

Mediastinal Hodgkin lymphoma arising from cystic lymphangioma: case report in a child

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SUMMARY: Büyükkapu-Bay S, Çorapcıoğlu F, Gürkan B, Erçin MC, Anık Y, Karadoğan M. Mediastinal Hodgkin lymphoma arising from cystic lymphangioma: case report in a child. Turk J Pediatr 2012; 54: 298-300.

Mediastinal disease is a frequent clinical presentation in children with Hodgkin lymphoma. It is usually due to mediastinal lymphadenopathy or involvement of the thymus gland. Cystic lymphangiomas are benign tumors of the lymphatic system, and less than 1% present as a solitary mediastinal mass. To our knowledge, there has been no report in the literature describing Hodgkin lymphoma arising from cystic lymphangioma. In this report, we describe a patient with mediastinal cystic lymphangioma, from which Hodgkin lymphoma was determined to have arisen.

Key words: Hodgkin lymphoma, cystic lymphangioma, mediastinum, child.

Mediastinal disease is a frequent clinical presentation in children with Hodgkin lymphoma (HL). It is usually due to mediastinal lymphadenopathy or involvement of the thymus gland and rarely to thymic cysts¹. To our knowledge, there has been no report in the literature describing HL arising from cystic lymphangioma. This is the first case in childhood with mediastinal HL arising from cystic lymphangioma.

Case Report

A 16-year-old adolescent male was admitted to our center with a two-week history of chest pain during deep inspiration and night sweat. His physical examination showed no remarkable findings. Initial investigations of the patient had revealed right-sided hilar and mediastinal mass on the chest X-ray (CXR). Hematological parameters, erythrocyte sedimentation rate and biochemical investigations were all within the normal range. Computed tomography (CT) of the thorax showed anterior and middle mediastinal cystic necrotic content and conglomerate lymphadenopathy measuring 5x9 cm. Abdominal ultrasonography and CT were normal. For diagnosis, an open thoracic biopsy was performed from the mass.

Macroscopically, the excised mass measured 2x1.5x1 cm. The external surface was irregular. On cut section, the specimen was white in color and soft in consistency with empty cyst appearance. Sections from the solid and cystic areas showed malignant neoplasm, which stained positively for CD15 and CD30 + typical and lacunar type Reed-Sternberg (RS) cells in dense fibrotic connective tissue. Nodular sclerosing HL infiltration consisting of RS cells in lymphoid-rich stroma was observed within the cystic structure (Fig. 1). The cyst wall lined by non-ciliated, single-layer cubic epithelium and cyst lumen was filled with clear proteinaceous fluid containing lymphocytes and homogeneous eosinophilic materials. There was no necrotizing change, cavitation or solitary lesion. Based on the typical morphological features, the case was diagnosed as HL in the wall of mediastinal cystic lymphangioma.

Because of the lymphangioma findings, diagnostic magnetic resonance imaging (MRI) of the thorax was performed in the postoperative period, which showed the mass lesion as high signal intensity due to contrast enhancement, while the lymphangioma component was seen as hypointense due to cystic content (Fig. 2).

Baseline positron emission tomography (PET) scan showed signs of increased uptake of

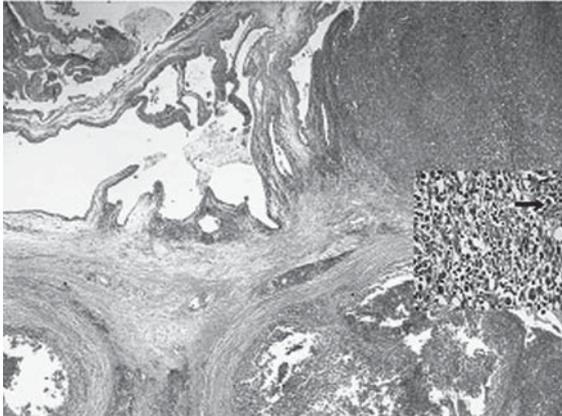


Fig. 1. Cystic lymphangioma is seen with homogeneous eosinophilic materials and lymphocytes in the lumen of cystic structures and Reed-Sternberg (RS) cells (arrow, hematoxylin-eosin stain [H&E], original magnification x400) in dense fibrotic connective tissue (H&E stain, original magnification x20).

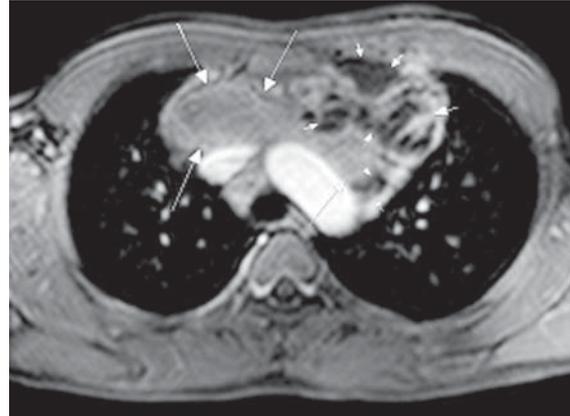


Fig. 2. Before treatment, thorax MRI in axial T1-weighted fat-suppressed contrast-enhanced image. The mass lesion is seen as high signal intensity due to contrast enhancement (long arrows), while the lymphangioma component is seen as hypointense due to cystic content (small arrows).

18-fluorodeoxyglucose in the region of the anterior mediastinal lymphadenopathy. He was staged as IIB HL, and four courses of ABVD (doxorubicin, bleomycin, vinblastine and dacarbazine) combination chemotherapy and involved-field radiotherapy were delivered. After completion of treatment, repeated thoracic MR showed total regression of the solid mass lesion, and the lymphangioma component was seen as a hypointense multiloculated lesion (small arrows) (Fig. 3); the PET scan showed no uptake of 18-fluorodeoxyglucose. He has been well for 19 months since completion of the radiotherapy and is disease-free.

Discussion

Cystic lymphangiomas are benign tumors of the lymphatic system. They usually occur in the neck (75%) or axillary region (20%), and less than 1% present as a solitary mediastinal mass. In the thorax, they represent 0.7% of all mediastinal tumors in adults². Mediastinal lymphangiomas are usually underdiagnosed because of lack of symptoms. Symptomatic cases are presented due to an infection, hemorrhage or compression of adjacent structures⁴. Malignant transformation of cystic lymphangiomas has not been described^{4,5}.

Combination of CT and MRI can be helpful in characterizing the cystic contents and anatomic relationships of the cysts. However, MRI delineates the tumor lesion extension

more clearly than CT scans³. On MRI, cystic lymphangioma is typically seen as a mass of low T1- and high T2-weighted signal intensity, with variable enhancement from septae⁷. In our patient, we repeated the thoracic MRI after completion of treatment, and the lymphangioma component was seen to be the same component and size as in pretreatment.

In 1983, Smith et al.⁶ reported a case in which the microscopic focus of HL was discovered in the wall of a large thymic cyst. However, there has been no report in the literature describing

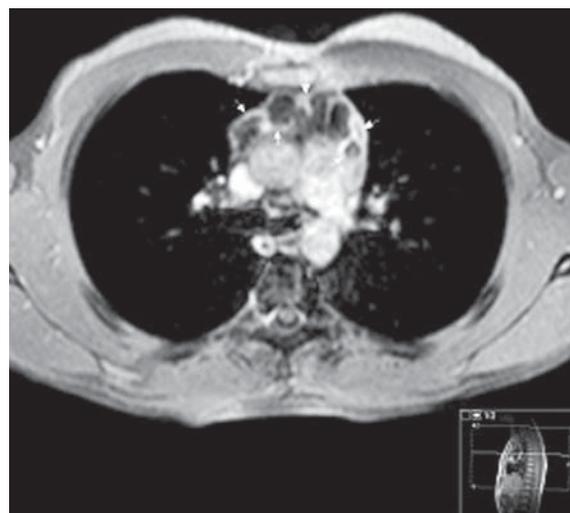


Fig. 3. After treatment, axial T1-weighted fat-suppressed contrast-enhanced thorax MRI shows lymphangioma component as a hypointense multiloculated lesion (small arrows).

HL in cystic lymphangioma. Here, we describe a patient in whom HL was identified in the wall of the cystic lymphangioma. In our case, the mediastinal mass was not excised completely, so we could not differentiate whether the HL originated primarily from the wall of the cystic lymphangioma or was only invasion. However, the final histopathological diagnosis was HL arising from cystic lymphangioma.

If MRI is performed in the cases without mediastinal biopsy who were accepted as undefined complete response after treatment for HL and PET-negative (especially nodular sclerosing HL subtype), we wonder about the probability of the diagnosis of cystic lymphangioma of the residual tissues without viable tumor.

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