Radiology Case Reports 18 (2023) 3729–3734

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Primary large B-cell lymphoma involving the cerebellopontine angle mimic acoustic schwannoma: Role of MR Spectroscopy in differential diagnosis. A case report ☆

Pier Paolo Arcuri, MD[a,*], *Vincenzo Aiello, MD*[b], *Simonetta Antonelli, MD*[a], *Giuseppe Lucio Cascini, MD*[d,f], *Marco Rossi, MD*[c], *Domenico Laganà, MD*[e,g]

[a] *Radiology Unit "De Lellis", Department of Radiology, Dulbecco University Hospital, 88100, Catanzaro, Italy*
[b] *Rheumatology Clinic "Madonna dello Scoglio" Cotronei, Crotone, Italy*
[c] *Department of Hematology-Oncology, Dulbecco University Hospital, 88100 Catanzaro, Italy*
[d] *Nuclear Medicine Unit, Dulbecco University Hospital, 88100, Catanzaro, Italy*
[e] *Department of Experimental and Clinical Medicine, Magna Graecia University of Catanzaro, 88100 Catanzaro, Italy*
[f] *Department of Experimental and Clinical Medicine, 'Magna Graecia' University of Catanzaro, Catanzaro, Italy*
[g] *Radiology Unit, Dulbecco University Hospital, 88100 Catanzaro, Italy*

ARTICLE INFO

Article history:
Received 1 May 2023
Revised 24 July 2023
Accepted 28 July 2023

Keywords:
Large B-cell lymphoma
Acoustic neuroma
Cerebellopontine angle
Meningioma
MR Spectroscopy
Primary CNS lymphoma

ABSTRACT

Primary central nervous system (CNS) lymphoma is a very rare aggressive non-Hodgkin disease that originates in CNS (brain, leptomeninges, spinal cord, or eyes). It seems to have increased over the last two decades in both immunocompromised and immunocompetent patients. Primary large B-cell lymphoma involving the cerebellopontine angle (CPA) is extremely rare: only 15 cases of large B-cell lymphoma of the CPA have been reported worldwide; based on our knowledge, no cases studied with MR Spectroscopy. Primary large B-cell lymphoma of the CPA must be differentiated from other cerebellopontine angle diseases, such as acoustic neuroma and meningioma. An early and accurate diagnosis of this neoplasm is necessary for the best management because it is a radiosensitive and chemosensitive tumor.

Herein, we report a rare case of B-cell lymphoma involving the left CPA in a 65-year-old man who presented with 3 months of hearing loss on the left, illustrated by MR and TC imaging, highlighting how the MR Spectroscopy, thanks to their greater specificity, is decisive in achieving the correct diagnosis of primary lymphoma and differentiating it from acoustic schwannoma or meningioma. Therefore, in the suspicion of a malignant heteroplastic lesion of the CPA, we suggest including Spectroscopy in the MR study protocol.

☆ Competing Interests: The authors have no conflicts of interest or potential conflicts to disclose.

* Corresponding author.

https://doi.org/10.1016/j.radcr.2023.07.069

Introduction

Central nervous system (CNS) lymphoma is a rare and aggressive tumor that may affect the brain, meninges, spinal cord, and eyes [1]. It is classified into primary CNS lymphoma and secondary CNS lymphoma [1–3,13]. Secondary CNS lymphoma may be CNS localization of systemic lymphoma or an isolated recurrence [3]. It is often not detectable on Imaging Diagnostic tools and cerebrospinal fluid cytology allows for diagnoses this disease more readily [3]. Primary CNS lymphoma is a very rare aggressive non-Hodgkin disease that originates in CNS (brain, leptomeninges, spinal cord, or eyes), it seems to have increased over the last two decades in both immunocompromised and immunocompetent patients [2–8]. An early and accurate diagnosis of this neoplasm is necessary for the best management because it is a radiosensitive and chemosensitive tumor [2]. Primary CNS lymphoma is often localized in the corpus callosum, basal ganglia, thalami, and paraventricular region, while its localization in cerebellopontine angle is rare and only 15 cases have been described thus far [4,5,8], based on our knowledge, no cases studied with MR Spectroscopy. Cerebellopontine angle lymphoma must be differentiated from acoustic neurinomas [9], meningiomas, and epidermoid tumors: all these other diseases are more common than cerebellopontine angle lymphoma [4,8]. All these tumors can cause hearing loss, a condition that is considered an otologic emergency requiring an immediate assessment and therapy when it appears suddenly [6]. Magnetic resonance imaging (MRI) is the gold standard imaging diagnostic tool to evaluate cerebellopontine angle masses and it is considered the method of choice to diagnose and follow-up primary CNS lymphoma [2,4,8,10,11]. Radiological features of this tumor are not specific in conventional MRI and advanced imaging techniques, such as diffusion-weighted imaging (DWI), diffusion tensor imaging (DTI), magnetic resonance perfusion (MRP), and MR spectroscopy (MRS) have revealed themselves as useful to differentiate primary CNS lymphomas from other neoplasms [2,3,7]. In our case, MR Spectroscopy played an important role to diagnose a primary lymphoma of CPA and differentiate it from an acoustic schwannoma or meningioma.

Case presentation

We present the case of a 65-year-old man who came to our Hospital after about 3 months of hearing loss on the left. The ENT consultation recommends, in agreement with the neurologist, suspecting a pathology involving the vestibulocochlear nerve, an MRI examination of the brain with a particular study of the pontocerebellar angle, before and after contrast media. The patient was not immunocompromised nor affected by current or previous oncological pathologies.

We performed a brain MRI scan completed with medium postcontrast sequence (Gadobenate dimeglumine, 1.0 mol/L) on a 1.5 Tesla MRI scanner. Obtained Spin Echo (SE) T1-weighted sequence, Turbo Spin Echo (TSE) T2-weighted sequence, FLAIR (Fluid-Attenuated Inversion Recovery) sequence, DWI sequence, Fast Field Echo (FFE) T2-weighted sequence, isotropic FFE T1-weighted sequence after contrast medium administration and a single-voxel MR spectroscopy using Two-dimensional pointed resolved spectroscopy (2D PRESS) sequence. MRI scan showed the presence of a focal lesion, maximum diameter = 15 mm localized in the left cerebellopontine angle extending into the cisternal portion of the left trigeminal nerve, showing an apparent solution of continuity, especially in DWI (Fig. 2A), with the ipsilateral vestibulocochlear nerve (VIII). The lesion was weakly hypointense on T1-weighted images (Fig. 1A), it showed intermediate signal intensity on TSE T2 (Fig. 1B), FFE T2 (Fig. 1C) and FLAIR (Fig. 1D).

No mass effect or perilesional edema was observed. The lesion showed restricted diffusion with hyperintensity on Diffusion-Weighted Images (Fig. 2A) and low ADC values ($0.62 \times 10^{-3} mm^2/s$) on ADC maps (Fig. 2B). MRS (Fig. 3) showed a high choline peak, slightly high N-acetylaspartate (NAA) peak, and low levels of creatine and lactate. NAA/Cr = 1.57, Cho/Cr = 7.09, NAA/Cho = 0.22, Cho/NAA= 4.50. After IV contrast agent injection, the lesion showed intense and homogeneous enhancement (Figs. 4A and B). Spectroscopy features and intense and homogeneous enhancement suggested the diagnosis of lymphoma rather than the diagnosis of acoustic schwannoma or meningioma. After MRI scan the patient

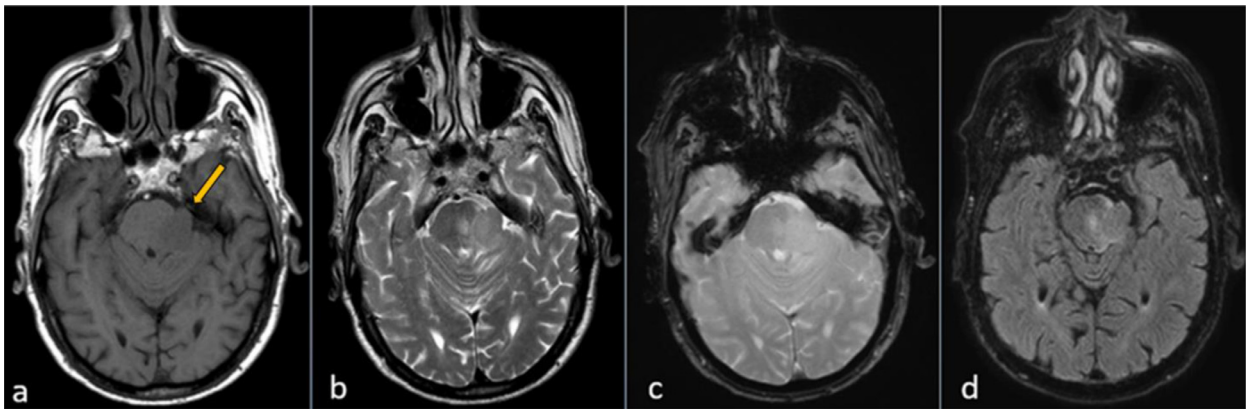

Fig. 1 – MRI detected a lesion in the left cerebellopontine angle, moderate hypointense on T1-weighted images (A), intermediate signal intensity on T2-weighted images (B) and on FFE T2-weighted image (C), intermediate/high signal intensity on FLAIR (D). No significant mass effect was detected.

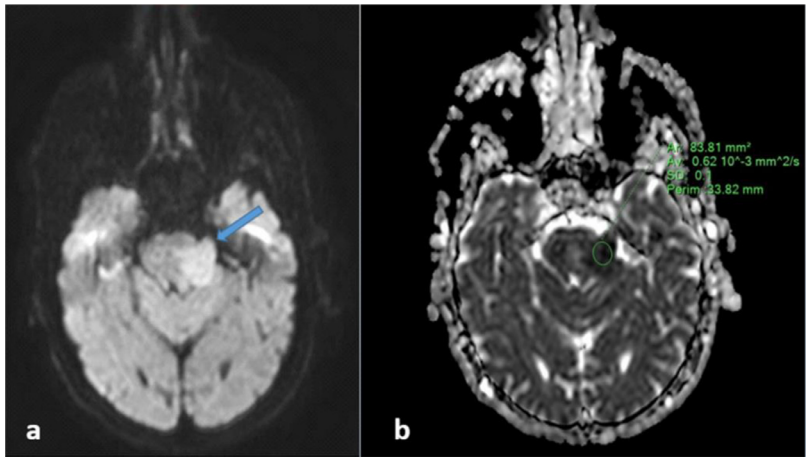

Fig. 2 – The lesion showed restricted diffusion with hyperintensity on diffusion-weighted images (A) and low ADC values ($0.62 \times 10^{-3} mm^2/s$) on ADC maps (B).

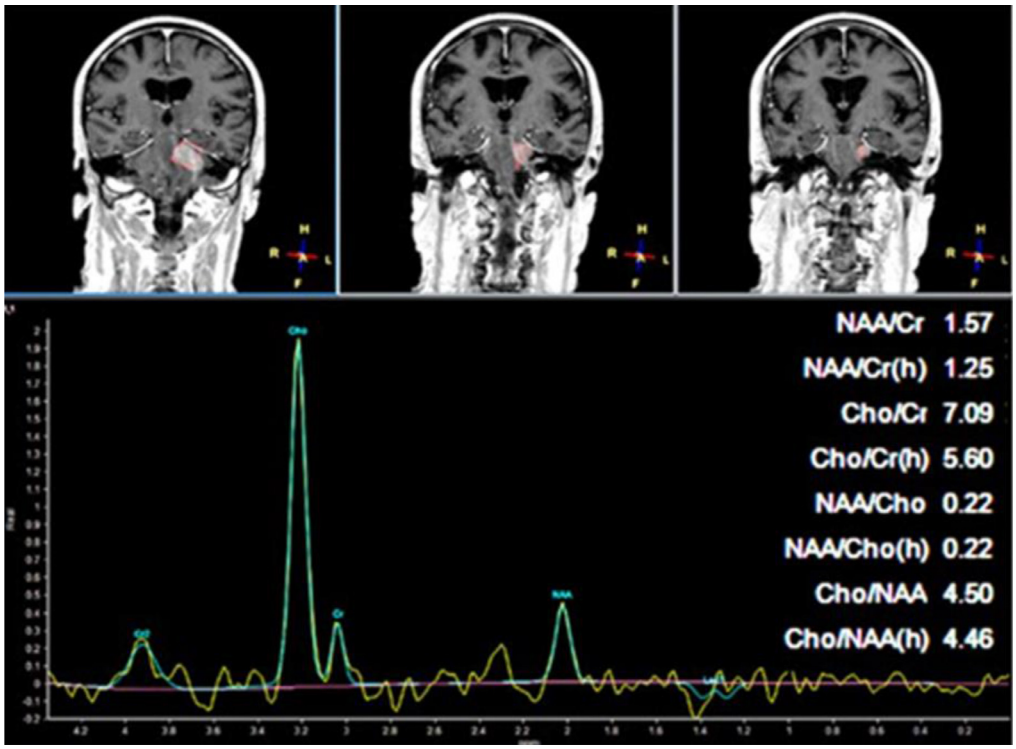

Fig. 3 – MR Spectroscopy demonstrated high choline peak, slightly high NAA peak and low values of creatine and lactate: these features suggested the diagnosis of lymphoma rather the diagnosis of acoustic schwannoma or meningioma.

underwent a whole-body computer tomography (CT) scan and a lumbar puncture. Cerebrospinal fluid analysis was negative, and CT revealed no other organs with tumors, except for the presence of the known brain lesion, localized to the left PCA, and the presence of multiple enlarged latero-cervical and axillary lymph nodes. Therefore, a biopsy of a latero-cervical lymph node (level 3) was performed.

The biopsy revealed the presence of diffuse proliferation of large lymphoid cells. Scattered mitoses with small reactive lymphocytes are seen (Fig. 5A): the picture appeared indicative of metastatic lesions of diffuse large B-cell lymphoma (aggressive). In the immunohistochemical study (CD 20+) cytoplasmic positivity confirmed the diagnosis of large B-cell lymphoma (Fig. 5B).

The patient was therefore referred to the hematologist who administers a therapy consisting of rituximab, methotrexate, cytarabine, and dexamethasone.

After 3 months from the beginning of the therapy, to the reduction of the symptoms, a CT examination of the brain was performed. Follow-up CT scan showed a significant

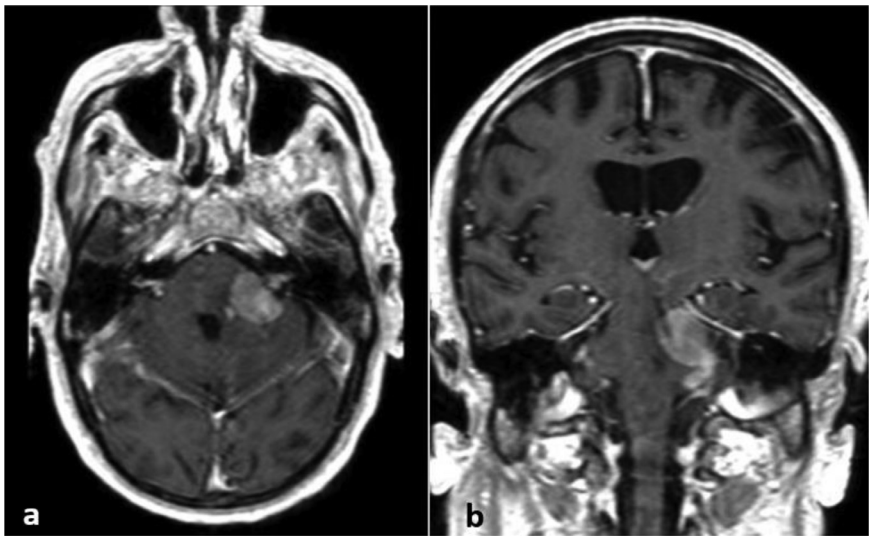

Fig. 4 – FFE T1-weighted sequences after contrast medium administration in the axial plane (A) and coronal plane (B): the lesion was homogenously contrast-enhanced.

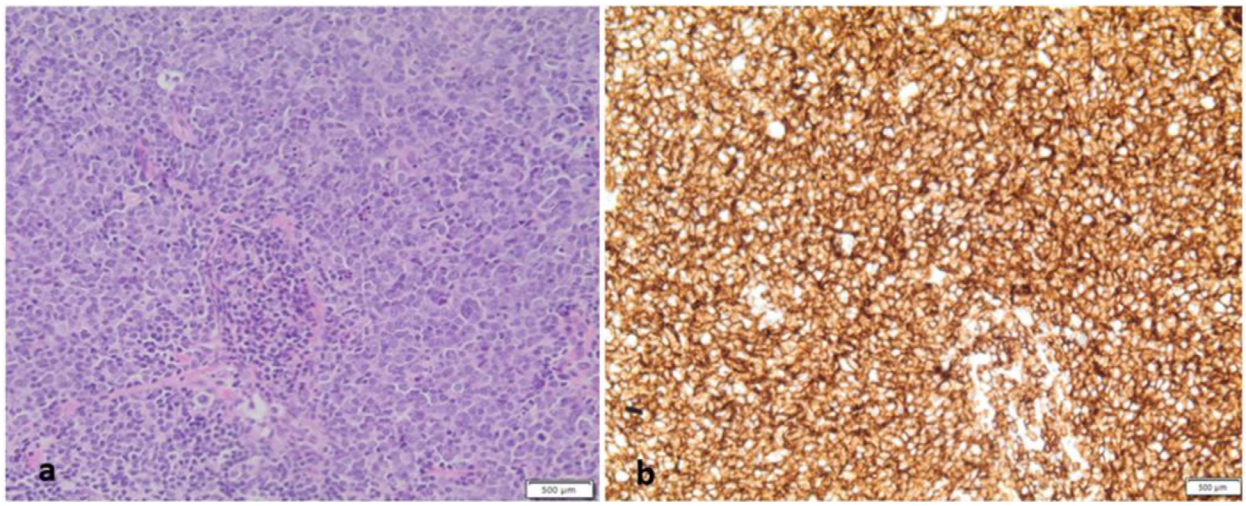

Fig. 5 – Histopathology. Hematoxylin-eosin stain (A) x 100 magnification, shows diffuse proliferation of large lymphoid cells. Scattered mitoses with small reactive lymphocytes are seen. Immunohistochemistry (B) x 100 magnification, CD 20+. Cytoplasmic positivity confirms the diagnosis of diffuse large B-cell lymphoma (aggressive).

decrease in the brain lesion, demonstrating the therapeutic efficacy (Fig. 6). The patient progressed to undergoing autologous hematopoietic stem cell transplant with noted febrile neutropenic episodes. The patient remains alive 20 months after diagnosis.

Discussion

Cerebellopontine angle tumors are a varied group of neoplasms: the most common CPA tumor is acoustic neuroma, also called vestibular schwannoma. The second most common CPA neoplasm is meningioma [7,12]. Primary CNS lymphoma is a rare tumor and primary cerebellopontine angle lymphoma is much rarer, only 15 cases of large B-cell lymphoma at the CPA have been reported worldwide [4,6,8], and no cases studied with MR Spectroscopy. The lesion we observed presented restricted diffusion on DWI. Acoustic neuroma is generally isointense on DWI, so it does not show restricted diffusion [12]. Meningiomas do not generally show restricted diffusion on DWI, except grade II-III meningiomas [3]. CNS lymphoma shows restricted diffusion in DWI and low apparent diffusion coefficient (ADC) values because this neoplasm is highly cellular, but restricted diffusion may also be observed in acute ischemic stroke, central necrosis of brain abscesses, solid portion of high-grade gliomas, some metastases and grade II-III meningiomas [2,3,7,10]. In the few cases of diffuse large B-cell lymphoma of the CNS studied with MR Spectroscopy (not specifically of the PCA) MRS revealed a

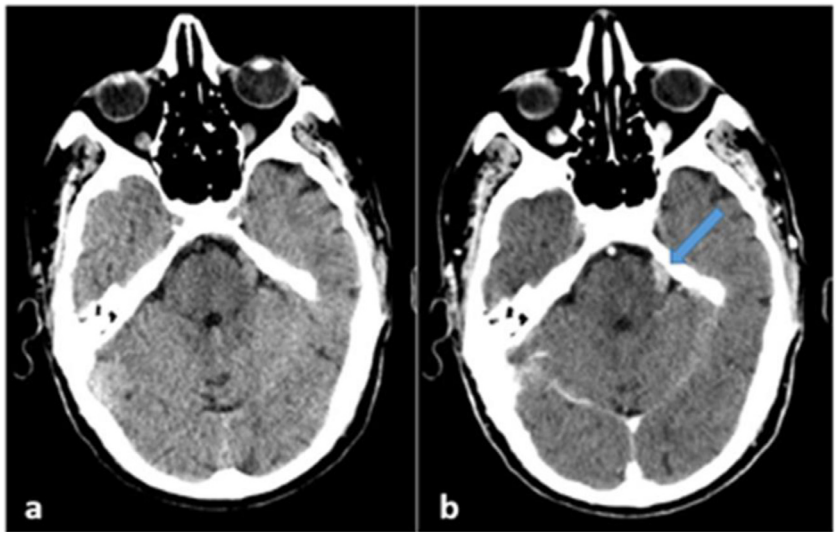

Fig. 6 – CT scan after chemotherapy: before contrast, agent administration (A) showed a significant dimensional reduction of the lesion in the left cerebellopontine angle. In a CT scan after contrast agent administration (B) the lesion maintains the characteristic postcontrast enhancement.

decrease in N-acetyl aspartate (NAA) peaks, as well as an inversion of the choline/creatinine and choline/NAA ratios and the presence of Choline peaks, indicating cell proliferation.

In our case, MR Spectroscopy revealed itself very useful to suggest the possible diagnosis of lymphoma: spectra of the lesion were marked by a high choline peak, slightly high NAA peak, and low values of creatine and lactate. The ratios between the various metabolites were: NAA/Cr = 1.57, Cho/Cr = 7.09, NAA/Cho = 0.22, Cho/NAA = 4.50. These MRS findings were quite similar to primary CNS lymphoma magnetic resonance spectroscopy (MRS) features, which are a significant rise of choline, NAA, and creatine decrease, high Choline/Creatine (Cho/Cr) and Choline/N-acetylaspartate (Cho/NAA) ratios and lipidic peak [2,3,5,7]. MRS helps to differentiate primary CNS lymphoma from other lesions, such as acoustic neuroma and meningioma: in fact, acoustic neuroma shows myo-inositol peak and high choline peak; myo-inositol peak is maybe related to microglial activation and astrogliosis, a feature that is observed in this lesion [2,3,11]. On MRS meningioma spectra are marked by an alanine peak, high choline peak, glutamate (Glu) peak, and low NAA peak; its higher alanine/creatine ratios are useful to differentiate it from other intracranial neoplasms [3,11]. MRS findings may not be useful to distinguish primary CNS lymphoma from glioblastoma and metastases, but some authors reported higher Glu/Cr, Cho/Cr, and glutamate/glutamate+glutamine (Glu/Glu+Gln) ratios in primary CNS lymphoma than in glioblastoma [2,5]. MRS researches to assess differences between primary CNS lymphoma and metastases are not available [5]. Besides studies have not found relevant differences between primary CNS lymphoma and secondary CNS lymphoma in MRI features [1].

This rare case presented, showed metabolite values very similar to those found in the primary CNS lymphoma MRS study [7], except for the absence of high lipid and lactate values. In our case, the absence of the lipid peak can be justified by the absence of significant phenomena of damage to the brain tissue, as occurs during brain tumor growth and the absence of Lactate (Lac) production are most likely due to the absence of necrotic phenomena contextual to the lesion.

Another peculiarity of our case is the absence of extensive perilesional vasogenic edema, contrary to the case presented by Seevaratnam et al. [14]. Therefore, the presence of perilesional edema does not represent a pathognomonic aspect of primary PCA lymphoma.

Conclusions

MRS is useful to differentiate primary CNS lymphoma from glioblastoma and other cerebellopontine angle lesions, such as acoustic neuroma and meningioma. In the case we presented, MRS proved to be decisive in orienting the diagnosis for lymphoma of the CPA, indeed MRS showed many of the metabolic characteristics of the primary large B-cell lymphoma of the CNS. Therefore, in the suspicion of a malignant heteroplastic lesion of the cerebellopontine angle, we suggest including MR Spectroscopy in the study protocol to increase the specificity of the examination.

Patient consent

Written informed consent was obtained from the patient.

REFERENCES

[1] Malikova H, Burghardtova M, Koubska E, Mandys V, Kozak T, Weichet J. Secondary central nervous system lymphoma:

spectrum of morphological MRI appearances. Neuropsychiatr Dis Treat 2018;14:733–40. doi:10.2147/NDT.S157959.
[2] Joshi A, Deshpande S, Bayaskar M. Primary CNS lymphoma in immunocompetent patients: appearances on conventional and advanced imaging with review of literature. J Radiol Case Rep 2022;16(7):1–17. doi:10.3941/jrcr.v16i7.4562.
[3] Lyndon D, Lansley JA, Evanson J, Krishnan AS. Dural masses: meningiomas and their mimics. Insights Imaging 2019;10:11. doi:10.1186/s13244-019-0697-7.
[4] Yan C, Kong X, Yang L, Ma W. An uncommon case of lymphoplasmacytic lymphoma in cerebellopontine angle region. Medicine (Baltimore) 2016;34:e4627. doi:10.1097/MD.0000000000004627.
[5] Chiavazza C, Pellerino A, Ferrio F, Cistaro A, Soffietti R, Rudà R. Primary CNS lymphomas: challenges in diagnosis and monitoring. Biomed Res Int 2018;2018:3606970. doi:10.1155/2018/3606970.
[6] Wang Y-T, Su H-H, Hou Y, Chu S-T, Lai P-H, Tseng H-H, et al. Diffuse large B-cell lymphoma of the cerebellopontine angle in a patient with sudden hearing loss and facial palsy. J Chin Med Assoc 2007;70(7):294–7. doi:10.1016/S1726-4901(07)70008-4.
[7] Housni A, Boujraf S, Alami B, Benzagmout M, Maaroufi M. Assessment of primary brain lymphoma using multimodal magnetic resonance imaging and proton magnetic resonance spectroscopy. Asian J Neurosurg 2018;13(4):1205–8. doi:10.4103/ajns.AJNS_137_17.
[8] Wen C-Y, Hao-Wuang P, Chang C-J. Primary cerebellopontine angle lymphoma. Kaohsiung J Med Sci 2015;31:55–6. doi:10.1016/j.kjms.2014.05.002.
[9] Raikwar KR, Ghodke MH, Deshmukh VB, Garde JB, Suryavanshi RK. Schwannoma of the lower lip mucosa: an unexpected finding. Int J Head Neck Surg 2015;6(1):35–7.
[10] Li L, Rong JH, Feng J. Neuroradiological features of lymphomatosis cerebri: a systematic review of the English literature with a new case report. Oncol Lett 2018;16:1463–74. doi:10.3892/ol.2018.8839.
[11] Khaled M, Moghazy K, Elsaadany W, Eissa L. Additional diagnostic role of MRI spectroscopy, diffusion and susceptibility imaging in differentiation of CPA masses: our experience with emphasis on schwannomas and meningiomas. Egypt J Radiol Nucl Med 2020;51:137.
[12] Kabashi S, Ugurel MS, Dedushi K, Mucaj S. The role of magnetic resonance imaging (MRI) in diagnostics of acoustic schwannoma. Acta Inform Med 2020;28(4):287–91. doi:10.5455/aim.2020.28.287-291.
[13] Da Silva AJ, Castro Pinheiro Gomes FE, Barros Pimentel Tenório GM, Pinto Almeida LM. Diffuse large B-Cell lymphoma in bilateral basal ganglia: a rare case report. Cureus 2022;14(12):e32743. doi:10.7759/cureus.32743.
[14] Seevaratnam V, Li Y, Loong Keegan Lee S, Olsson G. Primary central nervous system lymphoma at the cerebellopontine angle mimicking a trigeminal schwannoma: a unique case report and literature review. J Clin Neurosci 2018;52:115–19. doi:10.1016/j.jocn.2018.01.016.