Orbital Pseudotumor Associated With Retrobulbar Hematoma

—Case Report—

Satoshi TSUTSUMI, Takuma HIGO, Akihide KONDO, Yusuke ABE,
Yukimasa YASUMOTO, and Masanori ITO

Department of Neurological Surgery, Juntendo University Urayasu Hospital, Urayasu, Chiba

Abstract

A 75-year-old male suffered sudden onset of retroorbital pain, visual loss, periorbital ecchymosis, and double vision without preceding trauma, paranasal sinus surgery, or infectious signs. Neuroophthalmological inspection also revealed marked restriction of the extraocular movements, visual defect, and exophthalmos on the affected side. Neuroimaging showed an irregular-shaped retrobulbar mass centered in the inferolateral aspect and partially protruding between the inferior and lateral rectus muscles without enhancement by contrast medium. High dose steroid therapy provided little improvement in the visual symptoms. Needle aspiration biopsy revealed only fluid hematoma. Surgical exploration via the lateral wall of the orbit resulted in escape of chocolate-colored, liquefied hematoma during dissection between the inferior and lateral rectus muscles. No obvious vascular lesion was recognized. A small purplish elastic soft mass with irregular contours was recognized adherent to the inferior rectus muscle within the capsule of the hematoma. The mass was subtotally resected. The histological diagnosis was inflammatory pseudotumor with hemorrhagic change. No component of vascular malformation was found. Visual function and extraocular movement improved postoperatively and neuroimaging showed no abnormal structures in the affected orbit. Pseudotumor may bleed and form a retrobulbar hematoma which carries the risk of visual loss.

Key words: spontaneous intraorbital hematoma, periorbital ecchymosis, pseudotumor

Introduction

Intraorbital spontaneous hematoma is very rare, but clinical manifestation is associated with the potential risk of visual loss. Spontaneous retrobulbar hematoma has been induced by venous malformation, metastatic cancer, trauma, paranasal sinus surgery, sinusitis, lymphangioma, and hemangioma.^{3-7, 10,11,13,14)} In contrast, cases of unknown etiology are very rare,^{8,9)} although possibly associated with atherosclerosis, hypertension, anemia, labor, and von Willebrand's disease.⁷⁾ We present a case of spontaneous retrobulbar hematoma associated with retrobulbar pseudotumor manifesting as abrupt retroorbital pain, visual loss, periorbital ecchymosis, and double vision.

Case Report

A 75-year-old, previously healthy male suffered sudden onset of retroorbital pain, visual loss, and double vision in addition to left periorbital ecchymosis. His past medical history was unremarkable for hypertension, hematological disorder, thyroid disease, sarcoidosis, and paranasal sinus surgery. He was unaware of either recent head injury or preceding infectious signs. He consulted a local ophthalmology clinic where orbital tumor was suspected and he was referred to our hospital on October 11, 2006.

Examination identified mild proptosis, abduction, and periorbital ecchymosis predominant on the left (Fig. 1). His visual acuity was depressed to 0.3 on the affected side compared with 1.0 when examined previously. The extraocular movements were highly restricted on both lateral and inferior gazes in the ipsilateral side, but the light reflex was preserved and isocoric. His retroorbital pain was exaggerated

Fig. 1 Photograph of the patient immediately after the onset showing marked periorbital ecchymosis, abduction, and mild proptosis on the left, but no obvious ocular subconjunctival chemosis.

Fig. 2 T₁- (A) and T₂-weighted (B) magnetic resonance (MR) images showing the irregular-shaped mass as hyperintense and hypointense, respectively, with poor demarcation from the inferior rectus muscle, and partially protruding between the inferior and lateral rectus muscles. The optic nerve is seen separated from the tumor near the posterior pole of the bulb. T₁-weighted MR images with gadolinium (C) revealing no vascular lesions. Arrow: inferior rectus muscle, arrowhead: lateral rectus muscle, T: tumor.

synchronous with ocular movement. He was afebrile, and blood examination showed white blood cell count of 6700/ μ l and C-reactive protein of less than 0.3 mg/dl, in addition to no findings suggestive of coagulopathy.

Computed tomography revealed a homogeneously hyperdense irregular-shaped retrobulbar mass which occupied most of the inferolateral compartment of the left orbit, and was attached to the optic

Fig. 3 Intraoperative photographs showing (A) escape of chocolate-colored, pressurized fluid hematoma during dissection between the inferior and lateral rectus muscles, and (B) a purplish elastic soft mass which was adherent to the inferior rectus muscle and resected piecemeal, but no obvious vascular lesion. Arrow: inferior rectus muscle, arrowhead: lateral rectus muscle, asterisk: hematoma cavity, T: tumor.

Fig. 4 Photomicrographs showing findings compatible with inflammatory granuloma with interstitial hemorrhagic change (A; arrow), with lymphocytic infiltration, atypia, or vascular malformation component. Hematoxylin and eosin stain, original magnification A: $\times 100$, B: $\times 400$.

nerve near the orbital apex but separate near the posterior pole of the bulb. The orbital bony structures were intact. Magnetic resonance (MR) imaging showed the mass as hyperintense on T₁-weighted imaging and hypointense on T₂-weighted imaging, poorly demarcated from the inferior rectus muscle, and partially protruding outward between the inferior and lateral rectus muscles, without enhancement by gadolinium administration. No vascular malformation was found (Fig. 2).

High dose steroid therapy was begun for the visual function deficits under a diagnosis of pseudotumor, but provided little improvement. Needle aspiration biopsy performed by an ophthalmologist obtained fluid hematoma which contained small numbers of neutrophils, lymphocytes, and histiocytes, but no erythrocytes and malignant cells (Class I).

Surgery was performed on October 30, 2006. Preceding the surgical intervention, an ophthalmologist secured the tendons of the inferior and lateral

Fig. 5 Postoperative T₁-weighted magnetic resonance images with gadolinium showing no remaining sizable mass lesion or findings suggestive of vascular lesions. Arrow: inferior rectus muscle, arrowhead: lateral rectus muscle.

rectus muscles using threads for intraoperative identification within the narrow and clouded operative field. A linear skin incision 4 cm in diameter was placed posterior to the lateral canthus parallel to the lateral orbital rim followed by an osteoplastic lateral orbitotomy. The lateral periorbital found intact, however, narrow operating field prevented the inspection over the orbital floor. Chocolate-colored, pressurized fluid hematoma escaped during the dissection between the inferior and lateral rectus muscles. A small purplish elastic soft mass with irregular contours was recognized adherent to the superior surface of the inferior rectus muscle within the capsule of the hematoma. Brisk arterial bleeding occurred when removing the mass from the inferior rectus muscle but no vascular lesion was found within the operating field (Fig. 3). The tumor was subtotally resected.

The histological diagnosis was inflammatory pseudotumor with hemorrhagic change, but no component of vascular malformation (Fig. 4). The postoperative course was uneventful. His depressed visual acuity improved to 0.6 and he no longer had double vision or retroorbital pain. The periorbital ecchymosis became light-colored. MR imaging indicated that the orbital mass had been eliminated with no residual enhanced lesions (Fig. 5).

Discussion

The patient presented with deteriorating visual function refractory to high dose steroid therapy. Spontaneous orbital subperiosteal hematoma may have peculiar clinical characteristics.^{1,2,12)} A case

of spontaneous orbital hematoma confined within the lateral rectus muscle resolved spontaneously without medication, suggesting that serial MR imaging follow up would be sufficient for patients with spontaneous intramuscular hemorrhage in the extraocular muscle.⁹⁾ The present retrobulbar hematoma was mostly confined within the muscle cone, without continuity with the periosteum surrounding the orbital contents. Moreover, the hematoma adjoined a pseudotumor which adherent to the inferior rectus muscle. We undertook surgical intervention because the retrobulbar mass might have been a malignant lesion.

Inflammatory pseudotumor associated with retrobulbar hematoma has never been described. We consider that the present retrobulbar hematoma was induced by the pseudotumor because of the close anatomical relationship, the absence of obvious vascular lesions on both pre- and postsurgical MR imaging with contrast medium, and the histological findings were compatible with inflammatory pseudotumor with remarkable hemorrhagic change, which was considered to be the origin of the easy bleeding in the absence of vascular anomaly. The hematoma was chocolate-colored and liquefied, which might suggest indolent growth of the hematoma. However, the patient experienced sudden onset of symptoms. We suppose that the liquefied hematoma, adjacent to the inflammatory granuloma with a rich vascular system, grew rapidly by an unknown mechanism and finally ruptured, went anteriorward through the inferior aspect of the inferior rectus muscle, and caused the various signs with periorbital ecchymosis. However, we could not confirm the correlation between the periorbital overlying the orbital floor and the hematoma cavity because of the narrow operating field.

Orbital pseudotumor may bleed spontaneously, even if very rarely. Neurosurgeons should be aware of the possibility of hematoma induced by pseudotumor in a patient with retrobulbar mass manifesting as sudden onset of abrupt orbital pain.

References

- 1) Dobben GD, Philip B, Mafee MF, Choi K, Belmont H, Dorodi S: Orbital subperiosteal hematoma, cholesterol granuloma, and infection. Evaluation with MR imaging and CT. *Radiol Clin North Am* 36: 1185-1200, 1998
- 2) Gillum WN, Anderson RL: Reversible visual loss in subperiosteal hematoma of the orbit. *Ophthalmic Surg* 12: 203-209, 1981
- 3) Gurelik M, Ozum U, Erdogan H, Aslan A: Orbital lymphangioma and its association with intracranial venous angioma. *Br J Neurosurg* 18: 168-170, 2004

- 4) Hart RH, Luthert PJ, Rose GE: Renal cell carcinoma metastasis masquerading as recurrent orbital haematoma. *Orbit* 24: 281–284, 2005
- 5) Holland D, Maune S, Kovacs G, Behrendt S: Metastatic tumors of the orbit: a retrospective study. *Orbit* 22: 15–24, 2003
- 6) Jovanovic M: [Surgical treatment of post-contusion orbital hematoma in the late phase]. *Srp Arh Celok Lek* 131: 396–399, 2003 (Ser, with Eng abstract)
- 7) Krohel GB, Wright JE: Orbital hemorrhage. *Am J Ophthalmol* 88: 254–258, 1979
- 8) Matsuura H, Baba M, Kudo M, Nakaoka T: Spontaneous intraorbital hematoma mimicking orbital neoplasm — case report. *Neurol Med Chir (Tokyo)* 35: 45–47, 1995
- 9) Onami H, Kunikata H, Hojo M, Nakagawa Y, Tamai M: Natural course of hematoma in lateral rectus muscle followed by magnetic resonance imaging. *Tohoku J Exp Med* 206: 361–364, 2005
- 10) Petrelli RL, Petrelli EA, Allen WE 3rd: Orbital hemorrhage with loss of vision. *Am J Ophthalmol* 89: 593–597, 1980
- 11) Schick B, Weber R, Keerl R, Draf W, Rochels R: [Orbital hematomas]. *Laryngorhinootologie* 75: 363–367, 1996 (Ger, with Eng abstract)
- 12) Takahashi K, Uyama M, Okada H, Izumi H: [A case of spontaneous orbital hematoma in a child]. *Nippon Ganka Gakkai Zasshi* 92: 182–187, 1988 (Jpn, with Eng abstract)
- 13) Wright JE, Sullivan TJ, Garner A, Wulc AE, Moseley IF: Orbital venous anomalies. *Ophthalmology* 104: 905–913, 1997
- 14) Yen MT, Hipps WM: Nasolacrimal sac hematoma masquerading as an orbital mass. *Ophthalm Plast Reconstr Surg* 20: 170–172, 2004

Address reprint requests to: Satoshi Tsutsumi, M.D.,
Department of Neurological Surgery, Juntendo
University Urayasu Hospital, 2-1-1 Tomioka,
Urayasu, Chiba 279-0021, Japan.
e-mail: neuro1969@yahoo.co.jp