Nasal Malignant Peripheral Nerve Sheath Tumor Resembling Cavernous Hemangioma: A Case Report

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Abstract.

Background: Malignant peripheral nerve sheath tumor (MPNST) is a malignant, locally invasive tumor that originates from Schwann cells in the peripheral nerves, and has a high probability of metastasize. It accounts for 5-10% of all soft tissue sarcomas and has an incidence of 0.001% in the general population. Because of its rarity and lack of specificity, little is known about its imaging features, and reports on the nasal region are uncommon. Case Report: A 43-year-old woman with symptoms of nosebleed from her right nostril and had a tampon inserted for 4 days, but the bleeding persisted. No history of trauma. Physical examination is inconclusive. CT Angiography (CTA) shows a hypervascular mass in the right nasal cavity extending into the ethmoidal sinus, suggesting a cavernous hemangioma. The patient underwent mass extirpation surgery. Histopathological and histochemical examination of the removed tissue revealed a malignant peripheral nerve sheath tumor. The patient then underwent radiotherapy. An MRI evaluation revealed no residual tumor. Discussion: Malignant peripheral nerve sheath tumor (MPNST) is a rare soft tissue sarcoma. This tumor can be aggressive by infiltrating the surrounding tissue, including bone structures, in contrast to hemangiomas. Computed tomography is useful for locating the tumor, identifying infiltration, and planning treatment. Because this reported case was believed to be in the early phases of the illness, the lesion found in CTA has regular margins and well-defined boundaries, thus it is more appropriate for cavernous hemangioma. As a multi-parameter modality, MRI can provide a wealth of information on characterizing various components in tumors, thus is used for further investigation and post-treatment evaluation. Conclusion: Because of the overlapping radiographic features, distinguishing MPNST from other more prevalent tumors, such as cavernous hemangioma, is challenging. It is important for radiologists to consider MPNST as a differential diagnosis of masses in the nasal cavity.

Keywords: *Neurofibrosarcoma; Hemangioma, cavernous and Computed Tomography Angiography.*

I. INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) are malignant and locally aggressive soft tissue sarcomas with nerve sheath differentiation and a high propensity to metastasize. Their incidence is relatively rare in the general population, estimated at 1 per 100,000 individuals (1). Overall, MPNSTs comprise 5-10% of soft tissue sarcomas and are one of the most common *nonrhabdomyosarcomatous soft tissue sarcomas* (NRSTS) in pediatric patients. Although approximately 10-20% of all MPNSTs are diagnosed in children, there is no difference between children and adults in tumor location, size, or histologic grade (2).The diagnosis of MPNST has been described as one of the most difficult among all soft tissue tumors due to its nonspecific presentation both clinically, histopathologically, and radiologically (3). Several studies on MPNST imaging have been reported, but due to its rarity and lack of specificity of radiological examination results, the characteristics found cannot be well formulated, and nasal lesions are rare. This case report presents an MPNST that on radiological examination showed the appearance of cavernous hemangioma, thus presenting a challenge in diagnosis.

II. CASE REPORT

A 43-year-old woman presented with a nosebleed from her right nostril that persisted for 3 hours. She was admitted to the hospital and had a tampon inserted. After 4 days, the tampon was removed, but the bleeding continued. She denied any history of trauma, any history of blood clotting disorders, or any family history of tumors/cancer. A physical examination of the ear, nose, and throat revealed bleeding in the right nasal cavity, but the source of the bleeding could not be identified. A CT angiography (CTA) scan revealed a

hypervascular mass measuring 2.2 x 1.4 x 2.8 cm in the right nasal cavity extending into the ethmoidal sinus, consistent with a cavernous hemangioma. The patient underwent surgical extirpation of the mass. Histopathological and histochemical examinations of the removed tissue revealed a *malignant peripheral nerve sheath tumor*. She underwent 30 rounds of radiotherapy. An MRI scan 6 months later showed no residual tumor.

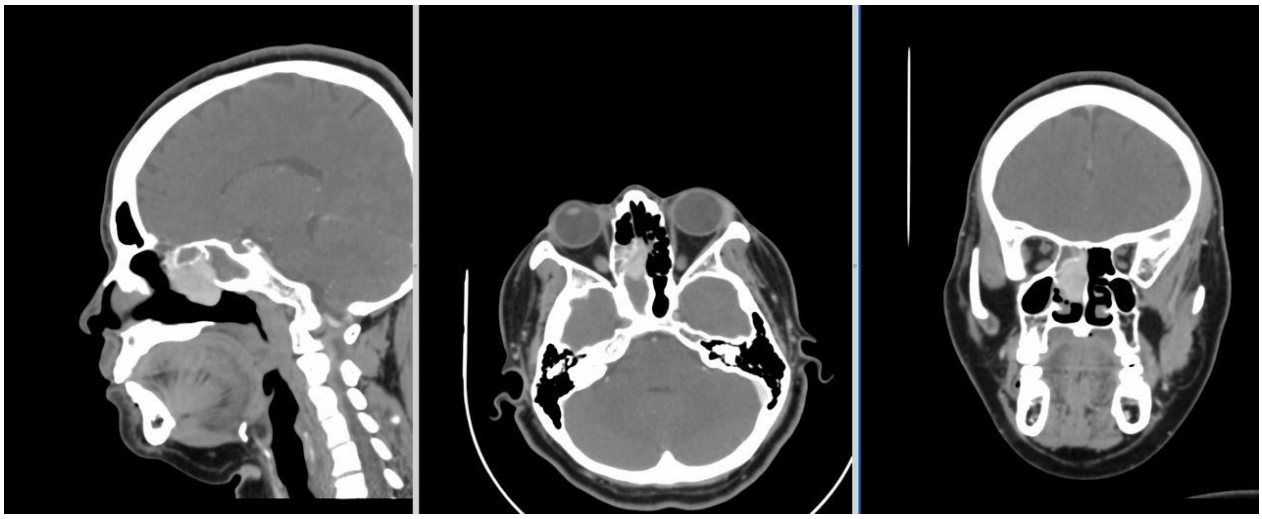

Fig 1 Hypervascular mass in the right nasal cavity extending into the ethmoidal sinus suspected to be a cavernous hemangioma.

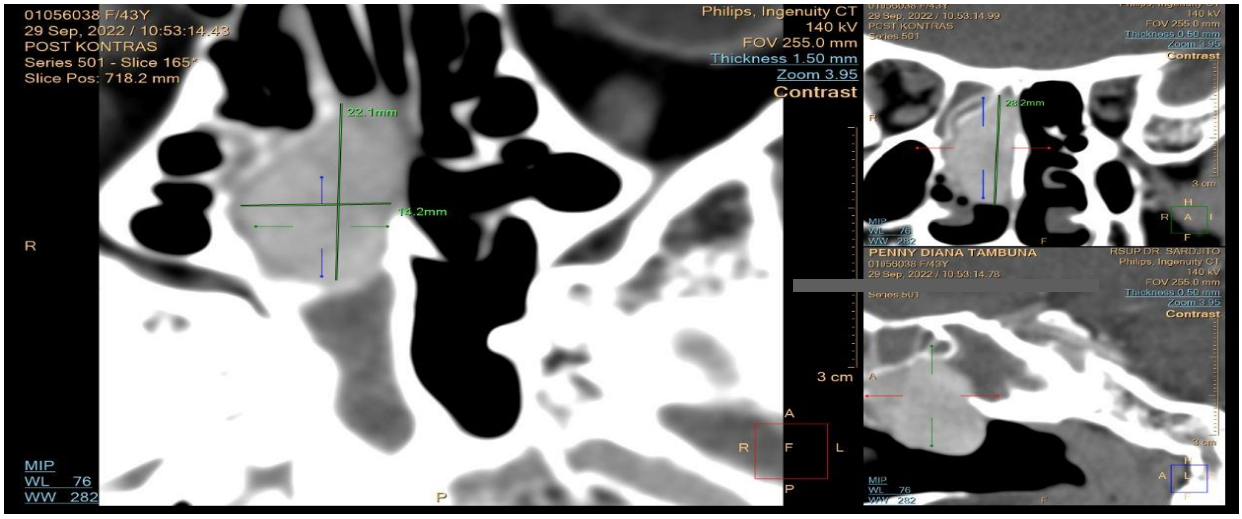

Fig 2 The size of the hypervascular mass in the right nasal cavity extending into the ethmoidal sinus is suspected to be a cavernous hemangioma.

III. RESULT AND DISCUSSION

Malignant peripheral nerve sheath tumors (MPNSTs) are a rare, aggressive neurogenic tumor, accounting for only 5-10% of soft tissue sarcomas (4). The most common anatomical sites are the proximal parts of the upper extremities, lower extremities, and trunk. Studies have shown that MPNSTs typically occur in major nerve trunks, such as the sciatic nerve, brachial plexus, and sacral plexus (5). However, our case of MPNST demonstrates a nasal tumor location, an entity that has not been previously reported.The size of the MPNST in our case is considered small, considering that the average size of MPNST in previous studies was reported to be more than 5 cm because this tumor can grow rapidly. (6)This raises the suspicion that the MPNST in our case was an early-stage MPNST that was discovered early, so its size had not yet reached its typical size. Another suspicion is the limited space in the nasal cavity with the presence of the osteomeatal complex that prevents the tumor from growing further and its nature that tends not to destroy/invade bone.Tumor margins also need to be considered in MPNST because these tumors can infiltrate the surrounding soft tissue and cause peritumoral edema, resulting in ill-defined margins.

(7)In our case, we had regular tumor margins, an atypical finding that led to our initial CTA diagnosis of cavernous hemangioma. In theory, benign neurogenic tumors tend to be well-defined and are usually surrounded by a capsule. However, some plexiform neurofibromas also have an infiltrative appearance. Indistinct margins and peritumoral edema can be useful but less specific signs for the diagnosis of MPNST (7,8). Both were not found in this case because the tumor was directly adjacent to the bone, making accurate assessment of the margin and peritumoral edema difficult.Cavernous hemangioma, a benign blood vessel tumor, can be identified well on a CT scan. This scan reveals a hyperdense lesion with homogeneous contrast enhancement and regular edges. (9,10). In our case, a well-defined, round, hyperdense lesion with a density similar to that of blood products was seen, without calcifications, and minimally enhanced homogeneously with contrast. The tumor did not cause edema or a mass effect due to its location in the nasal cavity, surrounded by a firm osteomeatal complex. This appearance is more consistent with the diagnosis of cavernous hemangioma, but histopathology suggested a diagnosis of MPNST.

IV. CONCLUSION

Differentiating MPNST from other, more common tumors is radiologically challenging due to the extremely rare incidence of MPNST in the nasal cavity and its radiological features, which can mimic cavernous hemangioma. It is important for radiologists to consider MPNST in the differential diagnosis of nasal masses.

REFERENCES

[1] Amirian ES, Goodman JC, New P, Scheurer ME. Pediatric and adult malignant peripheral nerve sheath tumors: an analysis of data from the surveillance, epidemiology, and end results program. ***J Neurooncol.*** 2014 Feb;116(3):609–16.

[2] Knight SWE, Knight TE, Santiago T, Murphy AJ, Abdelhafeez AH. Malignant Peripheral Nerve Sheath Tumors—A Comprehensive Review of Pathophysiology, Diagnosis, and Multidisciplinary Management. Children. 2022 Jan;9(1).

[3] Sharma S, Shah JS, Bali H. Malignant peripheral nerve sheath tumor: A rare malignancy. ***J Oral Maxillofac Pathol.*** 2020 Feb;24(Suppl 1):S86.

[4] Woo OH, Yong HS, Lee JB, Kim A, Koo BH, Kang EY. A giant malignant peripheral nerve sheath tumour of the breast: CT and pathological findings. ***Br J Radiol.*** 2007;80(950).

[5] Rafailidis V, Kaziani T, Theocharides C, Papanikolaou A, Rafailidis D. Imaging of the malignant peripheral nerve sheath tumour with emphasis on ultrasonography: correlation with MRI. ***J Ultrasound***. 2014;17(3):219.

[6] Soldatos T, Fisher S, Karri S, Ramzi A, Sharma R, Chhabra A. Advanced MR imaging of peripheral nerve sheath tumors including diffusion imaging. Semin Musculoskelet Radiol. 2015;19(2):179–90.

[7] Yu YH, Wu JT, Ye J, Chen MX. Radiological findings of malignant peripheral nerve sheath tumor: reports of six cases and review of literature. ***World J Surg Oncol***. 2016 May;14(1).

[8] Sperandio M, Poce I Di, Ricci A, Trapano R Di, Costanzo E, Cello P Di, et al. Malignant Peripheral Nerve Sheath Tumour: CT and MRI Findings. Case Rep Radiol. 2013;2013:1–5.

[9] Al-Sharydah AM, Al-Suhibani SS, Al-Jubran SA, Al-Abdulwahhab AH, Al-Bar M, Al-Jehani HM, et al. Endoscopic management of Atypical sellar cavernous hemangioma: A case report and review of the literature. ***Int J Surg Case Rep***. 2018;42:161.

[10] Pan X, Shen J, Ma Y, Lou H, Weng Y, Zhan R. Imaging characteristics of Intrasellar cavernous hemangioma: A case report. Medicine (Baltimore). 2020 Nov;99(47):e23405.