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## Case Report

# A Rare Case of Primary Central Nervous System Lymphoma Located in the Anterior Geniculate Ganglion Diagnosed by $^{18}$F-FDG PET/CT and MRI

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## Abstract

We presented a rare case of primary central nervous system lymphoma, a subtype of non-Hodgkin lymphoma confined to the brain, unusually occurring in the anterior geniculate ganglion, with no specific symptoms and without initial radiologic features on Magnetic Resonance Imaging (MRI). Following a full neurological evaluation and a Positron Emission Tomography/Computed Tomography (PET/CT) with $^{18}$F-FDG we suspected the presence of a lymphoma that was then confirmed with a biopsy.

**Keywords:** PET/CT; $^{18}$F-FDG; MRI; Primary central nervous system lymphoma; Anterior geniculate ganglion

**Abbreviations:** MRI: Magnetic Resonance Imaging; PET/CT: Positron Emission Tomography/Computed Tomography; CT: Computed Tomography; PNCSL: Primary Central Nervous System Lymphoma; AGG: Anterior Geniculate Ganglion; IAC: Internal Auditory Canal; PCF: Posterior Cranial Fossa; MCF: Middle Cranial Fossa; DWI: Diffusion-Weighted Imaging; DLBCL: Diffuse Large B-Cell Lymphoma; FNS: Facial Nerve Schwannoma; FN: Facial Nerve; FH: Facial Hemangiomas; FNM: Facial Nerve Meningioma; MPNST: Malignant Peripheral Nerve Sheath Tumor; SUV max: maximum Standardized Uptake Value; ADC: the Apparent Diffusion Coefficient

## Introduction

Tumors of the anterior geniculate ganglion are rare facial nerve neoplasms that manifest clinically with symptoms typically related to their brain location. Epidemiologically, the most frequent tumors include Facial Schwannoma, Hemangioma and Meningioma, although other entities can rarely occur.

Primary Central Nervous System Lymphoma (PNCSL) is a rare form of extranodal non-Hodgkin lymphomas, exclusively located in central nervous system, usually presenting as a supratentorial lesion [1]. When rarely occurs in geniculate ganglion, PNCSL can present non-specific neurological symptoms and have radiologic features that mimic, especially in the early stage, other pathologies more frequently involved in this area [2].

## Case Report

A 47 year-old women presented with severe neck pain, right frontal headache, right earache and postural instability. Physical examination revealed only right transmissive hypoacusis and exophoria so imaging was required. A temporal bone Computed Tomography (CT) revealed an enlarged fallopian canal with the presence of dense tissue, cause of restriction of epi/ mesotympanic space and minimal erosion of vestibulum (Figure 1). MRI showed an intra-axial lesion of neoplastic appearance in the Anterior Geniculate Ganglion (AGG), iso/hypointense in T1-weighted,

isointense in T2-weighted, hyperintense in FLAIR sequences, characterized by homogeneous contrast enhancement, extended through the Internal Auditory Canal (IAC). These features leads to the suspicion of Facial Nerve Schwannoma (FNS) and the patient was scheduled for surgical resection. After one month characterized by worsening of symptoms, a $^{18}$F-FDG PET/CT was performed in order to exclude other malignancy or other extracranial Schwannoma, which showed an intense, pathological and homogeneous uptake of the radiotracer located in petrous part of temporal bone, with a Maximum Standardized Uptake Value of 23.55, extended to Posterior Cranial Fossa (PCF) and Middle Cranial Fossa (MCF) (Figure 2). Thus, an MRI was repeated which confirmed the presence of a neoplastic lesion centered on AGG of the facial nerve, clearly increased in size with the involvement of PCF and MCF, compressing the right middle cerebellar peduncle, deforming the fourth ventricle, extended to extra-cranial carotid space and surrounding the proximal part of the internal carotid artery. Furthermore, Diffusion-Weighted magnetic resonance Imaging (DWI) revealed a restriction of signal, leading to the suspicion of a tumor characterized by high cellularity (Figure 3). For this reason, the patient underwent a biopsy, which described a tissue with diffuse lymphatic proliferation with a population of cell population CD20+, CD79a+, CD10-, bcl6+, bcl10+ and concluded with a diagnosis of Diffuse Large B-Cell Lymphoma (DLBCL).

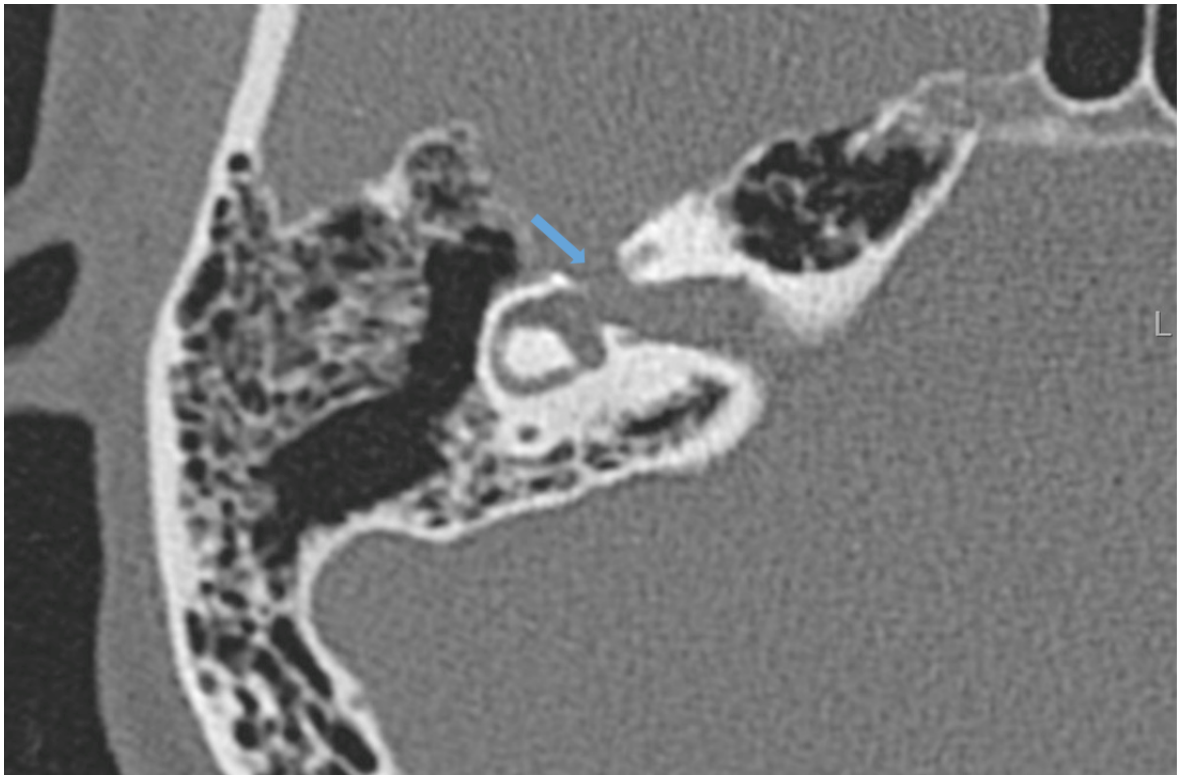


**Figure 1:** Axial image of high resolution of temporal bone computed tomography showing the enlargement of Fallopian Canal (blue arrow).

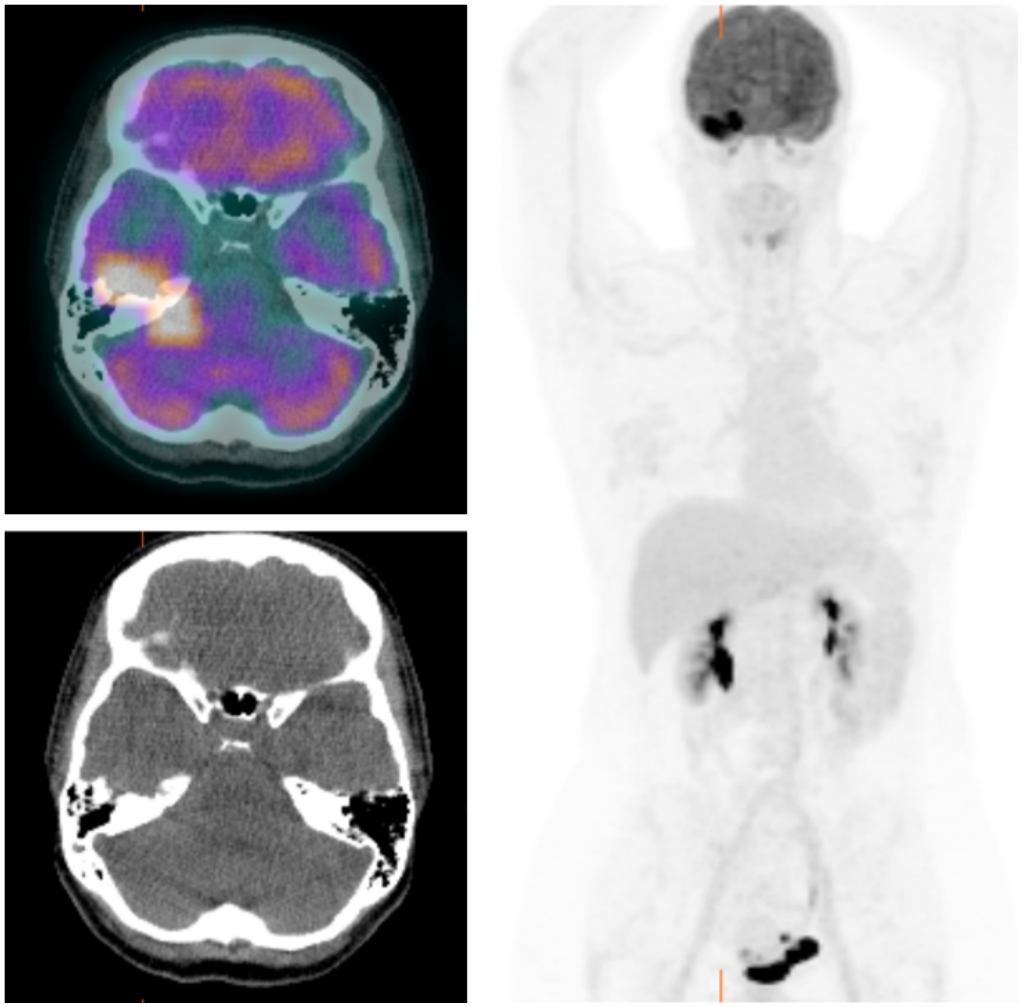

**Figure 2:** Pathological uptake of 18F-FDG in the well-known expansive lesion located on the right petrous rock, which determines (or with concomitant) osteolytic phenomena and extends, both anteriorly, to the middle cranial fossa, and posteriorly to the region of the ponto-cerebellar angle, in the posterior cranial fossa.

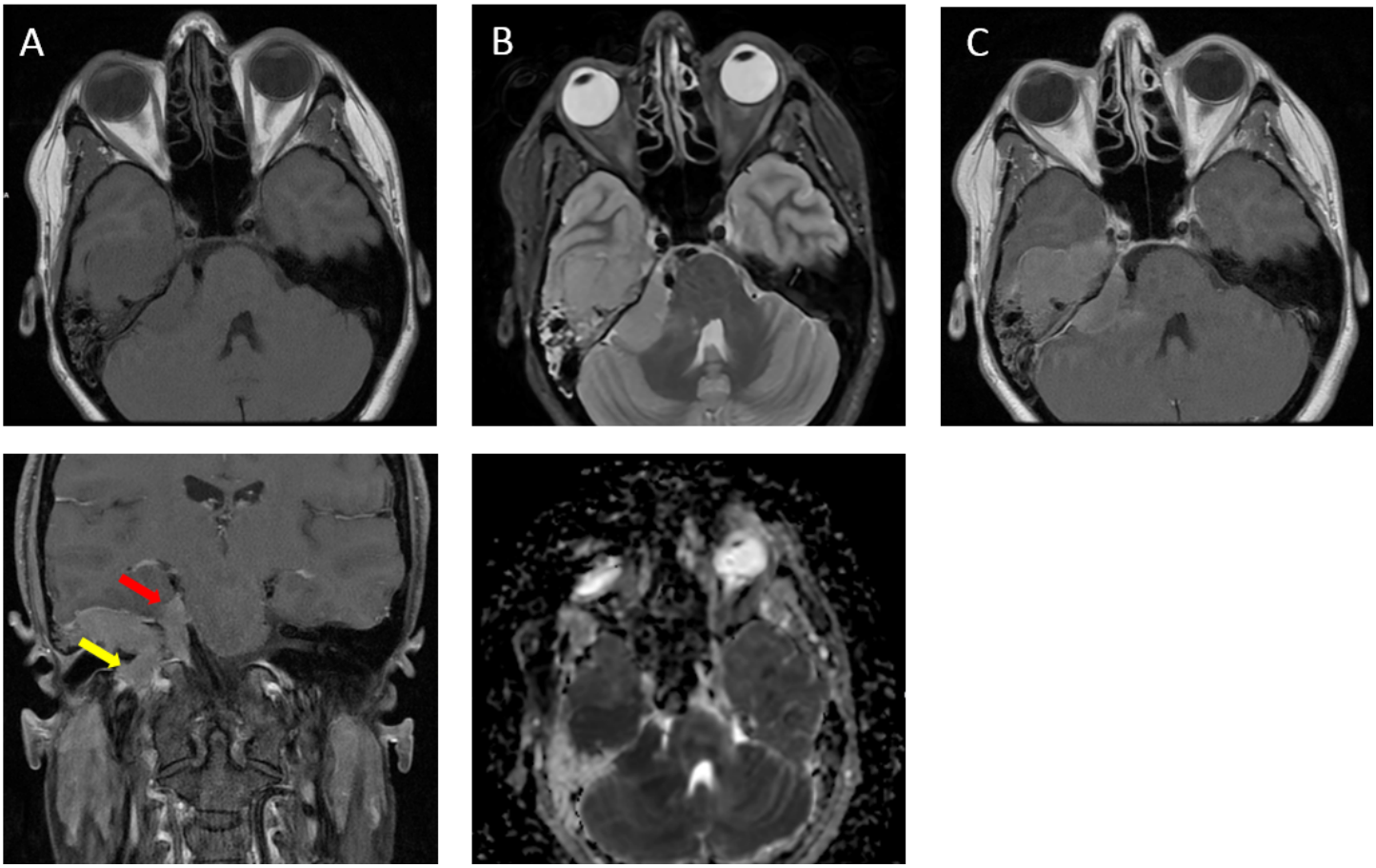


**Figure 3:** Neoplastic lesion involving the PCF and the MCF, isointense in T1-weighted (A), isointense in fat suppressed T2-weighted (B), with contrast enhancement (C,D) and ADC values extremely low in DWI MRI sequences (E), typical of tumor with high cellularity; in the coronal plan (D), the lesion clearly appears as infiltrative mass determining mass effect in the temporal lobe, imprinting the cerebellopontine angle cistern (red arrow) and extending caudally through the jugular bulb (yellow arrow).

## Discussion

Tumors located in the AGG are rare lesions characterized by a common clinical presentation, consisting of facial paralysis, herring loss, headache, postural instability and others nonspecific symptoms [3]. For this reason, imaging plays an important role to guide the diagnosis and the treatment. The most frequent neoplasms include Schwannoma, Hemangioma and Meningioma [3].

FNS can occur along any segment of Facial Nerve (FN) and they are often sausage-shaped expanding long segment of the FN [4]; there may be more than one "foci", for example, a component in the IAC and a component in MCF connected via a narrow waist through the labyrinthine FN. This feature allows the distinction between facial and acoustic schwannoma, indistinguishable especially if no extension into the labyrinthine segment of the facial nerve is present [5].

On MRI they are typically described as circumscribed fusiform enhancing mass, iso-hypointense relative to gray matter on T1-weighted images, hyperintense on T2-weighted series, and with homogeneous contrast enhancement [6].

Facial Hemangiomas (FH) are rare and slow-growing vascular malformations, representing the only 0.7% of petrous bone tumor, T1-iso or slightly hypointense and T2-hyperintense on MRI (more so than the typical schwannoma), avid contrast enhanced and with the pathognomonic presence of internal honeycomb ossific matrix on CT images, due to the capacity to form bone [6]; although FH is mostly located in the AGG, it can also occurs rarely in the IAC, involving the adjacent bone, with more aggressive bony changes, differently from FS [7].

Facial Nerve Meningioma (FNM) are extremely rare neoplasm that arise from arachnoid cells of meninges. To distinguish FNM and FS can be challenge, particularly if the tumor is confined

to IAC; their radiologic features are quite similar, including the hypo-isointensity on T1-MRI sequences, hyperintensity on T2-MRI sequence and enhancement with contrast [8]. At CT, FNM can present expansion of IAC and can have calcifications within the mass [9].

Other, less frequent, etiologies to be considered in the differential diagnosis of tumor of GGA are epidermoid cyst [10], primary brain tumor, metastases, perineural spread of other malignancies, lymphoproliferative disorders [2].

In the first MR study the imaging was not characteristic for any of the above described tumors but, considering the symptoms and the epidemiological frequency, the first hypothesis was FNS and the patient was scheduled for surgical resection.

Schwannoma is a benign encapsulated slow-growing lesion, arising from the abnormal proliferation of Schwann cell. The most frequent regions involved are the acoustic and the facial nerve, followed by the trigeminal nerve [6].

Despite Schwannomas are usually solitary lesions, multiple schwannomas can rarely develop sporadically or in genetic disorders like neurofibromatosis type 2 and schwannomatosis (an autosomal dominant genetic syndrome characterized by the presence of two or more schwannomas) [11].

Furthermore, even if it is a rare event, a benign Schwannoma can also degenerate in a more aggressive tumor called Malignant Peripheral Nerve Sheath Tumor (MPNST), with a consequent poorer prognosis [12].

The rational of using $^{18}$F-FDG PET/CT relies on the possibility to detect synchronous schwannomas, both intra- and extra-cranial tumors; moreover, even if the degree of FDG uptake is not useful to discriminate benign Schwannoma and MPNST, the maximum Standardized Uptake Value (SUV max) can reflects more aggressive behavior and consequently can guide to take further decision to change the treatment [11,13].

Schwannomas have a wide variation in FDG uptake; Miyake KK, et al. retrospectively analyzed 22 cases of schwannoma before surgery, reporting a SUVmax range from 1.5 to 17.3 [14]. In 2015, a study conducted by Ahlawat analyzed the metabolic characteristics of MPNST in patients with schannomatosis and reported a median SUVmax value of 6 (range 2.1-11.7) at early time point, to 10 (range 2.7-15.3) at later time point) [15].

Beaulieu, et al. studying the FDG behavior of nine patients scheduled for surgery, reached that the FDG uptake can be variable and that a SUV max of 6.0 cannot exclude Schwannoma from the differential diagnosis [16].

Our PET/TC study, requested for a rapid progression of symptoms, showed an intense FDG uptake, with a SUVmax of 23.5 that, together with the fast increase in size at MRI (less than 2 months) and temporal bone erosion, indicate a more aggressive tumor, than a Schwannoma suggesting a MPNST or primary brain tumor or cerebral lymphoma.s

Primary Central Nervous System Lymphomas (PCNSL) are aggressive non-Hodgkin lymphomas (NHL) confined to the brain, eyes, spinal cord or leptomeninges [17]. In immunocompetent patients are rare and symptoms are usually related to the site of CNS involvement (1 in the majority of cases, PCNSL presents as a single brain lesion, usually supra-tentiorial, consisting of highly proliferative cells. The most common histologic subtype is DLBCL (90%), characterized on MRI images by homogeneous contrast enhancement, presence of edema surrounding the mass lesion on FLAIR sequences and restricted diffusion within the tumor on DWI. In particular, the extremely high DWI signal intensity is typical in PCNSL, according to the cell density of this tumor, and the Apparent Diffusion Coefficient (ADC) value obtained from DWI, correlates well with tumor cellularity and represents a predicting factor of response to chemotherapy [18].

On $^{18}$F-FDG PET/TC, PCNSL is typically FDG avid, with homogeneous glucose uptake higher than other brain tumors [9]. A recent meta-analyses showed an high performance of FDG in the pre-treatment phase [10] and an important role for excluding other whole body localizations [19].

## Conclusion

Even if PCNSL is a tumor that can rarely occur in the GGA, it has to be considered in the differential diagnosis of facial nerve tumors, mainly in the early stage of the disease when it can mimic other lesions typically located in this area, such as FNS that can be surgically approached. $^{18}$F-FDG PET/TC can be useful to guide the diagnosis and for therapy decision making, particularly in patients with rapid progression of symptoms.

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## References

1. Grommes C, DeAngeli LM (2017) Primary CNS lymphoma. Journal of Clinical Oncology 35: 2410-2418.
2. Jayashankar N, Kodur S, Patkar D, Verma M (2021) Primary Lymphoma of Internal Acoustic Meatus Mimicking Vestibular Schwannoma-A Rare Diagnostic Dilemma. J Neurol Surg Rep 82: e1-e5.
3. Lahlou G, Nguyen Y, Russo FY, Ferrary E, Sterkers O, et al. (2016) Geniculate ganglion tumors: clinical presentation and surgical results. Otolaryngol Head Neck Surg 155: 850-855.
4. Mundada P, Purohit BS, Kumar TS, Tan TY (2016) Imaging of facial nerve schwannomas: diagnostic pearls and potential pitfalls. Diagn Interv Radiol 22: 40-46.

**5.** Eshraghi AA, Oker N, Ocak E, Verillaud B, Babcock T, et al. (2019) Management of facial nerve schwannoma: a multicenter study of 50 cases. J Neurol Surg B Skull Base 80: 352-356.

**6.** Wiggins RH, Harnsberger HR, Salzman KL, Shelton C, Kertesz TR, et al. (2006). The many faces of facial nerve schwannoma. AJNR Am J Neuroradiol 27: 694-699.

**7.** Yue Y, Jin Y, Yang B, Yuan H, Li J, et al. (2015) Retrospective case series of the imaging findings of facial nerve hemangioma. Eur Arch Otorhinolaryngol 272: 2497-2503.

**8.** Gao W, Zi D, Lu L (2020) Facial Nerve Meningioma: A Case Mimicking Facial Nerve Schwannoma. Ear Nose Throat J 145561320962582.

**9.** Magliulo G, Alla FR, Colicchio G, Trasimeni G (2010) Geniculate ganglion meningioma. Skull Base 20: 185-188.

**10.** Czernicki T, Kunert P, Nowak A, Wojciechowski J, Marchel A (2016). Epidermoid cysts of the cerebellopontine angle: Clinical features and treatment outcomes. Neurol Neurochir Pol 50: 75-82.

**11.** Sérézal IG, Ferkal S, Lerman L, Mulé S, Funalot B, et al. (2021) $^{[18F]}$ FDG Positron emission tomography with whole body magnetic resonance imaging ($^{[18F]}$ FDG-PET/MRI) as a diagnosis tool in Schwannomatosis. Orphanet J Rare Dis 16: 49.

**12.** Wong WW, Hirose T, Scheithauer BW, Schild SE, Gunderson LL (1998) Malignant peripheral nerve sheath tumor: analysis of treatment outcome. Int J Radiat Oncol Biol Phys 42: 351-360.

**13.** Halac M, Cnaral F, Sait S, Ylmaz S, Kerim S, et al. (2008) FDG PET/CT findings in recurrent malignant schwannoma. Clin Nucl Med 33: 172-174.

**14.** Miyake KK, Nakamoto Y, Kataoka TR, Ueshima C, Higashi T, et al. (2016). Clinical, morphologic, and pathologic features associated with increased FDG uptake in schwannoma. AJR Am J Roentgenol 207: 1288-1296.

**15.** Ahlawa S, Baig A, Blakeley JO, Jacobs MA, Fayad LM (2016) Multiparametric whole□body anatomic, functional, and metabolic imaging characteristics of peripheral lesions in patients with schwannomatosis. J Magn Reson Imaging 44: 794-803.

**16.** Beaulieu S, Rubin B, Djang D, Conrad E, Turcotte E, et al. (2004) Positron emission tomography of schwannomas: emphasizing its potential in preoperative planning. AJR Am J Roentgenol 182: 971-974.

**17.** Batchelor TT (2019) Primary central nervous system lymphoma: A curable disease. Hematol Oncol 37: 15-18.

**18.** Huang WY, Wen JB, Wu G, Yin B, Li JJ, et al. (2016). Diffusion-weighted imaging for predicting and monitoring primary central nervous system lymphoma treatment response. AJNR Am J Neuroradiol 37: 2010-2018.

**19.** Guedj E, Varrone A, Boellaard R, Albert NL, Barthel H, et al. (2021) EANM procedure guidelines for brain PET imaging using [$^{18}$F] FDG, version 3. Eur J Nucl Med Mol imaging 49: 632-651.

**20.** Gupta T, Manjali JJ, Kannan S, Purandare N, Rangarajan V (2021) Diagnostic performance of pretreatment 18F-fluorodeoxyglucose positron emission tomography with or without computed tomography in patients with primary central nervous system lymphoma: updated systematic review and diagnostic test accuracy meta-analyses. Clin Lymphoma Myeloma Leuk 21: 497-507.