

Primary B-Cell Lymphoma of the Central Skull Base

Case Report

Sotirios Bisdas¹, Mehran Baghi², Ahmed Abdelkader¹, Wolfgang Gstoettner², Sebastian Fetscher³, Thomas J. Vogl¹

Introduction

Primary non-Hodgkin lymphomas (NHL) of the upper airway are extremely rare neoplasms and are encountered more frequently in Asian than in Western countries. In the Western world, 0.2–2.0% of all NHL appear in the sinonasal tract [1]. Recently, atypical lymphoma manifestations in the upper airway have been increasingly reported [2]. NHL in this site cannot be easily diagnosed and differentiated as its clinical features can be similar to those of other tumorous and inflammatory diseases in their early stages [3]. Furthermore, the imaging features of lymphoma can often be atypical and hamper the differential diagnosis [4].

Case Report

A West-European immunocompetent 83-year-old male patient presented with sinusitis symptoms, chronic nasal obstruction, and headache. No epistaxis was reported in the medical history. Physical examination showed an alert and oriented patient without fever or weight loss. Positive findings were limited to a mass observed in the nasopharynx, initially evaluated as lymphoid hyperplasia.

A contrast-enhanced CT scan was performed and demonstrated a large nasopharyngeal mass involving the central skull base (Figures 1a and 1b). The CT perfusion imaging revealed a well-vascularized tumor in

this site (Figure 2). Contrast-enhanced MR imaging (1.5 T MRI unit) corroborated the initial diagnosis, showing a slightly enhancing tumor in the sphenoid sinus with extended infiltration of the nasopharynx and central skull base structures (Figures 3a to 3c). The subsequently performed proton MR-spectroscopy of the tumor site demonstrated elevated choline implying a neoplastic process with increased cell density (Figure 4). MRI of the neck soft-tissue revealed only a 10 × 12 mm lymph node on the level II on the right side with gadolinium enhancement. The subsequently performed PET examination demonstrated a vivid fluoro-deoxy-glucose (FDG) uptake in the nasopharynx without any other foci.

The further clinical course after the imaging studies was eventful; the patient demonstrated diplopia and ptosis on the right side. The partial operative decompression of the sphenoid sinus with biopsy of the tumor site showed a primary highly malignant B-cell lymphoma of the plasmoblastic-plasmocystic type with L-chain restriction. The Epstein-Barr-virus analysis was negative. The patient received intrathecal therapy and immunotherapy (Rituximab) as well as four cycles of CHOP chemotherapy after which showed a complete remission. A radiation therapy as well as further chemo-/immunotherapy followed the admission imaging.

Clin Neuroradiol 2006;16:258–61
DOI 10.1007/s00062-006-6033-x

¹Department of Diagnostic and Interventional Radiology, Johann Wolfgang Goethe University Hospital Frankfurt, Frankfurt, Germany,

²Department of Otorhinolaryngology, Johann Wolfgang Goethe University Hospital Frankfurt, Frankfurt, Germany,

³Department of Hematology and Oncology, Krankenhaus Süd, Lübeck, Germany.

Submitted: August 28, 2006, Accepted: November 3, 2006

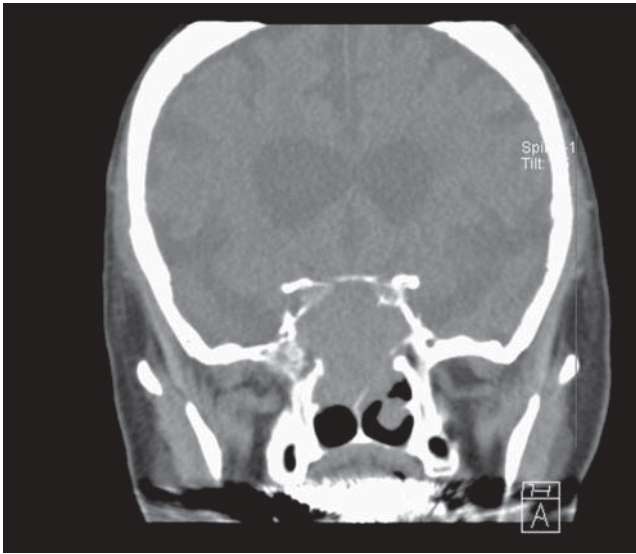


Figure 1a

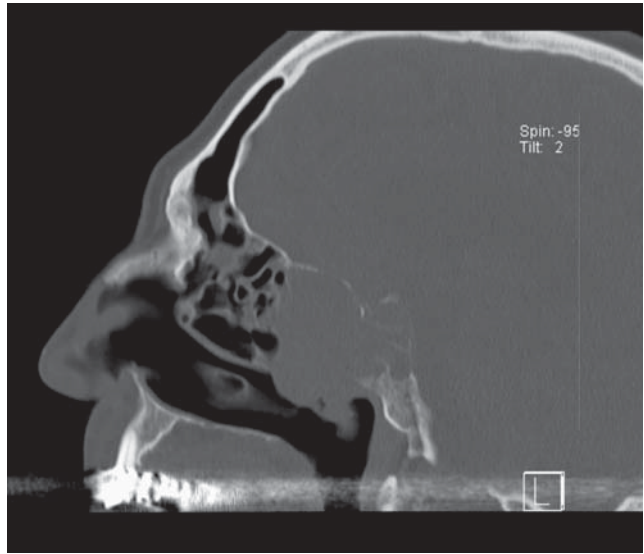


Figure 1b

Figures 1a and 1b. Non-enhanced CT imaging of a primary B-cell non-Hodgkin lymphoma of the sphenoid sinus. Coronal reconstruction of the multi-slice CT dataset shows a mass extending from the inferior nasal turbinate to the roof of the sphenoid sinus. Note the completely eroded floor of the central skull base. The roof and lateral walls of the sphenoid sinus show bone remodeling (a). Sagittal midline view of the mass in bone algorithm shows the pronounced infiltration and rarefaction of the adjacent skull base. The clivus is partially eroded. The mass causes a ballooning of the sphenoid sinus and extends in the nasopharynx (b).

Discussion

Lymphomas comprise 3–5% of all malignancies, with NHL accounting for 60% of cases [5]. Primary NHL of the sinonasal tract is a rare tumor and its B-cell subtype is more common in elderly men in the West, as distinct from the Far East and South America manifestation of the disease, which presents at younger age and is predominantly of the T-cell subtype [6, 7]. B-NHL from mucosa-associated lymphoid tissue often arises in gastric mucosa and is extremely rare in the upper aerodigestive tract. Few cases with marginal-zone lymphoma and (more frequently) diffuse large B-cell lymphoma in the upper aerodigestive tract have been described [2, 6, 8–10].

NHL in the upper airway manifests typically as a submucosal mass accompanied by polypoid, bulky masses with a smooth mucosal surface. On gross appearance lymphoma differs from squamous cell carcinoma, which is usually ulcerative. The presenting symptoms may masquerade as other nasal and head and neck findings such as nasal obstruction, hearing loss, acute sinusitis, rhinorrhea, epistaxis, visual disturbances, and pain. Nasopharyngeal lymphoma has an expansive growth with involvement of the parapharyngeal space and, normally, does not infiltrate the adjacent skull

base. However, clinically aggressive lymphomas such as Burkitt lymphoma and diffuse large B-cell lymphoma are characterized by bone destruction and infiltration

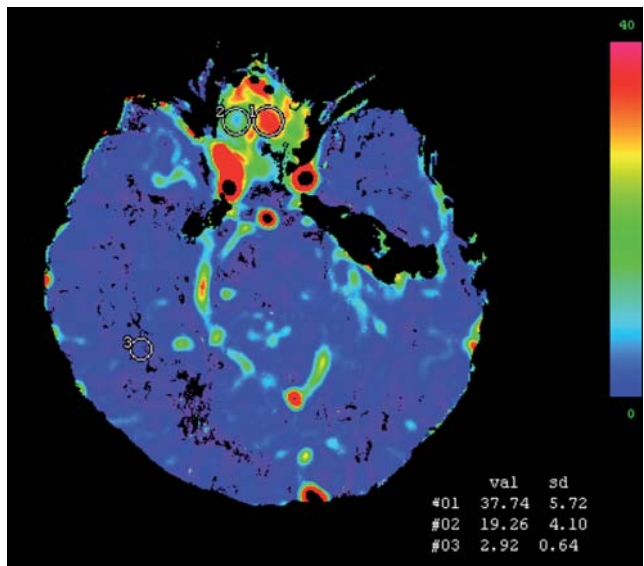


Figure 2. CT perfusion imaging of a primary B-cell lymphoma of the sphenoid sinus. Visual inspection of the blood volume parametric map demonstrates a marked hyperperfusion of the mass in the sphenoid sinus. ROI analysis shows elevated blood volume values of the tumor in comparison to the brain parenchyma.

of the dura with intracranial invasion. Thus, a classification of the lymphoma as extra- or atypical intra-axial in such complex situations may encounter difficulties [4, 11, 12]. The imaging features of primary lymphoma

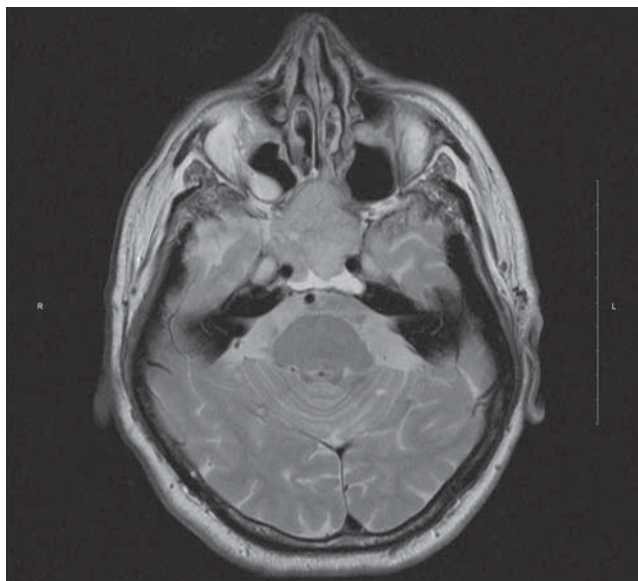


Figure 3a

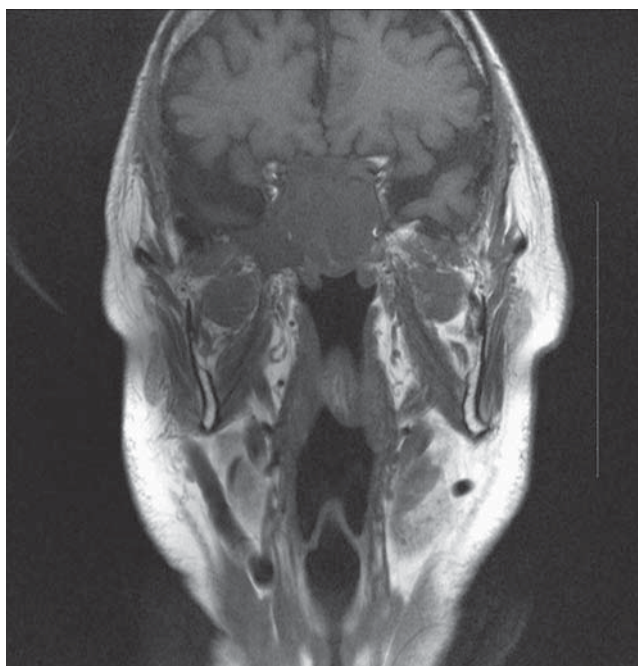


Figure 3b

may be indistinguishable from imaging characteristics of other neoplastic lesions in this region [6]. Differential diagnosis includes nasopharyngeal carcinoma, chordoma, intraosseous meningioma, esthesioneuroblastoma, pituitary adenocarcinoma, and metastasis. Multiplanar contrast-enhanced CT imaging characteristics are considered tumor non-specific. Similarly, perfusion CT imaging, which may reveal foci of hypervascularity and permeability disorders [13], thus supporting the tumor diagnosis, giving information about the tumor's extent, and therapy planning, is also tumor nonspecific. MRI with T2-weighted multiplanar imaging, which distinguishes tumor from thickened mucosa or fluid collections. Precontrast T1-weighted imaging can excellently depict bone involvement while the T1-weighted fat-saturated imaging helps defining the extent of bone and soft-tissue tumor infiltrations. However, fat-saturated postcontrast T1-weighted imaging may result in a slight loss of anatomical information and in susceptibility artifacts. Cranial nerves infiltrated by tumor are frequently seen, and infiltration of the pituitary gland may occur. Bone involvement without hyperostosis, calcifications, and cavernous sinus invasion without narrowing of the

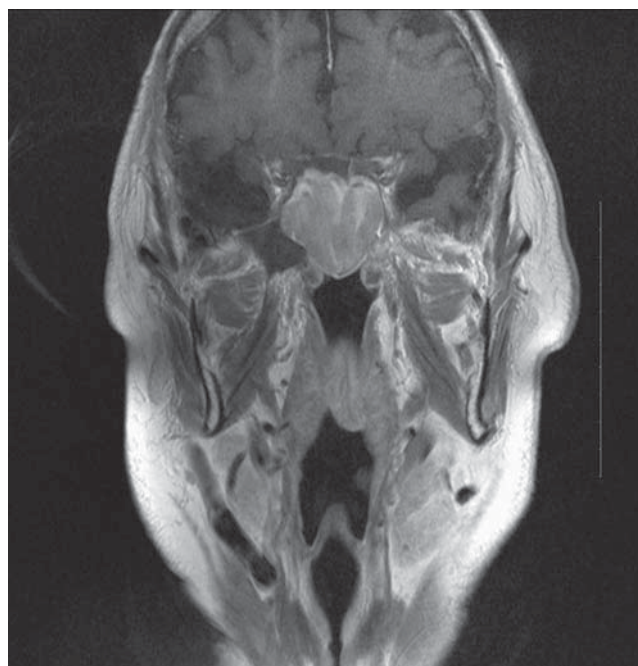


Figure 3c

Figures 3a to 3c. MR imaging of a B-cell lymphoma of the sphenoid sinus. T2-weighted imaging of a soft-tissue mass in the nasopharynx slightly hyperintense to brain parenchyma. Note the infiltration of the central skull base and the partial encasement of the internal carotid arteries without lumen narrowing (a). Non contrast coronal T1-weighted MR imaging shows a slightly lobulated mass isointense to the grey matter (b). The corresponding contrast-enhanced coronal scan demonstrates a heterogeneously enhancing mass with partial lobulation and septation within. Note the engulfment of the sella turcica and the extension into the parasellar regions without involvement of the cavernous sinus (c).

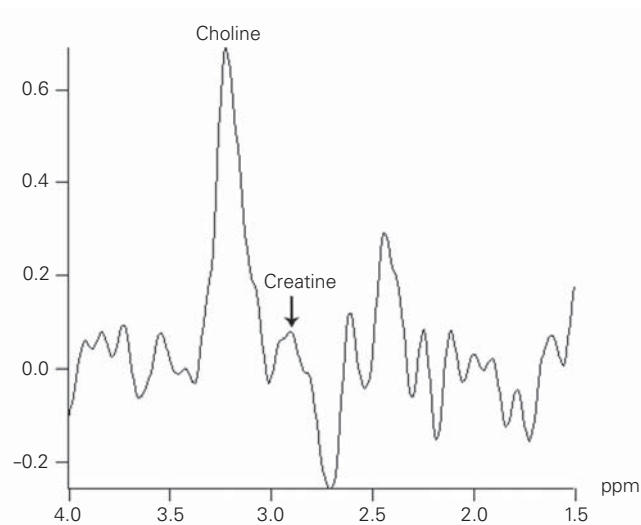


Figure 4. Single-voxel proton MR-spectroscopy (TE 135 ms, voxel size 20 mm³) of the B-cell lymphoma shows elevated choline. The choline/creatine ratio is 5.9.

carotid artery lumen may be helpful in the differentiation of lymphoma from meningioma. Dense cellular deposits of lymphoma do not show high signal intensity on proton density-weighted images; rather, the signal intensity of the tumor deposit typically remains hypo- to isointense in all pulse sequences. However, proton MR-spectroscopy can depict high cellularity of head and neck lesions by showing markedly elevated choline to creatine ratio [14]. There is initial evidence that proton MR-spectroscopy can differentiate between healthy soft tissue and tumor in the extracranial head and neck, while the benign tumors tend to have higher choline to creatine ratios than the malignant ones [15]. Our case report is the first patient in literature where a proton MR-spectroscopy is performed in a sinus sphenoidalis lymphoma.

Conclusion

Lymphomas have a high cure and survival rate and early diagnosis and staging with appropriate treatment is essential for the achievement of optimal treatment results. In patients with symptoms of sinusitis and nasal obstruction, it is imperative that the radiologist be familiar with findings of a nontypical mass in the sinuses and the nasopharynx. The diagnosis of NHL cannot be made from clinical findings solely and thus, imaging and biopsy of the lesion is mandatory prior to any treatment.

References

1. Cleary K, Batsakis J. Sinonasal lymphomas. *Ann Otol Rhinol Laryngol* 1994;103:911-4.
2. Hans FJ, Reinges MHT, Nolte K, Reipke P, Krings T. Primary lymphoma of the skull base. *Neuroradiology* 2005;47:539-42.
3. Liang R, Todd D, Chan TK, Chiu E, Choy D, Loke SL, Ho FC. Nasal lymphoma. A retrospective analysis of 60 cases. *Cancer* 1990;66:2205-9.
4. Roman-Goldstein SM, Jones A, Delashaw JB, McMenomey S, Neuwelt EA. Atypical central nervous system lymphoma at the cranial base: report of four cases. *Neurosurgery* 1998;43:613-5.
5. Boring CC, Squires TS, Tong T. Cancer statistics. *Cancer* 1993;43:7-26.
6. Weber AL, Rahemtullah A, Ferry JA. Hodgkin and non-Hodgkin lymphoma of the head and neck: clinical, pathologic, and imaging evaluation. *Neuroimaging Clin N Am* 2003;13:371-92.
7. Onakoya PA, Adeyi OA, Nwaorgu OG, Ojemakinde KO, Thomas JO. Primary extranodal non-Hodgkin's lymphoma of the upper aerodigestive tract - a descriptive analysis of the pattern seen in the University College Hospital, Ibadan. *Afr J Med Med Sci* 2003;32:59-63.
8. Prades E, Alobid I, Alos L, Guilemany JM, Bernal-Sprekelsen M, Mullol J. Extranodal lymphoma originating from mucosa-associated lymphoid tissue of the nasopharynx. *Acta Otolaryngol* 2003;123:1098-101.
9. Han MH, Chang KH, Kim IO, Kim DK, Han MC. Non-Hodgkin lymphoma of the central skull base: MR manifestations. *J Comput Assist Tomogr* 1993;17:567-71.
10. Singh S, Cherian RS, George B, Nair S, Srivastava A. Unusual extra-axial central nervous system involvement of non-Hodgkin's lymphoma: magnetic resonance imaging. *Australas Radiol* 2000;44:112-4.
11. Jung CS, Zimmermann M, Seifert V. Clivus lymphoma. *Acta Neurochir (Wien)* 2004;146:533-4.
12. Nakamura K, Uehara S, Omagari J, Kunitake N, Kimura M, Makino Y, Murakami J, Jingu K, Masuda K. Primary non-Hodgkin's lymphoma of the sinonasal cavities: correlation of CT evaluation with clinical outcome. *Radiology* 1997;204:431-5.
13. Glenn LW. Innovations in neuroimaging of skull base pathology. *Otolaryngol Clin North Am* 2005;38:613-29.
14. King AD, Yeung DK, Ahuja AT, Leung SF, Tse GM, van Hasselt AC. In vivo proton MR spectroscopy of primary and nodal nasopharyngeal carcinoma. *AJNR Am J Neuroradiol* 2004;25:484-90.
15. Bisdas S, Baghi M, Huebner F, Mueller C, Knecht R, Vorbuchner M, Ruff J, Gstoettner W, Vogl TJ. In vivo proton MR spectroscopy of primary tumours, nodal and recurrent disease of the extracranial head and neck. *Eur Radiol* 2006, May 16; [Epub ahead of print].

Address for Correspondence

Sotirios Bisdas, MD
 Department of Diagnostic and Interventional Radiology
 Johann Wolfgang von Goethe University Hospital
 Frankfurt
 Theodor-Stern-Kai 9
 60590 Frankfurt
 Germany
 Phone (+49/69) 6301-7292, Fax -83682
 e-mail: sbisdas@yahoo.com
 s.bisdas@med.uni-frankfurt.de