• Letter to the Editor •

Spontaneous rupture of ocular surface squamous neoplasia–a case report

Jun Hyuk Son[1], *Su-Ho Lim*[1,2]

[1]Department of Ophthalmology, Yeungnam University College of Medicine, Daegu 42415, Republic of Korea
[2]Department of Ophthalmology, Daegu Veterans Health Service Medical Center, Daegu 42835, Republic of Korea
Correspondence to: Su-Ho Lim. Department of Ophthalmology, Daegu Veterans Health Service Medical Center, #60 Wolgok-Ro, Dalseo-Gu 42835, Republic of Korea. mdshlim@gmail.com
Received: 2020-03-09 Accepted: 2021-09-29

DOI:10.18240/ijo.2022.02.25

Citation: Son JH, Lim SH. Spontaneous rupture of ocular surface squamous neoplasia–a case report. *Int J Ophthalmol* 2022;15(2):357-359

Dear Editor,

Ocular surface squamous neoplasia (OSSN) typically occurs adjacent to limbus or in inter-palpebral fissures, over a preexisting pinguecula[1]. The term OSSN includes mild dysplasia at one end of its disease spectrum and invasive squamous cell carcinoma at the other and is the most common non-pigmented malignancy of the ocular surface[2-3]. The tumor can involve tarsal conjunctiva or it can be associated with other conditions, and these presentations can make correct diagnosis challenging[4]. However, spontaneous rupture of OSSN is rare, and no previous report has been issued in Korea. Herein, we describe a case of active bleeding associated with spontaneous rupture of OSSN.

Ethical Approval This case study was conducted in accordance with the Declaration of Helsinki. Written informed consent was obtained from the patient for publication of this case report with any accompanying images.

CASE REPORT

An 87-year-old Korean male patient presented with a protruding ocular mass and ptosis in his right eye of several months duration. External photograph revealed a tumor 5×4 cm^2 sized tender, lobulated cystic mass and restricted extraocular movements (Figure 1A). Slit lamp examination demonstrated adome-shape, non-movable, and protruding ocular surface mass, originating from superior bulbar conjunctiva and limbus in his right eye. The patient also had Alzheimer dementia, lobar pneumonia, a newly developed splenic infarction, and pulmonary artery thromboembolism. He had a nasogastric tube because of his cognitive and functional status. He was a retired veteran who served in Korean War and Vietnam War. There was no other lesion suspicious of human papillomavirus (HPV) infection including common warts, genital or anal wart by physical examination and medical history. Preoperative serologic test revealed leukocytosis, elevated C-reactive protein, erythrocyte sedimentation rate, elevated liver enzyme and creatinine above 2.9 mg/dL (estimated glomerular filtration rate, eGFR=22 mL) due to systemic illness. And routine preoperative human immunodeficiency virus (HIV) antibody testing revealed the negative result.

Thus, palliative surgery was planned for biopsy and tumor size reduction. Two days after the first visit to the ophthalmic clinic, he came back due to active bleeding associated with spontaneous rupture of the tumor (Figure 1B). Although magnetic resonance imaging (MRI) is usually preferred to evaluate soft tissues and intraocular involvement, we performed the computed tomography (CT) scan considering his general condition and economic status. A contrast enhanced-CT axial cut demonstrated an ocular surface lesion with homogenous enhancement and no visible intraocular invasion (Figure 1C). Under the sub-Tenon anesthesia with 2% lidocaine, we performed the palliative surgery. Under the surgical microscope, the tumor was located at or near the limbus (overhanging conjunctivo-corneal mass) and demonstrated fleshy lobulated characteristics, engorged feeder vessels, and was immobile and firmly fixed to underlying episcleral or scleral tissues (Figure 1D). We removed almost all visible protruding ocular surface mass and left most of the lesion in a bare sclera state without ocular surface reconstruction such as conjunctival flap or other donor graft tissues. To minimize exposure of extraocular muscles, we performed minimally sliding conjunctival advancement for superior conjunctiva at 12 o'clock direction using 8-0 vicryl anchoring suture.

Histopathologic examinations showed full-thickness cell atypia, loss of polarity, prominent mitotic figures and malignant epithelial cells in substantia propria forming "keratin pearls", which suggested OSSN (Figure 2A, 2B). OSSN was managed by wide surgical excision under local

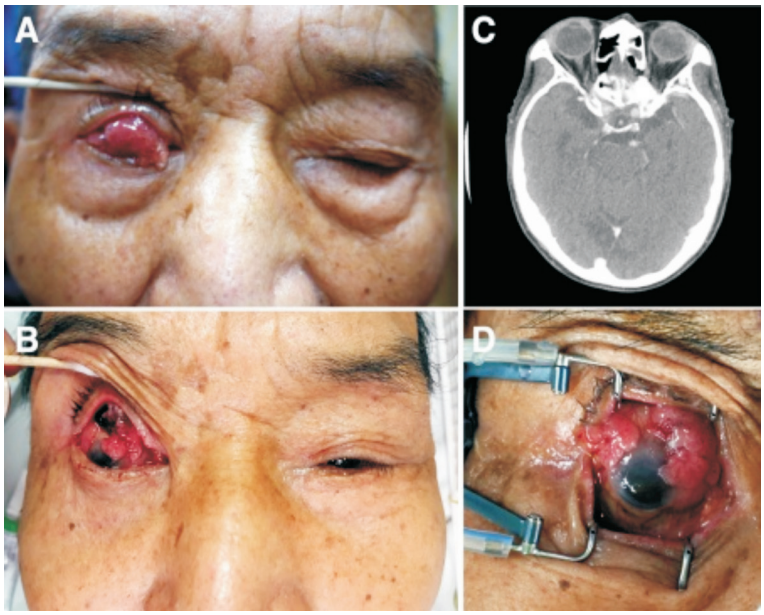

Figure 1 External photographs and computed tomography scan A: Preoperative photograph showing a tumor 5×4 cm^2 sized lobulated cystic mass; B: Photograph taken two days after first examination showing active bleeding associated with spontaneous tumor rupture; C: Contrast enhanced-CT axial cut showing a homogenous enhanced ocular surface lesion with no visible intraocular invasion; D: Under the surgical microscope, the tumor, which was located at or near the limbus (overhanging conjunctivo-corneal mass), demonstrated fleshy lobulated characteristics, engorged feeder vessels, and was immobile being firmly fixed to underlying episcleral or scleral tissues.

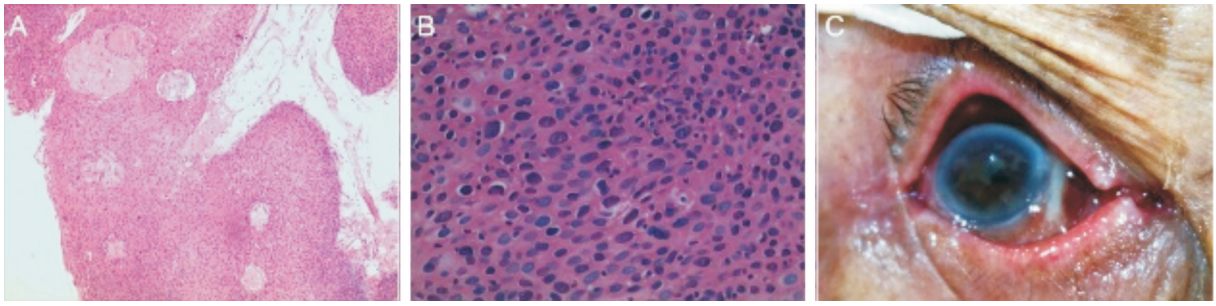

Figure 2 Histopathologic examinations and external photograph A: Histopathologic examination demonstrated full-thickness cell atypia, loss of polarity, prominent mitotic figures, and malignant epithelial cells in the substantia propria forming "keratin pearls", which suggested OSSN (hematoxylin & eosin stain, ×100). B: At high magnification, the neoplastic cells showed pleomorphism, with hyperchromatic nuclei. A mitotic figure is present with intercellular bridge (hematoxylin & eosin stain, ×400). C: After 2nd cycle of adjuvant topical chemotherapy with 0.04% MMC, the tumor was markedly reduced.

anesthesia; it was followed by adjuvant topical chemotherapy with mitomycin-C (MMC) 0.04% to reduce residual tumor. We planned the 4 cycles of postoperative MMC (0.04% four times daily regimen), each cycle consisting of 1wk on and 1wk off treatment. After 2nd cycle of adjuvant chemotherapy, the tumor was markedly reduced (Figure 2C). However, the patient was lost to follow-up due to his systemic conditions and did not complete planned chemotherapy.

DISCUSSION

OSSN is typically characterized by epithelial thickening, a prominent "corkscrew" vascular pattern, or a gelatinous or leukoplakic surface, indicative of surface keratinization. In particular, adjacent conjunctiva might contain a lesion feeder vessel[3-5]. However, in the described case, the initial presentation was a protruding ocular mass without the typical characteristics of OSSN, and thus, accurate diagnosis was challenging before histopathologic evaluation. Recently, technical developments such as *in vivo* confocal microscopy and anterior segment optical coherence tomography (OCT) can provide additional information about superficial dysplastic lesions[6-7], but they cannot be used to assess the potential invasive growth of lesion or protruding nature. In our patient, after spontaneous OSSN rupture, the surface of the tumor exhibited the typical characteristics of OSSN. Active bleeding associated with spontaneous rupture, as was observed in this case, is rare and has not been previously reported in Korea.

The etiology of OSSN appears to be multifactorial, and likely involves a variety of environmental factors in a susceptible

host[2,4]. These multifactorial etiologies include exposure to ultraviolet radiation, chemical carcinogen, mutation of tumor suppressor gene p53, and viral infections including human papilloma virus and HIV infection associated with immunosuppression[2,4,8]. In this patient, chronic sun exposure (ultraviolet) associated with his occupation (retired soldier), and current immunosuppressive condition related to general illness may contribute to the risk of malignancy.

The management of OSSN includes surgical resection with topical chemotherapy, topical/local interferon alpha-based immunomodulation, cidofovir, and/or photodynamic therapy. A "no touch" technique is preferred to avoid the potential risk of seeding and to provide a tumor-free margin[8]. In the present case, the patient had several severe systemic diseases. The palliative surgical treatment strategy employed achieved a relatively favorable result.

To the best of our knowledge, this case is the first case report issued of spontaneous OSSN rupture in Korea. Our experience highlights that initial presentation of OSSN may not exhibit typical characteristics of a "corkscrew" vascular pattern or a gelatinous or leukoplakic surface, and that active bleeding associated with OSSN rupture might occur in elderly patients on anticoagulants.

ACKNOWLEDGEMENTS

Foundation: Supported by a VHS Medical Center research grant, Republic of Korea (No.VHSMC 22017).

Conflicts of Interest: Son JH, None; **Lim SH,** None.

REFERENCES

1 Cicinelli MV, Marchese A, Bandello F, Modorati G. Clinical management of ocular surface squamous neoplasia: a review of the current evidence. *Ophthalmol Ther* 2018;7(2):247-262.

2 Mittal R, Rath S, Vemuganti GK. Ocular surface squamous neoplasia—review of etio-pathogenesis and an update on clinico-pathological diagnosis. *Saudi J Ophthalmol* 2013;27(3):177-186.

3 Oellers P, Karp CL, Sheth A, *et al*. Prevalence, treatment, and outcomes of coexistent ocular surface squamous neoplasia and pterygium. *Ophthalmology* 2013;120(3):445-450.

4 Shields CL, Chien JL, Surakiatchanukul T, Sioufi K, Lally SE, Shields JA. Conjunctival tumors: review of clinical features, risks, biomarkers, and outcomes—the 2017 J. Donald M. Gass Lecture. *Asia Pac J Ophthalmol* (*Phila*) 2017;6(2):109-120.

5 Xu Y, Zhou Z, Xu Y, Wang M, Liu F, Qu H, Hong J. The clinical value of *in vivo* confocal microscopy for diagnosis of ocular surface squamous neoplasia. *Eye* (*Lond*) 2012;26(6):781-787.

6 Atallah M, Joag M, Galor A, Amescua G, Nanji A, Wang JH, Perez VL, Dubovy S, Karp CL. Role of high resolution optical coherence tomography in diagnosing ocular surface squamous neoplasia with coexisting ocular surface diseases.*Ocul Surf* 2017;15(4):688-695.

7 Ip MH, Robert George CR, Naing Z, Perlman EM, Rawlinson W, Coroneo MT. Topical cidofovir for treatment-refractory ocular surface squamous neoplasia.*Ophthalmology* 2018;125(4):617-619.

8 Sayed-Ahmed IO, Palioura S, Galor A, Karp CL. Diagnosis and medical management of ocular surface squamous neoplasia. *Expert Rev Ophthalmol* 2017;12(1):11-19.