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RESEARCH ARTICLE

SURGICAL RESECTION OF MULTIPLE ANGIOMATOUS LESIONS OF THE LIMBS IN AN 11-YEAR-OLD GIRL WITH BEALS SYNDROME: A CASE REPORT

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ABSTRACT

Background: Beals syndrome, or congenital contractural arachnodactyly (CCA), is a rare autosomal dominant connective tissue disorder caused by pathogenic variants of FBN2. The association with diffuse vascular malformations is exceptional. Airway management during anesthesia is challenging due to joint contractures and restricted cervical mobility. **Case presentation:** We report the case of an 11-year-old girl with genetically confirmed Beals syndrome who underwent resection of multiple angiomatous lesions of the limbs. Preoperative evaluation revealed a marfanoid habitus, arachnodactyly, joint contractures, and limited neck extension. Cardiac function was normal. A difficult airway was anticipated, and mult
iple devices were prepared (videolaryngoscope, flexible bronchoscope, supraglottic airways). **Management and Outcome:** Induction was performed with preservation of spontaneous ventilation. Endotracheal intubation was successfully achieved using a videolaryngoscope. Surgical dissection proceeded cautiously due to tissue fragility, and no major hemorrhage occurred. The patient was extubated safely in the recovery unit, and her postoperative course was uneventful. **Conclusion:** This case emphasizes the importance of structured anesthetic planning for airway management and vigilant perioperative monitoring to prevent complications in children with Beals syndrome.

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INTRODUCTION

Beals syndrome, also known as congenital contractural arachnodactyly (CCA), is an autosomal dominant connective tissue disorder linked to pathogenic variants of the FBN2 gene. The phenotype typically includes arachnodactyly, congenital joint contractures, scoliosis, and occasionally cardiovascular involvement. Association with diffuse vascular malformations is extremely rare. We present the case of a child with Beals syndrome undergoing surgical resection of angiomatous lesions, highlighting anesthetic challenges and perioperative management strategies.

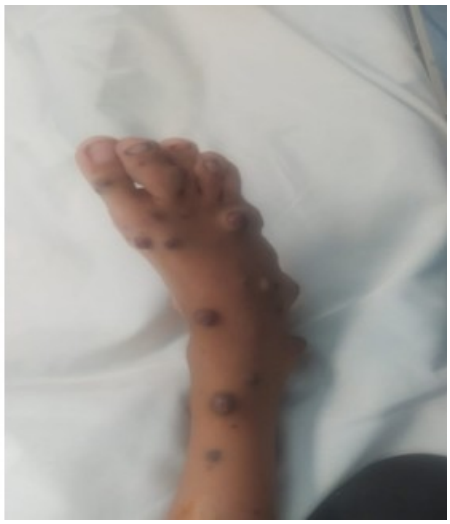
CASE PRESENTATION

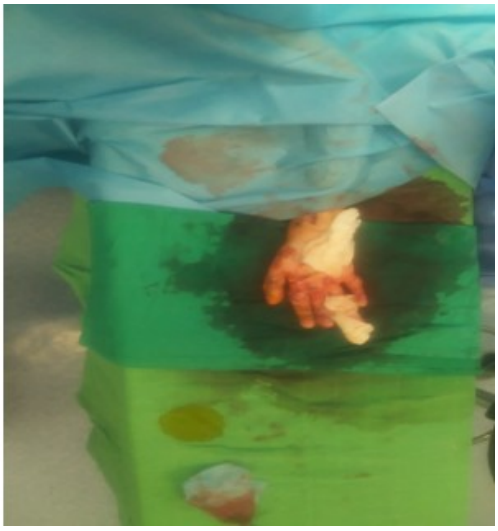
An 11-year-old girl with genetically confirmed CCA was referred for surgical excision of multiple angiomatous lesions of the limbs. Examination revealed arachnodactyly, marfanoid habitus, large-joint contractures, and limited cervical extension. Subcutaneous angiomatous masses were noted in the lower limbs. Preoperative echocardiography was normal. Hematological and coagulation profiles were within normal limits. Given the risk of a difficult airway, anesthetic planning included preparation of a videolaryngoscope, flexible bronchoscope, supraglottic airways, and an intubation bougie. An experienced pediatric anesthesia team was involved, with blood products and a cell salvage device readily available.

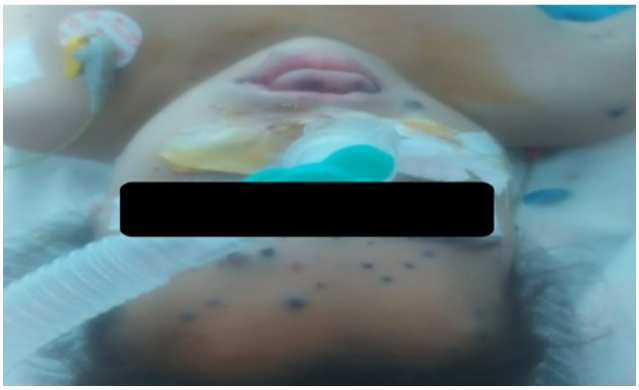
Anesthetic Management: Induction was performed with inhalational anesthesia, preserving spontaneous ventilation. Optimal preoxygenation and invasive hemodynamic monitoring were established. Tracheal intubation was achieved on the first attempt using a videolaryngoscope after an initial cautious maneuver. Surgical dissection was meticulous to avoid tissue injury. Blood loss was limited due to careful hemostasis. Extubation was performed in the post-anesthesia care unit with stable respiratory and hemodynamic parameters.

DISCUSSION

Congenital contractural arachnodactyly is caused by pathogenic FBN2 variants. Severe cases may involve cardiovascular abnormalities, warranting systematic preoperative echocardiographic evaluation (1,2,7). Recent studies suggest that specific FBN2 mutations, particularly within exons 31–35, may be associated with increased cardiovascular risk (7).







Airway management remains the most significant anesthetic challenge in Beals syndrome. Contractures, cervical rigidity, and possible craniofacial anomalies contribute to difficult intubation. Published pediatric cases describe various successful approaches including videolaryngoscopy, fiberoptic intubation, bougie-guided techniques, and blind nasal intubation, depending on expertise and resources (3–6,9). The OrphanAnesthesia guidelines recommend thorough airway assessment, preparation of backup plans, and caution with regional anesthesia in patients with spinal deformities (4).

General pediatric difficult airway guidelines, such as those of the Difficult Airway Society (DAS), stress prioritizing oxygenation at first attempt, stepwise escalation across devices, and structured pauses after failed attempts (8,10,11). In our case, maintaining spontaneous ventilation until airway control, early use of videolaryngoscopy, and immediate access to rescue options contributed to a safe outcome. Although diffuse vascular malformations are not typical of CCA, they increase the risk of intraoperative bleeding. Blood conservation strategies, availability of blood products, and careful monitoring are recommended. Our case confirms that extensive resection is feasible when anesthetic and surgical planning is meticulous.

CONCLUSION

Children with Beals syndrome present unique anesthetic challenges due to musculoskeletal deformities and potential cardiovascular involvement. Anticipatory, multimodal airway management strategies and adherence to pediatric difficult airway algorithms are essential. The coexistence of vascular malformations further necessitates vigilant perioperative hemostatic management.

Informed Consent: Written informed consent was obtained from the patient's family for publication of this case report.

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