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Multiple Myeloma Presenting as a Parotid Mass

ABSTRACT

Objective: To present the case of a patient with left facial swelling as the primary manifestation of Multiple Myeloma and discuss the surgical management, diagnostic dilemma and subsequent medical management done for this unusual presentation.

Methods:

Design: Case Report
Setting: Tertiary Government Hospital
Patient: One

Results: A 55-year-old man with an enlarging left pre-auricular mass of one (1) year duration underwent superficial parotidectomy with facial nerve preservation and selective lymphadenectomy for pleomorphic adenoma based on initial clinical and cytologic findings. Histopathologic examination showed plasmacytoid proliferation and subsequent work-ups finally revealed Multiple Myeloma.

Conclusion: Emphasized in this case report is the thorough work-up and targeted therapy needed for the timely diagnosis and treatment of a patient with Multiple Myeloma.

Keywords: *Multiple myeloma, plasmacytoma, parotid gland, pleomorphic adenoma*

Myeloma is a neoplasm of plasma cells that usually causes bony lesions, blood abnormalities, and other potentially fatal complications. It may present as a solitary intramedullary lesion (plasmacytoma) or may involve multiple sites (multiple myeloma).¹ Extramedullary myeloma is an unusual presentation; its occurrence in the salivary glands is rare with only 24 published cases as of 2017.²⁻⁶ We report a case of Multiple Myeloma with unilateral pre-auricular swelling as the initial presentation of disease, its diagnostic work-up and the eventual approach and management done to treat the patient.

CASE REPORT

A 55-year-old widower from Taguig City, Philippines consulted at our Ear, Nose, Throat (ENT) out-patient department for a 1-year history of slowly enlarging left pre-auricular mass. On physical examination, a 7 cm x 7 cm mass on the left side of his face extended anteroposteriorly from the left tragal area to the left malar area and superoinferiorly from the left zygomatic arch to the left mandible. The mass was firm, non-tender, movable and non-erythematous with no intra-oral

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involvement on bimanual palpation. Several teeth were missing with some dental caries. No cervical lymphadenopathies were palpated. The rest of the ENT examination was unremarkable. (Figure 1)

A fine-needle aspiration biopsy (FNAB) of the pre-auricular mass revealed Pleomorphic Adenoma confirming the primary consideration of a benign parotid disease. (Figure 2) On performing superficial parotidectomy with facial nerve preservation & selective neck dissection, the parotid gland was noted to be almost normal in size with several adherent slightly enlarged lymph nodes. Histopathology of the excised specimens revealed atypical plasmacytoid cell proliferation. (Figure 3)

With a strong suspicion of a plasma cell pathology (mainly due to the findings from the periparotid lymph nodes), work-up became specific into ruling-out the possibility of a plasmacytoma or myeloma. A histopathologic slide review showed plasmacytic proliferation – considerations were plasmacytoma or myeloma. Immunohistochemical stain with CD138 was strongly positive for cells of interest. Complete blood count, electrolytes, liver and kidney function tests, fasting blood

sugar and metabolic panel were within normal limits as were the total urine and blood protein. Although urine protein electrophoresis showed no detectable immunoglobulins, serum protein electrophoresis was consistent with a monoclonal gammopathy (monoclonal peak concentration of 20.3% or 17.9 g/L at the Beta 1 region).

While these examinations were being conducted, an enlarging mass was noted in the previous surgical site. A facial CT scan revealed a left hemi-mandibular expansile lytic mass with some aggressive features. (Figure 4) A chest CT scan showed lytic lesions at vertebral bodies T7, T11 and T12 suggesting probable metastasis although this was not corroborated by bone scintigraphy (that did not show any evidence of a bony metastatic process).

Bone marrow aspirate biopsy yielded histologic and immunohistochemical findings consistent with a plasma cell neoplasm. Focal large aggregates of plasma cells, comprising 10%-20% of cell population with abnormally large and binucleated forms were present. The entire clinical picture and diagnostic exams satisfied the



Figure 1. Facial photographs of the patient (published with permission) showing the left facial swelling.

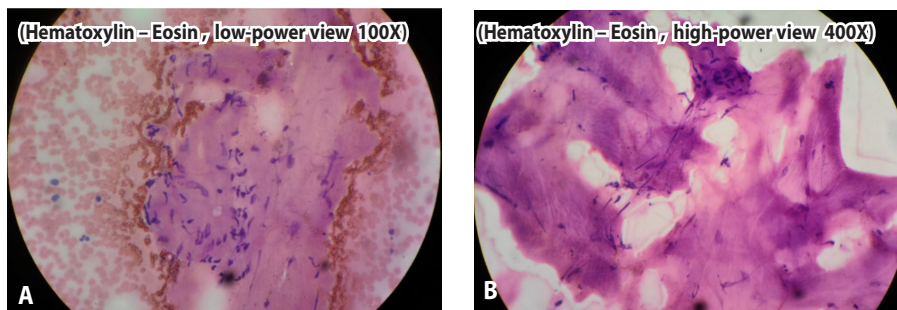


Figure 2. Fine-needle aspiration biopsy (Hematoxylin-Eosin). **A.** Low-power view (100x), interpreted as negative for malignant cells, cytomorphologically suggestive of pleomorphic adenoma; **B.** High-power view (400x), showing several spindle-shaped myoepithelial cells amidst a bloody chondromyxoid stromal background.

International Myeloma Working Group (IMWG) criteria⁷ for a diagnosis of Multiple Myeloma.

The patient underwent a treatment protocol consisting of eight (8)

cycles of Bortezomib-Melphalan-Prednisone with good tolerance and no adverse side effects. The previous left facial swelling was no longer palpable on post-treatment out-patient follow up. (Figure 5)

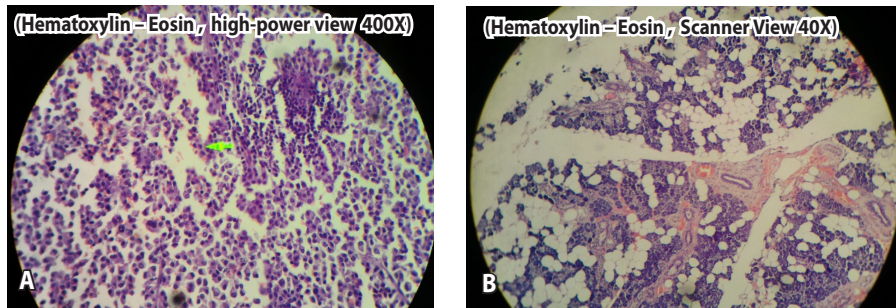


Figure 3. Histopathologic slides (Hematoxylin-Eosin). **A.** High-power view, 400x showing lymph nodes diffusely infiltrated with increased number of uniform-sized plasmacytoid cells with eccentric nuclei; **B.** Scanner view (40x) showing parotid tissue with scattered mature adipocytes within unremarkable salivary gland acini, parenchyma and stroma.

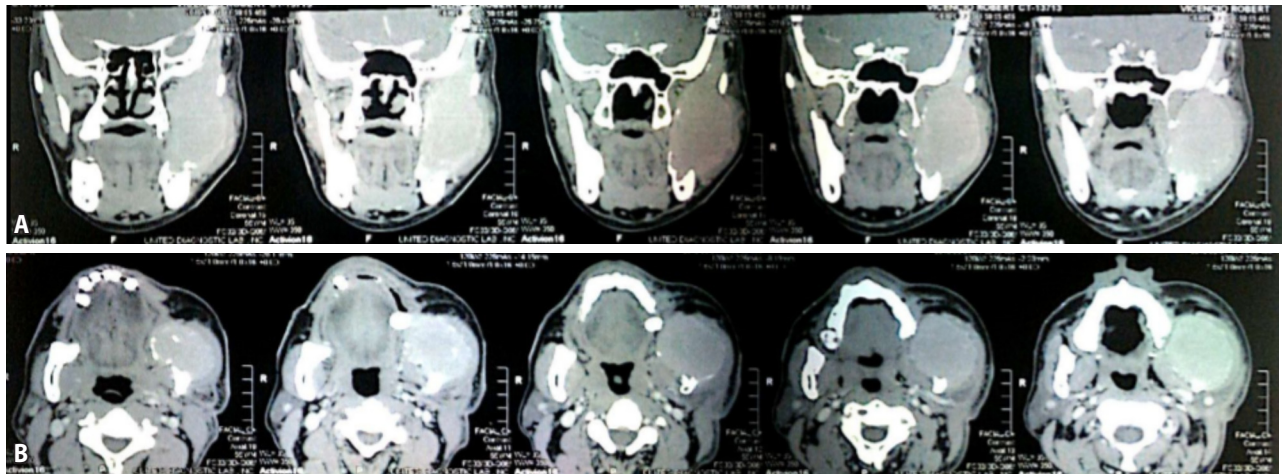


Figure 4. Contrast CT scans. **A.** Coronal sections **B.** Axial sections (bottom), showing a large well-margined expansile lytic enhancing solid mass appearing to arise from the anterior segment of the left mandibular ramus (7.3 x 4.7 x 5.9 cm). Anteriorly, the mass partially erodes the posterolateral wall of the left maxillary sinus. Superiorly, it is difficult to delineate from the insertion of the temporalis muscle. Laterally, the masseter is displaced outward but appears uninvolved. Medially, the lateral pterygoid shows no signs of invasion.



Figure 5. Post-chemotherapy photos (published with permission); the previous facial swelling is no longer appreciable.

DISCUSSION

Plasmacytoma, a tumor of plasma cells within soft tissue is traditionally divided into medullary and extramedullary types which in turn could either be solitary or multiple; the most common form is the generalized medullary also known as Multiple Myeloma.¹ With a male predilection, the average age of Multiple Myeloma at diagnosis is around 60 years old.⁵

Normally, plasma cells become B-cells when exposed to pathogens and produce antibodies. However in Multiple Myeloma, plasma cells generate clones of itself and form tumors eventually interfering with normal cell production and function.⁸ Since the malignant cells come from a common precursor, the antibodies produced are identical - monoclonal immunoglobulins (M-proteins) are then released into the blood and urine giving the symptoms and becoming targets for diagnostic examinations.⁵

Multiple myeloma usually present with bony or intramedullary lesions. Extramedullary plasmacytoma is an uncommon presentation with predilection for the upper respiratory tract; if ever it occurs in the head and neck region, soft tissue plasmacytomas tend to involve the nasal cavity or nasopharynx.¹ Its occurrence in the salivary glands is singularly rare⁹ with only 24 published cases since it was first reported in 1965 up until 2017.²⁻⁶

The patient presented with a one (1) year history of a slowly enlarging left pre-auricular mass, with no other accompanying symptoms and no intra-oral involvement – the common presentation of pleomorphic adenoma which is a benign parotid tumor. Routine imaging in patients with well-defined superficial lobe masses is not warranted because the result will not change the treatment plan. Excision of the parotid was performed after FNAB. In the same institution, FNAB has a sensitivity of 46% and a specificity of 100%.¹⁰ This relatively low sensitivity may explain why the initial benign finding of the pre-auricular mass FNAB turned out to be malignant. It could also be that the parotid gland was biopsied even if the tumor was in the mandible.

Intraoperatively, the parotid was noted to be almost normal in size with several adherent lymph nodes. The parotid together with 5 nodes were excised and sent to the laboratory for further investigation. Histopathologic findings of atypical plasmacytoid cell proliferation then warranted further investigation of a plasma cell pathology.

The diagnosis of Multiple Myeloma was confirmed after satisfying the IMWG criteria.⁷ For our patient, this includes 1: clonal bone marrow plasma cells >10% and 2: >1 lytic lesion on the vertebral body; other diagnostics which point to Multiple Myeloma include the confirmed monoclonal gammopathy on protein serum electrophoresis and the plasma cell morphology on histopathology.

The negative bone scan is attributed to the fact that scintigraphy is of

limited use in Multiple Myeloma. Detection of bone involvement using technetium 99-m relies on the osteoblastic response and activity of the skeletal system for uptake. Multiple Myeloma, however, is primarily an osteolytic neoplasm.¹¹ The patient's lytic lesions on the vertebra, therefore, would have been more identified in MRI or CT scan.

For therapy, the standard treatment for Multiple Myeloma has been Melphalan and Prednisone. The patient's chemotherapy protocol included these plus Bortezomib – a new proteasome inhibitor which prevents protein breakdown in Multiple Myeloma. This protocol significantly prolongs overall survival compared to Melphalan and Prednisone alone.¹²

This report highlights the presentation, diagnostic work-up and management done in a case of an unusual facial swelling. It shows a rare initial presentation of Multiple Myeloma which can be addressed medically to improve overall health outcomes. Although financially burdened, the patient is currently contented with his state, highly optimistic and hopeful for his complete remission and recovery from the disease.

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