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Perioperative Considerations for a Patient with Juvenile Idiopathic Scoliosis and Kaposiform Lymphangiomatosis Undergoing Spinal Fusion: A Case Report

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Learning Point of the Article:

Posterior spinal fusion is an appropriate and routine means of management for patients with juvenile idiopathic scoliosis; however, in patients with kaposiform lymphangiomatosis, such a routine procedure should not be performed without adequate pre-operative and intraoperative planning given the high risk of post-operative complications.

Abstract

Introduction: Kaposiform lymphangiomatosis is a lymphatic anomaly that, when symptomatic, presents in children at a median age of 6.5 years. Symptoms may include respiratory issues, palpable masses, and hemostatic abnormalities. Kaposiform lymphangiomatosis can be life-threatening due to diffuse tissue expansion and subsequent invasion of surrounding organs and tissues. Initial detection of this rare condition can be difficult due to the varied nature of its presenting symptoms and the overall lack of familiarity of clinicians with the condition. Consequently, misdiagnoses can occur, such as pneumonia, cancer, or other vascular anomalies. When considering operative treatment in patients with kaposiform lymphangiomatosis, meticulous pre-operative planning and multidisciplinary care are required due to the high risks of morbidity and death from blood loss. To the best of our knowledge, this is the first reported case of kaposiform lymphangiomatosis in a patient with juvenile idiopathic scoliosis.

Case Report: We present the case of a 13-year-old boy diagnosed with kaposiform lymphangiomatosis at age 8 who, after being followed for several years in our orthopedic clinic for worsening juvenile idiopathic scoliosis despite brace wear, underwent posterior spinal fusion with minimal complications (i.e.., minor cerebrospinal fluid leak). The patient had also been followed for several years before his kaposiform lymphangiomatosis diagnosis for unexplained thrombocytopenia, fatigue, and joint pain. Interdisciplinary care involved multiple specialist teams to choose appropriate pre-operative medications, induction protocol, and bone graft.

Conclusion: Successful operative treatment in a patient with kaposiform lymphangiomatosis can be achieved with the involvement of an interdisciplinary team, anticipation and preparation for cardiac and pulmonary complications via chest tubes and pericardial windows, setting goal parameters to guide intraoperative monitoring, and ceasing medications such as sirolimus to prevent wound-related complications. Given the lack of a current standard of care for managing patients with kaposiform lymphangiomatosis who have spinal deformity, this report can serve as a guide that sets a benchmark for the management of similar cases.

Keywords: Juvenile idiopathic scoliosis, kaposiform lymphangiomatosis, pre-operative planning.

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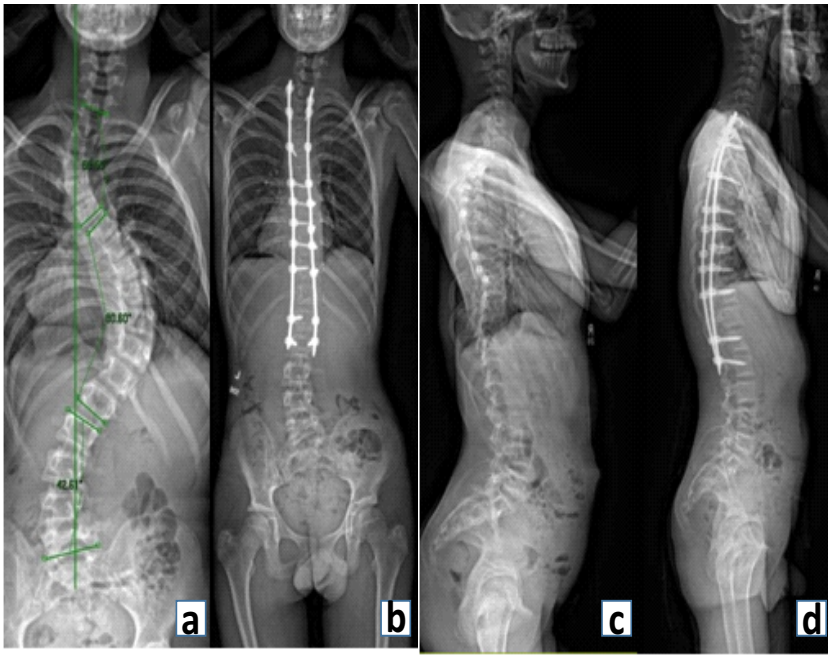

Figure 1: Radiographs of a 6-year-old boy with kaposiform lymphangiomatosis and juvenile idiopathic scoliosis. Anteroposterior images were taken before and after posterior spinal fusion from T2 to L2 and show (a) pre-operative scoliosis and (b) post-operative deformity correction. Lateral images show (c) pre-operative scoliosis and (d) post-operative deformity correction.

Introduction

Kaposiform lymphangiomatosis (KLA), a rare disorder in which lymphatic fluid vessels expand and interconnect throughout the body, is potentially life-threatening due to involvement with surrounding tissues, bones, and organs [1]. Symptoms present at a median age of 6.5 years [1, 2]. The most common symptoms are hemostatic abnormalities, respiratory symptoms, and palpable masses, though fewer than half of patients exhibit all three [3].

The vague symptoms of KLA often delay diagnosis. Essential diagnostic tools include computed tomography and magnetic resonance imaging, which typically show pleural and pericardial effusions, interstitial thickening, mediastinal infiltration, osteolytic lesions, and thorax, abdomen, and skeleton lesions, as well as histopathological analysis, which typically shows dilated lymphatic vessels [4, 5]. Treatments include chemotherapy, thoracic duct embolization, or shunts to divert lymphatic fluid from surrounding organs. In addition, sirolimus can be effective for patients with complicated vascular anomalies [4].

Common comorbidities include pericardial and pleural effusions, as well as consumptive coagulopathies [4]. The leading causes of death are cardiopulmonary, respiratory, or multiorgan system failure; others include tension pneumothorax, bleeding, bloody effusions, and pulmonary hemorrhage [4, 6]. New treatments have achieved a 5-year survival rate of 79% and a mean time from disease onset to death of 4.9 years [6].

Management of KLA is challenging due to vague symptoms, lack of standard multidisciplinary treatment protocol, and clinicians' unfamiliarity with the condition. In addition, consumptive coagulopathies increase the risk of hemostatic abnormalities and hemorrhage during surgery, including scoliosis surgery in pediatric patients [7]. Although posterior spinal fusion (PSF) is a standard surgical intervention for patients with adolescent idiopathic scoliosis, complications occur in approximately 6.3% of patients [8]. In those with underlying lymphatic malformations such as KLA, risks may be higher secondary to complications, including lymphatic fluid fistulas, which are rare but serious concerns in posterior lumbar procedures [9]. The increased risk for these patients is not quantified in the literature, but documented lymphatic fluid complications underscore the importance of a multidisciplinary approach to care. Performing PSF for a patient with KLA requires detailed pre-operative planning to prevent morbidity and death [10].

We present the case of a 13-year-old boy with KLA who underwent successful PSF for juvenile idiopathic scoliosis. The patient and his parents provided informed consent for publication of this case report.

Case Report

A 6-year-old boy with underlying thrombocytopenia presented to our orthopedic clinic for evaluation of low back and hip pain. At his initial visit to our clinic, his weight and height had dropped from the 90th to 40th percentile. On physical examination, he described tenderness over the right paraspinal muscles and sacroiliac joint; no apparent abnormalities in leg length, leg alignment, leg rotation, or pain with range of motion were noted. He walked with an in-toeing gait with abductor lurching bilaterally and no apparent increases in spasticity. Anteroposterior and lateral radiographs of the spine revealed osteopenic vertebral bodies and 23° of right convex scoliosis from T6 to L1, confirmed by dual-energy X-ray absorptiometry.

During the next several months, he underwent testing by multiple specialists for symptoms including fever, rash, bone pain, decreased appetite, fatigue, easy bruising, and scrotal edema. The differential diagnosis included parvovirus, immune thrombocytopenic purpura, unspecified rheumatologic conditions, and aplastic anemia. Initial treatment with steroids

was followed by intravenous immunoglobulin, high-dose steroids, and hydroxychloroquine due to concerns for antiphospholipid antibody syndrome after he sustained a subgaleal hemorrhage from minor head trauma. Discoloration of the scrotum, magnetic resonance imaging of the abdomen and pelvis showing diffuse microcytic lymphangiomatosis, and a biopsy of the malformation in the paraspinal region revealed lymphatic channels consistent with KLA. He began taking sirolimus.

He was followed in our orthopedic clinic annually for scoliosis, which worsened despite brace wear. In patients with severe scoliosis, such as an 80° thoracic curve, non-operative management is unlikely to prevent progression, given the natural history of large curves worsening over time and leading to restrictive pulmonary disease, spinal imbalance, and decreased quality of life. Consequently, at age 13, PSF was recommended to decrease the risk of worsening spinal curvature and secondary neurologic complications. At his last pre-operative clinic visit, he had a left upper thoracic curve of 58°, a right main thoracic curve of 80°, and a left lumbar curve of 42°.

Pre-operative recommendations were made by an interdisciplinary team of specialists from hematology, anesthesiology (involved both preoperatively and intraoperatively), oncology, and orthopedics. The oncologist recommended discontinuing sirolimus for 1–2 weeks before surgery until 2 weeks postoperatively due to concerns about impaired wound healing. The patient completed a course of zoledronic acid for the treatment of osteopenia. On examination, he had no petechiae or bruising, a normal airway with Mallampati Class II view, adequate peripheral pulses and perfusion, and clear breath sounds. He underwent general endotracheal anesthesia after an uneventful intravenous induction with a standard protocol of propofol, fentanyl, diazepam, ketamine, and rocuronium. Tranexamic acid was used to minimize bleeding. Fibrinogen and angiopoietin-2 levels were measured before, during, and after surgery to monitor for KLA flare. In the event of excessive bleeding, cryoprecipitate was determined to be the first-line treatment, with second-line treatment of transfusion of red blood cells.

The patient underwent successful PSF from T2 to L2. Bone grafts of the iliac crest were avoided due to areas of KLA disease identified on imaging. Estimated blood loss was 700 mL, 245 mL of which was replaced with the use of cell salvage. The patient was extubated and admitted to the pediatric intensive care unit, where pediatric hematology monitored him and provided recommendations in the event of consumptive coagulopathies. The plan at that time included serial scrotal examinations every 12 h given his history of KLA and risk of lymphangiomas; close monitoring for bruising, petechiae, and bleeding; and treatment with steroids and vincristine with an oncology consultation if he showed signs of a flare during the immediate post-operative period. First-line treatment of acute issues, such as bruising, respiratory difficulty, diffuse petechiae, or excessive bleeding would consist of steroids, with second-line treatment being vincristine. He had previously responded well to vincristine. Per his parent's request, angiopoietin-2 level was measured serially. He experienced a minor cerebrospinal fluid leak, but otherwise tolerated the procedure well. Pre- and post-operative radiographs are shown in Fig. 1.

At 1-month follow-up with his oncologist, he had no acute concerns and planned to return to school. He did not report excessive fatigue or pain and had no infection. His appetite had returned to baseline and he was gaining weight. At 1-year follow-up with his primary care physician, he had comfortably returned to school with no noted limitations in his daily activities.

Discussion

This patient's high risk of consumptive coagulopathy made managing intraoperative blood loss critical. He did not experience excessive intraoperative or post-operative bleeding, suggesting the effectiveness of tranexamic acid in patients with KLA, with cryoprecipitate as the first-line treatment to reduce the risk of hemorrhagic complications [1].

It was important to monitor for a potential intraoperative KLA flare preoperatively and intraoperatively. The following "goal" parameters were set for laboratory values: Intraoperative fibrinogen concentration of 150–200 mg/dL, post-operative hemoglobin concentration of 8–10 g/dL, and post-operative platelet count of >75,000 cells/mm3. Such thresholds may be considered for monitoring and avoiding flares intraoperatively, given the lack of current standard-of-care management for KLA [1, 3, 4].

Although comparative data on intraoperative monitoring strategies for KLA are limited, the management of coagulopathy in similar lymphatic anomalies has been described. A study on anticoagulation in patients with vascular anomalies highlights the challenges of severe coagulopathy in rare disorders like KLA, emphasizing the need for careful monitoring and tailored therapeutic approaches [11]. In addition, research on multimodal therapy for KLA has shown improvements in coagulopathy and reductions in blood angiopoietin-2 levels, suggesting that vigilant intraoperative monitoring of coagulation parameters is crucial [12].

Our patient responded well to zoledronic acid, which produced higher height-adjusted z-scores at the spine, hip, and total body

(less head), stabilizing his osteopenia before surgery. Zoledronic acid also decreased c-telopeptide, a marker of bone resorption, to a normal level, further indicating its effectiveness in stabilizing the osteolytic effects of KLA.

Pleural and pericardial effusions were of concern; however, our patient did not experience adverse cardiopulmonary events. Chest tubes and pericardial windows have no clear contraindications in patients with KLA and can be considered when addressing cardiopulmonary complications if they arise. Out of concern for infection due to the patient's history of immunosuppressive medications and overall condition, he was prescribed trimethoprim/sulfamethoxazole for *Pneumocystitis jirovecii* prophylaxis.

Only sirolimus has been shown to be effective for managing patients with high-risk, symptomatic KLA [1,3,4,6,10,12,13]. Along with steroids and bisphosphonates, sirolimus decreases markers of the disease, such as angiopoietin-1 and -2, by stabilizing the lymphatic overgrowth with partial responses to the mTOR inhibitor [1,3,4]. Due to the immunosuppressive properties of sirolimus, it was discontinued 2 weeks before our patient's surgery and for 2 weeks afterward to avoid impaired wound healing. Our patient did not experience wound-related complications, suggesting that avoiding the use of sirolimus during the perioperative period may be beneficial.

Although we are aware of no published descriptions of the surgical management of adolescent idiopathic scoliosis in patients with KLA, similar surgical challenges have been encountered in other rare lymphatic disorders that involve the spine. Case reports and case series of patients with Gorham-Stout disease, generalized lymphatic anomaly, and spinal lymphangiomas have highlighted the complexities of spinal deformity correction in the setting of progressive osteolysis, diffuse lymphatic involvement, and multisystem disease [5,14,16].

What makes the surgical management of these lymphatic conditions even more remarkable is the advancement of treatment options that have occurred over time. Historically, management of such rare lymphatic disorders focused on symptom management, with interventions such as radiotherapy and surgical resection being common [17]. Over time, medical therapies have been introduced, including bisphosphonates and immunosuppressants, which can slow or reverse the disease process [18]. These historical approaches underscore the challenges clinicians faced in treating such conditions and highlight the advancements that have led to multidisciplinary care models. Ultimately, an analysis of these cases provides context for the feasibility and considerations of posterior spinal fusion in patients with KLA, emphasizing the multidisciplinary approach needed to optimize outcomes.

Although our case highlights the benefits of a multidisciplinary team approach, challenges such as logistical coordination, financial constraints, and resource limitations – particularly in lower-resource healthcare settings – may limit the feasibility and consistency of such comprehensive care, warranting further exploration [19].

It is important to consider long-term outcomes after PSF in patients with KLA. However, we have found no literature describing long-term prognosis in this patient population. Given that KLA has a 5-year survival rate of approximately 51%, with overall survival at 34% due to complications like cardiorespiratory failure, the long-term prognosis of patients undergoing PSF remains uncertain, as the physiological burden of both the disease and the surgical intervention may affect morbidity and functional outcomes, necessitating further investigation [3].

Conclusion

The primary takeaway from this case is the necessity of comprehensive, multidisciplinary management for surgical patients with complex underlying conditions such as KLA. Future research should help establish guidelines and standard-of-care treatment for medically and surgically managing this unique patient population. Our patient's successful post-operative course can help guide orthopedic spine procedures for patients with complicated lymphatic and vascular anomalies. Specific recommendations for patients with KLA undergoing orthopedic procedures include establishing a comprehensive diagnostic workup, collaboration with care teams of various disciplines, thorough surgical planning, and streamlined post-operative monitoring and planning.

The diagnostic workup should involve blood tests, dual-energy x-ray absorptiometry, biopsy, and imaging of the spine and scrotum. Collaboration with specialists in hematology, oncology, orthopedics, anesthesiology, pediatric intensive care, radiology, rheumatology, pediatrics, pharmacy, nutrition, and social work is essential both before and after surgery. Surgical planning entails special considerations, such as preparing for cardiac or pulmonary complications through chest tubes and pericardial windows, monitoring for specific complications or decompensation (e.g., coagulopathies, bleeding), and setting goal parameters to guide intraoperative monitoring. In addition, restriction of certain medications such as sirolimus may help decrease wound-related complications. Finally, post-operative care should involve a regimented treatment plan, including the use of steroids, bisphosphonates, and sirolimus, as well as consistent follow-up.

Clinical Message

When considering surgical treatment in patients with kaposiform lymphangiomatosis, pre-operative planning, and multidisciplinary care are required to mitigate the high risk of morbidity and death from blood loss.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient and his patients have given consent for his images and other clinical information to be reported in the journal. The patient and his parents understand that his name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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