

An Anaesthetic Experience With A Rare Case Of Cutis Laxa Syndrome

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Abstract

Cutis Laxa is a rare, inherited or acquired connective tissue disorder in which the skin becomes inelastic and hangs loosely in folds like loose dress. These patients usually come to the Pediatric, Orthopedic or Plastic surgeons for the treatment of this disorder or its associated problems. An anaesthetic experience is reported here with such a case that came to the hospital for the correction of deformity of foot.

INTRODUCTION

Cutis Laxa comprises a heterogeneous group of congenital and acquired conditions characterized by a distinctive lax and inelastic skin. The clinical features of Cutis Laxa are thought to be due to a defective elastin synthesis that results in sparse and abnormal elastic fibers in the affected tissues [1].

CASE REPORT

An eleven (11) years old male boy weighing 40 kilograms was presented to our hospital for the correction of the deformity of foot under general anaesthesia. On the pre-operative anaesthesia visit, he was well oriented in time, space and person. He has senile looking face (as shown in figure 1), loose and thin skin.

Figure 1

Figure 1: The face reveals an antimongoloid slant, slightly averted nostrils, long upper lips, long philtrium and short columella, prominent low set ears and prominent epicanthic folds (covered)



There was no cyanosis, jaundice or edema and he was afebrile. Pulse was 100/minute, regular with good volume and Blood Pressure was 100/50 mm Hg. Examination of chest revealed normal shaped chest. Trachea was central and apex beat on its normal position. Respiratory rate was

20/minute and regular. Auscultation of chest showed vesicular breathing with equal intensity and vocal resonance on both sides. There were no added sounds. First and 2nd heart sounds were normal in intensity but here was a pansystolic murmur 2-3/6 heard all over the precordium.

Airway examination showed Mallampati Class I, adequate anterior mandibular space and good range of movements at atlanto-occipital and cervical joints.

Complete blood examination showed WBC $8.2 \times 10^9 /L$, RBC $4.3 \times 10^{12} /L$ and Platelets $218 \times 10^9 /L$. Haemoglobin level was 11.7 g/dl with Haemocrit of 34.5%. Prothrombin time was 13.9 seconds and APTT 31.7 seconds. Blood sugar, liver and renal function tests were all within normal limits.

X-ray Chest PA and Lateral view revealed mild hyperinflation of both lung fields and normal shaped heart shadow as shown in figure 2.

Figure 2

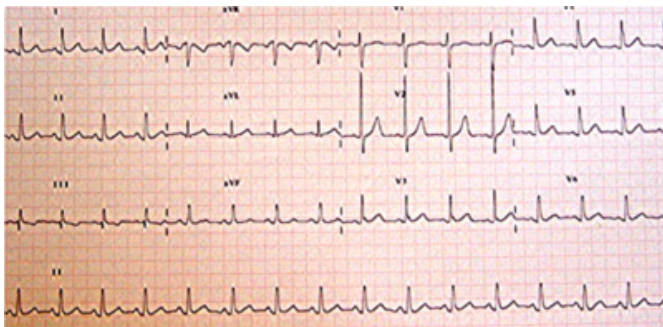
Figure 2: X-rays chest PA and Lateral view showing mild hyperinflation of both the lung fields. Cardiac shadow seems to be normal.



A standard 12 leads ECG showed Right Ventricular Hypertrophy by voltage criteria as shown in figure 3.

Figure 3

Figure 3: A standard 12 leads ECG showing sinus rhythm with rate of 94/minute. There is Right Ventricular Hypertrophy (RVH) by the voltage criteria.



On the basis of clinically detected pansystolic murmur an Echocardiography revealed:

- No Atrial Septal Defect (ASD), Ventricular Septal Defect (VSD) or Patent Ductus Arteriosus (PDA)
- Good ventricular function (Ejection fraction 65%).
- Mild to moderate mitral regurgitation with a pressure gradient of 70 mm Hg.
- Mild mitral valve prolapse.
- Mild tricuspid regurgitation with a pressure gradient of 18 mm Hg.
- Segmental hypertrophy of interventricular septum.

Currently he was taking Propranolol 5 mg orally twice daily.

FAMILY HISTORY

The patient is the third child of first cousin, consanguineous healthy parents. His elder sister has same skin disorder but she has Congenital Dysplasia of Hip joint (CDH) as well. The first male brother is normal.

PAST MEDICAL/SURGICAL HISTORY

This child is a product of full term normal vaginal delivery. Pregnancy was uneventful. His birth weight was 3.2 kilograms. In addition to laxity of skin, he had bilateral Congenital Talipes Equinovarus (CTEV), bilateral inguinal hernias and undescendent testes. On the basis of clinical examination and skin biopsy just after his birth he, was labeled as a case of Congenital Cutis Laxa autosomal recessive (AR) type.

During his early infancy, he had history of hospitalization for epistaxis, acute bronchopneumonia and pancytopenia. During his stay in hospital Ultrasonography (USG) was done which revealed:

ABDOMINAL USG

- Normal liver, pancreas, gall bladder, both kidneys and spleen.
- Normal common bile duct and portal vein.
- Partially filled urinary bladder.

CRANIAL USG

- Mild to moderate dilation of the lateral ventricles,

III ventricle and IV ventricle.

- Bilateral inter-hemispheric ratio – 0.56
- Transverse diameter of III ventricle – 1.2 cm
- Transverse diameter of IV ventricle – 3.2 cm

Conclusion: mild to moderate Hydrocephalus

But the patient did not require any intervention for this Hydrocephalus. During these days, he was diagnosed as a case of mitral regurgitation as well by Echocardiography in another hospital.

At the age of 5 ½ months he had surgery (Elongation of Achilles Tendon and posterior capsulotomy) under general anaesthesia for right foot.

One week later, he had similar operation under general anaesthesia for the left foot.

At the age of 2 ½ years he had surgery for bilateral hernias, undescended testes and circumcision with Plasti-bell under general anaesthesia.

He had normal developmental milestones. Now he started pain on walking and came to the hospital for its cure.

ANAESTHETIC MANAGEMENT

On the preoperative anaesthesia visit, the patient was advised to take tablet Propranolol 5 mg orally with sips of water at 6.00 a.m. on the day of surgery and he followed. Moreover, he also received 2 grams of injection Ampicillin intravenously after skin test before induction of anaesthesia for the prophylaxis of endocarditis. He came to the operating room with a 22 G intravenous canula with running fluid. Pulse Oximeter, ECG, Nerve stimulator and Non-invasive BP monitors were attached and induction started with preoxygenation with 100% O₂, injection Fentanyl 80 ug and Propofol 80 mg. Endotracheal intubation was facilitated with injection Rocuronium 40 mg. After correct placement of the endotracheal tube with EtCO₂ monitor, it was fixed and mechanical ventilation started. Pressure points of the body were properly padded and protected. Right radial artery was cannulated for Invasive BP monitoring and another 20 G intravenous cannula was passed on the left dorsum of hand. A nasopharyngeal probe was inserted for Temperature monitoring. The patient also received injection Cephazoline 1 gram intravenously before the inflation of tourniquet. Anaesthesia was maintained with N₂O, O₂ and Sevoflurane.

Lactated Ringer's solution continued at the rate of 80 ml/hour with infusion pump. Incremental doses of Fentanyl 20 ug and Rocuronium 10 mg were also given. The procedure (Elongation of Achilles tendon, elongation of tendon tibialis posterior and calcaneal-cuboid fusion) took about 90 minutes. Injection Atropine 0.8 mg and Neostigmine 2.0 mg were given to antagonize the residual effect of Rocuronium. Trachea was extubated in lateral position and then he was sent to the recovery room where he received injection Pethidine 40 mg intramuscularly for post-operative pain. During the procedure he remained haemodynamically very stable. Intra-arterial cannula was removed in recovery room and the patient was sent to ward. The intra and postoperative course was uneventful.

DISCUSSION

Cutis Laxa (Latin for loose or lax skin) Syndrome or simply Cutis Laxa (also known as Elastolysis, Dermatomegaly, Cutis Pedula, etc) is a rare, inherited [2] or acquired [3] connective tissue disorder in which the skin becomes inelastic, loose and hangs in folds. Presentation of this disorder and mode of inheritance shows considerable heterogeneity.

Primary Cutis Laxa is present since birth (congenital) is hereditary. Secondary Cutis Laxa arises later in the life and may be either hereditary or acquired. Acquired Cutis Laxa also arises later in life and is not hereditary [4]. In both the hereditary or acquired types, the internal organs are frequently involved [5,9]. Hereditary disorders with Cutis Laxa are presently divided into five types: an autosomal dominant type, an X-linked recessive type and autosomal recessive type that is further divided into three sub-types [6]. Differential diagnosis of Cutis Laxa includes Costello Syndrome and Ehlers-Donlas Syndrome type III C [7]. A few dozen cases of Cutis Laxa have been documented in the literature [8]. How these patients react to our anaesthetic drugs is even extremely rare in the literature related to anaesthesia. Karapurkar SA, Borkar JD and Bermole BJ [9] reported a baby out of five who underwent surgery (lobectomy) for congenital lobar emphysema and lung cyst under general anaesthesia. This baby had severe wheeze and intercostal in drawing after extubation. Immediately trachea was reintubated. The baby was sedated and ventilated with injection Pethidine and Pancronium but the wheeze continued and Pediatrician's opinion was sought. He started Hydrocortisone and Aminophylline but there was no response. There was no response to terbutaline as well. After 48 hours, endotracheal tube was blocked and removed. The

Neonatologist who was consulted, noted other anomalies namely hypertelorism, short columella, hook nose, large philtrium, large upper lip with loose skin on dorsum of hands and back of the neck. The diagnosis was "Cutis Laxa" with systemic involvement. There are also case reports of association of Cutis Laxa with pulmonary emphysema [3].

If we see our patient, he was exposed to anaesthetic drugs three times in the past.

First time, he was induced and intubated with Isoflorane in Oxygen and then maintained with N₂O, O₂ and Isoflorane. Second time, he was induced and intubated with Halothane in Oxygen and then maintained with N₂O, O₂ and Isoflorane. Third time, he was induced N₂O, O₂ and Halothane and then maintained with N₂O, O₂ and Isoflorane with the use of LMA. He had also caudal block with Bupivacaine. All the three times no muscle relaxant was used and breathing was spontaneous.

Presently we used intravenous induction agent, muscle relaxant and controlled mechanical ventilation. In all the four events mentioned above, the patient tolerated anaesthetic agents very well. The perioperative course all the time was uneventful. Thus, this normal response of these patients to anaesthetic drugs might be the one of the reasons that our anaesthesia literature is very scanty regarding Cutis Laxa Syndrome.

CONCLUSION

Cutis Laxa Syndrome is an extremely rare disorder in the practice of Anaesthesiology. if someone comes across such a case, he/she should look for other anomalies and deal with

them accordingly.

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