Sturge-Weber-Syndrome with Extreme Ocular Manifestation and Rare Association of Upper Airway Angioma with Anticipated Difficult Airway

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SUMMARY

We report a rare case of an 18 year old girl with Sturge-Weber syndrome, she had extensive facial port wine stains, right bupthalmos and advanced glaucoma involving both eyes. She underwent right eye glaucoma drainage device surgery under general anaesthesia, and had a difficult intubation due to extensive angiomatous like soft tissue swelling at her upper airway. This report highlights the importance of being aware of the need for continuous follow-up in Sturge-Weber syndrome patients as this syndrome can lead to blindness due to advance glaucoma and the awareness of possible difficult intubation for this group of patients.

INTRODUCTION

Sturge Weber Syndrome (SMS) belongs to a group of disorders collectively known as phakomatoses, disorders of the central nervous system that results in lesions on skin, brain and eyes. This syndrome consists of congenital hamartomatous malformation that may affect the eye, skin and central nervous system at different times. There is no sexual or racial predilection. Vascular angiomas may involve the mouth and airway (nose, palate, gingival, tongue, larynx and trachea) leading to difficult mask ventilation, laryngoscopy and intubation, apart from bleeding caused by perforation or rupture. Leptomeningeal angiomas may cause hemiparesis, stroke like episodes, seizures, developmental delay, learning disabilities and mental retardation. Cardiac anomalies which may be associated with this syndrome include septal defects, valvular stenosis, great vessels transposition and deep arteriovenous malformation (rare) which may lead to considerable shunt and lead to cardiac hypertrophy and failure [1-2]. It is postulated that failure of regression of residual embryonal blood vessels results in formation of angiomata, which will lead to increased permeability, stasis, thrombosis, ischemia, infarction or anoxia. SWS based on Roach scale can be divided into Type I – Both facial and leptomeningeal angiomas; may have glaucoma, Type II – Facial angioma alone; may have glaucoma and Type III – isolated leptomeningeal angiomas; usually no glaucoma.

CASE REPORT

The patient was born with a bigger right eye and purplish birth marks on her face. At one month old she started to developed seizures and was first seen by an ophthalmologist. Her parents were told that their daughter had 'some eye problem' in her right eye and needed to put eye drops. However, the parents assumed that their daughter was well as her eyes remained white and not in pain. The patient was lost to follow up till the age of 18 years old. She was said to be mentally 'slow' but able to take care of herself.

Upon her visit to us at the age of 18, she had extensive facial port wine stains involving both sides of her face, which extended to her neck and upper trunk. Her right eye was buphthalmic, with right exotropia (Figure 1,2).There was a right relative afferent papillary defect (RAPD) indicating severe right optic nerve dysfunction. Her right eye vision was reduced to hand movement with good light projection to superior and temporal visual field only. The vision in the left eye was 6/24, improved to 6/12 (pin hole) and good near vision of N6. The intraocular pressure (IOP) was 50 mm Hg in the right eye and 28 mm Hg in the left eye (normal 10-21 mm Hg). Fundus examination of the right eye revealed a pale optic disc with advanced cupping and left eye also showed glaucomatous disc changes with thinning of the nasal and inferior neuroretinal rim. Gonioscopy examination of the anterior chamber angles was open without any blood seen in the schlemm canal. A diagnosis of bilateral secondary open angle glaucoma associated with SWS was made on clinical grounds, supported by Humphrey visual field findings.

In order to control the intraocular pressure, 4 types of topical anti-glaucoma were started to the right eye and 1 type of topical anti-glaucoma medication to the left eye. Despite that her right IOP remained high (>30 mm Hg). She underwent right glaucoma drainage surgery (Ahmad valve implant) under general anaesthesia within 2 months.

The anaesthetist was consulted on the potential airway problems that may be associated with this syndrome. From the history, she had been intubated twice and admitted to the Intensive Care Unit for status epilepticus and intubation was said to be difficult.When assessed, she was classified as Mallampati 3 – 4. This assessment made more difficult in

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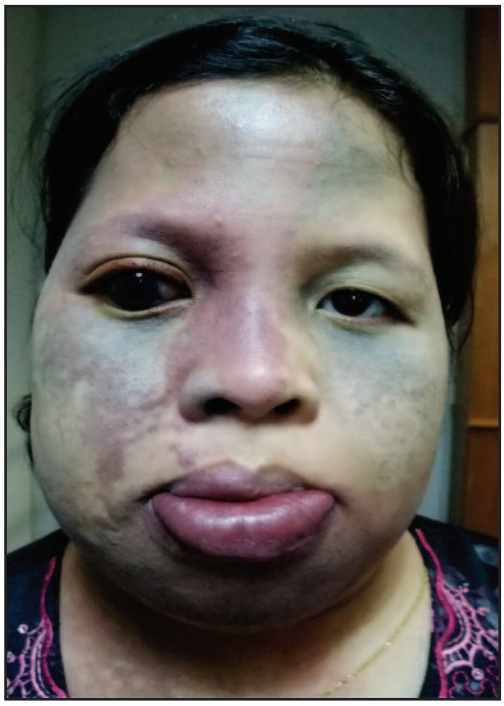

Fig. 1: Extensive facial port-wine stain with right bupthalmous

view of her large tongue. Thyromental distance was approximately 3 fingerbreaths. Difficult mask ventilation was anticipated in view of the gross vascular hypertrophy of lips, buccal mucosa, gums, tongue. In view of her generalized port wine stains on her face, neck and upper chest, the possibility of angiomatous lesions within the oral cavity and airway was also anticipated. The patient was preoxygenated and subsequently anaesthetized with inhalational induction using 100% oxygen and sevoflurane in increasing incremental doses. Facemask ventilation was possible and a nondepolarising muscle relaxant was given. Once an adequate depth of anaesthesia was achieved, the Glidescope was inserted gently and carefully. The vocal cords were visualised with cricoid pressure assistance however there was difficulty in manoeuvring the endotracheal tube (ETT) through the vocal cords. After 2 attempts in inserting the ETT, there was contact bleeding and the patient desaturated briefly to 94%. After mask ventilation and oxygen was saturation back to 100%, oral fibreoptic intubation was attempted as planned if intubation with the Glidescope failed. Two attempts at fibreoptic intubation were required before successful intubation was achieved as the patient desaturated during fibreoptic manipulation made difficult by the oedematous oral tissues. IV dexamethasone 8 mg was given in view of the numerous manipulations on the airway to help reduce potential airway oedema. Subsequently, anaesthesia was uneventful throughout the surgery.The patient was extubated when she had a good gag reflex, responded to call and was able to protect her airway. No further bleeding was detected on oropharyngeal suctioning prior to extubation .She was monitored in recovery and given 100% via facemask, where she remained stable with no signs of airway obstruction.

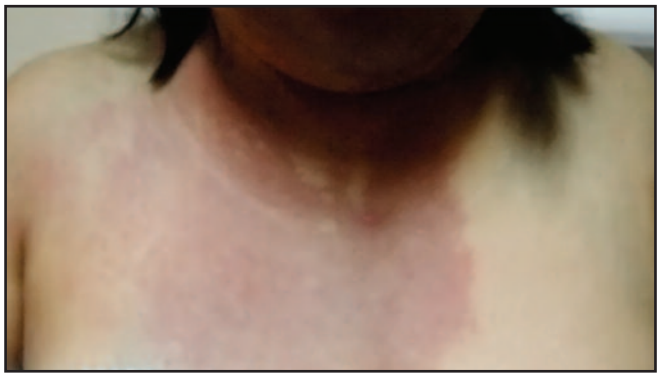

Fig. 2: Extensive port-wine stain extending to her neck and upper chest.

The right eye IOP normalized at post-operation 1 week, however her right visual acuity did not show improvement.

DISCUSSION

Glaucoma is one of the sight threatening complications associated with SWS. This syndrome has been linked to increased episcleral venous pressure leading to disruption of normal aqueous humor outflow from the eye, resulting in high IOP. Histopathological examination of the eye in SWS also found abnormal trabecular meshwork which may contribute to raise IOP[3]. The glaucoma can present anytime from infancy to adulthood. In this case, the girl was born with a right big eye indicating that she had congenital glaucoma leading to bupthalmos.

The most recognizable clinical feature of SWS is the facial angioma or port wine stain. It has been reported to occur in up to 70% of SWS patients[4]. Interestingly glaucoma is found to be closely associated with the location of facial angioma. Glaucoma has been shown to occur most often on the side where the facial angioma is located, especially with the involvement of the ophthalmic (V1) and maxillary (V2) division of the trigeminal nerve. 25% of SWS patients developed glaucoma in the eye without cutaneous involvement. In our case, the facial angiomas involve both side of her face with more extensive involvement of her right side. It correlates with the severity of glaucoma involvement in both her eyes. Management of glaucoma in SWS is challenging and surgical management is fairly common for adequate intraocular pressure control. In this case, we would like to highlight the upmost importance of education to parents with a SWS child regarding the disease process and its associated ocular complication which can be devastating and leads to irreversible visual lost. The parents need to be committed and understand the need of long term follow up for SWS child. Due to poor understanding, the child ocular problem was dismissed by the parents as being 'born with it' and benign. Even though she had multiple visits to doctors, she was not referred to an ophthalmologist till the age of 18. Medical personals also need to be alerted of the close association of SWS with glaucoma.

Patients with Sturge-Weber Syndrome should be carefully evaluated for associated anomalies. In this patient there were no cardiac abnormalities. Neurologically, apart from epilepsy, she was deemed to be mentally 'slow'. Though these patients generally tolerate anaesthesia well, mask ventilation, laryngoscopy and intubation may be difficult due to angiomas involving the nose, lips, oral cavity, tongue, palate, larynx and trachea [1-2]. Uncontrolled haemorrhage may result from perforation or rupture of these vascular lesions [1]. Therefore, intubation should be done using a soft, well lubricated, non-styleted cuffed ETT and careful oropharyngeal and tracheobronchial suction is crucial in avoiding trauma to these lesions [1-2]. A smooth induction, intubation and extubation of these patients is also mandatory to limit the rise in intraocular and intracranial pressures as a light plane of anaesthesia, bucking, straining and airway obstruction during these periods of time are detrimental [5]. Drugs such as a suxamethonium and ketamine which cause a rise in intraocular pressure should also be avoided [1]. Perioperative hypoxemia, hypoglycaemia, hypotension and hyperthermia may precipitate status epilepticus and should be avoided [2]. Because these vascular lesions also have abnormal autoregulation, intraoperative blood pressure should also be well controlled [3]. Concommitant chronic use of antiepileptics can also affect the metabolism of certain anaesthetic agents.

Despite the history of a difficult airway, we did not to do an awake fibreoptic intubation for her as the patient was deemed mentally 'slow' and we did not want to risk causing stress and agitation as emotional stress can cause increased blood pressure and consequent swelling of angiomas. Our plan was to induce the patient with an inhalational induction agent, test ventilate the patient and assess if mask ventilation was possible and then give a non-depolaring muscle relaxant to ensure a smooth intubation and avoid any further rises in intraocular pressure. Two anaesthetists were in the operating theatre to manage her airway. The plan was to use a Glidescope to visualize the cords and perform careful intubation failing which oral fibreoptic intubation would be performed. A GlideScope® Video Laryngoscope is designed for difficult and unpredictable airways to help provide a clear, real time view of the patient's airway and endotracheal tube placement (Figure 3).By using the Glidescope, we would be able to see where we were inserting the intubating blade of the scope and the passage of the ETT as it was inserted during intubation thereby avoiding trauma to the surrounding tissues and anticipated vascular lesions intraorally. After failing to intubate the patient with the Glidescope, oral fibreoptic intubation was performed successfully after 2 attempts in which there was difficulty manipulating the scope due to contact bleeding and oedematous soft tissues intraorally. We felt it was best not to use a supraglottic airway as it may cause trauma to the intraoral angiomas during insertion and would not be able to prevent aspiration of blood if rupture of the angiomas should occur. Airway angiomas in SWS is rare but failing to recognize the potential risk of torrential bleeding during airway management will undoubtedly endanger the patient's life

CONCLUSION

Glaucoma is closely related to SWS especially when there is facial angioma involving V1 And V2 trigeminal distribution. However prompt and early surgical intervention can provide adequate IOP control. Early intervention can save sight. Anaesthetic management is challenging due to the varied presentation and extent of the syndrome, ranging from localized superficial skin lesions to extensive systemic and airway involvement.

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