Case Report

Perioperative Anaesthetics Management of a Girl with Byars-Jurkiewicz Syndrome for Mammoplasty

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Abstract

Byars-Jurkiewicz syndrome is a cutaneous- facial -genital disorder characterized by sporadic congenital macrogengivae hypertrichosis, massive bilateral enlargement of breast after puberty and Kyposis. It is an extremely rare condition as only four to five patients diagnosed throughout the world till now. Miss Bithi, aged 12 years was admitted in plastic surgery in BSMMU with hypertrichosis on the face from birth, huge gingival fibromatosis and bilateral excessive rapid enlargement of breast since last year which is almost half of her 38 kg body weight. She was scheduled for mammoplasty. No abnormalities detected in all routine tests. Small mouth opening with hypertrophied gum obscured visibility of the posterior pharyngeal wall, thyromental distance and cervical spine mobility was found normal. General anesthesia was planned with anticipation of profuse blood loss during procedure and ICU bed was booked for prolonged surgery, .Difficult airway trolley was kept ready, two wide bore canula were inserted and all routine monitoring were applied before induction. Laryngoscopic view was equivalent to Cormac Lehan grade 2. Surgery was completed uneventfully except huge bleeding and prolonged duration (8 hours). She was transferred to ICU, mechanically ventilated overnight and extubated in the next morning and sent the patient to the ward.

Key words; *Byars-Jurkiewicz-syndrome, gingival fibromatosis anaesthetics management, mammoplasty*

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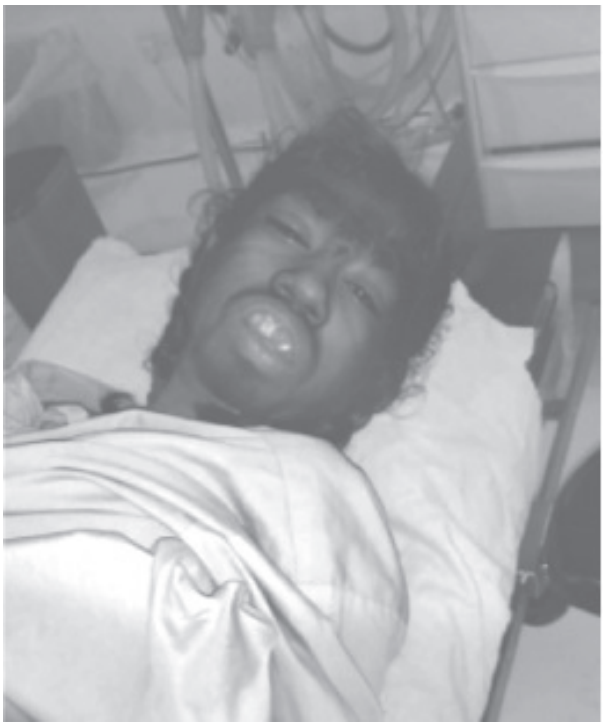
Introduction:

Byars-Jurkiewicz syndrome is a cutaneous - facio-genital disorder characterized by sporadic congenital macrogengivae, hypertrichosis, and massive bilateral enlargement of breast and kyphosis after puberty[1]. Total 50 persons were affected since middle age. It is primarily dermatological disease, manifested as long lanugo hair covered entire body except palms of the hands and sole of the feet, hence it is also known as Werewolf syndrome[2]. It is a heterogeneous disease and exact cause and mechanism of development of this disorder is still unknown.

Case report:

A 12 years old girl was admitted in plastic surgery unit of BSMMU presented with the complaints of huge bilateral enlargement of breasts for one year which was rapidly increased in size, generalized hypertrophy of gum that buried her teeth for 5 years and whole body covered by long hair. She yet to attained menarche and no history of galactorrhoea. Her school performance and intellectuality was normal. Her mothers antenatal and postnatal period was uneventful, she was delivered vaginally at home. There was no history of parental consanguineous marriage and no use of drug during pregnancy. No family member affected such illness.

On general examination: Body mass index was 19.47kg/m^2, generalized gingival overgrowth which is firm, pink and painless. Her breast were massively enlarged which were extended up to inguinal region, Small ulceration were seen on the undersurface of both breasts. dark terminal hair covered her body involving face, neck, both extremities and entire back. Her Pulserate-80/min,



blood pressure- 110/70mmof Hg.There was no abnormalities in cardio- respiratory system . Hematological and biochemical hormonal profiles were normal. Normal X-ray chest, ECG and whole abdomen of ultrasonograpy were reported. Echocardiography revealed left ventricular ejection fraction (LVEF) 60%.

The patient was scheduled for bilateral reduction mammoplasty of breast as ASA grade II. Difficult intubation was predicted due to small mouth opening, gum hypertrophy and huge of enlargement of breast. Surgery was planned with an arrangement of ICU bed because of anticipated intraoperative complication and long duration of surgery. The proposed procedure, its outcome and risks were explained with her guardian. Oral anxiolytic and antiulcerant was given before surgery.

In operation theatre, her heart rate 84/min, blood pressure 110/70 mm of Hg, and SpO2- 97% were recorded. Two 20G cannula were inserted in both hand. Preoxygenation was done with 100% O2 for 5 minutes. Anaesthesia was induced by inj. fentanyl-50 microgram, propofol 80mg; she was intubated with 6.0 mmID endotracheal tube and facilitated by suxamethonium 80mg. Anaesthesia was maintained with 0.5% halothane in 30% oxygen and 70% nitrous oxide. Vecuronium bromide was used as muscle relaxant.

Fluid balance was maintained by isotonic crystalloid solution. First two hour of the procedure, her hemodynamic status was stable then she developed mild hypotension; one unit of fresh blood transfused. After removal of the huge mass(breast mass) again patient developed severe hypotension. Another 3 units of fresh blood were transfused and inj vasopressor also given to maintain haemodynamically stable .Duration of total surgery was six hours. After completion of the surgery she was transferred to ICU without extubation .She was mechanically ventilated and closely observed overnight. Extubation was done next day morning after fulfilling of all criteria of extubation and sent to the ward.

Discussion:

Byars-Jurkiewicz syndrome is an extremely rare condition as only four to five patients diagnosed globally. For the first time this case is diagnosed in Bangladesh. The patient suffering from this conditions manifested as gingival fibromatosis, hypertrichosis, giant fibroadenoma of breast and kyphosis.1,2. Sometimes it's also called "Werewolf syndrome" for their external appearance. The patient presented with huge enlargement of breast, generalized hypertrophy of upper gum, excessive growth of hair in the whole body since birth3, 4. All these features are challenging to the attending anesthetists. In this case, all laboratory and radiological investigations found normal. Surgical intervention was done due to overweight of hugely enlarged breasts which was extended to the inguinal region bilaterally. The key factor is to remember when preparing patient for surgery is to get medically stabilized .Airway assessment revealed that difficult intubation may occur due to small mouth opening with gum hypertrophy and huge enlargement of breast. Difficult airway trolley and all resuscitation equipment were kept ready. Intubation was done smoothly. During preoperative assessment it was anticipated that huge bleeding, prolonged surgery and development of unknown life threatening consequences due to paucity of this rare pathologic state. Therefore, four units of fresh blood and booking of an ICU bed were advised. It was assumed that abnormally growing breast might have high vascularity. During intraoperative period ,four units fresh blood was transfused due to profuse bleeding and severe hypotension. The patient was kept in ICU for 24

hours and then extubated and transferred to the general ward. A case of 12 years old female child presented with generalized severe gingival over growth and had full set of teeth remaining invisible within the confinement of gingival tissue. Conventional gingivectomy was done under general anesthesia. The post operative result was uneventful and the patient appearance improved considerably.

Conclusion

Byar- jurkiewicz syndrome is an uncommon disease not only in our country but also globally. although four to five case diagnosed in the world but the pathophysiology yet not known. Early involvement of anaesthesiologists with skilled anesthetist, optimum monitoring and continuous vigilance make the surgery uneventful. Improved technique of anaesthesia and skilled surgery was played a key role in successful management of mammoplasty in this young girl with rare Byar-jurkiewicz syndrome.

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