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Stage II multiple extramedullary plasmacytoma, synchronous the nasal polyps of the head & neck. A case report and review of the literature

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Extramedullary plasmacytoma (EMP) is the term for plasma cell tumours which present outside of bone. The plasma cell lesions affecting the head and neck are multiple myeloma, EMP, plasma cell granuloma, and solitary plasmacytoma. EMP has been rarely met in the form of multiple lesions (10 per cent) (1). EMP could be staged according to the spread of the disease: Stage I is disease confined to one site; Stage II includes tumors with local spread or local node involvement; and Stage III cases show metastatic spread (i.e. disseminated disease or myelomatosis) (2).

Microscopically, EMP tissue consist of sheets of plasma cells which may be monomorphous or pleomorphic. Since the plasma cells are derivated as a clonal proliferation from B-lymphocytes, they may be characterized by immunohistochemical demonstration of cytoplasmic immunoglobulin determinants in the neoplastic cells. Immunoperoxidase methods are convenient routinely processed histological specimens. By immunohistochemical demonstration of a monoclonal staining pattern, consisting of one light chain type and one heavy chain class, most EMP's can be distinguished from reactive plasma cell infiltrates with polyclonal staining pattern (3). Localized disease may be treated by surgery or radiotherapy to which it responds well (1). Also disseminated disease is treated with chemotherapy.

A rare case of multiple EMP, localized in the nasal cavity, nasopharynx, and cervical area and synchronous the nasal polips was presented in this manuscript.

## Case report

A 50-year-old man was admitted to our clinic due to a four-month history of bilateral nasal obstruction and the left-sided neck region swelling. Clinical examination revealed a fleshy, yellow-gray to red polypoid masses in

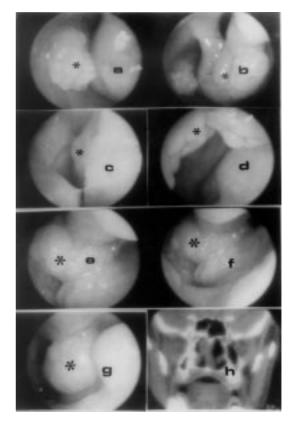


Figure 1. Endoscopic view of the nasal cavity and the nasopharynx shows polypoid masses (small stars shows *multiple extramedullary plasmacytoma*) in a) the postero-inferior region of the right nasal septal mucosa, b) the posterior region of the right inferior conchae, c) the right nasopharyngeal lateral wall, and d) the right choana. Also, endoscopic view of the nasal cavity and the nasopharynx shows polypoid masses (large stars shows *nasal polyps*) in e, f) the postero-inferior region of the left inferior conchae, and g) the left nasopharyngeal lateral wall. In aditional, h) shows coronal CT scan of the paranasal sinuses.

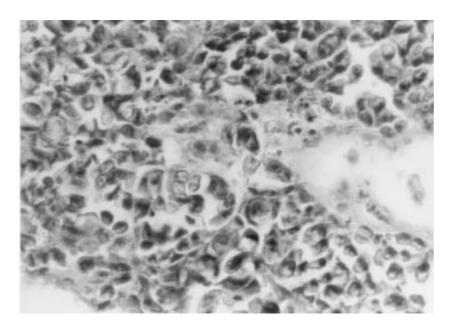


Figure 2. Light microscopically, the biopsy specimen shows solid cell groups formed from atypical plasma cells of eccentric nucleus and eosinophilic cytoplasm (H&E

the both nasal cavity and a gross, fixed left-sided mass in his upper profound neck region. There was no other present or past history of Ear-Nose-Throat disease. On endoscopic examination, the polypoid masses arising from posterior part of the inferior concha, the septal mucosa, and the nasal floor were seen in both sided. These masses were extended up to the lateral walls of nasopharynx. The endoscopic examination findings was revealing axial computed tomography scan of the paranasal sinuses and nasopharynx (Figure 1). Axial computed tomography scan of the cervical region showed the hypodence lymph nodes extended up to left lateral side of the cricoid cartilage from the left submandibular region. One of those lymph nodes was 4x5x5 cm in dimension, with central necrosis, consistent with invasion of the major vascular formations, and also had adjacent two lymph nodes, 3 cm in diameters and hypodence. Iliac crest bone marrow biopsy, bone scan, serum protein electrophoresis, urine analysis, and chest X-rays showed no other sytemic lesions.

Multiple EMP was diagnosed by transnasal endoscopic punch biopsies from the right nasal cavity and the right lateral wall of nasopharynx and by FNAC from the left upper profound jugular lymph nodes. In addition, nasal polips were diagnosed by transnasal endoscopic punch biopsies from the left nasal cavity. Light microscopical examination of specimen obtained from right side showed that consisting of a pure population of plasma cells in a sparse capillary stroma containing no inflammatory cell, which exhibit atypical features including large irregular nuclei, reversal of the nuclear cytoplasmic ratio, large coarse nuclear chromatin clumps and mitotic activity.

Also, immunohistochemistry demonstrated an Ig A-kappa phenotype and lambda light chain. Methyl green pyronine (MGP) staining for plasma cells was positive (Figure 2). A light microscopical examination of biopsies from the left side were showed nasal polips. This specimen had an edematous submucosa infiltrated by inflammatory cells, especially eosinophils and plasma cells, and overlying respiratory mucosa. The nasal polyp was extirpated by an endoscopic surgery. In addition, he received irradiation of total 4500 CGY in 22 fractions over 32 days to the neck, nasal, and nasopharyngeal regions, 5 days later punch biopses and FNAC. No recurrence has occured in the sixmonth and twelve-month follow-up period by repeated CT scan, endoscopic examination, transnasal biopsy.

Our case was interesting by stage II EMP i.e. localized in the right nasal cavity, the right nasopharyngeal lateral wall, and the left upper profound jugular lymph nodes, synchronous the nasal polyps localized in the left nasal cavity. According to this state, it was considered that is interrelation between EMP and nasal polyp by etiologically. This case may be value to delineate the nature of this disease. For this purpose, the patient should be currently under close follow-up. In addition, we understand that the polyps can be EMP or associated with EMP by chance.

In the paranasal sinuses, orbits, nasal cavity and nasopharynx, plasmacytoma is often bulky and is characterized by a combination of expansion with remodelling of bone and lytic bone destruction (4). The polypoid form of the EMP appears to behave in a less aggressive manner. The aggressive ones are usually soft

and friable and invade the underlying bone (1). Also, our case is characterized by expansion with remodelling of bone and lytic bone destruction. The symmetry of behavior of the disease on both sides is unusual (5). Our case come to an agreement with the literature, because of the masses in the left nasal cavity and lateral wall of nasopharynx rather than in the right of patient. Multiple myeloma (MM) will developed in 8 to 33 per cent of EMP cases and the necessity for lifelong follow-up is emphasized (6). But, MM did not developed in his twelvemonth follow-up. Histologically it must be differentiated from multiple myeloma and polyclonal infiltrates of plasma cells. A monomorphous population of well differtiated myeloma cells in FNAC was showed (7). In our case, plasmacytoma has showed a monoclonal origin by immunohistochemisry.

In the literature review, that is relation with paranasal sinuses and nasal cavity EMP was encountered the following studies: Koike et al., a man showed swelling of left maxillary plasmacytoma (Stage I) reported. The tumor disappeared after radiotherapy. After 18 months, they found left cervical and abdominal paraaortic lymph node swelling by computed tomography (Stage III). In this stage, he died inspite of chemotherapy. Autopsy showed that plasma cells invased in all gastrointestinal tract (8). Suzuki et al., a case of EMP of nasopharynx reported (Stage I). A woman was presented with complaint of postnasal drip. In this case, physical examination revealed a mass lesion in nasopharynx without cervical lymph node swelling. She received radiotherapy, followed by transnasal endoscopic resection (9). Parrot, a study presented benign appearing nasal polyp with the surprising diagnosis of EMP (Stage I) (10). But, none of these cases had not multiple EMP, synchronous nasal polyps. In our knowledge, our case might be interesting by stage II multiple EMP, synchronous the nasal polyps. The same of our case was not encountered in the literature review.

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