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Case Report

Diffuse large B-cell lymphoma in the sphenoid sinus: A case report and review of literature

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ABSTRACT

Background: Non-Hodgkin lymphomas (NHLs) in paranasal sinus are uncommon, accounting for 0.17–2% of all NHL cases; it is especially rare in the sphenoid sinus. In this report, we describe a case of NHL in the sphenoid sinus.

Case Description: A 66-year-old man presented with a sudden left eye movement disorder. His head computed tomography and gadolinium-enhanced magnetic resonance imaging (Gd-MRI) showed a mass lesion extending around the left sphenoid sinus. However, the tumor regrowth about twice was observed during 2 weeks, partial removal of tumor was performed by the endoscopic trans-nasal transsphenoidal surgery, then histologically proved it to be diffuse large B-cell lymphoma (DLBCL). After R-THP-COP regimen (rituximab 375 mg/m^2, cyclophosphamide 750 mg/m^2, epirubicin 50 mg/m^2, vincristine 2 mg/day, and prednisolone 100 mg/day) and two courses of intrathecal methotrexate therapy for DLBCL, the symptoms and the lesion of enhanced Gd-MRI and fluorodeoxyglucose-positron emission tomography were completely disappeared.

Conclusion: NHLs in the sphenoid sinus is very rare disease, however, it is important to be diagnosed pathologically as soon as possible for being in remission state by the chemotherapy.

Keywords: Chemotherapy, Endoscopic trans-nasal transsphenoidal surgery, Non-Hodgkin lymphoma, Sphenoid sinus

INTRODUCTION

Non-Hodgkin lymphomas (NHLs) in paranasal sinus are uncommon, accounting for 0.17–2% of all NHL cases; it is especially rare in the sphenoid sinus.[3] It is difficult to diagnose them correctly by imaging modalities. In this report, we describe a case of NHL in the sphenoid sinus.

CASE PRESENTATION

A 66-year-old man was referred to our hospital with a sudden onset of headache, diplopia, and left ptosis. He was suffered from rheumatoid arthritis for several years and had taken methotrexate (MTX) tablets regularly. Neurological examinations revealed left oculomotor and trochlear nerve palsy without other cranial nerve disorders. Head computed tomography (CT) and gadolinium-enhanced brain magnetic resonance image (Gd-MRI) showed the mass lesion around the left sphenoid bone extending both to the left temporal middle fossa and to sphenoid sinus [Figure 1a and b]. Laboratory findings were white blood cell counts 7700/μl, red blood cell counts 516 × 10^4/μl, platelet counts 52.8 × 10^4/μl, lactate dehydrogenase 391U/l, soluble

interleukin-2 receptor (sIL-2R) 808 U/ml, Epstein–Barr virus-viral capsid antigen antibody, immunoglobulin G (EB-VCA-IgG) over 20 times, EB-VCA-IgM under 10 times, and EB EBV nuclear antigen (EBNA) over 20 times.

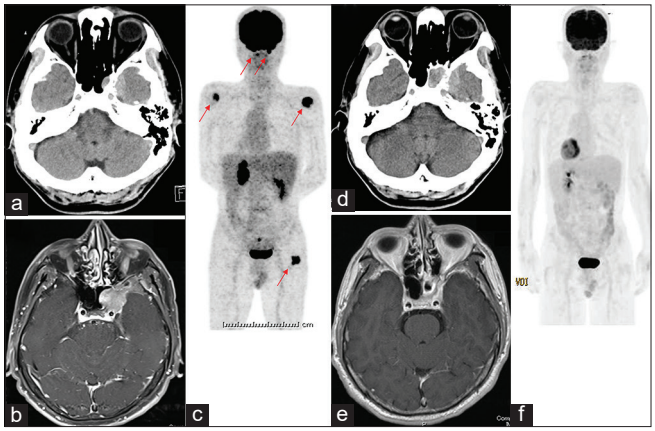

Figure 1: Image examinations of the case. Head computed tomography examinations before operation showed the mass lesion around the left sphenoid bone extending to lateral to the left temporal middle fossa and medial to sphenoid sinus (a). The mass lesion was heterogeneously enhanced with the gadolinium-enhanced magnetic resonance image (Gd-MRI) (b). Fluorodeoxyglucose-positron emission tomography (FDG-PET) showed multiple lesions in the left sphenoid bone, nasal cavity, bilateral humeri, and left femur (c). Two weeks after the first presentation to our hospital, the tumor size was increased about the twice size compared as the previous study (d). After the removal of tumor and chemotherapy, his Gd-MRI and FDG-PET showed the disappearance of the lesion (e and f).

We thought that pathological examination was necessary to rule out malignant tumors. Fluorodeoxyglucose-positron emission tomography (FDG-PET) showed multiple lesions in the left sphenoid bone, nasal cavity, bilateral humeri, and left femur [Figure 1c]. He was suspected as MTX-induced nonspecific lymphoproliferative disorder with rheumatoid arthritis or idiopathic lymphoma. Two weeks later, the tumor volume was increased compared as those of the previous study [Figure 1d], so we performed partial removal of tumor by endoscopic trans-nasal transsphenoidal surgery under general anesthesia.

In spite of the cessation of oral MTX for rheumatoid arthritis for 2 weeks after the removal of tumor, the tumor regrowth was observed on the head CT. The patient was treated with R-THP-COP regimen (rituximab 375 mg/m^2, cyclophosphamide 750 mg/m^2, epirubicin 50 mg/m^2, vincristine 2 mg/day, and prednisolone 100 mg/day) and two courses of intrathecal MTX therapy for central nervous system prophylaxis. After the eighth course of R-THP-COP, his Gd-MRI and FDG-PET showed the disappearance of the lesion [Figure 1e and f], and his symptoms completely disappeared. The patient has stayed recurrence free after the start of the treatment at 3-year follow-up.

Histopathological examinations showed that large B cells with nuclei display prominent nucleoli that diffusely

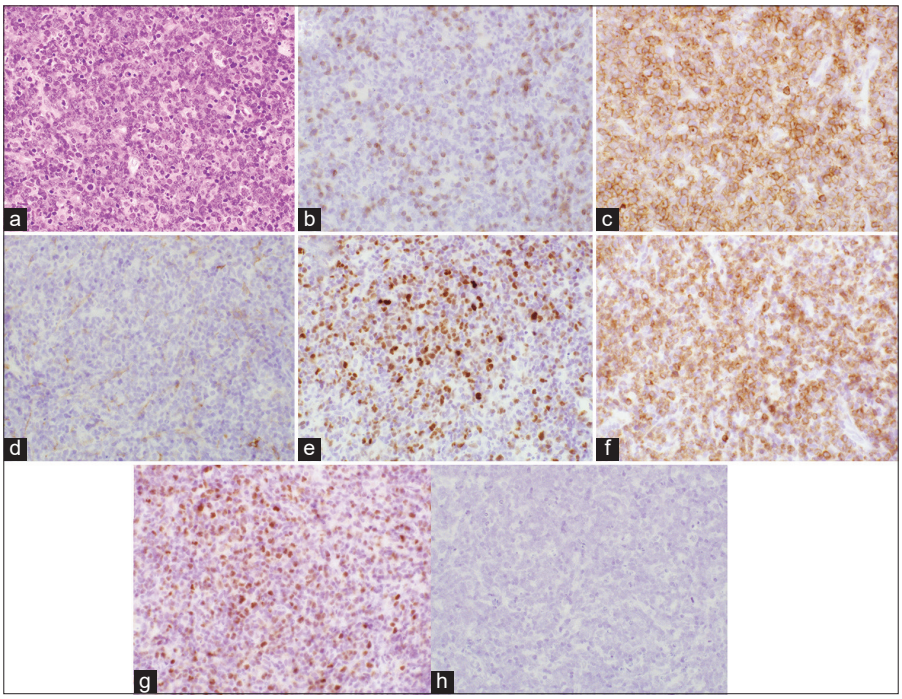

Figure 2: Histopathological examinations including immunohistochemistry and flow cytometry showed that large cells with nuclei display prominent nucleoli that diffusely infiltrate the brain tissue in hematoxylin-eosin staining (a), cluster of differentiation (CD) 3(−) (b), CD5(+) (c), CD10(−) (d), multiple myeloma oncogene (MUM)-1(+) (e), Bcl-2(+) (f), Bcl-6(+) (g), and EBER (EBV-encoded small RNA) *in situ* hybridization (EBER-ISH) (−) (h), thus the diagnosis was confirmed as diffuse large B-cell lymphoma.

infiltrate the brain tissue in hematoxylin-eosin staining [Figure 2a], cluster of differentiation (CD) 3(−) [Figure 2b], CD5(+) [Figure 2c], CD10(−) [Figure 2d], multiple myeloma oncogene (MUM)-1(+) [Figure 2e], Bcl-2(+) [Figure 2f], Bcl-6(+) [Figure 2g], and EBER (EBV-encoded small RNA) *in situ* hybridization (EBER-ISH) (−) [Figure 2h], thus the diagnosis was confirmed as diffuse large B-cell lymphoma (DLBCL). The tumor is classified as ABC type using the Hans classification based on MUM1, CD10, and Bcl6.

DISCUSSION

Primary malignancies of the sphenoid sinus, especially the lymphoma, are rare. As far as we have reviewed, 15 cases of sphenoid lymphoma including this case have been reported with detailed clinical history [Table 1].[1,2,4-8,10-14] Nasal obstruction or discharge appeared only in 5 patients (33.3%), unlike its high occurrence in other paranasal sinus tumors. Eleven patients (73.3%) initially presented with ptosis or diplopia caused by cranial nerve II, III, IV, and VI palsy. Nine patients experienced headache or facial pain (60%). Tumor extension was most common in the cavernous sinus (33.3%).

In our case, the patient had oral MTX tablets regularly for rheumatoid arthritis. We suspected the MTX-associated lymphoproliferative disorders; however, the regrowth after the cessation of oral MTX was observed. In addition, we diagnosed clinically this case as the DLBCL in the sphenoid

Table 1: Reported cases of sphenoidal lymphoma.

Author	Age	Symptoms	Sex	Histology	Cranial nerve disturbance	Local Invasion	Hans classification
Weber and Loewenheim[13]	39	Nasal congestion, facial pain, epiphora	Male	DLBCL	V	Sellar turcica	N/A
Deleu *et al*.[4]	44	Diplopia, esotropia ocular movement disorder	Male	DLBCL	V, VI	Clivus, cavernous sinus, ethmoidal sinus	N/A
Roth and Siatkowski[10]	5	Sudden visual loss, optic neuropathy	Male	DLBCL	II	Suprasellar, cavernous sinus	N/A
Van Prooyen Keyzer *et al*.[11]	65	Nasal obstruction	Unknown	NHL (B cell)		Erosion of the anterior wall of the sphenoid	N/A
Lewis *et al*.[5]	4	Ptosis, headache, lethargy, vomiting	Male	Burkitt	III	Cavernous sinus, infratemporal fossa	N/A
Mra *et al*.[6]	33	Ocular motility disorder, diplopia, ptosis	Female	NHL (B cell)	III	Cavernous sinus	N/A
Re *et al*.[8]	62	Diplopia, headache	Male	Burkitt-like	III	Cavernous sinus	N/A
Vedrine *et al*.[12]	68	Ocular motility disorder, diplopia	Female	DLBCL	II, III, V, VI	Cavernous sinus	N/A
Park *et al*.[7]	53	Ocular motility disorder, headache, diplopia, ptosis, mydriasis, exotropia	Female	DLBCL	III	Cavernous sinus	N/A
Bisdas *et al*.[1]	83	Ocular motility disorder, nasal obstruction, sinusitis, headache	Male	NHL (B cell)	II, III, IV, VI	Nasopharynx, skull base	N/A
	79	Ocular motility disorder, nasal obstruction, sinusitis, headache	Male	NHL (B cell)	II, III, IV, VI	Dura	N/A
	64	Ocular motility disorder, nasal obstruction, sinusitis, headache	Female	NHL (B cell)	II, III, IV, VI	Nasopharynx, cavernous sinus	N/A
Chennupati *et al*.[2]	66	Left-sided headache	Female	Burkitt	V	No invasion	N/A
Yoshihara *et al*.[14]	70	Ocular motility disorder, diplopia, ptosis, mydriasis	Male	DLBCL	III, IV	Cavernous sinus	ABC type
Our case	66	Ocular motility disorder, diplopia, ptosis, headache	Male	DLBCL	III, V, VI	Sphenoid sinus, cavernous sinus, temporal middle fossa	ABC type

NHL: Non-Hodgkin lymphoma, DLBCL: Diffuse large B-cell lymphoma

sinus. Pathologically, many cases of MTX -associated lymphoproliferative disorders have been positive in EBER immunostaining;[9] however, EBER-ISH was negative in our case. So finally, we diagnosed DLBCL. It was not definitive about the origin of DLBCL in our case, however, we consider the nasal cavity as the origin, because there were some reports about cases, whose origin was nasal cavity, and other parts (humerus and femur) were not. In addition, many cases have been checked no detailed immunostaining study, however, two cases including our case are ABC type in Hans classification.

In clinical practice, it is known that definite histological diagnosis is required before the initiation of any treatment.[3] It remains controversial whether or not patients with a very strong clinical suspicion of lymphoma should receive aggressive lymphoma treatment to prevent or recover certain complications. Therefore, the establishment of the optimal treatment option for such patients is required.

CONCLUSION

NHLs in paranasal sinus are very rare, however, this should be considered as a differential diagnosis of paranasal sinus tumors.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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