Anesthesia Management of a Patient with Gorlin-Goltz Syndrome: Risk of Difficult Airway: Case Report

Gorlin-Goltz Sendromlu Hastada Anestezi Uygulaması: Zor Hava Yolu Riski

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ABSTRACT Gorlin-Goltz syndrome is an inherited disease which affects the skin, bones, ocular and dental structures. Gorlin-Goltz syndrome have possible skeletal defects, central nervous system abnormalities, internal neoplasms, mental retardation and endocrin disorders. The characteristic skeletal anomalies of this syndrome are mandibular prognathism, bossing of frontal and parietal bones, high arched palate, cleft palate and lip, significance of supraorbital ridges, bifid rib and vertebral anomalies. A characteristic feature of Gorlin-Goltz syndrome is the formation of giant papillomas on the skin and mucous membranes. This syndrome is characterized by many alterations, some of which is preventing especially intubation at a normal conduct of anesthesia. In the present case report, after induction of anesthesia, difficult airway management of the patient with Gorlin-Goltz Syndrome who took mass excision on the right upper eyelid is presented.

Key Words: Focal dermal hypoplasia, basal cell nevus syndrome , anesthesia, larynx, papilloma, airway obstruction

ÖZET Gorlin-Goltz sendromu cilt, kemik, göz ve dişin yapısını etkileyen kalıtsal bir hastalıktır. İskelet ve santral sinir sistemi anomalileri, mental retardasyon, endokrin anomaliler ve internal maligniteler Gorlin-Goltz sendromunda bulunabilmektedir. Mandibular prognatizm, frontal ve parietal şişkinlik, yüksek arklı damak, dudak ve damak yarığı, supraorbital sırtların belirginliği, kaburgalarda çatallanma ile vertebra anomalileri bu sendromdaki gözlenebilen iskeletsel anomalilerdir. Gorlin-Goltz sendromunun belirgin özelliği cilt ve mukoz membranlarda büyük papillomlar oluşturmasıdır. Bu sendrom pek çok değişikliklerle karakterize olup bunlardan bazıları anestezi uygulamasında özellikle entübasyona engel olmasıdır. Bu olgu sunumunda sağ üst göz kapağında kitle eksizyonu yapılan Gorlin-Goltz sendromlu hastada anestezi indüksiyonu sonrası zor havayolu yönetimini sunuldu.

Anahtar Kelimeler: Fokal dermal hipoplazi, bazal hücreli nevus sendromu, anestezi, larinks, papillom, hava yolu obstrüksiyonu

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Gorlin-Goltz syndrome (focal dermal hypoplasia, Gorlin syndrome) is an inherited group of multiple defects involving the skin, nervous system, eyes, endocrine system, and bones.[1] First described in 1960, Gorlin-Goltz syndrome is an autosomal dominant condition that can cause unusual facial appearances and a predisposition for skin cancer.[2] Histologically, the skin lesions are characterized by a marked decrease in dermal connective tissue.[3] This syndrome is characterized by many

alterations, some of which can interfere with a normal conduct of anesthesia, especially at intubation (prognathism, kyphoscoliosis, incomplete segmentation of cervical-thoracic vertebrae).[4,5] In the present study, difficult airway management of a patient with Gorlin-Goltz Syndrome after induction of anesthesia is presented.

CASE REPORT

A 28 year-old man with Gorlin-Goltz Syndrome underwent a mass excision procedure in his right upper eyelid under general anesthesia. Gorlin-Goltz Syndrome diagnosed when he was 6 years-old and endocrine treatment administered. He was short in stature with the height of 158 cm. In preoperative evaluation, the patient was in good condition and active, but all his body was covered with hypoplastic, atrophic and hyperpigmentatic skin changes. Skin desquamation, teleangiectasias, papillomas in perioral and oral cavity were detected (Figure 1). Hypoplastic nails with longitudinal splitting and circumscribed atrophic alopecia were noted. Visual impairment of 15 % was diagnosed in the left eye. Many verrucous lesions in the hypopharynx and the tonsils were determined in otolaryngologic examination. No respiratory, gastrointestinal system and renal system abnormalities were detected. Serum electrolytes, creatinine, alanine aminotransferase (ALT), aspartate aminotransferase (AST), haemoglobin, platelet and differential blood count were normal. Informed consent was obtained from the patient. The patient fasted for 6 hours prior to surgery and was pre-medicated with atropine (0.5 mg) intramuscularly 30 minutes before anaesthesia. Atropine was especially used to dry secretions in premedication. The patient was taken to the operating room. Peripheral oxygen saturation (SpO_2), electrocardiogram (leads II, V1), cutaneous temperature (T), noninvasive blood pressure (NIBP) of the patient was monitored. Difficult airway management without fiberoptic bronchoscopy was planned as no fiberoptic broncoscope was available. We planned oro-tracheal intubation or fast-track laryngeal mask airway placement as an alternative to fiberoptic bronchoscopy for difficult airway. The equipment was ready for immediate tracheotomy. Anesthesia was induced by using 1 μg kg^{-1} remifentanil, 2 mg kg^{-1} propofol and 1mg kg^{-1} succinylcholine intravenously. During the assisted ventilation by mask, persistent partial upper airway obstruction was occured. Direct laryngoscopy showed variously sized verrucous lesions in the hypopharynx and supraglottic larynx, and the vocal cords and trachea could not be visualized. Therefore, gently blind nasal entubation was performed successfully. Meanwhile, peripheral oxygen saturation was not reduced below 90%. After nasotracheal entubation, the patient was ventilated with air/O_2 and anaesthesia was maintained with sevoflurane, fentanyl and vecuronium to obtain stable haemodynamic parameters. The duration of the surgery was 90 minutes. After recovery of muscle tone, spontaneous breathing was adequate and trachea was extubated. No respiratory or hemodynamic problems were occurred. The patient was admitted to recovery room with stable vital signs. Thirty minutes later he was sent to service with stable vital signs.

DISCUSSION

Gorlin-Goltz syndrome is a disease of mesoectodermal development. Specifically, patients exhibit pigmentary and atrophic skin changes, nail dystrophy, thin hair, syndactyly, bone hypoplasia, and small teeth with dental hypoplasia.[6,7] Cutaneous manifestations included hypoplastic, hyper-pigmentatic and atrophic skin changes, teleangiectasias, skin desquamation, papillomas of the oral and perioral cavity (Figure 1).

There are papillomas on lip, buccal mucosa, and gingivae. Laryngeal papillomas can obstruct the glottic opening. Airway obstruction is so severe in some patients that a tracheostomy is required. On the basis of the present experience, it could be said that patients with Gorlin-Goltz syndrome should have a thorough preoperative otolaryngologic examination. Teeth should be examined preoperatively for number and stability in patients. Gastroesophageal reflux is common therefore precautions should be taken to minimize the risk of aspiration.[6,8,9]

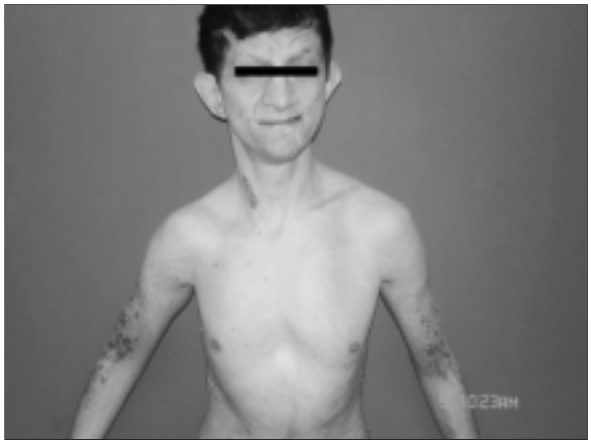

FIGURE 1: Clinical presentation of the cutaneous abnormalities in the patient with Gorlin-Goltz syndrome. The skin lesions are characterized by atrophy, telangiectasia, hyperpigmentation, and papillomas of the perioral.

Gorlin-Goltz syndrome is characterized by many alterations, some of which can cause difficulty during a normal conduct of anesthesia, especially at insertion of the tracheal tube due to laryngeal and subglottic papillomas. Postoperative withdrawal of the laryngeal tube may be difficult due to strongly resistive papillomas. Therefore severe obstruction of the larynx is anticipated, and extubation may not seem feasible. Endoscopy reveals obstruction due to papillomas of the larynx above the vocal cords and on both sides of the hypopharynx down to the sinus piriformis.[9]

Although periorificial papillomas are common, papillomas of the laryngeal mucous membranes have rarely been reported. Tracheotomy may be needed in some of patients with laryngeal papillomas.[6,9,10] The airway must be handled gently, as mucosal papillomas are highly friable. Multiple attempts of laryngoscopies should be avoided.[8]

The endoscopic resection of the papillomas by laser-surgery is a well established procedure and is considered to be the method of choice. The papillomas could be removed successfuly by endoscopic CO_2-laser surgery and electroresection therefore extubation became possible without tracheotomy.[9] Gordjani et al presented a case of 14 years with papillom-induced obstructive symptoms in the hypopharynx and the larynx. Papilloma-masses of a diameter of up to 5 cm were excised, thus tracheotomy was avoided. After surgical intervention the dysphagia, dyspnoe, cough and hoarseness disappeared promptly. The patient fully recovered within four months.[9]

Rhee et al[8] reported that patient developed airway obstruction during mask ventilation in anesthesia. They partially visualized the larynx and intubated the trachea. In order to prevent postoperative airway obstruction, the larger papillomas were excised. Holzman et al[11] presented a patient with persistent partial upper airway obstruction, during the administration of inhalation anesthesia by mask. They showed variously sized verrucous lesions in the hypopharynx and supraglottic larynx at direct laringoscopy, however the vocal cords and trachea could not be visualized. These verrucous lesions were excised by cup forcep and laser. Prior reports stated that the vocal cords are not affected by laser-surgery.[9,11]

We succesfully and gently performed blind nasal entubation. No information is available in literature about application of laryngeal mask airway and gently blind nasal entubation in Gorlin-Goltz syndrome. As no respiratory disorder such as dispne, obstructive symptoms were detected prior to, it is not necessary to remove papillomas via electroresection. No severe obstruction was anticipated postoperatively, and extubation seemed feasible so that extubation became possible without tracheotomy. The equipment was ready for immediate tracheotomy.

CONCLUSION

The present case report suggests airway involvement and anesthetic management in Gorlin-Goltz syndrome. We succesfully and gently performed blind nasal entubation in the first attempt, and it was not necessary to remove papillomas via electroresection. We suggest that patients with Gorlin-Goltz syndrome should have a thorough preoperative otolaryngologic examination. The airway must be handled gently, as mucosal papillomas are highly friable. Multiple attempts of laryngoscopies should be avoided. Fiberoptic

bronchoscopy can be an alternative, as is tracheotomy in the event of significant compromise. A good monitorization is necessary and all precautions should be taken including tracheotomy in case of difficult airway in Gorlin-Goltz syndrome.

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