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


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 ¹⁸F-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.
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
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.

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-tentorial, 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 18F-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. 18F-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


