Vascular anomaly in the levator aponeurosis of neurofibromatosis type 1

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Dear Editor,

I am Satoru Kase, from the Department of Ophthalmology, Faculty of Medicine and Graduate School of Medicine, Hokkaido University, Sapporo City, Japan. I write to present a case of neurofibromatosis type 1 (NF1) showing massive hemorrhage during involutional blepharoptosis surgery. NF1 is a genetic disorder caused by mutations in the NF1 tumor suppressor gene, characterized by its cutaneous manifestations called “café au lait” spots, lentigines, and neurofibroma. Ophthalmologically, NF1 complicates various tumors involving the uvea and eyelid. A previous pathological study verified that hematopoietic-stromal interactions underpin the neurofibroma. In addition to the occurrence of the tumors, vascular abnormalities such as aneurysms, disruption of the elastic lamina in the vessel walls, and proliferation of vascular smooth muscles, have been reported histologically, which could occur in the arteries and veins of the human kidney. Vascular involvements in the organs other than the kidney to cause serious sequela, however, have been underestimated in patients with NF1. Indeed, little is known about the hemorrhagic risks associated with the vascular abnormalities in oculoplastic surgeries of NF1 patients. We herein report an NF1 patient with marked hemorrhage during blepharoptosis surgery, and the histological findings in the levator aponeurosis tissue obtained during the operation, proving causative vascular abnormalities.

A 64-year-old female complained of blurred vision and eyelid drooping in both eyes. She has been diagnosed with NF1 when she was young. Her medical history included clipping of an unruptured brain aneurysm in 1998. She had been wearing hard contact lenses (HCL) bilaterally for over 30 years because of high myopia. She stopped using HCL after cataract surgery with intraocular lens implantation in July 2012. Her visual acuities were 20/20 bilaterally with a normal intraocular pressure in February 2014. Marginal reflex distance was 2 mm bilaterally, and levator functions were 13 mm bilaterally (Figure 1A). Slit-lamp examination revealed bilateral multiple iris nodules. She had not taken any anti-coagulation agents or anti-platelet agents. Fundus examination showed nothing of note. Based on the diagnosis of aponeurotic blepharoptosis, she underwent levator aponeurosis suturing to the superior tarsus margin as a blepharoptosis surgery bilaterally.

Marked hemorrhage appeared during the surgery on isolation of the orbicularis muscle, tarsal plate, and levator aponeurosis, as well as suturing aponeurosis with the tarsal plate. The origin of hemorrhage was not determined. Bipolar coagulations were frequently required to regulate intraoperative bleeding. During the operation, the levator aponeurosis tissue was partially resected and submitted for histopathological analyses.

Histologically, the aponeurosis tissue contained arteries and veins with irregularly dilated vessel walls, where a few capillaries were intermingled (Figure 2A, 2B). Moreover, a variety of α-smooth muscle actin (SMA)-positive spindle cells were present around the vessel walls (Figure 2C). S100-positive neurofibromas were not observed in the tissue. After the surgery, icing of the bilateral upper eyelid was conducted all day for one day. While mild swelling of the upper eyelid was noted one week after blepharoptosis surgery (Figure 1B), it showed favorable correction without swelling 3 months after the surgery (Figure 1C).

The present patient, who had suffered from NF1 since she was young, presented with aponeurotic blepharoptosis, which probably resulted from the long-term use of HCL and/or cataract surgery. Blepharoptosis surgery was conducted;
however, massive bleeding occurred during the operation. Although the bleeding was stopped by coagulation, ruptured vessels were not detectable. Adekeye et al.[5] reported surgical excision of plexiform neurofibromas affecting the eyelids in 6 patients with NF1, and described intraoperative bleeding during the excision of eyelid neurofibromas associated with NF1. This was very similar to the situation in our patient during the operation, suggesting that patients with NF1 have vascular system vulnerability during an operation.

Interestingly, the histological findings in the present patient showed vascular abnormalities in the levator aponeurosis tissues without nodular proliferations; the tissue contained a variety of abnormally dilated arteries and veins, while capillaries were rarely observed. On the contrary, the aponeurosis in involutional blepharoptosis with no history of NF1 contains striated and smooth muscles[4], fibroblasts and fat, where capillaries are intermingled (Figure 2D). The present histological findings also proved altered distributions of arteries and veins located in the aponeurosis of NF1. Immunohistochemically, SMA-positive smooth muscle cells formed several layers in the vessel wall, strongly indicating the proliferation of vascular smooth muscle. This histological findings are almost identical to those reported in the kidney of an NF1 patient[3]. Thus, patients with NF1 may have uniform vascular abnormalities consisting of an altered distribution of arteries and veins as well as vascular smooth muscles in the systemic vascular system. These vascular pathologies may be responsible for the appearance of massive hemorrhage during the operation. Therefore, this report indicates that oculoplastic surgeons should pay attention to intraoperative hemorrhage in blepharoptosis surgery with/without eyelid tumors, which is possibly caused by vascular anomalies in patients with NF1.

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REFERENCES