

Primary Central Nervous System Lymphoma of the Cerebellopontine Angle: Epidemiological Characteristics and Imaging Features may be Helpful for Making an Accurate Preoperative Diagnosis

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Abstract

Purpose

Primary central nervous system lymphoma (PCNSL) of the cerebellopontine angle (CPA) is rare, thus the preoperative diagnosis is still a challenge.

Methods

We searched the database at our institution and performed a search of English literature in PubMed and Google Scholar. Keywords used were as follows: “primary central nervous system lymphoma”; “cerebellopontine angle”; “lymphoma”. Only cases in the English language that were located in the CPA and contained adequate clinical information pertinent to the analysis were included.

Results

297 cases of pathologically confirmed PCNSLs were recorded between January 2009 and October 2020 at our institution. 6 cases were located in the CPA, accounting for 2.0% of all PCNSLs. 26 cases meeting the above criteria were found in the literature. Including ours, a total of 32 patients were analyzed. Females were more frequently affected (F/M ratio, 2.2:1). There was a preponderance of left-sided lesions in the PCNSLs of the CPA (L/R ratio, 1.5:1). On CT, 80.0% of them presented as hyperdense lesions. On T1-weighted image, 66.7% appeared isointense. While on T2-weighted image (T2WI), 68.4% appeared isointense/hypointense. After contrast administration, 86.2% presented intense homogeneous enhancement.

Conclusion

PCNSL of the CPA is extremely rare, accounting for 2.0% of all PCNSLs in our study. There is a preponderance of females and left-sided lesions in this disease. Contrast-enhanced magnetic resonance imaging with T2WI is very helpful in the preoperative diagnosis of the CPA PCNSL. Although rare, lymphoma should be included in the differential diagnosis of CPA lesions.

Introduction

Primary central nervous system lymphoma (PCNSL) is a rare malignant extranodal non-Hodgkin's lymphoma that arises in the craniospinal axis [20, 40]. Most PCNSLs are supratentorial in a central location, including the corpus callosum, basal ganglia, thalamus, and paraventricular region [21, 25, 30]. Infratentorial PCNSL is rare, accounting for only 10%-20% of all PCNSLs [2]. PCNSL of the cerebellopontine angle (CPA) is extremely rare and would probably be mistaken for other CPA lesions such as vestibular schwannomas or meningiomas [2, 21, 30]. To the best of our knowledge, only 26 cases of CPA PCNSLs have been previously reported in the English literature [1–4, 9, 11, 13, 14, 17–19, 21, 22, 25–32, 34, 36–39]. We present six new cases of CPA PCNSLs and discuss the presentation, radiologic characteristics, preoperative differential diagnosis and treatment.

Materials And Methods

The neurosurgical database at the Second Affiliated Hospital of Zhejiang University School of Medicine, was searched for all surgical cases of primary central nervous system lymphomas between January 2009 and October 2020. A retrospective chart review was then performed with research ethics board approval. The age, sex, image

presentation, location, preoperative diagnosis, surgical management and pathological examination were recorded. All patients underwent computed tomography (CT) or magnetic resonance imaging (MRI) for diagnosis and surgical planning. Postoperative clinical information and neuroradiological presentation were evaluated.

Additionally, for the review, a search was conducted of the English literature in PubMed and Google Scholar for every case report, series, letter to the editor, original article and literature review related to PCNSL of the CPA. In addition, the reference lists of the obtained articles and previous reviews were examined. Keywords used were as follows (single word or combination): “primary central nervous system lymphoma”; “cerebellopontine angle”; “lymphoma”. Only cases in the English language that were located in the CPA and contained adequate clinical information pertinent to the analysis were included.

Statement of human and animal rights

All procedures performed involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. For this type of study, formal patient consent is not required. Ethical approval for this study was obtained from the Ethics Committee of the Second Affiliated Hospital, Zhejiang University School of Medicine (ID: 0182/2021).

Results

Two hundred and ninety-seven cases of pathologically confirmed primary central nervous system lymphoma were recorded between January 2009 and October 2020 at our institution. Six of them were located in the CPA, accounting for 2.0% of all PCNSLs in the present study. They comprised two men and four women with an average age of 60.8 years (range 51–69 years). Headache and vomiting were the most common presenting symptoms, and were seen in four cases. Other symptoms and signs included hearing loss, dizziness, dysphagia, hoarseness, facial palsy and facial hypoesthesia. PCNSL of the CPA was frequently found on the left than on the right side (L/R ratio, 2:1). In two patients, tumors were resected via suboccipital retrosigmoid craniotomy. In the other four patients, stereotactic biopsy was performed.

The English medical literature in the PubMed and Google Scholar databases was reviewed, and 26 cases of CPA PCNSLs meeting the above-mentioned criteria were included. The characteristics of the 26 cases were summarized in Table 1. Including our six patients, a total of 32 cases were analyzed. The reported age range was between 21 and 82 years, with a median age of 56.5 years. Females were more frequently affected (F/M ratio, 2.2:1). Regarding the locations of PCNSLs of the CPA, 18 cases (56.3%) were in the left side, 12 cases (37.5%) were in right side, and 2 cases (6.3%) were bilateral. The most frequent initial symptom of CPA PCNSL was hearing disturbance (64.5% of all the patients), and cerebellar signs (ataxia and unsteady gait) were the second most common symptom (41.9% of all the patients). Facial palsy, headache and vomiting were also common symptoms. CT examinations were performed in 15 cases, 80.0% presented as hyperdense lesions (12 cases), 13.3% appeared as isodense masses (2 cases), and only 6.7% showed hypodense masses (1 cases). Contrast-enhanced CT examination were performed in 6 cases, and all appeared significant enhancement. On T1-weighted image (T1WI), 66.7% appeared isointense and 33.3% presented hypointense. While on T2-weighted image (T2WI), 42.1% appeared isointense, 31.6% presented hyperintense and 26.3% showed hypointense. After contrast administration, 86.2% presented an intense homogeneous enhancement, 10.3% showed a heterogeneous enhancement, and only

3.4% appeared no enhancement. Craniotomy for tumor resection was performed in 80.6% of the patients (25 cases), and stereotactic biopsy was achieved in 12.9% of the patients (4 cases, all in the present study). Cerebrospinal fluid (CSF) cytology was used to diagnose pathologically in one patient. One patient was diagnosed at autopsy. Most of the preoperative diagnoses were schwannoma and meningioma, other preoperative diagnoses included metastatic tumor, hemangioma and tuberculosis meningitis. Only three cases were accurately diagnosed preoperatively. Diffuse large B-cell lymphoma was the most common type of PCNSLs of the CPA.

Table 1

Reported cases of primary central nervous system lymphoma of the cerebellopontine angle

Case	Authors/ Reference	Age/sex	Side	Clinical presentation	CT/MRI Characteristics	Preoperative diagnosis	Surgery
1	Yan et al. [38]	43 y/F	L	Hearing loss, headache, vomiting	MRI: isointense on T2 and T1, homogeneous enhancement	Schwannoma	Craniotomy
2	Danchaivijitr et al. [11]	21 y/F	R	Hearing loss, facial palsy, facial dysesthesias, headache, nausea, vomiting	CT: hyperdense. MRI: hypointense on T1 and T2, heterogeneous enhancement	Meningioma, schwannoma	Craniotomy
3	Nishimura et al. [28]	63 y/F	L	Dizziness, nausea, ataxia, nystagmus	CT: hyperdense, homogeneous enhancement. MRI: hypointense on T1, isointense on T2, no enhancement	Meningioma, schwannoma	Craniotomy
4	Hill et al. [14]	76 y/F	R	Hearing loss, facial palsy, ataxia	MRI: homogenous enhancement	N.A.	Craniotomy
5	Yang et al. [39]	50 y/M	L	Hearing loss, ataxia, dizziness	CT: isodense, homogeneous enhancement	N.A.	Craniotomy
6	Wang et al. [36]	57 y/M	L	Hearing loss, facial numbness, facial palsy, ataxia	MRI: isointense on T1 and T2, homogeneous enhancement	Schwannoma	Craniotomy
7	Bonneville et al. [3]	49 y/M	R	N.A.	MRI: homogeneous enhancement	N.A.	N.A.
8	Jaiswal et al [19]	36 y/M	B	Hearing loss, facial palsy, tinnitus, headache, ataxia, nystagmus	MRI: isointense on T1, hyperintense on T2, homogenous enhancement	N.A.	Craniotomy
9	Wen et al. [37]	75 y/F	L	Facial palsy, facial hypoesthesia, dizziness	MRI: homogenous enhancement	N.A.	Craniotomy
10	Garth et al. [13]	68 y/F	B	Hearing loss, ataxia, diplopia, dysarthria,	CT: hyperdense. MRI:	N.A.	Craniotomy

				nystagmus, reduced corneal reflexe	homogenous enhancement		
11	Ierokomos et al. [17]	82 y/F	L	Hearing loss	CT: hypodense, enhancement	N.A.	Craniotomy
12	Itoh et al. [18]	28 y/F	L	Hearing loss, tinnitus, nausea, vomiting, nystagmus	MRI: isointense on T1, slight hyperintense on T2, homogenous enhancement	Schwannoma	Craniotomy
13	Berciano et al [4]	39 y/F	L	Hearing loss, facial paraesthesiae, headache, vomiting, otalgia	CT: hyperdense, enhancement	N.A.	Autopsy
14	Angeli et al. [1]	56 y/F	L	Hearing loss, facial palsy, ataxia	MRI: isodense on T1, homogenous enhancement	Schwannoma, hemangioma	Craniotomy
15	Kariya et al. [22]	63 y/M	R	Hearing loss, tinnitus, ataxia, dizziness, nausea	CT: enhancement. MRI: heterogeneous enhancement	N.A.	Craniotomy
16	Taskin et al. [34]	43 y/F	R	Hearing loss, facial palsy, dizziness, ataxia	CT: enhancement. MRI: homogenous enhancement	N.A.	Craniotomy
17	Shuangshoti [31]	27 y/F	R	Hearing loss, facial palsy, ataxia; corneal reflex decreased; hoarseness	CT: enhancement. MRI: isodense on T1, homogenous enhancement	N.A.	Craniotomy
18	Seevaratnam et al. [32]	60y/F	L	Hearing loss, headache, vomiting, ataxia	CT: hyperdense. MRI: isointense on T1, hypointense on T2, homogeneous enhancement	Schwannoma	Craniotomy
19	Phang et al. [29]	48y/F	R	Facial pain, unsteadiness, facial hypoesthesia, corneal reflex decreased	MRI: hypointense on T2, homogeneous enhancement	Schwannoma, meningioma	Craniotomy
20	Cederberg et	48y/F	L	Headache,	MRI:	Tuberculosis	CSF

	al. [9]			facial palsy, facial hypoaesthesia	homogeneous enhancement	meningitis	cytology
21	Berrocal et al. [2]	60y/M	L	Hearing loss, facial hypoaesthesia, ataxia	MRI: hypointense on T1, moderately hyperintense on T2, homogeneous enhancement	Schwannoma, meningioma,	Craniotomy
22	Shin et al. [30]	47y/F	R	Facial palsy	CT: slightly hyperdense, homogeneous enhancement MRI: hypointense on T1, isointense on T2, homogeneous enhancement	Meningioma, Schwannoma	Craniotomy
23	Mori et al. [26]	50y/M	R	Dizziness, hearing loss	MRI: homogeneous enhancement	N.A.	Craniotomy
24	Lin et al. [25]	67y/F	R	Dysphagia, dizziness	MRI: slightly hypointense on T1, slightly hypointense on T2, homogeneous enhancement	Metastasis	Craniotomy
25	Mielczarek et al. [27]	65y/F	L	Hearing loss, tinnitus, vertigo, facial palsy	MRI: isointense on T1, hypointense on T2, homogeneous enhancement	Schwannoma	Craniotomy
26	Kommu et al. [21]	24y/M	L	Headache, vomiting	CT: slightly hyperdense. MRI: isointense on T1 and T2, heterogeneous enhancement	Schwannoma	Craniotomy
M, male; F, female; y, year; N.A., not available; L, left; R, right; B, bilateral; MRI, Magnetic Resonance Imaging; T1, T1-weighted imaging; T2, T2-weighted imaging.							

Illustrate case presentation

A 63-year-old man presented with left sided hearing loss for 3 months, and continuous headache accompanied by nausea and vomiting for 1 month. His past medical history was unremarkable. On admission, he was alert and cooperative. Visual fields and acuity were normal. Complete physical examination revealed no lymphadenopathy

or organomegaly. Neurological examinations revealed left facial palsy (House-Brackmann Grade 4) and moderate degree of hearing loss on the left side. The rest of the neurological and general examinations was unremarkable. Routine hematological parameters including tumour markers were within normal limits, and the serology tests for HBV, HCV, and HIV were negative.

CT scan revealed a slight hyperdense mass in the left CPA (Fig. 1). No adjacent bony abnormality was present. The lesion was isointense on T1WI and T2WI (Fig. 2a, b). The fourth ventricle and adjacent brainstem were mildly compressed by the lesion. Perifocal edema was detected on the T2WI (Fig. 2b). Contrast-enhanced MRI demonstrated a well-enhanced mass lesion in the left CPA, extending into the left internal auditory canal (Fig. 2c-e). FDG-PET/CT examination showed an intensely hypermetabolic lesion in the left CPA (Fig. 3), thus excluding the possibility with systemic tumor. Lumbar puncture was performed, CSF analysis revealed an increased protein level (protein 321.8 mg/dL), with a normal glucose level and a pleocytosis with a preponderance of lymphocytes (98% lymphocytes). Additionally, CSF IgG was significantly elevated (377.0 mg/L). No bacteria, acid-fast bacilli, or fungal organisms were seen on smears or cultures. CSF cytology revealed no malignant cells. On the basis of these imaging findings, a malignant pathology such as high-grade glioma, metastatic tumor or lymphoma were considered preoperatively. We recommended craniotomy or stereotactic biopsy with pathological examination. Finally, the patient and his family chose stereotactic biopsy. No steroid treatment before the surgery was provided.

Stereotactic biopsy was carried out to obtain tumor tissue. The frozen section was suggestive of a lymphoma. Haematoxylin and eosin (H&E) staining showed diffuse infiltration of large anaplastic cells with irregular nuclei and scanty cytoplasm, with frequent mitoses and apoptosis (Fig. 4a). The immunohistochemical study yielded positive results for CD20 and CD79a, and negative results for CD3, CD43 and glial fibrillary acidic protein (GFAP) (Fig. 4b). These findings were compatible with primary diffuse large B-cell lymphoma (DLBCL) of the central nervous system.

Postoperatively, the patient's hearing loss and facial palsy were unchanged and no additional neurological deficits were developed. He was then transferred to the hematology department for chemotherapy.

Discussion

Primary central nervous system lymphoma is a rare and aggressive extranodal non-Hodgkin lymphoma that restricted entirely to the brain, leptomeninges, spinal cord or eyes, without systemic involvement [28, 33]. PCNSL accounts for approximately 1-4% of all primary brain tumors [32, 40], but its incidence has been increasing over the past 20 years in both immunocompetent and immunocompromised individuals [14, 36]. Diffuse large B-Cell Lymphomas are the most common type (~90%), while the remaining 10% are poorly characterized by Burkitt's lymphomas, T-Cell lymphomas and low-grade lymphomas [5, 12, 15, 33]. PCNSL most commonly locates in the cerebral hemispheres (38%), thalami/basal ganglia (16%), corpus callosum (14%), periventricular region (12%), or cerebellum (9%) [21, 25], while it rarely occurs in the CPA. We reviewed 32 cases of PCNSLs of the CPA, including the reported cases in the English literature and six patients in our hospital, trying to figure out the accurate preoperative diagnosis of this rare disease.

The incidence of PCNSL of the CPA has never been previously reported in the literature. In the present study, we found that PCNSL of the CPA accounts for 2% of all PCNSLs. In contrast to those with PCNSLs (M/F ratio, 2:1) [11, 23], there was a preponderance of females in those with PCNSLs of the CPA (M/F ratio, 1:2.2) according to our study. With regard to the laterality of the lesions, there was a preponderance of left-sided lesions in the PCNSLs of

the CPA (L/R ratio, 1.5:1). Clinical manifestations of the PCNSLs of the CPA are related to the anatomical structures surrounding the lesion and volumes of the tumors. PCNSLs of the CPA typically present with trigeminal, facial, audiovestibular and cerebellar dysfunction, including hearing loss, tinnitus, facial palsy, vertigo and ataxia [14, 19, 33, 38]. Other neurological symptoms include headache, nausea, vomiting, hoarseness and nystagmus [18, 31, 32].

Given that CPA PCNSLs can be very non-specific with the presenting symptoms, accurate preoperative diagnosis of the lesion is mainly based upon imaging. On computed tomography (CT) scan, PCNSLs of the CPA generally present as hyperdense lesions with homogenous enhancement after contrast administration [12, 22, 28, 32]. Bony erosion and expansion of the internal auditory canal is rare in the PCNSL of CPA, which can be differentiate from acoustic neurinoma [28]. In the present study, we found that 80.0% of all the CPA PCNSLs presented as hyperdense lesions on CT scans. On MRI, PCNSLs of the CPA are typically homogeneously isointense on T1WI [19, 36, 38], and homogeneously isointense/hypointense with mild perifocal edema on T2WI [11, 28, 32]. After contrast administration, they show an intense homogeneous enhancement [22, 35]. In a review of the MRI appearance of CPA PCNSLs in the present study, 66.7% appeared isointense on T1WI, and 68.4% appeared isointense/hypointense on T2WI. After contrast administration, 86.2% presented an intense homogeneous enhancement.

Accurate preoperative diagnosis of CPA PCNSLs can be very difficult due to the rarity and clinical similarities to other common CPA tumors. However, increasing experience with CT and MRI studies allow greater accuracy in the preoperative differential diagnosis [10]. The common differential diagnosis includes schwannoma (70%-90%), meningioma (5%-10%), epidermoid cyst (5%-7%), metastatic tumors and glomus jugulare tumors [2, 6, 10, 11, 19, 21]. Kendall et al analyzed 208 cases of CPA tumours and found bony erosion of the internal auditory canal in 98 cases on CT scan, with 93.9% of the of these being acoustic neuromas [24]. However, bony erosion and expansion of the internal auditory canal is rarely observed in CPA PCNSLs, which can be differentiate from acoustic neurinomas [28]. Characteristic T2-wighted MRI may play an important role in the preoperative differential diagnosis [11, 28, 38]. In the present study, we found that 68.4% of all the CPA PCNSLs appeared as isointense to hypointense on T2WI, which is in contrast to most other CPA lesions [16, 32]. This signal intensity is attributable to the high cellularity and high nuclear to cytoplasmic ratio of the PCNSL, which also correspondingly explains the high signal intensity of PCNSLs usually observed on diffusion-weighted imaging (DWI) [3, 32]. In our study, we found that only three case were accurately diagnosed preoperatively, and all the three cases were identified at our institution. The accurate preoperative diagnosis is mainly because of the accumulated experience from the previous surgery cases of CPA PCNSLs.

When PCNSL of CPA is suspected by CT/MRI, conclusive diagnosis should be made by histological or cytological examination of tumour [12, 22, 27]. CSF sampling by lumbar puncture can be performed at the time of initial assessment [12, 22]. If CSF cytology is successful to establish a definitive diagnosis of malignant lymphoma, surgery can be avoided [7, 22]. However, the positive rate of CSF cytology is low. Balmaceda et al. reported that the initial CSF cytological studies were positive in only 15% [8]. Serial CSF samples may result in increased diagnostic sensitivity [12]. If CSF cytology failed to confirm the diagnosis, tumor excision or biopsy should be performed. The aim of the surgery is to obtain a frozen section of CPA PCNSL during surgery [19, 37]. Since radical tumor resection has no advantage on survival and may cause more neurologic deficits, radical decompression should be discouraged [19, 36, 37]. It is advisable to proceed with stereotactic biopsy for brain lesions with a radio-graphic appearance consistent with PCNSL [15]. Once the diagnosis of CPA PCNSL is established, more effective

treatment should be delivered. However, the optimal treatment of PCNSL has yet to be defined [15]. High-dose methotrexate (HD-MTX)–based induction chemotherapy is considered standard for newly diagnosed PCNSL [15]. Through review of the literature and our cases (Tables 1, 2), CSF cytology was used to diagnose pathologically in only one patient, tumor resection was performed in 80.6% of the patients, and stereotactic biopsy was achieved in only 13.8% of the patients.

Table 2

Primary central nervous system lymphoma of the cerebellopontine angle in the present study

Case	Age/sex	Side	Clinical presentation	CT/MRI Characteristics	Preoperative diagnosis	Surgery
1	61 y/M	L	Hearing loss, headache, vomiting	MRI: isointense on T1 and T2, homogeneous enhancement	Meningioma	Craniotomy
2	69 y/F	R	Dysphagia, hoarseness	CT: hyperdense. MRI: hypointense on T1, isointense on T2, homogeneous enhancement	Meningioma	Craniotomy
3	51 y/F	L	Facial hypoesthesia, slurred speech	CT: hyperdense. MRI: isointense on T1, slightly hyperintense on T2, homogeneous enhancement	Lymphoma	Stereotactic biopsy
4	63 y/M	L	Hearing loss, headache, nausea, vomiting, facial palsy	CT: hyperdense. MRI: isointense on T1 and T2, homogeneous enhancement	Malignant tumor	Stereotactic biopsy
5	59 y/F	R	Headache, dizziness, vomiting	CT: isodense. MRI: isointense on T1, slightly hyperintense on T2, homogeneous enhancement	Lymphoma	Stereotactic biopsy
6	62 y/F	L	Headache, vomiting, facial hypoesthesia	CT: hyperdense. MRI: hypointense on T1, slightly hyperintense on T2, homogeneous enhancement	Lymphoma	Stereotactic biopsy
M, male; F, female; y, year; L, left; R, right; MRI, Magnetic Resonance Imaging; T1, T1-weighted imaging; T2, T2-weighted imaging						

Conclusion

In conclusion, PCNSL of CPA is extremely rare lesion, accounting for 2.0% of all PCNSLs in the present study. There is a preponderance of females and left-sided lesions in the PCNSLs of CPA. Contrast-enhanced MRI with T2WI is very helpful in the preoperative diagnosis of the CPA PCNSL. Although rare, lymphoma should be included in the differential diagnosis of CPA lesions.

Declarations

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Conflict of interest

The authors declare no competing interests.

Availability of data and material

The datasets generated during and/or analyzed during the current study are available from the corresponding author on reasonable request.

Code availability (software application or custom code)

Not applicable.

Author contribution

All authors contributed to the study conception and design. Also, all authors were involved in material preparation and data collection. Data analysis was performed by Jian Zheng, Buyi Zhang, Danchan Lu and Chongran Sun. The first draft of the manuscript was written by Jian Zheng and Buyi Zhang, and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Ethics approval

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the Ethics Committee of the Second Affiliated Hospital, Zhejiang University School of Medicine (ID: 0182/2021).

Consent to participate

Informed consent was obtained from all individual participants included in the study.

Consent for publication

Patients signed informed consent regarding publishing their data.

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Figures



Figure 1

CT showed a slight hyperdense mass in the left CPA.

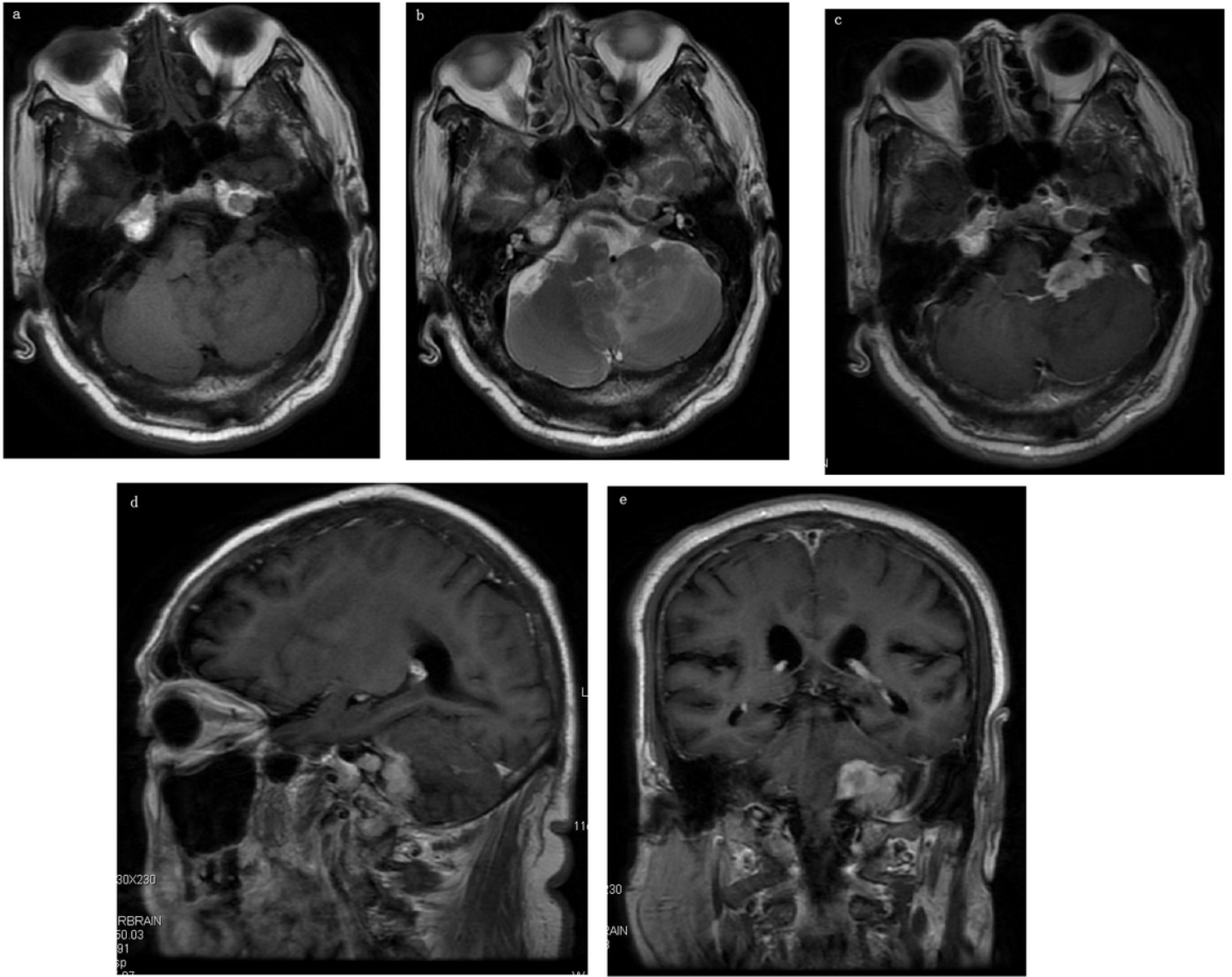


Figure 2

a An axial T1-weighted image showed an isointense mass. b An axial T2-weighted image showed an isointense mass, with perifocal edema. c A contrast-enhanced axial image revealed homogeneous enhancement of the lesion, extending into the left internal auditory canal. d A contrast-enhanced sagittal image revealed a well-enhanced mass lesion. e A contrast-enhanced coronal image revealed a well-enhanced mass lesion.

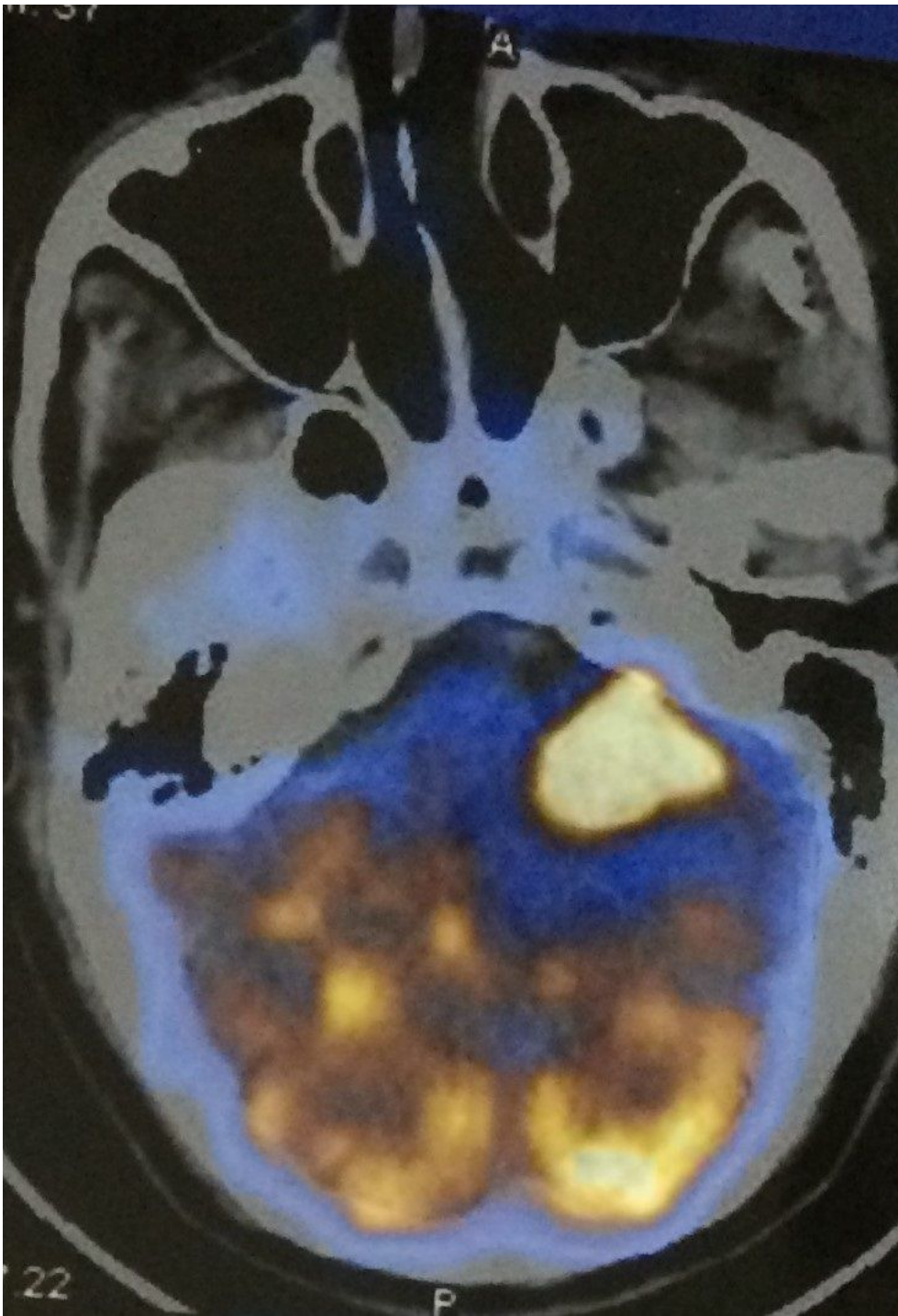


Figure 3

FDG-PET/CT examination showed an intensely hypermetabolic lesion in the left CPA.

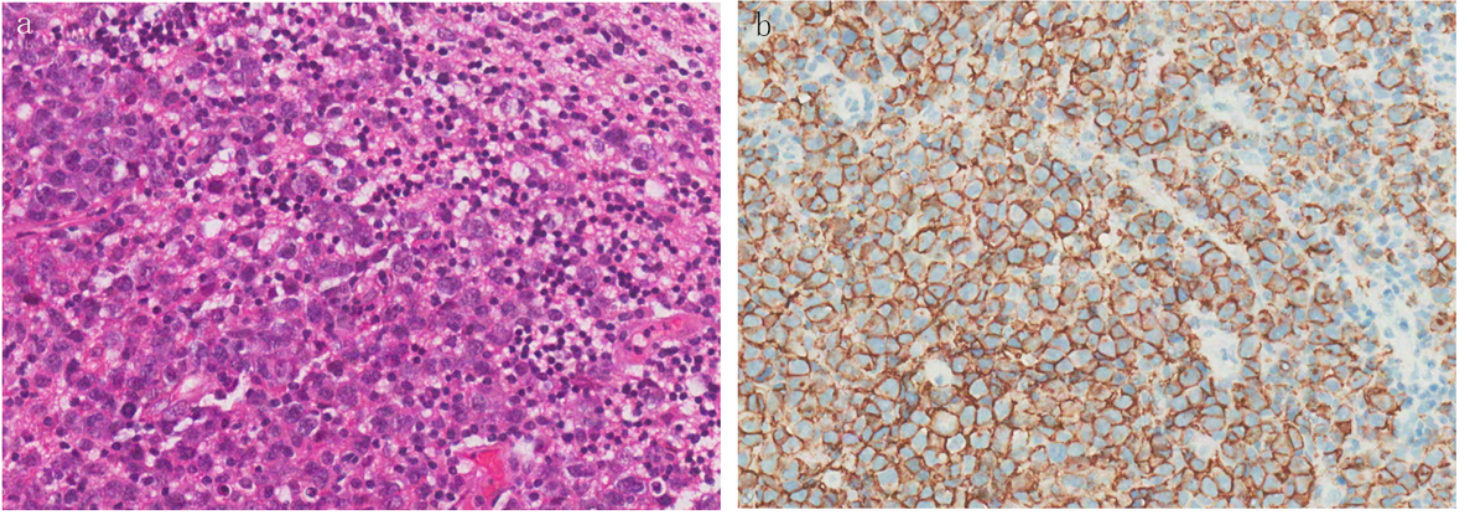


Figure 4

a Hematoxylin and eosin staining (magnification $\times 200$) showed diffuse infiltration of large anaplastic cells with irregular nuclei and scanty cytoplasm, with frequent mitoses and apoptosis. b CD-20 immunohistochemistry staining (magnification $\times 200$) demonstrating positivity.