Imaging features of Burkitt lymphoma in pediatric patients

Burkitt lymphoma (BL) is an aggressive subtype of non-Hodgkin lymphoma (NHL) with a doubling time of 24–48 h. Even with its aggressive character, BL is curable and highly sensitive to chemotherapy. Thus, early diagnosis and treatment are critical before life-threatening complications, such as airway compromise or spinal cord compression, set in. Knowledge about the imaging appearances assists in rapid detection and treatment. In this pictorial review, we aimed to identify the most common imaging features of Burkitt lymphoma in pediatric patients.

Head and neck involvement

In the head and neck region, the predilection of BL for certain sites varies among different subtypes. In the endemic form, jaw and other facial bones are commonly affected by osteolytic lesions. On the other hand, Waldeyer ring (palatine and lingual tonsils, nasopharyngeal adenoids) and nodal involvement are commonly observed in the sporadic form (Fig. 1). The clinical presentation may differ as per the site involved. The clinical features of endemic BL involving the jaws include acute swelling in or around the jaws or orbit, severely mobile teeth, displaced teeth, generalized lymphadenopathy and anesthesia or paresthesia when adjacent nerves are involved; pain is not a prominent feature. On the other hand, in Waldeyer ring involvement, airway compression may cause symptoms such as dysphagia, dyspnea, wheezing, and airway obstruction, which may be potentially life threatening (Figs. 2, 3) (2).

Ultrasoundography (US) may be the initial modality for enlarged lymph nodes in the neck. On the other hand, computed tomography (CT) or magnetic resonance imaging (MRI) not only identifies a possible Waldeyer ring involvement or bone destruction according to the sub-
type, but also enables a precise definition of the radiologic extent of the disease for staging (Figs. 4, 5). CT is widely available and inexpensive with a short examination time and no need for sedation. CT is superior for the imaging of bone destruction; however, it has poor soft tissue contrast and potential hazard of radiation. In contrast, MRI provides very detailed diagnostic pictures of most important sites in the neck, with superior soft tissue contrast that could demonstrate tumor extension. MRI does not use ionizing radiation, and is therefore, suitable for use in children. In addition, MRI is superior in showing the signal characteristics of the bone marrow. For highly cellular BL, diffusion-weighted MRI is a powerful tool that demonstrates restricted diffusion (Fig. 4c). However, the disadvantages of MRI over CT are the cost, longer scan time and requirement for sedation in a child, which is not preferred if airway obstruction is present.

In the sporadic form of BL, Waldeyer ring is the most common site of extranodal disease. On MRI, soft tissue masses and enlarged pharyngeal or neck lymph nodes are commonly observed. The enlarged lymph nodes and soft tissue masses demonstrate homogenous enhancement without any central necrosis. Mass lesions do not have a tendency to cause skull base erosions in the sporadic form of BL but a tendency to fill in the adjacent spaces, such as the nasal cavity, nasopharynx, or oropharynx. On T2-weighted sequences, the signal of a mass is isointense to hypointense because of its high cellularity. For the same reason, diffusion-weighted images show restricted diffusion. After contrast administration, the mass and the enlarged lymph nodes enhance homogeneously. Because necrotic lymph nodes and bone destruction are prevalent in carcinomas of the head and neck, their absence supports the diagnosis of a sporadic form of BL in a child (6).

Unlike the sporadic form of BL, the endemic form commonly presents with bone destruction around the jaws and orbit. CT shows osteolytic lesions in the maxillofacial bones, particularly in the maxilla and mandible. When the mass grows, it enlarges into the adjacent areas, erodes the adjacent bones, and invades the adjacent spaces (Fig. 6) (2, 4, 6).

Brain involvement is rare in BL, particularly in the sporadic form. However, in immunodeficiency-related type of BL, involvement of the central nervous system and calvarial bone marrow may be observed (Fig. 7a–7c) (2, 4).

**Main points**

- The predilection of Burkitt lymphoma for certain sites differs among different subtypes.
- In the endemic variant, head and neck involvement is more common; whereas abdominal involvement is commonly observed in the sporadic form.
- In the head and neck region, jaw and the other facial bones are commonly involved in the endemic form. Waldeyer ring (palatine and lingual tonsils, nasopharyngeal adenoid) and nodal involvement are commonly seen in the sporadic form.
- In the abdomen, ileocecal region (the Peyer’s patches in particular) is the most common site of involvement, as characterized by an asymmetric bowel wall thickening and aneurysmal dilatation of the bowel lumen.

**Abdominal involvement**

The abdominal and pelvic region, including the gastrointestinal tract, mesentery, peritoneum, retroperitoneum, and genitourinary tract, is commonly affected by BL, particularly by its sporadic form (7, 8). The ileocecal region, specifically the Peyer’s patches, is the most common site involved. The clinical presentation may differ according to the site involved. Mainly, obstruction or compression may cause various symptoms, such as palpable mass, abdominal pain, nausea, vomiting, distension, constipation, or intestinal obstruction (5, 9).
The initial imaging modality for abdominal BL may be US for palpable masses, but CT is an important modality to evaluate the severity and spread of the disease for tumor staging. Abdominal CT should be performed with oral and intravenous contrast agents to evaluate the bowel walls, as well as solid organs, retroperitoneum, and the mesenteric region. Recently, positron emission tomography (PET)/CT has been used for functional and anatomic assessment during tumor staging and follow-up after treatment (10, 11).

Along the gastrointestinal tract, ileum, cecum, and appendix are common sites of involvement, because the terminal ileum has the highest concentration of lymphatic tissue; colonic and gastric involvement...
is quite rare (Fig. 8). Although BL does not cause bowel obstruction due to lack of fibrosis or desmoplasia unlike that in gastrointestinal adenocarcinomas, it may trigger intussusception and lead to bowel obstruction (Fig. 9) (12).

Imaging findings on US include asymmetric bowel wall thickening, solid hypoechoic mass lesion, ascites, or lymphadenopathies. If intussusception occurs, the pseudo-kidney sign on longitudinal plane and the target sign on transverse plane appear on sonogram. On CT, asymmetric bowel wall thickening and aneurysmal dilatation of the bowel lumen are observed more clearly. Aneurysmal dilatation is thought to be a consequence of infiltration of the myenteric plexus (Fig. 8c). In addition to these local findings, CT may detect ascites, peritoneal and mesenteric thickening, discrete or conglomerated masses, widespread lymphadenopathies, and tumor involvement of other solid organs in the abdomen, which leads to upstaging of the disease (Fig. 10). In BL, ascitic fluid can accumulate as an exudate, which has a slightly higher density on CT, owing to its high protein content (Fig. 7d) (5, 9–11).

The typical mesenteric and retroperitoneal growth patterns in BL are infiltrative in nature. The involvement of mesenteric and retroperitoneal lymph nodes is observed as single or multiple abdominal or pelvic masses. Encasement of the major mesenteric vessels is a common feature (Fig. 10). Calcification and necrosis of the mesenteric and retroperitoneal masses may be observed in BL (5).

Although primary involvement of solid organs is rare, secondary involvement of the liver, spleen, or kidney is not uncommon. Radiologic findings include or-
ganomegaly and single or multiple focal hypoechoic (on US) or hypodense (on CT) masses (Fig. 11). Infiltrating masses in the periportal area of the liver or in the perihilar area of the kidney may be observed and cause biliary obstruction or ureteropelvic obstruction, respectively (Fig. 10) (5, 8, 10).

Testicular and ovarian involvement in BL has been reported. For testicular involvement, US is the primary imaging modality, which shows diffuse enlargement, parenchymal hypoechoigenicity-heterogeneity or focal hypoechoic mass in the testicular parenchyma. Ovarian involvement may be cystic or solid, or mixed, as a focal or large mass (5, 8).

Aside from abdominopelvic and head-neck involvement, BL may seldom be located primarily in the pleura, breast, soft tissues, skin, thyroid, pancreas, lung, or spinal extradural area (Fig. 12). In disseminated disease, these tissues may be secondarily involved (Fig. 11b).

Conclusion

As an aggressive tumor, BL has to be rapidly detected among radiologic abnormalities and diagnosed, so that treatment can be initiated immediately. Moreover, diagnosis of BL requires a thorough radiologic investigation for accurate staging of the disseminated disease. Awareness of the most common imaging appearances and sites of involvement leads to rapid detection and correct diagnosis.

Burkitt lymphoma in pediatric patients
Conflict of interest disclosure
The authors declared no conflicts of interest.

References