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Extramedullary Plasmacytoma of the Nasopharynx

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Extramedullary plasmacytoma (EMP) is a rare entity belonging to the category of non-Hodgkin lymphoma. EMPs make up 4% of all plasma cell tumors and occur mainly (80%) in the upper aerodigestive tract. We report a case of extramedullary plasmacytoma (EMP) of the nasopharynx presenting with nasal obstruction and nonspecific MR imaging features, which could not be distinguished from other more common lesions such as nasopharyngeal carcinoma and lymphoma. The diagnosis was established by histopathology, immunohistochemistry, and a systemic survey to exclude multiple myeloma. Our case highlights EMP may be one of the differential diagnoses of the nasopharyngeal tumor.

Key words: extramedullary plasmacytoma, nasopharynx, multiple myeloma

INTRODUCTION

Plasmacytoma is an immunoproliferative, monoclonal disease of the B-cell line and is classified as non-Hodgkin lymphoma. It originates as a clone of malignant transformed plasma cells.[1] They can occur as a solitary lesion outside the bone marrow [solitary bone plasmacytomas (SBP), solitary extramedullary plasmacytomas (EMP)], but can also be associated with multiple myeloma (MM).[2] EMPs make up 4% of all plasma cell tumors and chiefly occur (80%) in the upper aerodigestive tract (UAD), with the nasal cavity, paranasal sinuses, and nasopharynx being the most common sites. Along with these common sites, EMP has been noted to arise at any site in which lymphoid deposits are found, including the submucosal tissues of the pharynx, larynx, and oral cavity, as well as nonmucosal regions such as the thyroid gland.[1,3] EMP has a predilection for the male gender and occurs most frequently in patients 50 years and older.[4] Exclusion of multiple myeloma with dissemination is essential for diagnosing EMP. It should also be distinguished from other plasma cell malignancies and lymphomas due to their similar histopathologic findings. EMP of the nasopharynx is extremely rare and is presented here to highlight the clinical picture and histological features.

CASE REPORT

A 42-year-old healthy Taiwanese male was referred to our hospital with a six-week history of nasal obstruction. A biopsy of the nasopharynx suggested lymphoma. He denied any other subjective complaints such as otologic symptoms or headache. His past medical and family history were unremarkable.

Physical examination revealed a protruding mass of nasopharynx. The remaining head and neck examination was unremarkable. Nasopharyngoscopy showed a huge pink polypoid mass with intact surface filling the entire nasopharynx (Fig. 1). Magnetic resonance imaging (MRI) revealed a large enhancing tumor filling the entire nasopharynx and involving the right parapharyngeal space, without bony erosion, cervical lymphadenopathy, or metastasis of the neck (Fig. 2).

Biopsy of the nasopharyngeal tumor revealed submucosal infiltration with a monotonous population of neoplastic plasma cells with lambda light chains monoclonity. (Fig. 3). Pathologic diagnosis was plasmacytoma of the nasopharynx. Radiographic skeletal survey showed

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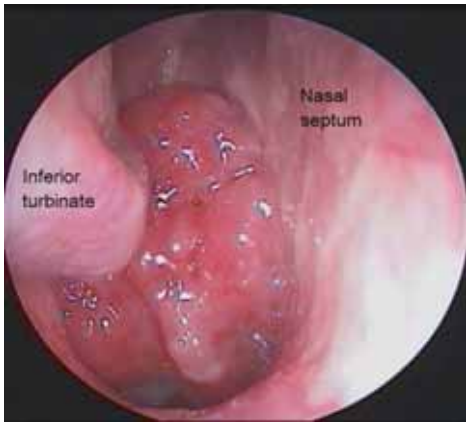

Fig. 1 Nasopharyngoscopy reveals a huge protruding pink polypoid mass, filling the entire nasopharynx without erosive or destructive surface.

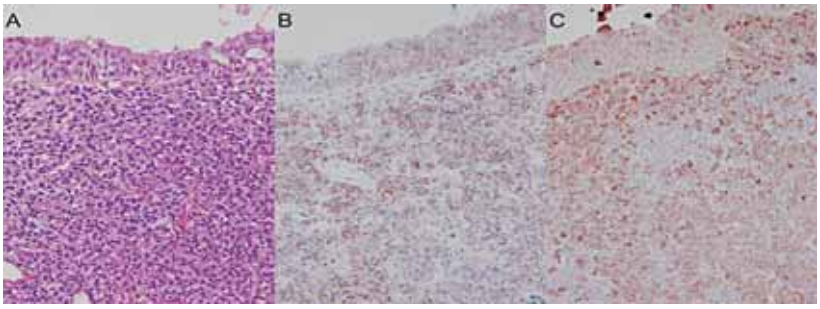

Fig. 3 Histological and immunohistochemical studies. (Original magnification ×400). (A) H&E stain. Submucosal generalizaed infiltration of a monotonous population of small-to medium sized neoplastic plasma cells with eccentric nuclei and abundant amphophilic cytoplasm. (B) CD 138 stain. Abundant diffuse staining for this plasma cell membranous protein. (C) Lambda light chains stain. Diffuse staining for a clonally expression of lambda light chains in the cytoplasm of the neoplastic plasma cells.

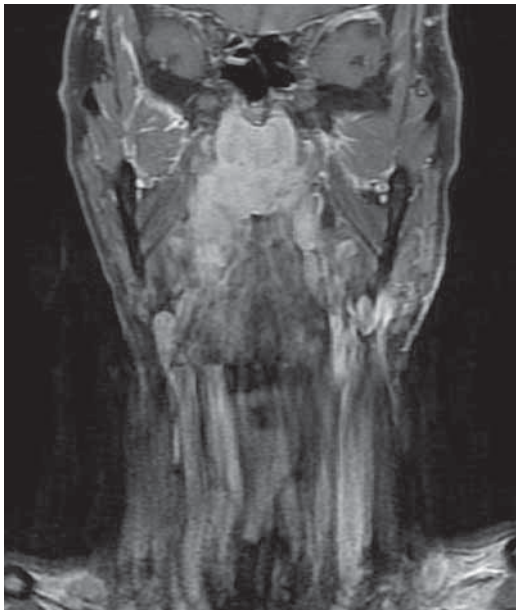

Fig. 2 MRI in a patient with extramedullary plasmacytoma of nasopharynx. Contrast-enhanced T1-weighted spin-echo image shows marked enhancement of the lesion without bony erosion, or cervical lymphadenopathy.

no evidence of lesions. Serum protein electrophoresis revealed a mildly elevated lambda light chains value of 767 mg/ dL (normal, 313-723 mg/dL). Urine protein electrophoresis was normal. A 24-hour urine collection revealed no Bence Jones proteinuria by immunofixation and immunodiffusion. Bone marrow examination revealed the lambda plasma cell population was within normal limits. He was diagnosed as having EMP of the nasopharynx, and subsequently received radiotherapy.

DISCUSSION

EMP is a rare monoclonal neoplastic proliferation of plasma cells characterized by an extramedullary tumor of clonal plasma cells with no or small amounts of monoclonal protein in the serum and/or urine, normal bone marrow, normal skeletal survey results, and no end-organ damage from the plasma cell proliferative process.[4] The EMP represents only 4% of all plasma cell neoplasms. Eighty percent of EMPs arise in the soft tissues of the head and neck region, with the nasal cavity, paranasal sinuses, and nasopharynx being the most common sites.[1]

The clinical presentation varies according to the involved region and mass effect. EMPs of the nasopharynx may present with nasal obstruction, otologic symptoms, or epistaxis.[5] The tumors may be red or pink polypoid lesions. In the presented case, the tumor has an intact surface with a polypoid appearance, consistent with its nature of submucosal growth. It does not resemble a nasopharyngeal carcinoma, which is a more vascular or erosive mucosa lesion.

The diagnosis of EMP is based on the morphologic and immunophenotype findings of a localized collection of monoclonal plasma cells without plasma cell proliferation at other sites, especially in the bone marrow, and no evidence of malignant lymphoma. Once a plasmacytoma is histologically confirmed, diagnostic procedures are carried out to exclude systemic involvement. These investigations include complete blood count (CBC), serum and urine protein electrophoresis/ immunoelectrophoresis, quantitative immunoglobulin (Ig) determination,

bone marrow biopsies, and skeletal survey. In EMP, laboratory findings reveal normal CBC, but may include abnormal monoclonal Ig (M-protein) or light chains in the blood and/or urine. Bone marrow biopsy is normal, and there are no osteolytic lesions noted on skeletal imaging.[3]

Knowledge of the imaging features of EMP in the literature is limited. EMP shows nonspecific CT and MR imaging features and cannot be distinguished from other more common lesions such as nasopharyngeal carcinoma and lymphoma.[6-8] However, features suggesting the diagnosis are a bulky soft-tissue mass or infiltrative lesion. The tumor does not usually become disseminated, but it may be locally aggressive and demonstrate marked involvement and destruction of the adjacent structures. CT and MR imaging are complementary techniques in evaluating the local extent of this rare plasma cell tumor, early detection of additional or recurrent lesions, and the presence of regional lymphadenopathy.[6]

The treatment of EMP consists of radiotherapy and/or surgery with a similar recurrence rate.[3] The review study of Aliexou et al. suggested surgery alone gives the best results in cases of EMP where resectability is good.[1] However, if complete surgical tumor resection is doubtful or impossible and/or if lymph node areas are affected, then combined therapy (surgery and radiation) is recommended.[1] There are also reports stating radiation provides excellent local and regional tumor control and survival in patients with EMP.[4,9,10] Although EMP of the nasopharynx is usually localized, surgery is used only to obtain adequate tumor tissue for pathologic diagnosis, not for curative treatment, because it is difficult to perform oncologically sound resection of nasopharyngeal tumors. In general, EMPs are considered to be radiosensitive; therefore, EMPs of the nasopharynx are usually treated with irradiation, with a local control rate of about 95- 100 %.[1,4]

Long-term follow-up is essential because local recurrence and development of disseminated disease into multiple myeloma or radiation-induced malignancy can occur many years after diagnosis.[3,4] The patients should be regularly monitored with radiologic imaging and circulating paraprotein of multiple myeloma.

In conclusion, EMP may be one of the differential diagnoses of the nasopharyngeal tumor and the diagnosis is made by histopathology and immunohistochemistry. The CT and MR imaging features of EMP are nonspecific. A thorough search for generalized myeloma is mandatory. Radiotherapy and/or surgery would be the main treatment of EMP.

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