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Case Report

Anaesthesia Section

Anaesthetic Management of Paediatric Patient with Klippel-Trenaunay Syndrome undergoing Excision of Scalp Cyst: A Case Report

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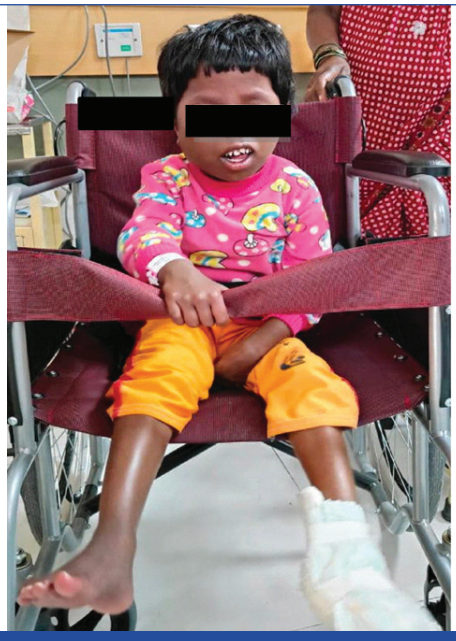
ABSTRACT

Klippel-Trenaunay Syndrome (KTS) is a sporadic condition characterised by the clinical triad of capillary malformation, atypical varicosity, and soft-tissue and bony hypertrophy. This triad was observed in the current case. Despite being initially described over a century ago, the exact incidence of this syndrome remains unestimated. The clinical presentation of KTS is highly diverse, ranging from asymptomatic cases to severe instances involving life-threatening bleeding and embolism. The authors present a case report of a 10-year-old girl with complaints of a cystic swelling in her left temporal region. She exhibited distinct features such as facial asymmetry, a high-arched palate, a prominent mandible, a port-wine stain on her cheeks, nasal bridge depression, and right-sided body hypertrophy. The child showed delayed developmental milestones, including holding her neck at 1.5 months, a bidextrous approach at seven months, and social smiling at six months, indicating global developmental delay. The patient was scheduled for excision of the swelling. Following the successful excision of the cyst, the child was extubated. The extubation was uneventful. In the postoperative period, the patient remained haemodynamically stable. Managing such a patient for a surgical procedure requires a meticulous approach involving accurate anaesthetic preparation and the prevention of complications.

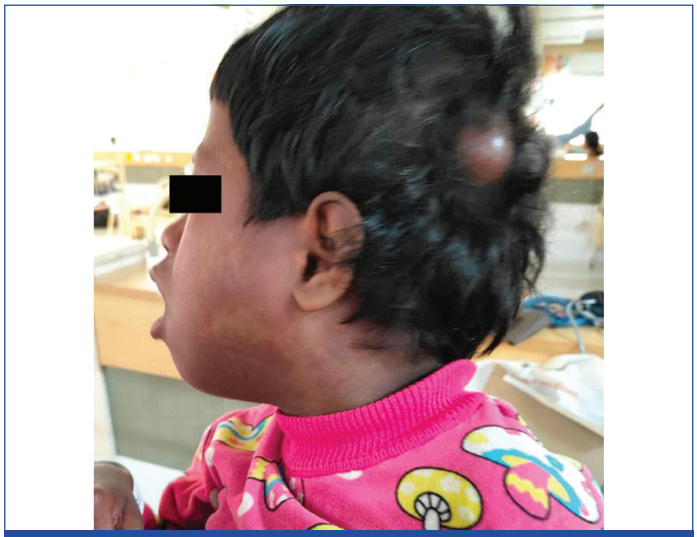
Keywords: Congenital syndromes, Haemangioma, Hemihypertrophy

CASE REPORT

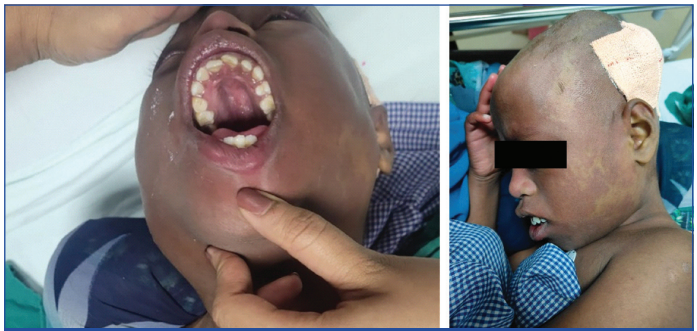
A 10-year-old girl [Table/Fig-1], weighing 20 kg, was brought by her parents with complaints of a cystic swelling in her left temporal region [Table/Fig-2]. The swelling had been growing for two months, was discharging pus, and she also experienced eyelid oedema and pain. Her medical history revealed a challenging start with Neonatal Intensive Care Unit (NICU) admission due to respiratory distress. For the past year, she had been experiencing recurrent seizures and was started on T. Sodium Valproate at 10 mg/kg, leading to developmental delays. She displayed distinct features like facial asymmetry, a high-arched palate [Table/Fig-3], a prominent mandible, a port-wine stain on her cheeks [Table/Fig-4], nasal bridge depression, and right-sided body hypertrophy. Her limbs showed reduced power and brisk reflexes, along with a noticeable gait difference. The child exhibited delayed developmental milestones, such as neck holding at 1.5 months, a bimanual approach at seven months, and social smiling at six months, indicating global developmental delay.



[Table/Fig-1]: Child with Klippel-Trenaunay Syndrome (KTS).



[Table/Fig-2]: Swelling on the occipital aspect of head.



[Table/Fig-3]: Patient showing high arched palate.
[Table/Fig-4]: Port-wine stain on cheeks. (Images from left to right)

An emergency incision and drainage were planned due to the swelling, but financial constraints and the child's irritable condition

prevented an Magnetic Resonance Imaging (MRI) from being conducted. However, based on the comprehensive clinical history and observed specific features, the paediatrician diagnosed Klippel-Trenaunay Syndrome (KTS).

The patient was scheduled for the excision of the swelling. Standard preoperative assessments, including a complete blood count, serum electrolytes, Prothrombin Time (PT)/International Normalised Ratio (INR), chest X-ray, and Electrocardiogram (ECG), were conducted and were within normal limits. On the morning of the surgery, the patient's nil by mouth status was confirmed, intravenous access was established, and informed consent was obtained from the parents for the high-risk procedure due to anaesthetic challenges. The Paediatric Intensive Care Unit (PICU) was reserved.

In the operating room, all necessary equipment, including the emergency drug trolley and difficult airway trolley, was prepared. Basic monitoring devices were connected, and due to the child's restlessness, Inj. Midazolam 0.4 mg i.v. was administered. Sodium valproate was readily available due to the history of seizures. Prior to anaesthesia induction, preoxygenation with 100% oxygen was conducted for three minutes. Premedication with Inj. Glycopyrrolate 0.08 mg i.v. and Inj. Fentanyl (1 mcg/kg) was given. Inj. Propofol 3 mg/kg was used for induction, followed by laryngoscopy with a Mcintosh blade no. 3. Considering the high-arched palate and anticipated difficult intubation, bougie-guided intubation was performed with a 5 mm cuffed Endotracheal Tube (ETT). Hydrocortisone 40 mg i.v. and Inj. Dexamethasone 2 mg i.v. were administered. Inj. Atracurium 0.5 mg/kg was used as a muscle relaxant. The intubation procedure proceeded smoothly without any complications.

There was significant blood loss due to abnormal capillary formation of the cyst, which was managed with intravenous fluids in the form of Ringer's lactate. Inj. Tranexamic acid (20 mg/kg) was also administered. Following the successful excision of the cyst, the child was extubated using Inj. Neostigmine (0.8 mg) and Inj. Glycopyrrolate (0.128 mg). The extubation process was uneventful.

During the postoperative period, the patient remained haemodynamically stable. The patient was monitored for two hours until full consciousness was regained, and the nil by mouth status was continued for an additional two hours.

DISCUSSION

Klippel-Trenaunay Syndrome (KTS), also known as angio-osteohypertrophy syndrome or hemangiectatic hypertrophy, is a rare congenital syndrome that presents at birth or during childhood with enlarged veins and arteries, limb hypertrophies, and capillary malformations [1]. The incidence of KTS is 2-4 per 100,000 live births, affecting both genders equally, with a slightly higher prevalence in males [2].

The exact cause of KTS is unknown. While many patients show a normal karyotype, sporadic translocations involving chromosomes 5 to 11 and 8 to 14, as well as ringed chromosome 18, have been observed [3]. KTS is classified into venous, arterial, and related venous dysplasia based on blood vessel dysplasia. The hypertrophy in the affected region results from excessive expansion of the soft tissue underneath it and adipose tissue hyperplasia caused by bone hypertrophy, leading to variation in limb length. A flat capillary haemangioma with a red or purple colour is often the initial sign of capillary abnormalities. Venous systems are affected by varicose veins that affect every patient. Deep vein anomalies can manifest as aneurysmal dilatation, agenesis, hypoplasia, and venous regurgitation [4].

Various organs, including the central nervous system, genitourinary tract, and gastrointestinal tract, can be involved. Rectal and bladder bleeding is a severe complication, with a reported occurrence of 1%. Additional clinical symptoms include:

1. **Digestive tract:** Splenic hemangiomas, oesophageal variceal haemorrhage [5].
2. **Genitourinary tracts:** Gross and recurrent haematuria [1].
3. **Skeletal symptoms:** Differences in leg length, ipsilateral hip and scoliosis dislocations, polydactyly, blue nevi, pulmonary vein varicosities, aneurysms, and pulmonary embolism [6].

The primary method of diagnosing KTS is clinical, as there is no definitive test. Non invasive imaging should be employed in the initial assessment of KTS patients to determine the extent of the condition and the distribution of both structural and functional venous abnormalities.

Usually, a Doppler ultrasonography of the extremities is performed to evaluate the deep vein system [7]. In the present case, the main concern was primarily focused on securing vascular access to avoid injury to superficial arteriovenous fistulous malformations. Intubation was challenging due to the hypertrophy on the right-side of the head, the presence of a high-arched palate, and soft tissue hypertrophy in the mouth. Excessive blood loss from swelling due to capillary malformation was managed effectively, considering the high-risk of venous thrombosis and pulmonary thromboembolism. Despite these challenges, the case was managed thoroughly and without any complications.

The optimal approach to managing KTS involves a multidisciplinary team, emphasising care coordination. Symptomatic care through medical management is essential, with surgical intervention reserved for refractory cases. Skincare is crucial in preventing superficial infections and bleeding resulting from scratching. In children, vigilant monitoring for limb length disparities is crucial, with referrals to orthopaedics for orthotics or surgical corrections when necessary. Interventions like compression stockings, limb elevation, and intermittent pneumatic compression devices are employed to mitigate lymphoedema and venous insufficiency. Sclerotherapy, including conventional and micro-foam techniques, is used to address capillary, venous, and lymphatic malformations. Laser treatment is considered for managing port-wine stains [8].

Surgical options, such as endovascular ligation of embryonic veins and stripping of severe varicose veins, are reserved for cases unresponsive to medical therapy. Rapamycin has emerged as a promising therapeutic option for KTS, capable of halting vascular malformation progression and improving patients' quality of life. Its mechanism of action involves inhibiting the PI3K/AKT/mTOR pathway, thereby arresting cell growth and preventing tissue overgrowth. Close monitoring for adverse effects, including haematological and lipid abnormalities, is crucial for patients undergoing rapamycin therapy [9].

CONCLUSION(S)

Anaesthetic management is crucial for KTS, a rare complex condition. Anaesthetists should always be vigilant about intraoperative blood loss during surgical procedures to avoid unnecessary complications, which are common due to capillary malformations. Imaging modalities such as MRI and angiography are critical for diagnosing, treating, and promoting the child's recovery. Good and prompt management followed by postoperative care can lead to a positive outcome. By following the available treatment modalities, the life expectancy of a KTS patient can be improved.

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