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Primary CD56-positive NK/T-cell Lymphoma of Median Nerve : A Case Report

Primary extranodal lymhomas of the central nervous system constitute 2% of all malignant lymphomas. The involvement of the peripheral nervous system is very rare. A solitary primary CD56-positive NK/T-cell lymphoma of the median nerve is described in a 70-year-old woman. On physical examination, a rubbery hard mass measuring 2.0 cm in diameter was palpated on the volar aspect of second to third finger of left hand. Excisional biopsy was performed. Under the fascia, a large fusiform tumor of the median nerve encapsulated with the epineurium was noted. Microscopically, the enlarged nerve showed extensive infiltration of atypical lymphoid cells. The lymphoid elements had abundant pale cytoplasm and large vesicular nuclei with peripheral prominent nuclei. The cells strongly expressed T-cell marker (UCHL-1) and natural killer cell marker (CD56). Gene rearrangement study showed rearrangement of T cell receptor (TCR-γ).

Key Words : *Lymphoma, malignant lymphoma; Killer cells, natural CD56; T-lymphocytes, NK/T-cell; Median nerve*

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INTRODUCTION

Primary extranodal lymphomas have been described in virtually every site of the body. Those of the central nervous system constitute 1% of all intracranial tumors (1) and 2% of all malignant lymphomas (2). The involvement of the peripheral nervous system (PNS) is rare (3). In most cases it is the result of systemic dissemination, the direct extension of an adjacent lymphoma (4) expressing itself or a "paraneoplastic phenomenon" (5). There are only five cases of primary malignant lymphomas involving the peripheral nervous system, and they occurred exclusively in the sciatic nerve (6-10). To our knowledge, no cases of primary malignant lymphomas of other peripheral nerves including the median nerve has been reported to date.

We report here one of the first cases of primary CD56-positive NK/T-cell lymphoma of the median nerve with immunohistochemical data and gene rearrangement study.

CASE REPORT

Clinical findings

A 70-year-old woman exhibited with a palpable hard mass with paresthesia on the left palm 20 days ago. On physical examination, a rubbery hard mass measuring 2.0 cm in diameter was palpated on the volar aspect of the second to third finger of the left hand and limitation of flexion of the involved two fingers was noted. Systemic examination was unremarkable, with no evidence of hepatosplenomegaly or systemic lymphadenopathy. Laboratory findings were normal. Excisional biopsy was performed. Under the fascia, a large fusiform tumor of the median nerve starting from the carpal tunnel and extending to the proximal phalanx of the second and third finger was noted. It was encapsulated with the epineurium. Partially the epineurium was ruptured and brownish friable tissue mass with adhesion to the lumbrical muscles and tendon sheaths was noted. The involved nerve and ill-defined soft tissue mass was curretted.

Pathologic findings

Grossly the specimen consists of a short segment of median nerve measuring 3.5 cm in length and 2.5 cm in diameter with a fish-flesh cut surface and several brown fragments of the invading tumor mass.

Formalin-fixed and paraffin-embedded sections of the enlarged nerve showed extensive infiltration of endoneurium, perineurium and epineurium of the nerve fibers by atypical lymphoid elements (Fig. 1, 2). The lymphoid

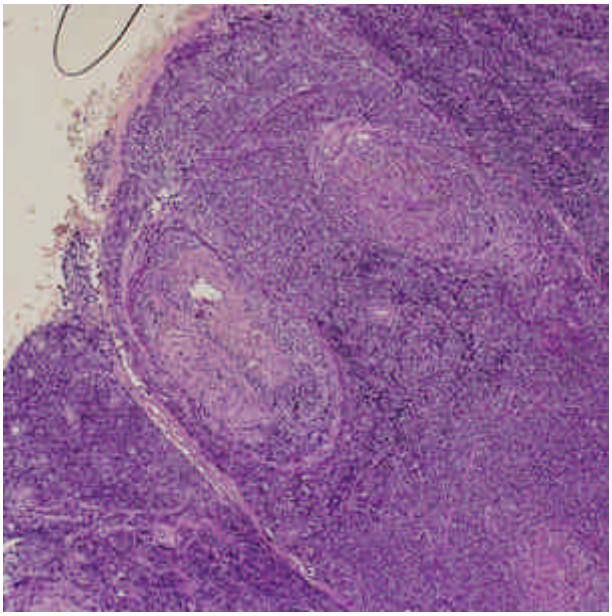

Fig. 1. Sections of the enlarged nerve showed extensive infiltration of epineurium, perineurium and endoneurium of the nerve fibers by atypical lymphoid elements (H & E).

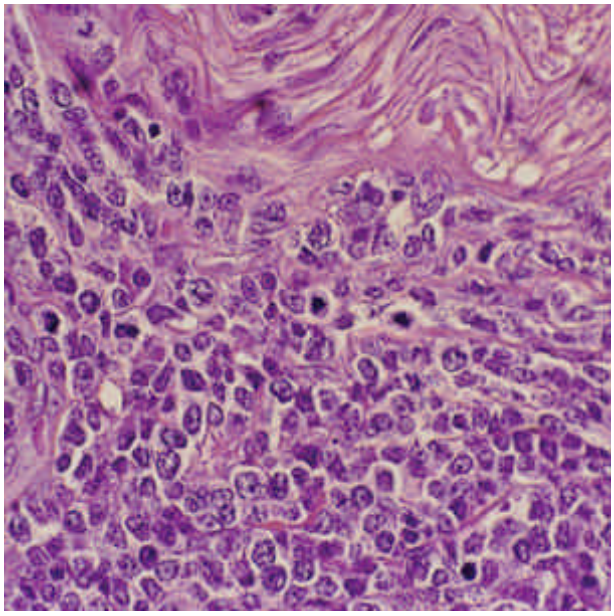

Fig. 3. The tumor cells infiltrating the nerve fibers are pleomorphic large lymphoma cells with large vesicular nuclei and prominent nucleoli (H & E).

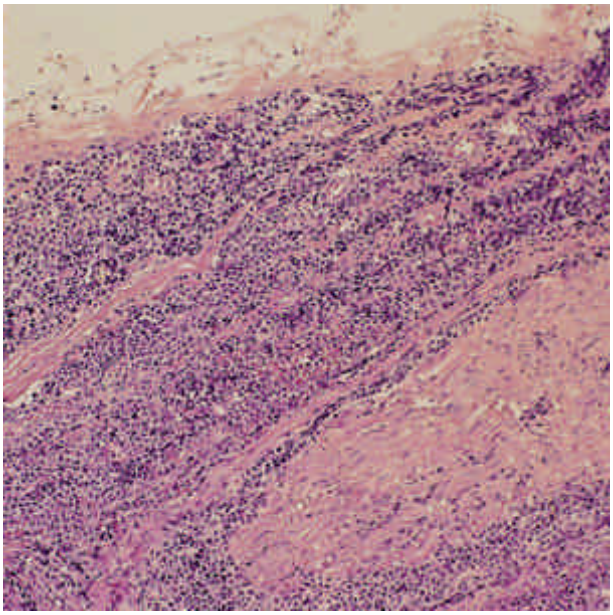

Fig. 2. Tumor cells infiltrating the perineurium and endoneurium of the nerve fibers (H & E).

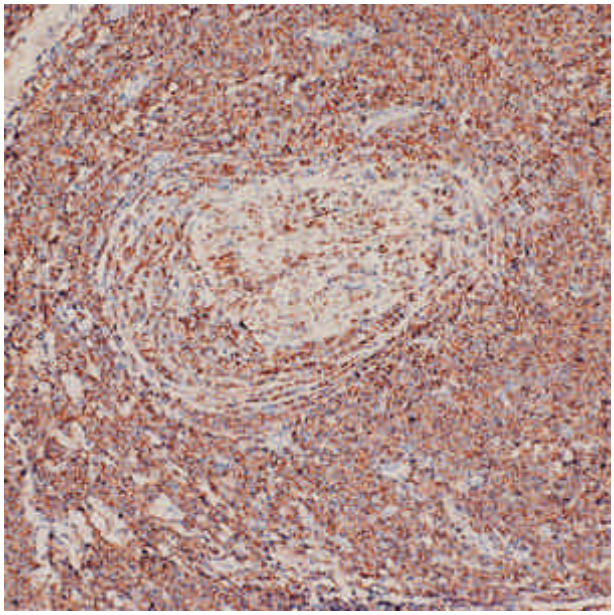

Fig. 4. The tumor cells infiltrating the nerve fibers are strongly positive for natural killer cell marker (CD56).

elements were pleomorphic large lymphoma cells with large vesicular nuclei and peripheral prominent nucleoli (Fig. 3). Mitoses were frequently seen. The tumor cells were positive for CD45 (leukocyte-common antigen), CD45Ro (UCHL1) and CD56 monoclonal antibodies (Fig. 4). Only small regular lymphocytes, but not the tumor cells, showed reactivity for CD20 (L26) monoclonal antibody. Gene rearrangement study using the PCR method showed rearrangement of T-cell receptor (TCR-γ) (Fig. 5). The tumor was diagnosed as primary CD56-positive NK/T-cell lymphoma.

DISCUSSION

The clinical and pathological findings suggest that this

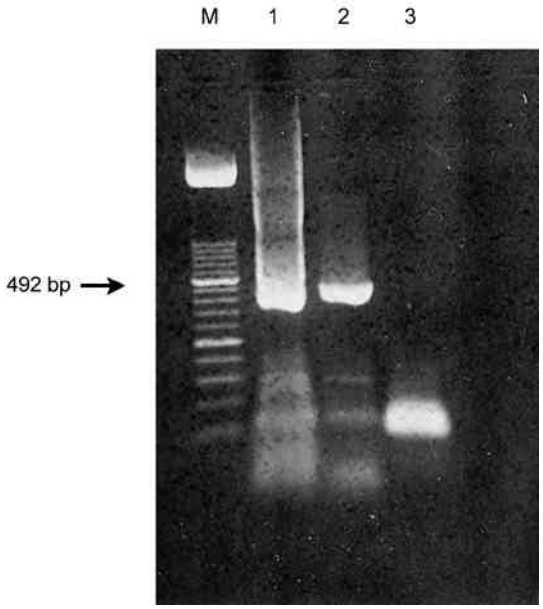

Fig. 5. Gene rearrangement study of TCR-γ gene. The rearranged band of TCR-γ was detected. M: size marker (50 bp Ladder marker), lane 1: positive control (Molt 3 cell line), lane 2: tumor, lane 3: negative control.

tumor is the first report of a solitary primary lymphoma of median nerve. It is known that malignant lymphomas may present at sites other than in the lymphoreticular system, and such nondisseminated (solitary) lymphomas of extranodal origin have been identified in a variety of organs. These include stomach and intestine, lung, skin, connective tissue, bone, testis, salivary gland, breast, thyroid and the central nervous system (11-12).

Only a few cases of involvement of the peripheral nerve are reported (6-10), and they all involved the sciatic nerve. Involvement of the peripheral nervous system by leukemias and malignant lymphomas is usually a manifestation of systemic dissemination, such as in the two cases of peripheral T-cell lymphoma described by Gherardi et al. (13). Both cases presented with peripheral neuropathy, due to involvement of the peripheral nerve, but this localization was found to be part of a systemic disease, also present in several lymph node regions. A few other cases of malignant lymphoma of the peripheral nerve system were because of a direct epineurial spread from adjacent tumors (14). Finally, peripheral neuropathy may be a "paraneoplastic" manifestation of lymphoproliferative disorders (5), mediated in some cases by antibodies to myelin proteins (15).

This case was characterized by involvement of the median nerve by a neoplastic proliferation that histologically proved to be CD56-positive NK/T-cell lymphoma. No evidence of disease was found elsewhere after thorough clinical staging workup. The patient was treated with radiotherapy and was disease free for 24 months after presentation.

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