PRIMARY PLASMACYTOMA OF THE CERVICAL LYMPH NODE: (A Case Report)

Servikal lenf nodunun primer plazmasitomu: (Olgu sunumu)

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Abstract: *Primary extramedullary plasmacytomas are rare disorders, and the majority arise in the upper respiratory system. Primary plasmacytoma of the lymph nodes is extremely rare, and there are fewer than 20 reported cases in medical literature. In this case report, we present a case of primary plasmacytoma of the cervical lymph node.*

Key Words: *Lymph node, Primary plasmacytoma*

Özet: *Primer ekstramedüller plazmasitomlar nadir hastalıklardır, çoğunluğu üst solunum sisteminden köken alır. Lenf nodunun primer plazmasitomları çok nadirdir ve literatürde 20'den az olgu bildirilmiştir. Bu olgu sunumunda, servikal lenf nodunda primer plazmasitom saptanan bir olgu sunduk.*

Anahtar Kelimeler: *Lenf nodu, Primer plazmasitom*

Plasmacytoma is an immunoproliferative, monoclonal disease of the B-cell line and is classified as non-Hodgkin lymphoma. It originates from a clone of malignant transformed plasma cells. The different types of plasma cell tumors are: 1) Extramedullary plasmacytoma (EMP): a) primary (true) plasmacytoma of the mucosa, unifocal/multifocal, with/without affected lymph nodes; and b) extramedullary manifestations of multiple myeloma (MM); 2) solitary plasmacytoma of the bone; 3) the multifocal form of MM; 4) MM; 5) plasmablastic sarcoma (modified classification of Bartl and Deicher, 1995) (3). Primary extramedullary plasmacytomas consist of proliferation of monoclonal plasma cell foci located outside the bone marrow. It is a rare disorder and the majority arise in the upper respiratory system (6,9,14). Primary plasmacytoma of the lymph nodes (PPLN) is very rare, and there are fewer than 20 reported cases in the medical literature (1,11,13). We report a case of primary plasmacytoma apparently confined to a single servical lymph node

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Case Report

A 41-year-old male patient was admitted for a painless swelling in his right cervical region. The swelling had been present for the last five years and had grown more rapidly in the last year. The blood formula showed $6.0x10^9$ /L white blood cells, with a normal differential count. The hemoglobin level was 15 g/dL and platelet count $150x10^9$/L. Total serum protein was 7.0 g/L. Bence Jones protein in the urine was negative. Serum concentrations of IgG, IgA and IgM were 993, 200 and 161 mg/dl, respectively. Bone marrow aspiration and biopsy showed normal hemopoiesis and no excess of plasma cells. A chest X-ray was normal. On physical examination there was a firm, easily movable lymph node swelling measuring 4 cm in diameter at the right cervical region. A right cervical lymph node biopsy was carried out. Roentgenographic examination displayed no lytic shadows in the bones. No other site of plasmacytoma was detected. Chemotherapy or radiotherapy could not be applied as the patient did not give consent. For the past two years, the patient has remained free of disease, with no evidence of local recurrence in the biopsy site.

The surgical specimen was fixed with 10% buffered formaldehyde and embedded in paraffin. Sections (4 μ thick) were stained with haematoxylin & eosin and Congo-red. Immunohistochemical analysis was

performed using the avidin-biotin peroxidase (ABC) method. Primary antibodies against the following antigens were used: CD20 (Neomarkers, 1:50 dilution), CD45 (Neomarkers, 1:50 dilution), kappa light chain (Zymed), lambda light chain (Zymed), immunoglobulin (Ig) E heavy chain (Dako, 1:25 dilution), IgM heavy chain (Zymed), IgA heavy chain (Biogenex) and IgG heavy chain (Zymed).

The specimen consisted of an encapsulated firm mass, which measured 6.5x3.5x3 cm in diameter, with a smooth greyish surface. The cut surface was uniform and grey-white in colour.

Microscopic examination revealed diffuse infiltration of atypical plasma cells in both cortex and medulla (Fig.1 and 2). Kappa light chain was evaluated as intensely positive (Fig. 3). Lambda light chain, CD45, CD20, and Ig heavy chains IgG, IgA, IgM, IgE were negative. The Congo-red staining performed for evaluation of amyloid was negative.

Histopathological and immunohistochemical examination revealed a kappa plasmacytoma.

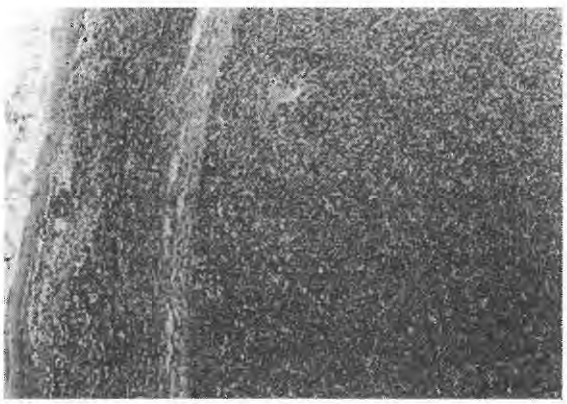

Figure 1. Sections of lymph node showing residual lymphoid tissuee and sheets of plasma cell infiltrates (H&E stain, x 50)

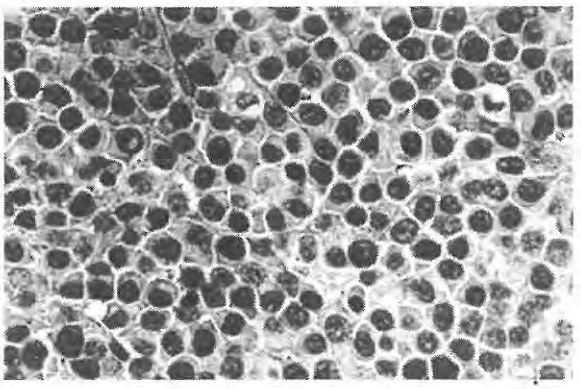

Figure 2. High-power view of the plasma cell infiltrates. (H&E stain, x 800)

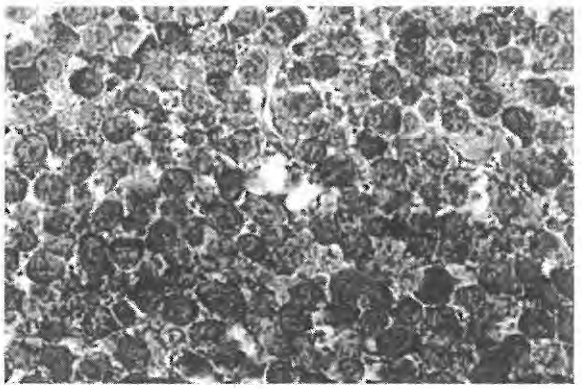

Figure 3. Plasmacytoma with positive immunoglobulin kappa light chain (Immunoperoxidase technique, kappa, x 800).

DISCUSSION

Extramedullary plasmacytomas, like multiple myeloma, consist of a proliferation of plasma cells showing varying degrees of differentiation. In order to diagnose PPLN, the first criterion is the establishment of the malignant nature of the tumor. Second, it must be proven that this is not a metastasis of myeloma or plasmacytoma of another organ. Plasmacytoma of the lymph node is occasionally seen in the course of multiple myeloma or metastasis from other sites of plasmacytoma (9,11,14). Therefore, the absence of bone marrow

infiltration of plasma cells and another primary plasmacytoma is an important factor in the diagnosis of PPLN. Histological and immunological findings are additional important factors in the diagnosis (13).

EMPs make up 4% of all plasma cell tumors and occur most commonly in the upper aerodigestive tract, and regional lymph nodes are sometimes involved (6,8,10,15). However, EMP has also been described in a wide array of locations including palatine tonsil, breast, lung, thyroid gland (2,8,14,15,17), gastrointestinal tract and skin. Primary plasmacytoma of the lymph nodes is very rare, and there are fewer than 20 reported cases in medical literature (6,11). The mean patient age in the previously reported cases is 50.6 years.

The immunophenotype of the primary nodal plasmacytomas is characteristic. According to Lin and Weiss, 7 of the 7 cases had light chain restriction, 4 of the 7 cases expressed monoclonal heavy chain (three with IgG and one with IgM), 1 of the 7 cases had nodal monoclonal IgG and serum monoclonal IgA elevation. None of the cases had CD20 or CD43 antigen expression, and 6 of the cases expressed CD79a (11). In the report by Menke et al, 17 of 20 cases had light chain restriction, 16 of 20 cases expressed monoclonal heavy chain (10 IgG, 4 IgM, 2 IgA), and 3 of 20 cases had CD20 expression (16). These results indicate that primary nodal plasmacytoma is an unequivocal monoclonal neoplastic proliferation. In our case immunohistochemical staining for kappa light chain was positive in the cytoplasm of tumor cells. Lambda light chain, IgG, IgA, IgM and IgE heavy chain, CD20, CD45 were negative.

In lymph nodes, a true plasmacytoma must be differentiated from nonneoplastic conditions in which larger numbers of plasma cells are found, such as the lymphadenopathy associated with rheumatoid disease and the plasma cell variant of giant lymph node hyperplasia, and neoplastic condition such as the plasmacytoid variant of malignant lymphoma. In these cases, the plasma cells are usually accompanied by many neoplastic small lymphocytes (1,7,11). The immunoperoxydase staining technique is the most useful method to achieve this goal (12).

Wiltshaw stressed that solitary extramedullary plasmacytoma has a natural history distinct from multiple myeloma and solitary myeloma of bone, with a more favorable prognosis (6,18). There appears to be a small number of patients in whom the tumor is confined to a solitary node or group of lymph nodes. In this group, local treatment is adequate and the prognosis is similar to that for cases of solitary plasmacytoma in other sites (1). Most cases respond well to excision with or without adjuvant chemotherapy or radiation therapy with no recurrence or progression (11). Our patient did not permit the application of chemotherapy or radiotherapy, and showed no recurrence or progression in the two years period of follow-up.

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