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Case Report

NK/T cell lymphoma involving mediastinum: report of a case and review of literature

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Abstract: Extranodal natural killer (NK)/T-cell lymphoma is a very aggressive malignant neoplasia with a poor prognosis. Herein we reported a case of NK/T cell lymphoma involving mediastinum. It was a 28-year-old Chinese male patient. The tumor cells were medium-sized, had irregularly folded nuclei, and inconspicuous or small nucleoli with coagulative necrosis. The tumor cells were positive for CD3ε, TIA-1, but negative for CD56. In situ hybridization revealed that tumor cells also expressed Epstein-Barr virus encoded RNA. To our knowledge, this is the first case of NK/T cell lymphoma involving mediastinum.

Keywords: NK/T cell lymphoma, mediastinum, Epstein-Barr virus

Introduction

Extranodal natural killer (NK)/T-cell lymphoma is a very aggressive malignant tumor that presents a poor prognosis and limited survival rates [1, 2]. It is characterized by vascular damage and destruction, prominent necrosis, cytotoxic phenotype and association with Epstein-Barr virus (EBV) [3]. In the mediastinum, malignant lymphoma may be primary or secondary. Mediastinal lymphoma arises in the mediastinal lymph nodes or in the thymus. The most common lymphoma in mediastinum is diffuse large B cell lymphoma, followed by MALT and T cell lymphoma. In spite of the importance of the thymus in T-cell ontogeny, mature T-cell neoplasms are very rare in the thymus.

Extranodal NK/T-cell lymphoma almost always shows an extranodal presentation. The upper aerodigestive tracts, including nasal cavity, nasopharynx, paranasal sinuses, and palate, are most commonly involved, with the nasal cavity being the most common site of involvement. Preferential sites of extranasal involvement include the skin, soft tissue, gastrointestinal tract and testis.

Case presentation

Here is a case of extranodal nasal NK/T-cell lymphoma that involved the mediastinum. A 28-year-old Chinese male patient was diagnosed as epididymitis with fever, cough, and dyspnea. He was treated with the antibiotic for a week at the local hospital, but the fever couldn't be controlled. Then He was transferred to Xiangya Hospital. In physical examination, his epididymis and testis showed mildly swelling. Computerized tomography (CT) scan showed that there was an oval tumor in the mediastinum, which size was about 95 mm * 65 mm. There were a lot of high-density shadows in the double lungs accompanied with the pleural and pericardial effusion and ascites (**Figure 1**). Type B ultrasonic inspection also showed a lot of ascites and pleural fluids. There was no obvious change in nasal cavity. The mediastinal mass, instead of epididymis, was chosen for biopsy because of its huge size.

Fine needle specimen was obtained under the guide of CT. The lymphoma was composed of medium-sized cells. The tumor cells often had irregularly folded nuclei, which could be elongated. Nucleoli were generally inconspicuous or

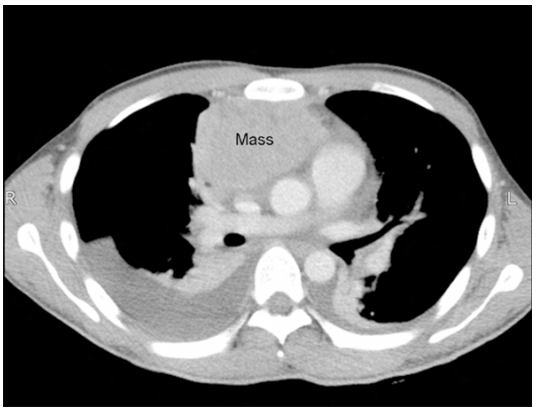

Figure 1. CT scan of mediastinal structure. Contrast enhanced chest CT (mediastinal windows) at the level of the right pulmonary artery showed homogeneous medium degree enhancement of the mass. There was obvious compression and distortion of mediastinal structure and bilateral pleural fluid.

small. The cytoplasm was moderate in amount and often pale to clear (**Figure 2A**). Mitotic figures were easily found. The angiodestruclive growth pattern was also present, accompanied by coagulative necrosis of tumor and fibrinoid changes in the blood vessels.

In situ hybridization showed that EB virus encoded RNA (EBER) was strong positive in tumor cells (**Figure 2B**). Tumor cells were positive for CD3 cytoplasmic staining, CD3ε membrane staining, CD45RO, TIA-1, and MUM-1, but negative for ALK, BCL2, BCL6, CD10, CD117, CD20, CD30, CD5, PAX-5, TdT, cytokeratin AE1/AE3, and CD56 in immunohistochemistry (**Figure 2C-E**). MIB index reached up to 90% (**Figure 2F**). The patient died 10 days later after admission. Autopsy was refused by his family.

Discussion

Although extranodal NK/T-cell lymphoma is more prevalent in China, there is no report of NK/T cell lymphoma involving mediastinum. Our case suffered from epididymitis. Testis and paratestis are the frequently involved sites by NK/T cell lymphoma. Although epididymis biopsy was not available, we think that primary NK/T cell lymphoma orignated in epidimitis. The NK/T tumor cells disseminated rapidly to various sites, e.g. mediastinal lymph node, pleura, pericardium, peritonium as the clinical course worsened. Therefore, our case showed mediastinal mass and polyserositis.

To date there are another two reported similar cases involved mediastinum in the literature. The first case was a 47-year-old Japanese man with CD56+ lymphoma with mediastinal and nasal involvement and an aggressive course [6]. Although lymphoma cells exhibited CD56+, cytoplasmic CD3 (cCD3) + with EBER negative, tumor cells was also positive for TdT. The second patient was a 63-year-old Japanese man with an anterior mediastinum tumor (7). This patient showed similar morphology with NK/T-cell lymphomas, with positive staining for CD56 and EBER but not T-cell antigens. Furthermore, TdT was also positive for this case. Since the natural killer cell marker, CD56, could be expressed in nonnasal lymphoma such as lymphoblastic lymphoma [8], we think lymphoblastic lymphoma is better for both cases. When lymphoma tumor cells show both CD56 and TdT positivity, whether it is NK/T cell lymphoma or lymphoblastic lymphoma? More cases should be collected, and more work need to be performed. Obviously, our case is the first case of NK/T cell lymphoma involving mediastinum in literature without any argument. Diagnosis of NK/T cell lymphoma is difficult in rare sites such as mediastinum, especially without the concomitant involvement of predilection sites. We still consider the possibility of NK/T cell lymphoma when lymphoma cells demonstrated the angiodestruclive growth pattern and coagulative necrosis in morphology.

However, we still cannot rule out the possibility that mediastinal lymph node was primarily involved by NK/T-cell lymphomas and then disseminated to other sites.

In conclusion, this case is a NK/T cell lymphoma arising from epididymis with involvement of mediastinum. Diagnosis of NK/T-cell lymphomas may be challenging, particularly in cases of unusual location.

Acknowledgements

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Disclosure of conflict of interest

None.

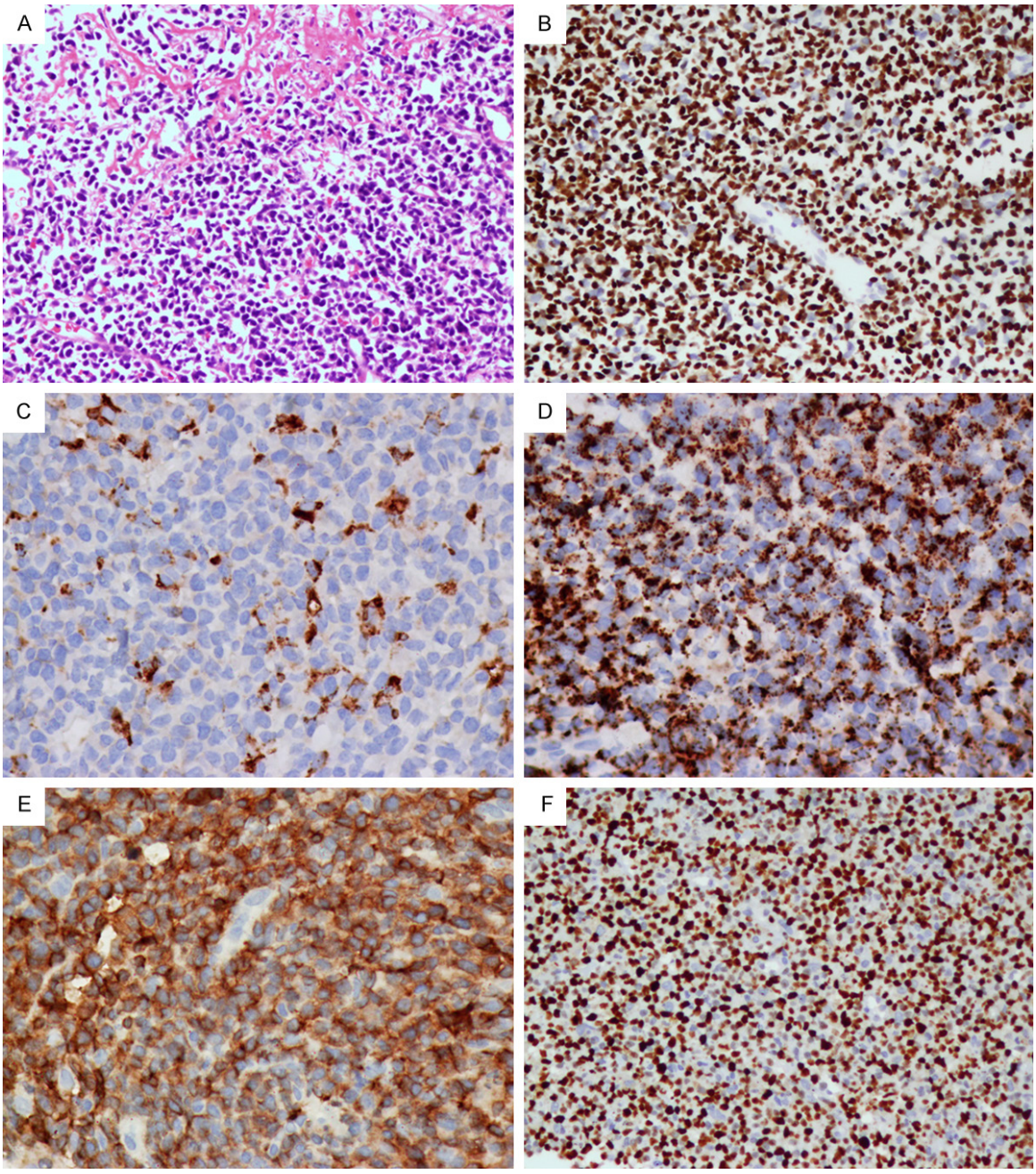

Figure 2. Pathological findings of the fine needle specimen from mediastinal mass. (A) H&E staining of mediastinal NK-T cell lymphoma with coagulative necrosis. *In situ* hybridization for EBER (B) and immunoreactivity for CD56 (C), TIA-1 (D), CD3ε (E) and Ki-67 (F). Magnification: ×200 for (A, B and F); ×400 for (C-E).

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