Case Report
Korean J Otorhinolaryngol-Head Neck Surg 2025;68(11):497-502 / eISSN 2092-6529
https://doi.org/10.3342/kjorl-hns.2024.00535

Malignant Peripheral Nerve Sheath Tumor Presenting as a Chronic Parotid Gland Mass: A Case Report

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Received November 6, 2024
Revised June 12, 2025
Accepted June 16, 2025
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Malignant peripheral nerve sheath tumors (MPNSTs) affecting the parotid gland represent only about 2.8% of all head and neck MPNSTs. We present this rare parotid tumor to contribute to published data, raise awareness of its clinical presentation and to improve surgical management. A 22-year-old male presented with a 10-year history of gradual right-sided hemifacial weakness, later developing a mass at the right preauricular area. Preoperative ultrasound showed a right parotid hypovascular mass exhibiting heterogenous echopattern and microcalcifications. A multidisciplinary team of surgeons from surgical oncology and plastic surgery performed total parotidectomy followed by a minimal access cranial suspension lift and supra brow lift to address the right sided facial drooping. Histopathology revealed a solid cluster of pleomorphic spindle cells which stained positive for S-100 and vimentin consistent with MPNST. There was improvement of the facial asymmetry and he did not undergo radiotherapy.
Korean J Otorhinolaryngol-Head Neck Surg 2025;68(11):497-502

Keywords Malignant peripheral nerve sheath; Multidisciplinary; Parotid tumor.

Introduction

Malignant peripheral nerve sheath tumor (MPNST) is a rare type of sarcoma representing about 2% of all sarcomas.[1,2] MPNSTs most commonly arise in the extremities and trunk, with tumors arising from the head and neck at only about 20% of all cases.[3] MPNSTs affecting the parotid gland represent only about 2.8% of all head and neck MPNSTs.[4] Here we present a case of MPNST of the right parotid gland in a 22-year-old male.

Involvement of the facial nerve in parotid tumors is not usual. It may be observed in advance and high grade malignancy. However, if the clinical presentation is such that the facial nerve paralysis became more evident before the appearance of the mass, this should raise the suspicion of a possible nerve involvement such as nerve sheath tumor and Inclusion of a plan on how to manage the facial nerve should be included. This case report aims to demonstrate the difference in the clinical presentation of this rare tumor and guide surgeons on how to manage not only the tumor but also its effect on the facial nerve so as to achieve good outcome.

Case

We present the case of a 22-year-old male who presented with a 10-year history of gradual right-sided hemifacial weakness, later developing a mass at the right preauricular area. The gradual enlargement of the mass prompted consultation and subsequent management in our tertiary government hospital.

On physical examination there was a 4×7 cm non-hard, slightly movable mass on the right inferior preauricular area (Fig. 1). Testing for actions of the facial muscles, the patient demonstrated right-sided weakness upon raising the eyebrows, closing the eyes, blowing out the cheeks and smiling (Fig. 2).

Ultrasonography showed a right parotid mass located inferolaterally to the gland, measuring 34.3×25.5×65.1 mm. The mass exhibited heterogenous echopattern containing hyperechoic and anechoic areas. It was hypovascular, with sol-

id and cystic areas, and microcalcifications (Fig. 3).

Ultrasound-guided fine needle aspiration of the right parotid gland revealed a salivary gland neoplasm of uncertain malignant potential and cellular basaloid neoplasm.

Considering the presence of facial paralysis and the preauricular mass, our clinical diagnosis was a parotid malignancy. Preparation for surgery included discussion of the extent of surgical resection with nerve sacrifice, planning for facial reanimation and counselling of the patient for possible permanent facial paralysis. A multidisciplinary team of surgeons from surgical oncology and plastic surgery worked together during the operation.

The patient underwent total parotidectomy performed by the surgical oncology team. During resection, the tumor was noted to arise from the deep lobe and under the facial nerve abutting the cervical and mandibular branches (Fig. 4A). The mass was dissected off from the nerve without breaking its capsule (Fig. 4B).

The plastic surgery team then proceeded to do a minimal access cranial suspension lift and supra brow lift to address the right sided facial drooping.

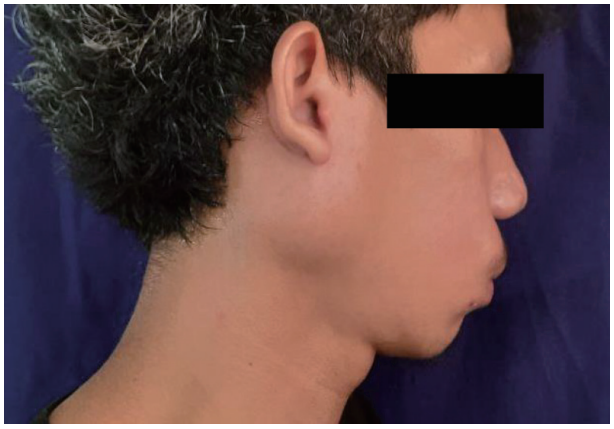

Fig. 1. Patient presenting with a right inferior pre-auricular mass.

Gross examination of the resected parotid specimen revealed a 7.5×4.3×3.2 cm parotid gland with a 3.7×3.4×2.9 cm, non-hard, well-encapsulated mass at the inferior portion (Fig. 5A). Cut section of the inferiorly located nodule revealed a 3.7×3.2×2.9 cm mass which was fleshy with cystic areas containing dark brown fluid (Fig. 5).

The patient's post-operative hospital stay was unremarkable, and he was discharged on the third post-operative day. However, his symptoms of right-sided facial weakness persisted.

Histopathologic examination of the resected specimen revealed a spindle cell neoplasm described as a solid cluster of pleomorphic spindle cells arrange in a storiform architecture, invading into the surrounding fibrous capsule (Figs. 6 and 7). Vesicular nuclei and prominent nucleoli admixed with numerous mitotic figures were also noted within the specimen. All lines of resection were negative.

Differential diagnoses at this stage included myoepithelial carcinoma, MPNST, and hemangiopericytoma. Further immu-

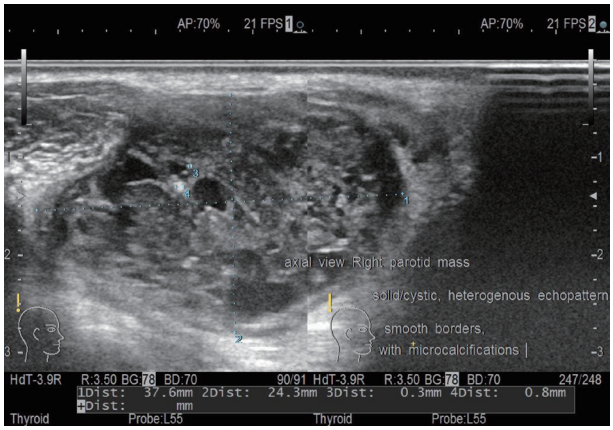

Fig. 3. Ultrasonography revealed a right parotid mass located inferolaterally to the gland, exhibiting heterogenous echopattern containing hyperechoic and anechoic areas, with hypovascularity, solid and cystic areas, and microcalcifications.

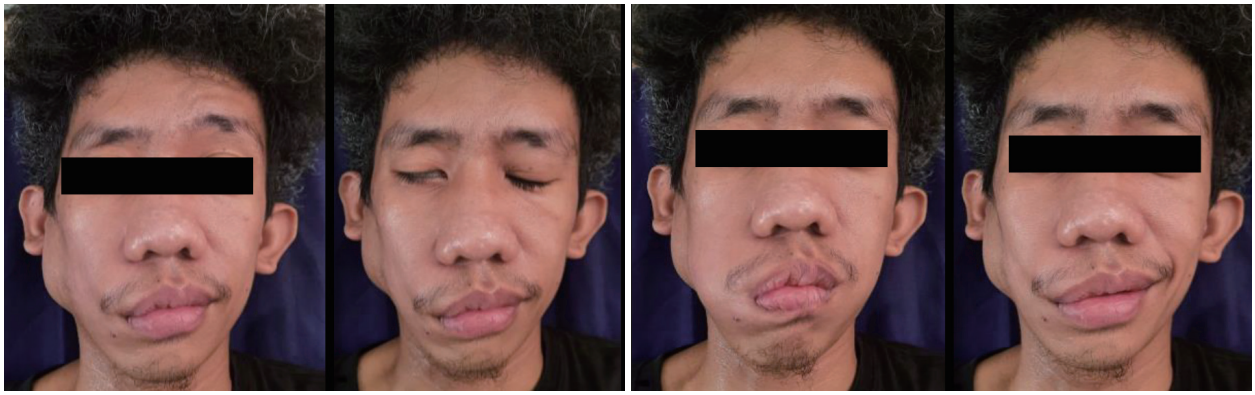

Fig. 2. Patient exhibiting right sided weakness upon testing of the branches of the facial nerve.

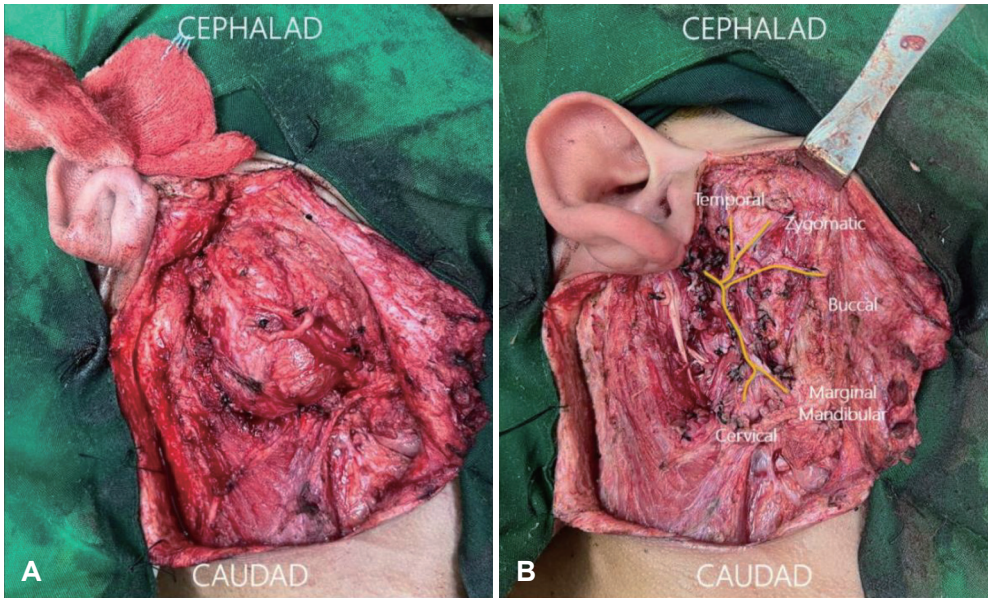

Fig. 4. The intraoperative findings showing the relation of the mass to the facial nerve and the gross pathologic appearance of the tumor. A: The tumor was noted to arise from the deep lobe and under the facial nerve abutting the cervical and mandibular branches. B: Intraoperative picture after removal of the tumor.

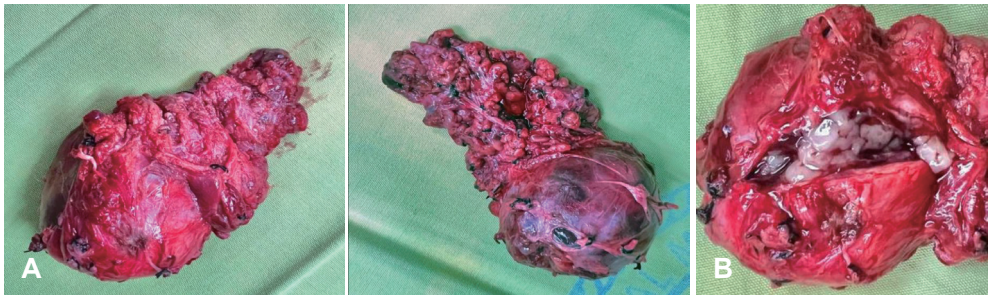

Fig. 5. A: Resected right parotid specimen measuring 7.5×4.3×3.2 cm. Inferiorly, it features a 3.7×3.4×2.9 cm, non-hard, well-encapsulated mass. B: Cut section of the mass located in the inferior part of the parotid was noted to be fleshy with cystic areas containing dark brown fluid.

nohistochemical staining with S-100, vimentin, p63, epithelial membrane antigen (EMA), smooth muscle actin (SMA), CD34, and cytokeratin allowed us to narrow down our diagnosis. Positive staining for S-100 and vimentin confirmed our diagnosis of MPNST (Figs. 8 and 9). There was marked improvement in his facial asymmetry as noted 12 months postoperatively (Fig. 10). He was advised to undergo radiotherapy but refused further treatment and follow up.

Discussion

Mesenchymal tumors represent approximately 2% to 5% of all salivary gland tumors, with over 95% of all tumors involving the major salivary glands. Malignant tumors occur less frequently with the ratio of benign to malignant tumors at 2.4:1 up to 18:1.[4] MPNSTs, tumors that arise from nerve sheaths of peripheral nerves, represent a small fraction of all sarcomas at about 2%. MPNSTs most commonly involve the nerve roots and bundles in the pelvis and extremities, with notable involvement of the sciatic nerve.[2] MPNSTs of the head and neck are very rare, with tumors involving the parotid gland even rarer still. Parotid gland MPNSTs comprised only 4 out of 142 head and neck MPNSTs in a review of literature spanning 13 years from 1990 to 2003.[3] According to this study, the usual clinical presentation of MPNST is that almost half arise in the context of neurofibromatosis (NF1) syndrome, usually in association with pre-existing plexiform neurofibromas which was not noted in our patient. MPNST present as rapidly enlarging mass which could either be painful or can cause local neurological symptoms such as weakness or paresthesias[3] as manifested by our patient.

A sarcoma is assumed to be MPNST if one of the follow-

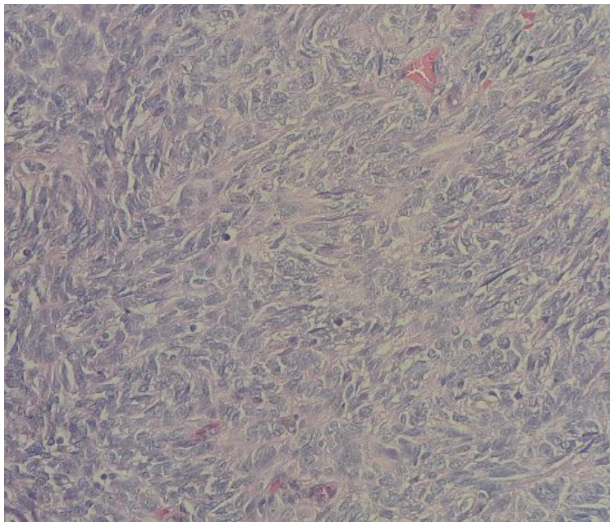

Fig. 6. Histopathologic examination revealing a solid cluster of pleomorphic spindle cells arrange in a storiform architecture. Hematoxylin and eosin stain high power magnification.

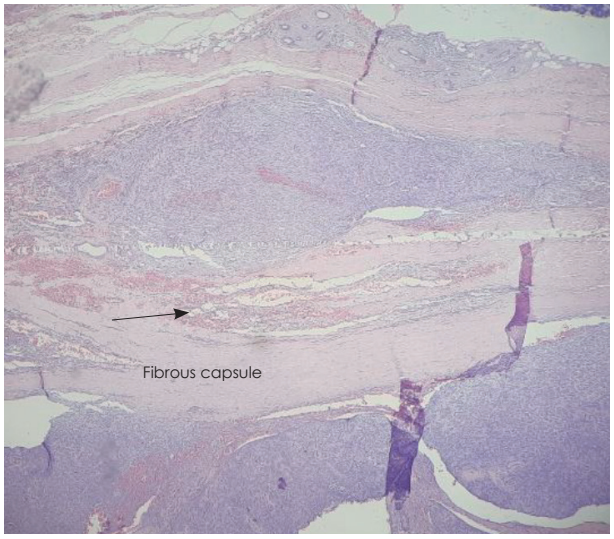

Fig. 7. Tumor cells (arrow) are seen invading into the surrounding fibrous capsule. Hematoxylin and eosin stain, low power magnification.

ing 3 criteria are met: 1) tumor arises from a peripheral nerve; 2) tumor arises from a pre-existing benign nerve sheath tumor or neurofibroma; and 3) tumor displaying constellation of histological features of Schwann cell differentiation.[4] In our patient, the tumor appears to arise from the facial nerve deep within the substance of the parotid.

Immunohistochemical studies were essential in confirming the diagnosis of MPNST. S-100 protein was the most widely used antigen for neural differentiation,[2] and approximately 50%–90% of MPNSTs are positive for S-100 protein.[5] Vimentin was also strongly positive in all reported cases of MPNST.[2] In our patient, the tumor stained positively for S-100 protein and vimentin on the cells of interest. The tumor did not stain positively for EMA, CD34 or cytokeratin marking the tumor as less likely of epithelial origin. Negative staining for SMA likewise ruled out hemangiopericytoma as a possible differential.

The prognosis of MPNST is generally poor. The 5-year survival rate varies from 40% to 50%.[2] The mainstay of treatment has been complete surgical resection. In our patient, the margins of resection were negative, however due to the rarity of these tumors further studies are needed to fully understand the extent of optimal surgical management of these tumors.[6] High quality studies evaluating adjuvant treatment with chemotherapy[7] and radiotherapy[8] likewise have not yielded promising results. MPNST is highly chemotherapy-resistant and adjuvant chemotherapy is usually only reserved for advanced, unresectable disease or metastatic disease however even in these populations the benefits are limited.[7] A comprehensive multidisciplinary approach to the management of the disease is necessary for a chance at a better outcome.

In conclusion, we present a rare case of MPNST arising from the parotid gland in a young male patient. Review of previous literature revealed that MPNST arising from the pa-

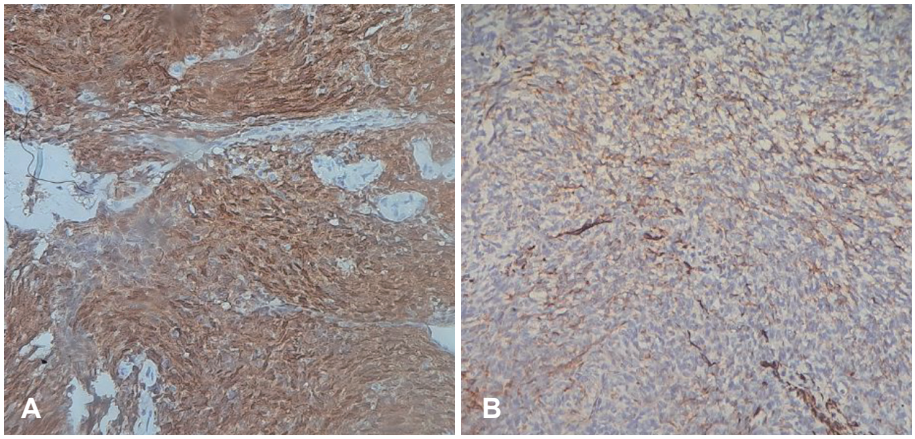

Fig. 8. Immunohistochemical staining for S-100 and vimentin on cells of interest (A and B, respectively). S-100 (A) and vimentin (B).

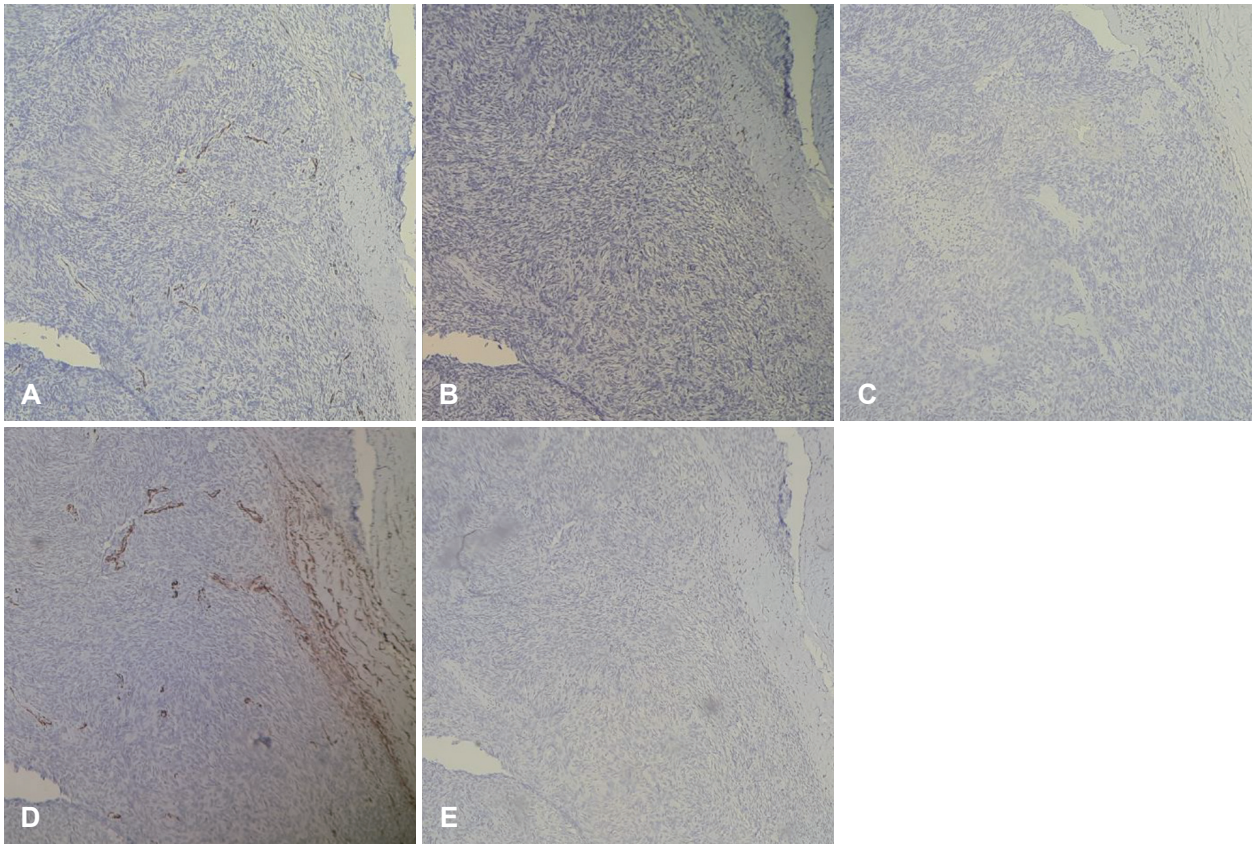

Fig. 9. No staining on cells of interest for p63 (A), CD34 (B), epithelial membrane antigen (C), cytokeratin and smooth muscle actin (D). p63 IHC low power magnification.

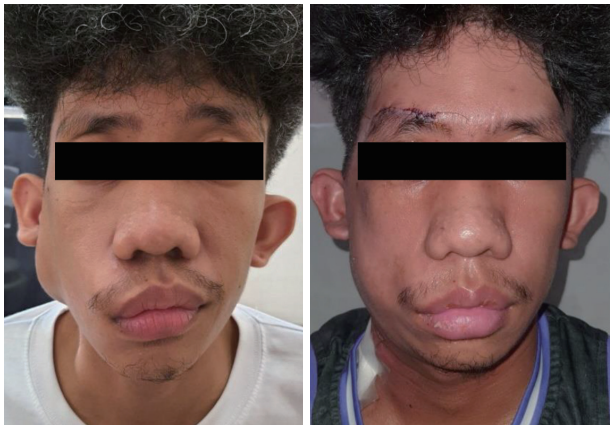

Fig. 10. Before and after photos of the patient showing improvement in right-sided eyebrow and right oral commissure drooping.

rotid gland is very rare and diagnosis relies heavily on confirmatory testing via immunohistochemistry. Prognosis of MPNST is generally poor and treatment relies on achieving complete resection of the tumor. After complete resection, reconstructive procedures can be considered to improve function and cosmesis. Options for adjuvant treatment are limited and a multidisciplinary approach to the patient is recommended to afford the best outcomes.

Acknowledgments

To our colleagues from the Department of pathology and the Department of Surgery, Section of Plastic & Reconstructive Surgery for helping in the management of the case. To our patient for allowing us to share his case for others may learn.

Author Contribution

Conceptualization: Wilson C. Zuñiga, Ida Marie Tabangay-Lim. Data curation: all authors. Formal analysis: Wilson C. Zuñiga, Ida Marie Tabangay-Lim. Funding acquisition: Ida Marie Tabangay-Lim. Investigation: all authors. Methodology: all authors. Project administration: Ida Marie Tabangay-Lim. Resources: all authors. Software: Wilson C. Zuñiga, Czarina Angelli L. Anastacio. Supervision: Ida Marie Tabangay-Lim. Validation: Wilson C. Zuñiga, Ida Marie Tabangay-Lim. Visualization: Wilson C. Zuñiga, Ida Marie Tabangay-Lim. Writing—original draft: Wilson C. Zuñiga, Czarina Angelli L. Anastacio. Writing—review & editing: Ida Marie Tabangay-Lim.

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