined radiolucent area around the right mandibular first premolar (Figure 2), but no other areas of abnormal bone resorption. Contrast-enhanced CT showed a well-defined subcutaneous mass with homogeneous soft-tissue density in the right mental region, measuring 32×22 mm (Figure 3a). The surrounding subcutaneous fatty tissue was considered almost normal. Bone window CT showed an area of cortical bone resorption on the buccal side of the first premolar (Figure 3b). However, the relationship with the subcutaneous mass was uncertain. MRI showed a subcutaneous mass on the buccal side of the right mandible, measuring 32×22 mm. The mass was well defined and showed signal hypointensity on T_1 weighted imaging (Figure 4a), and homogeneous signal hyperintensity on T_2 weighted imaging with fat suppression (Figure 4b). Homogeneous enhancement was evident on post-contrast T_1 weighted imaging with fat suppression (Figure 4c). Dynamic contrast-enhanced MRI

Figure 2 A panoramic image is showing a defined radiolucent area around the right mandibular first premolar (arrow).

(DCE-MRI) revealed early enhancement with a low washout ratio pattern. The mass had a low apparent diffusion coefficient (ADC) of $0.43 \times 10^{-3} \text{ mm}^2 \text{ s}^{-1}$ on diffusion-weighted MRI (DWI). After CT and MRI, FDG-PET/CT was also performed, showing multiple sites of increased uptake, including the right mental region, submental lymph node, bone marrow of the spine, pelvis and femur (Figure 5). Findings from multiple imaging modalities, such as a well-defined homogeneous mass on CT and MRI, a low ADC on DWI and multiple sites of increased uptake on PET/CT, strongly suggested malignancy rather than inflammation, including the possibility of NHL. After imaging studies, biopsy was performed from the right buccal mucosa. Histological examination revealed a tuberous, diffuse proliferation of intermediate to large-sized abnormal lymphoblasts with a high nuclear cytoplasmic ratio, absent to inconspicuous nucleoli, irregular nuclear contours and abnormal mitosis. Immunohistochemically, the tumour cells expressed terminal deoxynucleotidyl transferase (TdT) and B-cell antigens such as CD10, CD79a, and Bcl-2, with a Ki67 proliferative index of 80%. Tumour cells were negative for CD3, CD5, CD20 and Bcl-1. Based on the histological findings and results of immunohistochemical staining, B-LBL was diagnosed and the patient was referred to the National Cancer Center for chemotherapy.

Figure 3 Contrast-enhanced CT: a well-defined, subcutaneous, soft-tissue density mass in the right mental region is evident (arrow) (a). Cortical bone resorption can be seen on the buccal side of the first premolar (arrow) (b).

Figure 4 Contrast-enhanced MRI: axial T_1 weighted imaging is showing a hypointense signal mass in the right mental region (arrow) (a). Axial T_2 weighted imaging is showing a homogeneous hyperintense signal mass (arrow) (b). Axial post-contrast T_1 weighted imaging is showing homogeneous enhancement of the mass (arrow) (c).

Discussion

LBL arises from precursor lymphocytes with a B- or T-cell lineage. Approximately 90% of LBLs are of immature T-cell lineage, with B-LBL accounting for only about 10%.² The clinicopathological features of B-LBL have not been well defined because of the varied presentations and low frequency. Lin *et al*² reported the clinicopathological features of 25 cases with B-LBL. According to that report, the median age of patients was 20 years (range, 5–68 years). Sites of primary disease included the skin ($n = 9$), bones ($n = 5$), soft tissues ($n = 4$), lymph nodes ($n = 3$), breast

($n = 2$), stomach ($n = 1$), and colon and mediastinum ($n = 1$). Our case involved the head and neck region, representing an extremely uncommon site. Among such reported cases, two involved the parotid gland and Waldeyer's ring,⁷ and the remaining five cases presented in the oral cavity (four in the mandible and one in the maxilla).

In this case, the disease was initially diagnosed and treated as inflammation of the odontogenic process, despite a lack of supporting laboratory signs. Other reported cases^{3,4} were also initially diagnosed as inflammation of odontogenic origin, thus delaying appropriate treatment. Several imaging modalities were used in the present case, including contrast-enhanced CT, MRI and FDG-PET/CT. On CT, the lesion was revealed as a well-defined, homogeneous, soft-tissue density mass in the right mental region. Although partial bone resorption was shown in the buccal cortex of the mandible, whether this bone change was caused by tooth extraction or tumour invasion was uncertain. On MRI, the mass showed signal hypointensity on T_1 weighted imaging and homogeneous signal hyperintensity on fat-suppressed T_2 weighted imaging. A homogeneous enhancement was evident on post-contrast fat-suppressed T_1 weighted imaging. We also performed DCE-MRI and DWI. The former revealed an early enhancement pattern, considered to be corresponding to a high microvessel count.⁹ The latter revealed a low ADC of $0.43 \times 10^{-3} \text{ mm}^2 \text{ s}^{-1}$, which might reflect the high cellularity of the tumour.^{10,11} To the best of our knowledge, imaging findings for B-LBL have not been well covered in the literature. However, NHLs in the head and neck region are known to demonstrate homogeneous density or signal intensity with few areas of necrosis on CT or MRI. The case of B-LBL in this report showed similar CT and MR findings. With regard to DCE-MRI and DWI, Yabuuchi *et al*¹² reported that NHLs of the salivary gland showed

Figure 5 An axial fusion image of fluorine-18 fluorodeoxyglucose-positron emission tomography/CT is showing a mass with a maximum standardized uptake value of 3.7 in the right mental region (arrow).

a dynamic curve of an early enhancement with a low washout pattern and low ADC ($0.88 \pm 0.07 \times 10^{-3} \text{ mm}^2 \text{ s}^{-1}$). Our case showed a similar dynamic curve and low ADC ($0.43 \times 10^{-3} \text{ mm}^2 \text{ s}^{-1}$), consistent with their study.

FDG-PET/CT showed multiple foci located in the right mental region, bone marrow of the spine, pelvis and femur, which was useful to exclude an inflammatory disease. In this case, the submental lymph node also showed increased uptake. Lin et al² also found such regional lymphadenopathy in 7 of 25 cases with primarily extranodal LBL.

LBLs are currently most effectively treated using acute lymphoblastic leukaemia-based therapies such as hyper-CVAD (fractionated cyclophosphamide, vincristine, doxorubicin, and dexamethasone).¹³ Oliveira et al¹⁴

reported a favourable prognosis of childhood LBL using an intensive chemotherapy regimen derived from acute lymphoblastic leukaemia therapy. Specifically, they reported that 22 (81%) of 27 patients were alive in first complete remission. They also reported disease stage as a major prognostic factor for children with precursor B-LBL, with significantly better overall survival and event-free survival rates observed in patients with Stage I–III disease, as compared with those with Stage IV disease.

In conclusion, we have reported a case of childhood B-LBL in the mental region, with attention to imaging findings. An early accurate diagnosis is important for better prognosis, and we therefore recommend radiologists include this rare tumour in the list of differential diagnoses for head and neck tumours.

References

- Kim JY, Kim YC, Lee ES. Precursor B-cell lymphoblastic lymphoma involving the skin. *J Cutan Pathol* 2006; **33**: 649–53. doi: <https://doi.org/10.1111/j.1600-0560.2006.00405.x>
- Lin P, Jones D, Dorfman DM, Mwdwiro LJ. Precursor B-cell lymphoblastic lymphoma: a predominantly extranodal tumor with low propensity for leukemic involvement. *Am J Surg Pathol* 2000; **24**: 1480–90. doi: <https://doi.org/10.1097/0000478-200011000-00003>
- Cox DP, Treseler P, Dong R, Jordan RC. Rare oral cavity presentation of a B-cell lymphoblastic lymphoma. A case report and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2007; **103**: 814–19. doi: <https://doi.org/10.1016/j.tripleo.2005.11.037>
- Cavalcante AS, Anbinder AL, Pontes EM, Carvalho YR. B-cell lymphoblastic lymphoma in the maxilla of a child: a rare case report. *Int J Oral Maxillofac Surg* 2009; **38**: 1326–30. doi: <https://doi.org/10.1016/j.ijom.2009.07.010>
- Sai S, Watanabe C, Okada S. A rare case of childhood precursor B-cell lymphoblastic lymphoma in the mandible. *Leuk Res* 2012; **36**: e37–8. doi: <https://doi.org/10.1016/j.leukres.2011.08.012>
- Kim HJ, Lee TJ, Choi YS. Primary B-lymphoblastic lymphoma of gallbladder involving mandibular bone. *Int J Hematol* 2014; **99**: 790–3. doi: <https://doi.org/10.1007/s12185-014-1582-2>
- Maitra A, McKenna RW, Weinberg AG, Schneider NR, Kroft SH. Precursor B-cell lymphoblastic lymphoma. A study of nine cases lacking blood and bone marrow involvement and review of the literature. *Am J Clin Pathol* 2001; **115**: 868–75. doi: <https://doi.org/10.1309/Q5GV-3K00-WAC6-BBUB>
- Iwamuro M, Kawai Y, Yamawaki Y, Takata K, Yamamoto K. Precursor B lymphoblastic lymphoma involving the stomach. *Case Rep Gastrointest Med* 2013; **2013**: 930918. doi: <https://doi.org/10.1155/2013/930918>
- Yabuuchi H, Fukuya T, Tajima T, Hachitanda Y, Tomita K, Koga M. Salivary gland tumors: diagnostic value of gadolinium-enhanced dynamic MR imaging with histopathologic correlation. *Radiology* 2003; **226**: 345–54. doi: <https://doi.org/10.1148/radiol.2262011486>
- Gupta RK, Cloughesy TF, Sinha U, Garakian J, Lazareff J, Rubino G, et al. Relationships between choline magnetic resonance spectroscopy, apparent diffusion coefficient and quantitative histopathology in human glioma. *J Neurooncol* 2000; **50**: 215–26. doi: <https://doi.org/10.1023/A:1006431120031>
- Guo AC, Cummings TJ, Dash RC, Provenzale JM. Lymphomas and high-grade astrocytomas: comparison of water diffusibility and histologic characteristics. *Radiology* 2002; **224**: 177–83. doi: <https://doi.org/10.1148/radiol.2241010637>
- Yabuuchi H, Matsuo Y, Kamitani T, Setoguchi T, Okafuji T, Soeda H, et al. Parotid gland tumors: can addition of diffusion-weighted MR imaging to dynamic contrast-enhanced MR imaging improve diagnostic accuracy in characterization? *Radiology* 2008; **249**: 909–16. doi: <https://doi.org/10.1148/radiol.2493072045>
- Kantarjian HM, O'Brien S, Smith TL, Cortes J, Giles FJ, Beran M, et al. Results of treatment with hyper-CVAD, a dose-intensive regimen, in adult acute lymphocytic leukemia. *J Clin Oncol* 2000; **18**: 547–61.
- Oliveira MC, Sampaio KC, Oliveira AC, Santos AD, Castro LP, Viana MB. Outcome of children and adolescents with lymphoblastic lymphoma. *Rev Assoc Med Bras (1992)* 2015; **61**: 417–22. doi: <https://doi.org/10.1590/1806-9282.61.05.417>