Kulak Burun Boğaz Uygulamaları 2014;2(3):135-138
doi: 10.5606/kbbu.2014.03522

Case Report / *Olgu Sunumu*

Burkitt's lymphoma of the palatine tonsil

Palatin tonsil kaynaklı Burkitt lenfoma

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Burkitt's lymphoma is an aggressive form of non-Hodgkin B-cell lymphoma. While Burkitt's lymphomas commonly originate in the abdominal region, some involve the head and neck region. Palatine tonsil is a very rare location for Burkitt's lymphoma. Two common findings in Burkitt's lymphoma are unilateral tonsil hypertrophy and color alteration in the palatine tonsil. Tonsillectomy is used for diagnosis while chemotherapy is used for treatment.

Keywords: Burkitt's lymphoma; head and neck neoplasms; palatine tonsil.

Burkitt lenfoma, B hücreli Hodgkin dışı lenfomanın agresif bir formudur. Burkitt lenfomalar yaygın olarak abdominal bölgeden kaynaklanırken bazıları baş ve boyun bölgesini tutar. Palatin tonsil Burkitt lenfoma için nadir bir yerdir. Burkitt lenfomada en yaygın iki bulgu tek taraflı tonsil hipertrofisi ve palatin tonsilde renk değişikliğidir. Tanıda tonsillektomi kullanılırken tedavide kemoterapi kullanılır.

Anahtar sözcükler: Burkitt lenfoma; baş boyun neoplazmları; palatin tonsil.

Burkitt's lymphoma was first described by Denis Burkitt in 1958 while he was working as a surgeon in Uganda. Burkitt's lymphoma is an aggressive form of non-Hodgkin B-cell lymphoma mostly seen in childhood.[1] Lymphomas are the most malignant neoplasms of the head and neck in children. Of the lymphomas the non-Hodgkin lymphoma is predominant.[2]

There are three subtypes of Burkitt's lymphoma; endemic (found in Africa), sporadic (found in North America and Europe) and immunodeficiency related.[3] The endemic (African) subtype of Burkitt's lymphoma is mostly seen between ages 5-7 and more than 50% of cases involve the maxilla or mandible. The sporadic (non-African) form of the disease may be seen in adults and at the age of 10-12 in children. The sporadic form of the disease presents with abdominal, medullary or lymphatic involvement rather then head and neck involvement. Only 5% of the cases involve Waldeyer's ring as an initial presentation.[4]

The clinical presentation includes unilateral palatine tonsil enlargement, alterations in the color and vascularization of the palatine tonsil, cervical lymphadenopathy, dysphagia, snoring and difficulty in speaking due to enlargement of the tonsil, weight loss, night sweating and weight loss.[5]

We present a rare case of Burkitt's lymphoma originating from the palatine tonsil in a child. To the best of our knowledge, this case is the first reported from Turkey and from Asia.

CASE REPORT

A five-year-old boy was referred to our clinic for recurrent tonsillitis of 7-8 times in a year for one year, progressive snoring and mouth breathing, weight loss and night sweating. Routine ear nose and throat (ENT) examination revealed left tonsil hypertrophy (grade 4) and alteration in the color of the palatine tonsil

Received: March 31, 2014 *Accepted:* May 26, 2014

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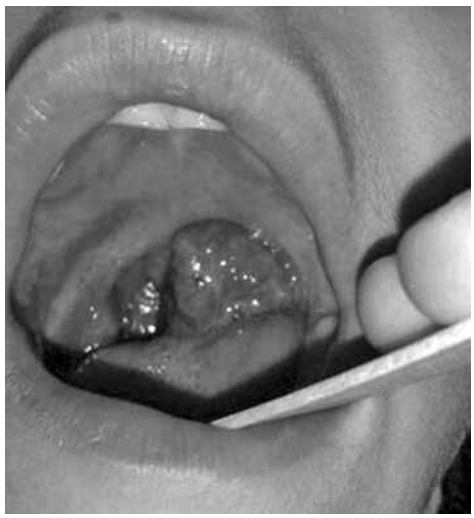

Figure 1. Note the left tonsil hypertrophy and alteration in the color of the palatine tonsil.

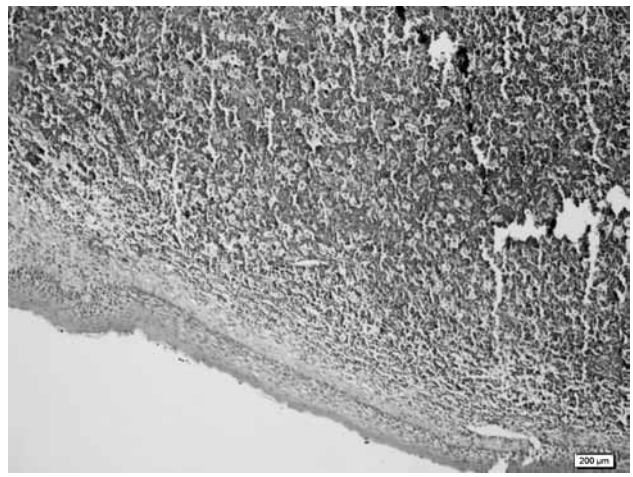

Figure 2. Diffuse and monotonous pattern of tumor infiltration in tonsil (H-E x 100).

(Figure 1). The right palatine tonsil was normal. Patient and family history were negative for systemic disease, medication use, radiation exposure and surgery. Routine blood examination of the patient was also insignificant. He underwent bilateral tonsillectomy for left tonsil hypertrophy under general anesthesia. The pathologic specimen was signed out as Burkitt's lymphoma of the left palatine tonsil.

The histopathologic examination read: "*The lymph node is involved with Burkitt lymphoma cells. The tumor cells are medium sized cells and a diffuse monotonous pattern of growth. The cells appear to be cohesive but some of them exhibit squared-off borders of retracted cytoplasm. The nuclei are round with finely clumped and dispersed chromatin, with multiple basophilic medium sized, paracentrally situated nucleoli. The cytoplasm is deeply basophilic and some of them contain lipid vacuoles. The tumor has an extremely high proliferation activity. A 'starry sky' pattern is present, which is imparted by numerous benign macrophages that have ingested apopitotic tumor cells*" (Figure 2-7).

The patient was referred to the pediatric oncology service for further management of the disease.

DISCUSSION

Most sporadic (non endemic) forms of Burkitt's lymphoma present with abdominal involvement. The distal ileum, cecum, stomach, kidney, testis and ovaries are regions where Burkitt's

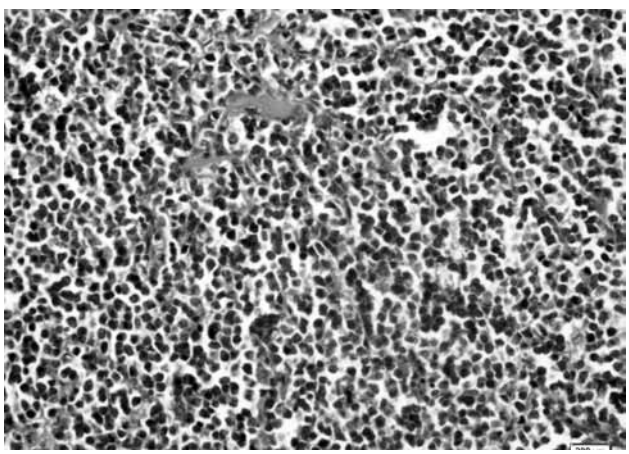

Figure 3. Burkitt's lymphoma cells are medium-sized, the nuclei are round with finely clumped and dispersed chromatin (H-E x 200).

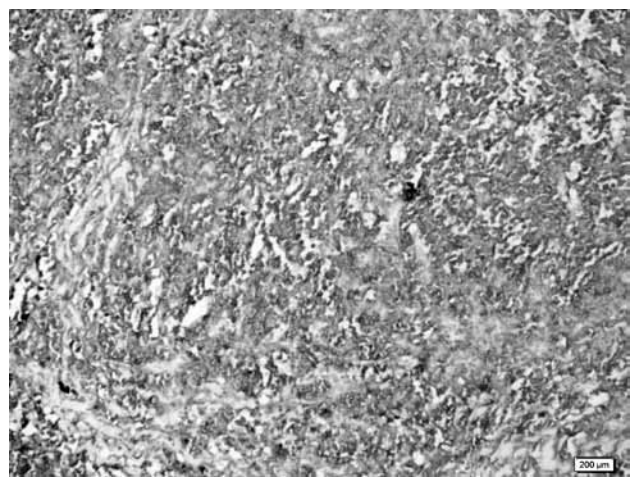

Figure 4. Tumor cells are showing strong membranous and cytoplasmic positivity with CD43 (x200).

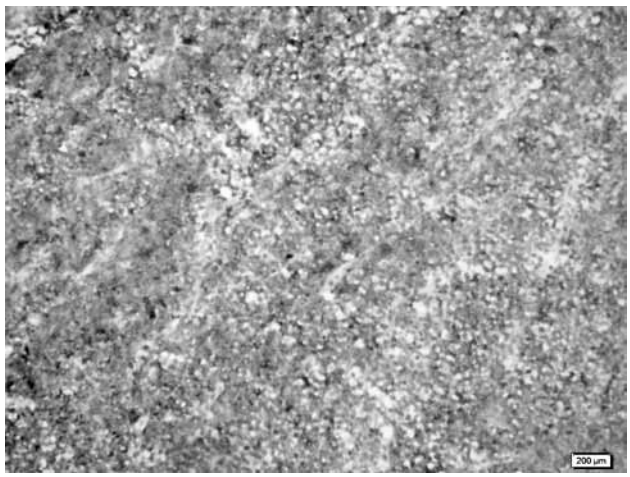

Figure 5. Tumor cells are showing strong cytoplasmic positivity with CD10 (x200).

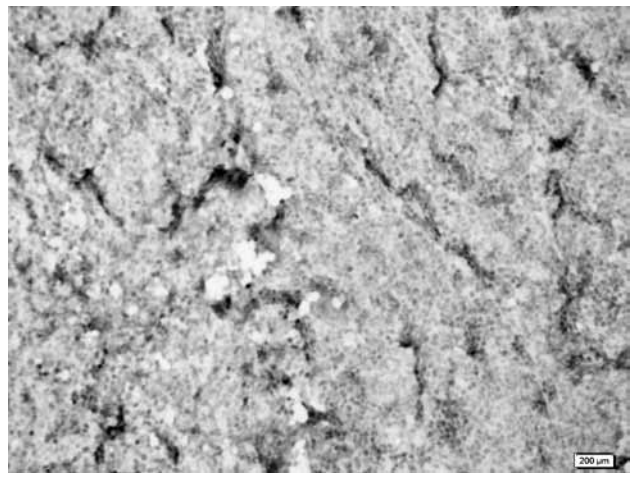

Figure 6. Tumor cells are showing strong nuclear positivity with Bcl-6 (x200).

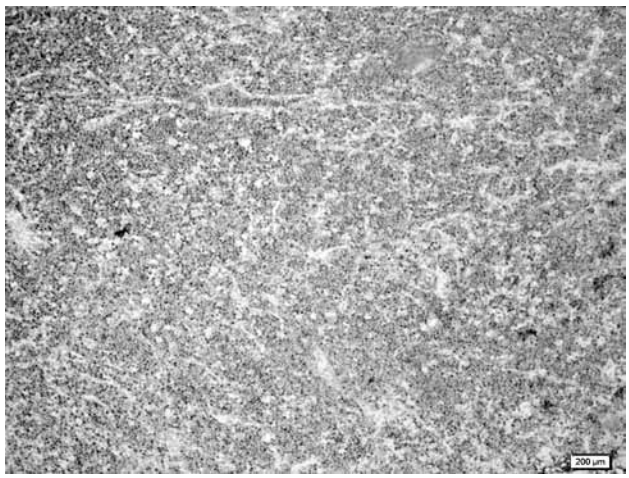

Figure 7. Immunohistochemical staining with Ki-67 (x200). Tumor cells have high proliferation index.

lymphoma usually originates.[4] Lymphadenopathy is the classic form of head and neck involvement. 4-25% of patients with head and neck Burkitt's lymphoma present with extranodal involvement of the nasopharynx, facial bone, palatine tonsil and lingual tonsil. Palatine tonsil involvement is reported at 2.9% of head and neck Burkitt lymphoma.[6] The endemic (African) form of Burkitt lymphoma originates from facial bones in 50-60% of patients.[7]

To the best of our knowledge, 33 cases have been reported in the English literature from 1996 to 2012. Our case is the 34th in the literature and first case reported from Turkey and from Asia according to our search of PubMed/Medline, LILACS, IBECS, Cochrane, SCIELO, BIREME and Scopus. Unilateral tonsil hypertrophy, alteration in the color of the palatine tonsil and cervical lymphadenopathy are the usual findings on routine ENT examination.[5] The patient in this paper presented with snoring, mouth breathing and recurrent tonsil infections. Unilateral tonsil hypertrophy, alteration in the color of the palatine tonsil and cervical lymphadenopathy were detected on the ENT examination.

The prognosis of Burkitt's lymphoma is related to early diagnosis and treatment. Although recurrent tonsillitis and tonsil hypertrophy may be frequently seen in childhood, the possibility of malignancy should always be kept in mind especially in the presence of unilateral tonsil hypertrophy and color alteration of the palatine tonsil.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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