CASE REPORT

Smallpox: An eradicated infection with persistent sequels – Case report and a brief on smallpox

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Abstract Sequelae of smallpox infection on the ocular surface are still seen, including corneal scars adherent leukoma and phthisical globes. This paper will report another sequel of smallpox infection causing inadvertent bleb in a 62-year-old diabetic female with no history of ocular surgery or trauma in either eye. The patient had smallpox infection during her childhood. Her follow up extended from May 1997 until August 2007 with a constant eye examinations including controlled intraocular pressure, avascular cystic inadvertent bleb, and up drown peaked pupil.

1. Introduction

Although World Health Organization (WHO) declared global eradication of smallpox in May 1980, its irreversible sequelae are observed in our daily ophthalmology practice.

Ocular complications after smallpox infection are uncommon but when occurs, it could be devastating. About 5–9% of patients with smallpox infection develop ocular complications, while 10–20 patients per million develop ocular complications from smallpox immunizations, usually through autoinoculation, in which the patient transfers vaccinia virus from the immunization site to the eye (Semba, 2003). Painful pustules on the bulbar conjunctiva develop with tremendous inflammatory reaction and purulent discharge often extending to the cornea causing inflammation, scarring, and perforation with loss of the eye in few patients. Ocular lesions can involve infection to the lids, conjunctiva, and cornea, iris, and optic neuritis (Semba, 2003). The most serious is corneal involvement; blindness results in approximately 35–40% of eyes affected with keratitis and corneal ulcer (Semba, 2003; Ray, 1984; Peters and Gilles, 1981; Ruben and Lane, 1970; Ellison, 2004).

Our concern with ocular complications of smallpox continuous as vaccinia immunization given to selected cases may produce similar generalized disease. Variola virus, the cause of smallpox, and vaccinia virus, used in smallpox immunization, are both orthopoxviruses that are associated with serious ocular complications (Semba, 2003; Ruben and Lane, 1970).

Symptoms include tearing or reduced vision but it can be asymptomatic. Signs vary and include observing a subconjunctival bleb, wound gape, leaking wound, hypotony with corneal folds and shallow anterior chamber, and macular edema. The bleb can be complicated by hyphema, choroidal signs of hypotony or endophthalmitis.
Inadvertent bleb formation is seldom seen after transient ocular infection. We report a case of inadvertent filtering bleb formation after ocular sequelae of childhood smallpox infection.

2. Case report

A 50-year-old Saudi woman was first presented to the King Khaled Eye Specialist Hospital in Riyadh, Saudi Arabia in May 1997 with corneal scar in her left eye (Fig. 1). She was referred for evaluation; the patient denied any past ocular surgical history or infections except of childhood smallpox, resulting in her left corneal scar with a drop of vision thereafter.

Right eye examination revealed vision of 20/60, corneal scar, and immature cataract. Clinical examination showed her vision in the left eye as counting fingers near face, a large superior avascular limbal bleb, elevated with hot spots by fluorescein staining, but no frank leak (Fig. 2). Corneal scarring was seen under the area of the bleb, the pupil was peaked and updrawn superiorly. Anterior chamber was deep and quiet. The lens showed signs of immature cataractous changes. Posterior pole examination was normal. The patient did not have any underlying systemic problems and the rest of ocular examination was unremarkable.

During her follow ups in 1998, she was found to have a leaking bleb. It was managed conservatively with antibiotics and patching. On her next follow up, she had stable clinical ocular exams with normal intraocular pressure (IOP) measurements and a functioning, non-leaking bleb.

The patient developed cataract and underwent an uneventful clear corneal temporal approach phacoemulsification with intraocular lens implantation in March 2003. At the end of the procedure a small air bubble was kept in the anterior chamber which was seen under the bleb in the first postoperative day, indicating a functioning fistula between the anterior chamber and the bleb through the sclera (Fig. 3).

Postoperative examination, vision improved to 20/40, intraocular pressure measurement (IOP) of 11 mmHg, quite conjunctiva with an elevated bleb, clear central cornea with secured wound, anterior chamber deep and quiet, posterior chamber intraocular lens was well centered in place.

3. Discussion

Smallpox infection has played a significant role in world history in relation to both serious epidemics and dangerous measures taken to prevent the spread of the infection. The virus is a large ovoid DNA virus that belongs to the poxvirus family. Derivatives of the virus are vaccinia and molluscum contagiosum. There are two types of the virus; variola major and variola minor (Alastrim) with fatality rates of 3–35% and <1%, respectively (Semba, 2003; Ray, 1984).

Global eradication of the disease was confirmed in 1979 and accepted by World Health Organization (WHO) in May 1980. However, the viruses were retained by only two reference laboratories – one in Atlanta, USA and another in Moscow, Russia. Viruses were destructed in all other laboratories (Peters and Gilles, 1981; Edward and Agwunobi, 2007).

Inadvertent blebs may occur, particularly, after scleral or limbal incision techniques for intraocular procedures and commonly after complicated cataract surgery with wound dehiscence (King, 1991). Causes of inadvertent blebs include wound dehiscence or leak under sealed conjunctiva, poor wound closure at the time of surgery, postoperative complications, blunt trauma to the globe, Scleromalacia Paralimbus (King, 1991; Nirankara et al., 1962), or it can even be spontaneous with peripheral corneal degeneration (Starr et al., 1981).

Prolonged use of topical steroid, more than 3 weeks after
surgery, increases the rate incidence of inadvertent bleb formation by 8.7% compared to 1.6% when corticosteroids are omitted (Harold and Kirk, 1974). Four out of five inadvertent bleb cases close spontaneously; the rest may require surgical intervention (Christensen and Rundle, 1970).

Infection by the virus usually occurs through the respiratory route by inhalation of infective material as droplets from the respiratory tract, dust containing dried droplets, or material derived from the skin lesions. Ocular infection after vaccinia immunization may occur after touching the vaccination site then auto-inoculating the virus into the eye (Semba, 2003; Ray, 1984; Peters and Gilles, 1981).

Ocular complications were uncommon but when it occurs, it could be devastating (Ray, 1984; Peters and Gilles, 1981; Ruben and Lane, 1970; Ellison, 2004). Frequently exanthematous watery conjunctivitis develops about 5 days after the onset of the clinical disease and clears without complications. Painful pustules on the bulbar conjunctiva develop with tremendous inflammatory reaction and purulent discharge often extend to the cornea causing inflammation, scarring and perforation with loss of the eye in few patients. Vaccinia may produce a similar systemic and ocular disease (Ruben and Lane, 1970).

In this case the possible mechanism for the bleb formation was intense inflammation and melting in the limbal area causing a pustule that perforated creating a fistula or a track between the anterior chamber and the sclera, resulting in the inadvertent bleb formation.

Inadvertent blebs can result from surgical, traumatic insult to the limbal area. The bleb can survive for many years without causing major trouble to the eye.

Smallpox sequelae are still seen and its corneal complications caused many patients their vision in the effected eye. From this report, we can observe unreported sequelae of smallpox infection on the eye, and the benign course of an inadvertent bleb. Continuous monitoring of IOP is crucial to avoid increase of the IOP if the functioning inadvertent bleb stops filtering.

Observation is usually enough with no treatment if there are no signs or symptoms indicating the possibility of complications. Many interventional ways have been described to treat these blebs including medical treatment with patching or autologous injection of blood into the bleb (Wong et al., 1995). Neodymium: YAG laser have been reported to manage complicated blebs (Geyer, 1998). Surgical repair of the wound, bleb excision and resuturing (Myers et al., 2000), or scleral grafts (Rao and Padmanaban, 1997; Clinch and Kaufman, 1992) can be done. Surgical intervention is required in some cases where the bleb is large cystic thin walled, gapping wound with or without iris prolapse, threatened infection, chronic irritation, foreign body sensation, persistent hypotony ± macular edema and inability to wear contact lenses (Myers et al., 2000; Rao and Padmanaban, 1997; Clinch and Kaufman, 1992).

Corneal complications of smallpox are seen in our daily practice requiring on occasions penetrating keratoplasty for visual rehabilitation if there is an acceptable visual potential.

References


