Primary posterior mediastinal Burkitt lymphoma

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A 14-year-old boy admitted to our clinic with a 20-day history of fever, cough and respiratory distress. Mediastinal enlargement was observed on chest radiograph. Computed tomography of the chest demonstrated a large posterior mediastinal mass. The histopathological examination of the mass revealed Burkitt's lymphoma. We report this case because primary posterior mediastinal involvement in Burkitt lymphoma is extremely rare in childhood.

Key words: Burkitt lymphoma, posterior mediastinum, childhood.

Lymphoma accounts for approximately 8–10% of childhood cancer in developed countries[1], while it is more common in Turkey and constitutes 20% of childhood malignancy[2]. The most common type of lymphoma in children and adolescents is non-Hodgkin lymphoma (NHL), which accounts for approximately 60% of all lymphomas[1]. NHL has four major pathological subtypes, and Burkitt lymphoma (BL) accounts for approximately 40% of NHLs[1]. Sporadic BL commonly presents with abdominal involvement while endemic BL commonly presents with head and neck involvement[1]. Primary BL of the posterior mediastinum is extremely rare in children. Only one case was reported in the medical literature[3]. In this report, a case with primary posterior mediastinal BL is presented.

Case Report

A 14-year-old boy was referred to our hospital with complaints of fever, cough and respiratory distress. Tachypnea, nasal flaring, dyspnea, and intercostal retraction were noted on his physical examination. Other system examinations were normal. The laboratory assessment revealed hemoglobin: 12.1 g/dl, white blood cells: 11.2 x 10^3/uL with normal differential count, thrombocytes: 76 x $10^3/\mu$L, erythrocyte sedimentation rate: 2 mm/h, lactate dehydrogenase (LDH): 4794 U/L, creatinine: 0.4 mg/dl, and uric acid: 5.6 mg/dl. Mediastinal enlargement was observed on chest radiograph. Vanillylmandelic acid and alpha fetoprotein levels were normal. A contrast-enhanced computed tomography of the chest revealed a 160x70x100 mm mass located in the posterior mediastinum that surrounded the esophagus and pushed the heart forward (Fig. 1). A biopsy was taken from the mediastinal mass. The histopathological examination revealed diffuse infiltration of the medium-sized lymphoblastic cells with small, round, noncleaved nuclei and deeply basophilic cytoplasm containing lipid vacuoles. Characteristic starry sky appearance was also seen. Numerous mitoses were present. Immunohistochemical staining was positive for CD20 (Fig. 2). The pathological diagnosis was BL. Chromosomal analysis was not studied due to the lack of the technical facilities. Twenty percent L3-type blasts were determined on bone marrow examination. Flow cytometric

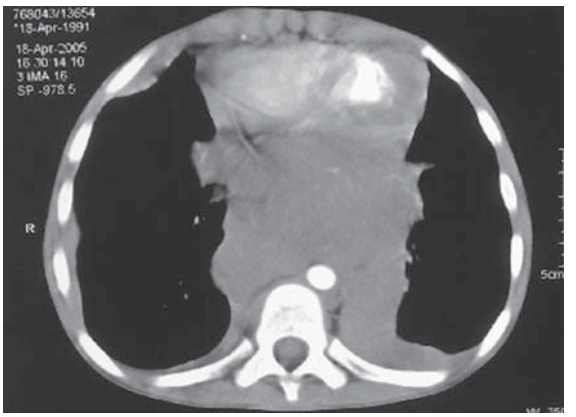

Fig. 1. Chest CT shows a homogeneous mass in the posterior mediastinum.

analysis was positive for CD20 and negative for CD3, which was consistent with B-cell lymphoma. Cerebrospinal fluid (CSF) studies were negative for lymphoma. The patient had stage IV disease. French Society of Pediatric Oncology LMB-B chemotherapy protocol was started. Recurrence has not been observed for five years.

Discussion

Lymphoma is the second most common malignant tumor in childhood and it constitutes 20% of childhood malignancy in our country[2]. NHL constitutes 10-18% of all mediastinal tumors in childhood and these tumors generally arise from the anterior and/or middle mediastinum[4-6]. Lymphoblastic and diffuse large B-cell lymphomas are the most common types of NHL originating from the mediastinum[1].

Burkitt's lymphoma (BL) constitutes approximately 40% of all childhood NHL[1]. The abdomen, head and neck are the most commonly involved areas in BL. BL rarely arises from the mediastinum[1]. Twenty to thirty percent of all mediastinal tumors are located in the posterior mediastinum[7]. The posterior mediastinum is an extremely uncommon location not only for BL but also for other histological subtypes of NHL. Neurogenic tumors such as ganglioneuroma and neuroblastoma are the most common posterior mediastinal tumors. The remaining posterior mediastinal tumors are constituted from a heterogeneous group of tumors including rhabdomyosarcoma and Ewing sarcoma[4-6]. Our case is interesting in view of its posterior mediastinal presentation. Lymphoma is malignant monoclonal proliferation of cells of the lymphocytic lineage present in lymph nodes and lymphoid tissue. Posterior mediastinal lymphoma may originate from paraesophageal lymph nodes lying adjacent to the wall of the esophagus or may arise from paravertebral lymph nodes along the nerve root and extending through the posterior mediastinum[1].

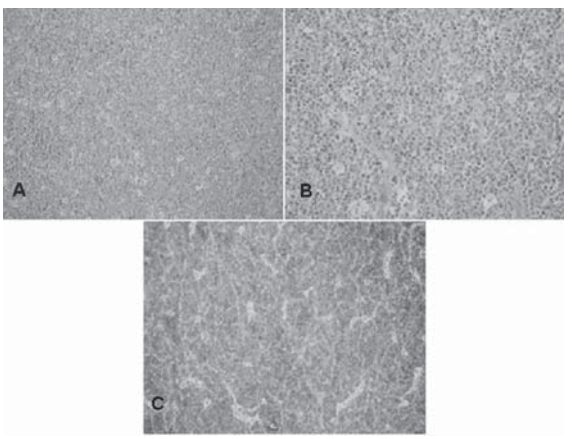

Fig. 2. Burkitt lymphoma arising from the posterior mediastinum. **A:** Diffuse infiltrate of atypical lymphoid cells with a prominent starry-sky appearance (hematoxylin & eosin [HE] X200). **B:** Same section as in Fig. A. (HE X400). **C:** Uniform CD20 immunoreactivity in Burkitt lymphoma.

We could find only a few reported cases with lymphoma arising primarily from the posterior mediastinum[3,8-12]. Most cases were Japanese adult patients and almost all of them had diffuse large B-cell lymphoma[9-12]. Franssila et al.[8] reported that 7% of BL cases in Finland had initial mediastinal involvement. However, we could find only one case report on posterior mediastinal BL in the English literature. In this case, the tumor was associated with bilateral cavernous sinus involvement[3]. To our knowledge, our case is the second BL case arising from the posterior mediastinum. Since a very limited number of cases with posterior mediastinal BL exist in the literature, the clinical and prognostic importance of this location is controversial. However, our patient completely responded to treatment and is alive after a five-year follow-up period.

Tumors located in the posterior mediastinum can reach a large diameter before being diagnosed. Symptoms of posterior mediastinal tumors are usually due to compression or direct invasion of the surrounding structures. The most common presenting symptoms are respiratory symptoms due to enlargement toward the trachea and neurologic symptoms due to spinal cord compression. Our case presented with respiratory distress and did not have any neurological sign or symptoms.

In conclusion, although lymphoma is rarely localized in the posterior mediastinum, it should be considered in the differential diagnosis of posterior mediastinal masses.

REFERENCES

1. Link MP, Weinstein HJ. Malignant non-Hodgkin lymphomas in children. In: Pizzo PA, Poplack DG (eds). Principles and Practice of Pediatric Oncology (5th ed). Philadelphia: Lippicott; 2006: 722-747.
2. Kutluk T, Yeşilipek A. On behalf of Turkish Pediatric Oncology Group and Turkish Pediatric Hematology Society Pediatric Tumor Registry for 2002-2005 in Turkey. J Pediatr Hematol Oncol 2007; 29: 12-13.
3. Huisman TA, Tschirch F, Schneider JF, Niggli F, Martin-Fiori E, Willi UV. Burkitt's lymphoma with bilateral cavernous sinus and mediastinal involvement in a child. Pediatr Radiol 2003; 33: 719-721.
4. Tansel T, Onursal E, Dayloğlu E, et al. Childhood mediastinal masses in infants and children. Turk J Pediatr 2006; 48: 8-12.
5. Sairanen H, Leijala M, Louhimo I. Primary mediastinal tumors in children. Eur J Cardiothorac Surg 1987; 1: 148-151.
6. Grosfeld JL, Skinner MA, Rescorla FJ, West KW, Scherer LR 3rd. Mediastinal tumors in children: experience with 196 cases. Ann Surg Oncol 1994; 1: 121-127.
7. Abdel Rahman AR, Sedera MA, Mourad IA, Aziz SA, Saber TK, Alsakary MA. Posterior mediastinal tumors: outcome of surgery. J Egypt Natl Canc Inst 2005; 17: 1-8.
8. Franssila KO, Heiskala MK, Rapola J. Non-Hodgkin's lymphomas in childhood. A clinicopathologic and epidemiologic study in Finland. Cancer 1987; 15: 1837-1846.
9. Nagashima O, Takahashi K, Sato K, et al. Case of malignant lymphoma arising from the posterior mediastinum, with pleural effusion. Nihon Kokyuki Gakkai Zasshi 2004; 42: 772-776.
10. Kawachi Y, Watanabe A, Nishihara T, Uchida T, Setsu K, Mori M. Chylothorax in a patient with 9-year remission of malignant lymphoma. Rinsho Ketsueki 1995; 36: 1311-1315.
11. Takamizawa A, Koizumi T, Fujimoto K, et al. Primary malignant lymphoma in the posterior mediastinum. Respiration 2004; 71: 417-420.
12. Yamanouchi J, Okada T, Hasegawa A, et al. Acute mitral regurgitation caused by malignant lymphoma of the posterior mediastinum. Rinsho Ketsueki 1998; 39: 606-610.