



Case Report

A case of intraorbital malignant lymphoma biopsied using an endoscopic transnasal approach

Masahiro Uchimura¹, Kentaro Hayashi², Tatsunori Sakamoto³, Hidemasa Nagai¹

¹Department of Neurosurgery, Shimane University Faculty of Medicine, ²Advanced Stroke Center, Shimane University Hospital, ³Department of Otorhinolaryngology-Head and Neck Surgery, Shimane University Faculty of Medicine, Izumo, Shimane, Japan.

E-mail: *Masahiro Uchimura - m_u.8953@med.shimane-u.ac.jp; Kentaro Hayashi - kentaro@med.shimane-u.ac.jp; Tatsunori Sakamoto - sakamoto_tatsunori@med.shimane-u.ac.jp; Hidemasa Nagai - h-nagai@med.shimane-u.ac.jp



***Corresponding author:**

Masahiro Uchimura,
Department of Neurosurgery,
Shimane University Faculty
of Medicine, Izumo, Shimane,
Japan.

m_u.8953@med.shimane-u.ac.jp

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ABSTRACT

Background: While most orbital tumors are primary, some are secondary, including extension or invasion from adjacent sites. The diagnosis varies widely, and the treatment strategy depends on the pathological diagnosis. Transcranial and transorbital surgical approaches are typically used. Recently, a transnasal endoscopic approach has emerged as a viable option. We report a case of an intraorbital tumor treated with endoscopic transnasal biopsy and compare the results with those of other surgical approaches.

Case Description: A 74-year-old woman visited a nearby hospital due to a right eye protrusion and decreased visual acuity. An intraorbital tumor was detected and the patient was referred to our hospital. Head computed tomography revealed a mass along the posterior wall of the right orbital apex. Contrast-enhanced magnetic resonance imaging showed a 37-mm lesion with a uniform contrast effect and no intracranial extension. Intraorbital lymphoma was considered a differential diagnosis, and a biopsy was performed using an endoscopic transnasal approach. The pathological diagnosis was B-cell lymphoma, and chemotherapy was administered.

Conclusion: The endoscopic transnasal approach for intraorbital tumors is less invasive, highly cosmetic, and useful, especially for medial and inferior orbital lesions.

Keywords: Biopsy, Endoscopic transnasal approach, Malignant lymphoma, Orbital tumor

INTRODUCTION

Approximately 90% of orbital tumors are primary; 10% are secondary tumors, including extension or invasion from adjacent sites.^[1,8] Lymphoproliferative diseases account for more than half of these tumors.^[4] However, preoperative and pathological diagnoses are indispensable because the diagnosis varies widely, and the treatment plan depends on the pathological type. The conventional surgical approach for such tumors is either transcranial or transorbital. However, the endoscopic transnasal approach has recently been considered a practical option.^[2,3] We present a case of an intraorbital tumor in which an endoscopic transnasal approach was used for biopsy and discuss the advantages of the endoscopic transnasal approach over the conventional surgical approach.

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CASE REPORT

A 74-year-old woman visited a nearby hospital with a 3-month history of right ocular protrusion and decreased right-eye visual acuity. She had no relevant medical history. Based on computed tomography (CT), an intraorbital tumor was suspected, and the patient was referred to our hospital. The patient was conscious and explicit during the initial examination. Right exophthalmos, conjunctival edema, and papillary edema of the optic nerve were observed. No intraocular hypertension was observed. Her corrected visual acuity was 0.3 on the right and 0.6 on the left. Head CT revealed a mass superior to the medial side of the right orbit [Figures 1a and b]. CT angiography revealed an ophthalmic artery outside the mass [Figure 1c], and magnetic resonance imaging (MRI) showed T1 isointense and T2 isointense lesions. Diffusion-weighted MRI revealed a hyperintense lesion, and T1-weighted Gd-enhanced MRI showed a homogeneously enhanced lesion [Figures 2a and b]. Constructive interference in steady-state sequencing showed that the internal rectus and superior oblique muscles were externally compressed by the tumor [Figures 2c and d]. Blood test results were negative for Immunoglobulin G (IgG), IgG4, and beta 2-microglobulin. All tumor markers tested negative. Therefore, malignant lymphoma was suspected. Because the tumor was located on the medial side of the right orbit, an endoscopic transnasal approach was used for a biopsy 2 weeks after admission.

Endoscopic transnasal approach

Bosmin (0.1%) was injected into the incision site of the nasal septal mucosa in the right nasal cavity, and the right maxillary sinus was opened by resecting the uncinat process of the ethmoid [Figure 3a]. The superior wall of the maxillary sinus is the landmark of the inferior wall of the orbit. Ethmoidectomy was performed to expose the medial side of

the orbit [Figure 3b]. The ethmoidal mucosa of the medial orbit was then dissected, and the lamina papiracea was removed to expose the periorbita [Figure 3c]. Based on the anatomic position of the internal rectus and inferior rectus muscles, the orbital periosteum was incised from the anterior to posterior end in the lower part of the orbit. Due to the increased intraorbital pressure caused by the tumor, a thin incision through the orbital periosteum caused the fat in the orbit to protrude. Therefore, we believed that there was no internal rectus muscle directly below the orbital periosteum incision site and made a deep incision in the orbital periosteum, and subsequently, the tumor was identified. However, distinguishing between the fat, tumor, and medial rectus muscles was difficult. The medial rectus muscle and tumor area were bluntly dissected, and a portion of the tumor was removed [Figure 3d]. The intraoperative diagnosis was malignant lymphoma; an additional permanent specimen was removed from the same site. To the extent possible, the exposed orbital contents were partially covered with the remaining ethmoidal mucosa [Figure 3e], and a silicone sheet was placed in an inverted U-shape to complete the surgery [Figure 3f]. The postoperative course was uneventful, and eye movement was fully restored. No new visual loss or diplopia was observed postoperatively.

Pathological study

Histopathological examination revealed proliferating lymphocytes with moderate cellular atypia [Figures 4a and b]. In addition, immunostaining showed positivity for CD5 [Figure 4c], CD20 [Figure 4d], and CD79a [Figure 4e], with a Ki67 index of 70% [Figure 4f]. The permanent pathological diagnosis was high-grade mucosa-associated lymphoid tissue (MALT) lymphoma (Ki67 index: 70%).

First, radiotherapy (40 Gy) was administered to the right intraorbital tumor and neck. We then administered three

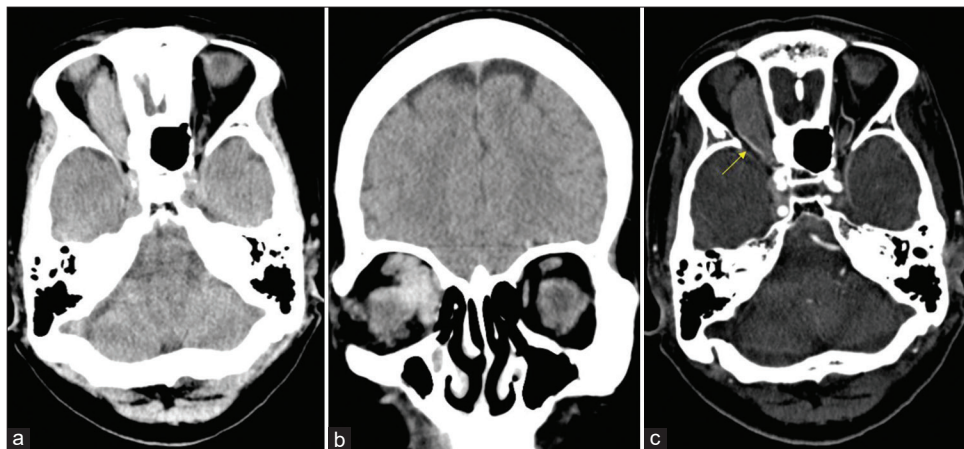


Figure 1: (a and b) Head computed tomography (CT) revealed a mass superior to the medial side of the right orbit. (c) CT angiography revealed an ophthalmic artery (arrow) outside the mass.

cycles of rituximab, cyclophosphamide, doxorubicin hydrochloride (hydroxydaunorubicin), vincristine sulfate (Oncovin), and prednisone. Six months after treatment, T1-weighted Gd-enhanced MRI showed that the tumor had shrunk, ocular protrusion had improved, and no evidence of visual acuity or visual field abnormalities was present.

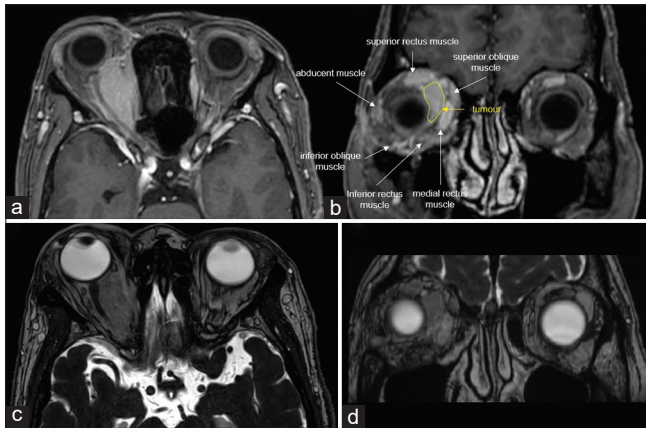


Figure 2: (a and b) T1-weighted Gd-enhanced magnetic resonance imaging showing a homogeneously enhanced lesion on the superior to medial side of the right orbit. (c and d) Constructive interference in steady-state sequence showing the internal rectus and superior oblique muscles compressed externally by the tumor.

DISCUSSION

Because the diagnosis of orbital tumors is diverse, and treatment strategies differ according to the type of disease, pathological diagnosis is essential. Approximately 90% of orbital tumors are primary; <10% are secondary, including extension or invasion from adjacent sites, and a few are reported to be metastatic.^[1,4,8] Lymphoproliferative diseases account for more than half of the cases; half of these are lymphomas. MALT lymphomas account for most lymphomas, followed by large diffuse B-cell, follicular, and mantle lymphomas. Other lymphoproliferative diseases include orbitocavernous hemangiomas, lacrimal gland tumors, meningiomas, schwannomas, and secondary tumors.

In addition to the transcranial and transorbital approaches for treatment, another alternative has been used recently: the endoscopic transnasal approach.^[2,3] The safety and efficacy of orbital tumor surgery may be improved by applying new imaging techniques and intraoperative devices, such as ultrasonography and neuronavigation.^[6,10] Considering the orbit as a clock face, tumors located between the 6 and 1 o'clock positions are suitable for the conventional transcranial approach. In contrast, tumors located medially (1–7 o'clock) are better suited for the endoscopic transnasal approach; tumors located from 1 to 6 o'clock are better suited for the transorbital approach [Figure 5].^[7] However, the transorbital

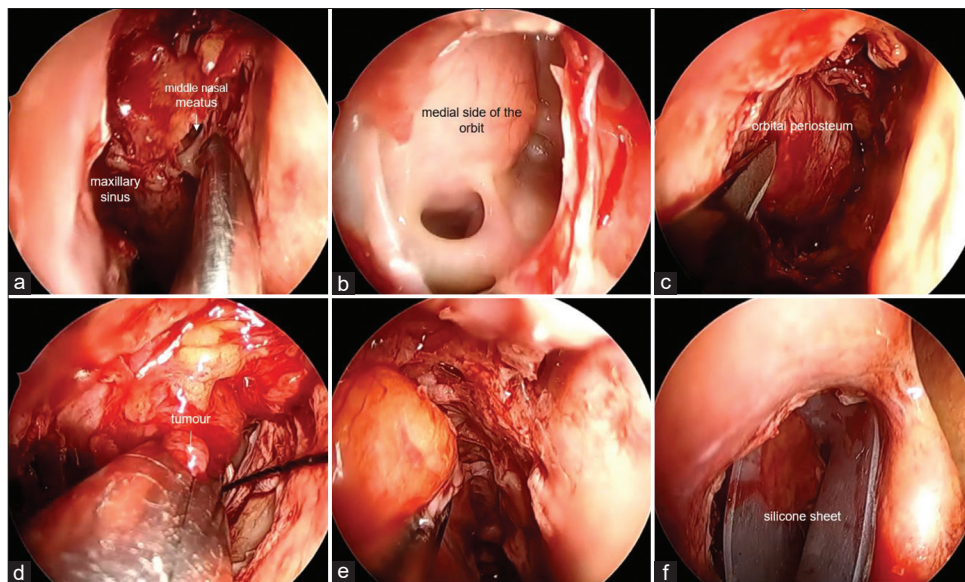


Figure 3: (a) The right maxillary sinus entered through the right nasal cavity and was freed by excision of the cribriform stalk. The superior wall of the maxillary sinus is an important landmark because it serves as the inferior wall of the orbit. (b) The middle nasal meatus is punctured to release the ethmoid sinus, exposing the medial side of the orbit. (c) Mucosa of the medial orbit was dissected, and the bone was removed to expose the orbital periosteum. (d) The orbital periosteum was incised, but distinguishing between the fat, tumor, and medial rectus muscle was difficult. The medial rectus muscle and tumor area were bluntly dissected, and the tissue was removed. (e) The exposed orbital contents were partially covered with mucosa. (f) A silicone sheet is placed in an inverted U-shape to complete the surgery.

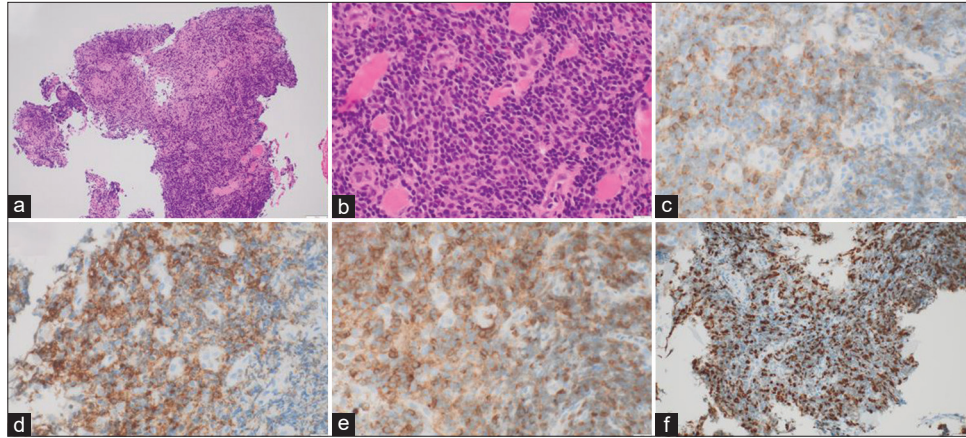


Figure 4: (a and b) Histopathology showing proliferating lymphocytes with moderate cellular atypia. (c-f) Immunostaining was positive for CD5, CD20, and CD79a, and the Ki67 index was 70%.

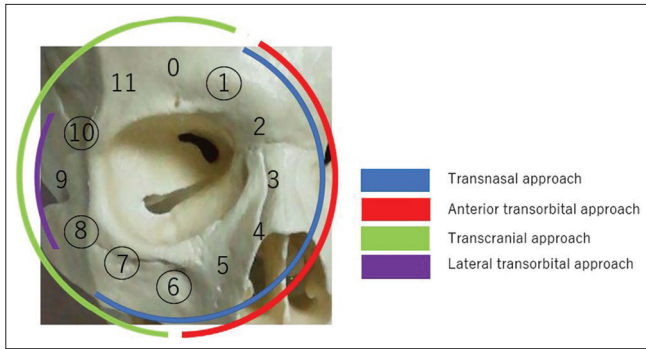


Figure 5: Schematic diagram created from the report by Paluzzi *et al.*^[7] If the orbit is a clock face, tumors located between 6 and 1 o'clock are good candidates for the conventional transcranial approach. In contrast, tumors located medially-from 1 to 7 o'clock-are better suited for the endoscopic transnasal approach, whereas those from 1 to 6 o'clock are better suited for the transorbital approach.

approach is limited to the tumors located anterior to the equatorial plane of the eye due to the obstructive effects of the eyeball. In our patient's case, the tumor was located at the 0–4 o'clock position, posterior to the equatorial plane of the eye; therefore, a nasal endoscopic approach was considered optimal.

The following is a comparison of each approach from a different perspective. The advantages of the endoscopic transnasal approach are that it requires no skin incision and is highly esthetic. Its disadvantages include less operability, limited field of view, difficulty performing the procedure if bleeding occurs, and risk of injury to the medial rectus and superior oblique muscles [Table 1].^[5,7] Zoli *et al.* reported that although diplopia occurred in 13% of endoscopic transnasal procedures, all were transient.^[12] Based on these findings, we believe that this approach is particularly suitable for tumors that are located in the medial orbit, do not bleed easily, and are intended for biopsy rather than total tumor resection.

Table 1: Comparison of approaches to intraorbital tumors.

Approach	Benefit	Risk
Transnasal	Excellent cosmetic appearance without the need for skin incisions	The operability and field of view may be limited, and the specimen volume may be reduced. Difficult to perform if bleeding occurs. Risk of injury to the internal rectus and superior oblique muscles. Inferior cosmetic appearance.
Transcranial	The ability to handle cases of intracranial extension. The ability to adapt to a wide range of lesions, including malignant tumors.	
Anterior transorbital	Incisions are inconspicuous and highly esthetic.	The eye is obstructed and restricted to lesions anterior to the equatorial plane of the eye.
Lateral transorbital	The lesions can be treated up to orbital apex lesions.	The external incision is extended laterally, making it less cosmetically pleasing.

In contrast, the transcranial approach should be considered for tumors that are located in the lateral and superior orbits, extend intracranially, and are intended for total resection. Recently, the total resection of intraorbital tumors has shown low complication rates.^[11] Shin *et al.* stated that gross-total resection of extraperiosteal tumors is generally possible without neurological complications, whereas surgical indications should be considered carefully for intraperiosteal tumors.^[9]

CONCLUSION

In addition to the transcranial and transorbital approaches, the endoscopic transnasal approach effectively treats intraorbital tumors. In particular, the endoscopic transnasal approach shows excellent safety and is minimally invasive in biopsy procedures for tumors located in the infraorbital region.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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