CASE REPORT
DEFORMITY
http://dx.doi.org/10.1590/S1808-185120242303288432

MITIGATING HIGH-RISK COMPLICATIONS: TENSION HEMOTHORAX IN NF-1 PATIENTS UNDERGOING SCOLIOSIS CORRECTION

MITIGANDO COMPLICAÇÕES DE ALTO RISCO: HEMOTÓRAX HIPERTENSIVO EM PACIENTES COM NF-1 SUBMETIDOS À CORREÇÃO DA ESCOLIOSE

MITIGANDO COMPLICACIONES DE ALTO RIESGO: HEMOTÓRAX HIPERTENSIVO EN PACIENTES CON NF-1 SOMETIDOS A CORRECCIÓN DE ESCOLIOSIS

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ABSTRACT

Objectives: This study aims to report and analyze two cases of tension hemothorax in patients with neurofibromatosis type 1 (NF1) undergoing scoliosis surgery, highlighting the incidence, etiology, and optimal treatment strategies. Methods: We present two detailed cases of tension hemothorax in NF1 patients who underwent scoliosis surgery. Clinical presentations, diagnostic findings, treatment interventions, and outcomes were meticulously documented. A literature review was conducted to contextualize these cases within the broader spectrum of tension hemothorax in NF1 patients undergoing similar surgeries. Results: Both cases involved severe complications requiring immediate surgical interventions. The first patient experienced arterial bleeding necessitating endovascular embolization, while the second patient required open thoracotomy. Postoperative management included intensive monitoring and multidisciplinary care. The literature review supported these findings, indicating that surgical interventions such as chest tube insertion, thoracoscopy, and angiography with endovascular embolization are common in such cases. Conclusion: These cases underscore the critical need for vigilance and prompt intervention in managing tension hemothorax in NF1 patients undergoing scoliosis surgery. A multidisciplinary approach is essential for optimizing treatment outcomes and improving understanding of this rare but life-threatening complication in this specific surgical context. ***Level of Evidence IV; Case Report.***

Keywords: Neurofibromatosis 1; Peripheral Neurofibromatosis; Hemothorax; Scoliosis.

RESUMO

Objetivos: Este estudo visa relatar e analisar dois casos de hemotórax hipertensivo em pacientes com neurofibromatose tipo 1 (NF1) submetidos a cirurgia de escoliose, destacando a incidência, etiologia e estratégias de tratamento ideais. Métodos: Apresentou-se dois casos detalhados de hemotórax hipertensivo em pacientes com NF1 que foram submetidos a cirurgia de escoliose. As apresentações clínicas, achados diagnósticos, intervenções terapêuticas e desfechos foram meticulosamente documentados. Uma revisão da literatura foi realizada para contextualizar esses casos dentro do espectro mais amplo de hemotórax hipertensivo em pacientes com NF1 submetidos a cirurgias semelhantes. Resultados: Ambos os casos envolveram complicações graves que necessitaram de intervenções cirúrgicas imediatas. O primeiro paciente apresentou sangramento arterial que necessitou de embolização endovascular, enquanto o segundo paciente precisou de toracotomia aberta. O manejo pós-operatório incluiu monitoramento intensivo e cuidados multidisciplinares. A revisão da literatura apoiou esses achados, indicando que intervenções cirúrgicas como inserção de tubo torácico, toracoscopia e angiografia com embolização endovascular são comuns em tais casos. Conclusão: Esses casos ressaltam a necessidade crítica de vigilância e intervenção rápida no manejo do hemotórax hipertensivo em pacientes com NF1 submetidos a cirurgia de escoliose. Uma abordagem multidisciplinar é essencial para otimizar os desfechos do tratamento e melhorar a compreensão dessa complicação rara, mas potencialmente fatal, nesse contexto cirúrgico específico. ***Nível de Evidência IV; Relato de Caso.***

Descritores: *Neurofibromatose 1; Neurofibromatose Periférica; Hemotórax; Escoliose.*

RESUMEN

Objetivos: Este estudio tiene como objetivo informar y analizar dos casos de hemotórax hipertensivo en pacientes con neurofibromatosis tipo 1 (NF1) sometidos a cirugía de escoliosis, destacando la incidencia, la etiología y las estrategias de tratamiento óptimas. Métodos: Se presentan dos casos detallados de hemotórax hipertensivo en pacientes con NF1 sometidos a cirugía de escoliosis. Se documentaron meticulosamente las presentaciones clínicas, los hallazgos diagnósticos, las intervenciones terapéuticas y los resultados. Se realizó una revisión bibliográfica para contextualizar estos casos dentro del espectro más amplio de hemotórax hipertensivo en pacientes con NF1 sometidos a cirugía similar. Resultados: Ambos casos presentaron complicaciones graves que requirieron intervención quirúrgica inmediata. La primera paciente presentó una hemorragia arterial que requirió embolización endovascular, mientras que la segunda necesitó una toracotomía abierta. El tratamiento postoperatorio incluyó vigilancia intensiva y atención multidisciplinar. La revisión bibliográfica respaldó estos hallazgos, indicando que las intervenciones quirúrgicas como la inserción de un tubo torácico, la toracoscopia y la angiografía con

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Received on 07/12/2024 accepted on 08/20/2024
Reviewed by: Helton Defino

embolización endovascular son frecuentes en estos casos. Conclusión: Estos casos destacan la necesidad crítica de vigilancia e intervención rápida en el tratamiento del hemotórax hipertensivo en pacientes con NF1 sometidos a cirugía de escoliosis. Un enfoque multidisciplinar es esencial para optimizar los resultados del tratamiento y mejorar la comprensión de esta complicación rara pero potencialmente mortal en este contexto quirúrgico específico. ***Nivel de Evidencia IV; Informe de Caso.***

Descriptores: *Neurofibromatosis 1; Neurofibromatosis Periférica; Hemotórax; Escoliosis.*

INTRODUCTION

Neurofibromatosis type 1 (NF1), also known as von Recklinghausen's disease, is the most common type of phacomatosis.1 NF1 is characterized by polymorphic clinical manifestations, with skin and neurological symptoms being most prevalent, along with involvement of the skeletal and vascular systems.

Skeletal deformities, particularly spinal deformities, occur in 10-30% of cases.[2] Two types of spinal deformities are observed in NF1. The non-dystrophic type of scoliosis is nearly indistinguishable from typical idiopathic scoliosis. Still, it tends to have a more severe course and poorer prognosis, with dystrophic changes potentially occurring as the patient grows. The second type of deformity, dystrophic or dysplastic, is unique to NF1 and exhibits characteristic features primarily identified through radiographic examination.

Vascular changes occur in only 1-3.6% of NF1 patients.[3,4] Most commonly affected are medium and large caliber vessels, with disruptions attributed to weakness in the vessel wall leading to the formation of aneurysmal dilations and stenoses.

The motivation for conducting this analysis stemmed from encountering similar complications twice in our practice: both patients with NF1 developed tension hemothorax following posterior spinal surgery without any vertebrectomies.

The main question of this study: what is the incidence and etiology of tension hemothorax in patients with NF1, particularly following spinal surgery or other intervention on the thorax and thoracic spine? What is the optimal approach to treating this condition?

We hypothesize that tension hemothorax is a rare but potentially fatal complication in NF1 patients, primarily attributed to vascular weakness related to NF1-associated vasculopathy.

METHODS

In the subsequent section of the article, we provide detailed descriptions of two of our cases, outlining the clinical course, diagnostic evaluations, treatment interventions, and outcomes for each case. All participants signed the free and informed consent form. The study was approved by the local ethics committee. It is retrospective and does not affect the quality of patient treatment. The number of the ethics committee protocol is N92024.

RESULTS

Description of cases

A 16-year-old patient was admitted to the hospital with progressive dystrophic kyphoscoliosis attributed to NF1 (Figures 1a and 1b). On MRI, intrathoracic pathological apical tissue, probably neurofibroma was visible on the right side of the deformity (Figures 1c, 1d, 1e). Following the data assessment, a treatment plan was established, comprising a two-stage surgical intervention targeting the upper and lower spine to correct scoliosis.

After a straightforward surgical correction of the upper thoracic curve using short pedicle screw construction without vertebrectomy, the patient regained consciousness and remained stable for 8 hours post-surgery. However, the patient's condition gradually worsened, presenting with oliguria, pallor, dyspnea, and dullness upon percussion of the right chest wall. A thoracic X-ray revealed a tense right-sided hemothorax (Figure 2a). Initially, a right pleural tube was inserted, but persistent copious blood discharge for over 30 minutes ensued, leading to hemodynamic instability and cardiac arrest. Consequently, an immediate decision was made to proceed with revision surgery on the spine. During repositioning on the operating table, another cardiac arrest occurred. However, resuscitative measures were swiftly administered, and cardiac function resumed within a minute.

The revision surgery involved removing a rod and one screw from the convex side, 5th right rib head removal. Diffuse bleeding from pathological tissue in the apical was observed. The presumed source of bleeding, identified as the 5th intercostal artery, was coagulated, and tamponade of the paravertebral vein was performed by oxidized cellulose, fibrin hemostatic sponge, and collagen hemostatic sponge.

The following morning, a CT scan with angiography revealed no apparent source of bleeding, although some blood was observed in the pleural space (Figure 2b). Subsequently, a thoracotomy was

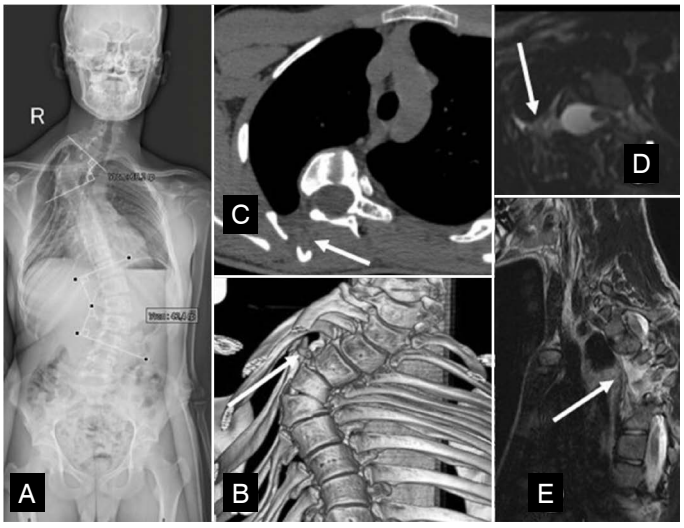

Figure 1. Preoperative Imaging: Case 1. (A) X-ray of the spine revealing scoliosis; (B) 3D CT reconstruction of the upper thoracic spine depicting features of dystrophic scoliosis; (C) CT slice through the apex of the deformity, displaying dystrophic bone changes and intrathoracic pathological tissue (indicated by the white arrow); (D) Axial MRI slice through the apex of the deformity highlighting dural ectasia and pathological intrathoracic tissue in the apex of the right lung (indicated by the white arrow); (E) Sagittal MRI slice at the level of the spinal deformity unveiling pathological intrathoracic tissue in the posterior apex of the right lung (indicated by the white arrow).

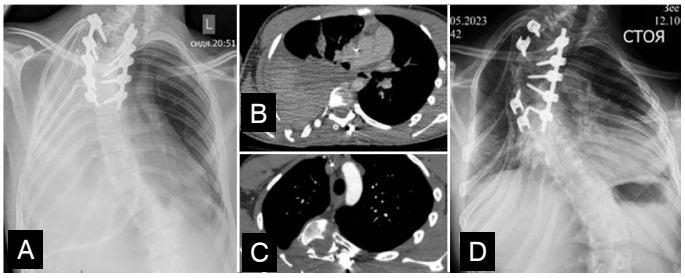

Figure 2. Postoperative Imaging: Case 1. (A) Postoperative chest X-ray 8 hours after scoliosis surgery, revealing a tense right-sided hemothorax; (B) Chest tomography following revision surgery for hemothorax, displaying a hematoma in the posterior regions of the right lung; (C) Chest tomography taken four days after thoracotomy; (D) Chest X-ray captured two years after surgery, illustrating the progression of dystrophic scoliosis.

performed, uncovering minor blood leakage from the paravertebral venous plexuses in the area of the 5th intercostal space. The pleural drains were removed on the 4th day post-surgery (Figure 2c). The patient was discharged satisfactorily on the 20th day post-surgery and remained well during the 2-year follow-up period (Figure 2d).

In the second case, a 24-year-old female patient underwent surgical intervention for kyphoscoliosis, spinal stenosis, and lower paraplegia associated with NF1 (Figure 3a-c). The surgical procedure involved a vertebral column resection (VCR) at the Th2--Th3 level, accompanied by posterior screw fixation from C5 to Th6 (Figure 3d-e). Initially, the patient was fine, but on the 4th day, she reported intensified pain and dyspnea, prompting a chest X-ray and CT scan. The radiography revealed a complete right-sided hemothorax, resulting in lung collapse (Figure 3d). Consequently, the patient underwent a chest tube, which was removed after two days. Following a comprehensive treatment regimen, she was discharged on the 21st day post-surgery in a stable condition. A year after surgery, the patient suffered from pelvic and hip osteomyelitis and lower extremities hip fracture, and therefore, she underwent multi-stage treatment. Over the course of 8 years of follow-up, she has persistent paraplegia without repeated hemothorax (Figure 3e).

DISCUSSION

Diagnosis of NF1-associated vascular disease may occur in both children and adults, with hemorrhage being a significant contributor to NF1-related mortality.

The pathogenesis of arterial lesions in NF1 is complex, involving factors such as the proliferation of spindle cells within vessel walls, fibrous thickening, cellular nodularity compromising vessel wall integrity (particularly in small vessels), Schwann cell proliferation within large vessel arterial walls, and dysplastic smooth muscle.[5] These abnormalities increase the vulnerability of arteries in NF1 patients, potentially leading to life-threatening hemorrhages, such as tension hemothorax.

The notion that surgical procedures involving the thoracic vertebrae or intrathoracic surgeries, such as vertebral resection or correction of deformities, can inevitably lead to tense hemothorax, particularly in patients with neurofibromatosis. Surgical interventions on the thoracic vertebrae or intrathoracic surgeries carry inherent risks of hemothorax in patients with NF1, especially in presence of apical or paravertebral neurofibromas. Postsurgical hemothorax is probably directly linked to the surgical site. This scenario considerably streamlines the process of identifying the bleeding source.

According to the literature review and our clinical experience, the presence of tension hemothorax in NF1 patients correlates strongly with elevated risks of cardiac arrest, mortality, and recurrent bleeding, both in the immediate and long-term postoperative periods.

Treatment options include traditional open thoracotomy, thoracoscopy, and endovascular treatment.

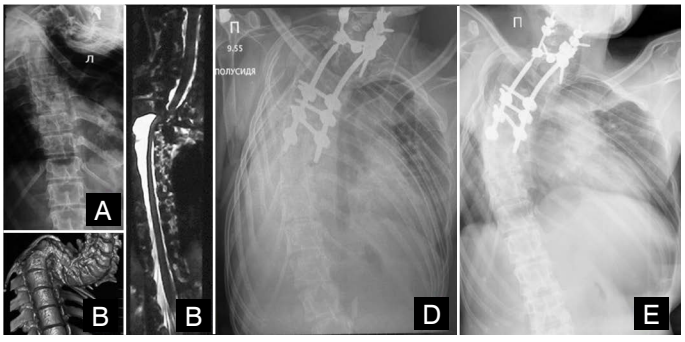

Figure 3. Preoperative and Postoperative Imaging: Case 2. (a) X-ray of the spine revealing scoliosis; (b) 3D CT reconstruction of the upper thoracic spine depicting features of dystrophic scoliosis; (c) MR slice through the apex of the deformity, displaying dural ectasia and stenosis; (d) Postoperative chest X-ray 4 days after scoliosis surgery, revealing a tense right-sided hemothorax; (e) Chest and spine X-ray captured eight years after surgery.

CONCLUSION

The analysis of two clinical cases of tension hemothorax in patients with neurofibromatosis type 1 (NF1) undergoing scoliosis surgery highlights the critical importance of timely and effective intervention. Both cases demonstrated that immediate surgical response, including techniques such as endovascular embolization and open thoracotomy, is essential for managing severe complications. The complexity of these cases underscores the necessity of a multidisciplinary approach to optimize patient outcomes. Continuous vigilance and careful monitoring are vital to address potential postoperative complications and ensure long-term recovery. Further research and clinical experience are needed to refine treatment protocols and improve the prognosis for NF1 patients facing similar challenges. The analysis of two clinical cases of tension hemothorax in patients with neurofibromatosis type 1 (NF1) undergoing scoliosis surgery highlights the critical importance of timely and effective intervention. Both cases demonstrated that immediate surgical response, including techniques such as endovascular embolization and open thoracotomy, is essential for managing severe complications. The complexity of these cases underscores the necessity of a multidisciplinary approach to optimize patient outcomes. Continuous vigilance and careful monitoring are vital to address potential postoperative complications and ensure long-term recovery. Further research and clinical experience are needed to refine treatment protocols and improve the prognosis for NF1 patients facing similar challenges.

All authors declare no potential conflict of interest related to this article.

CONTRIBUTIONS OF THE AUTHORS: Each author has made an individual and significant contribution to the development of this article. OMS contributed to Methodology and supervision; ARG contributed to formal analysis and data curation; DMS contributed to Validation and writing – proofreading, and editing; PVO made substantial contributions to the acquisition of data; AVB contributed to project administration and supervision; AVE contributed to conceptualization and funding acquisition.

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