ROTARY SUBLUXATION OF THE COLUMN IN NEUROFIBROMATOSIS TYPE I: APPLICATION OF THE INFRAAXILLARY ROUTE.

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Summary

Introduction

60% of patients with neurofibromatosis associate vertebral dystrophies and spinal deformities, most of them high and very angular kyphoscoliosis, where the vertebrae progressively dislocate, exerting shearing on the medullary canal that gives rise to severe neurological alterations. In these patients, the approach to the deformity through the usual routes continues to be a challenge; For this reason, we describe a recent technique performing an infraaxillary thoracotomy.

Material and methods

We present the case of a 9-year-old woman with NF type I and left convexity high thoracic kyphoscoliosis (double thoracic curve T1-T5 of 71º left and T5-T10 of 90º right, with an angular kyphosis of 59º), with normal neurological examination. . After a period of spinal traction, we prepared to perform a posterior arthrodesis, but when the patient was placed in the prone position on the surgical table, motor evoked potentials abolished, forcing the intervention to be suspended.

Results

The solution was to perform a left infraaxillary thoracotomy, approaching the curve from the convexity. The patient was placed in the right lateral decubitus at 70º and a rib resection was performed two or three levels above the apex of the deformity. After careful dissection keeping close to the chest wall, decompression and anterior discectomy were performed at the apex level. Two "strut" tibial allografts were then placed in the concavity as a palisade to withstand the compressive forces. In a second stage, a posterior instrumented approach was performed to achieve a circumferential arthrodesis and optimal stability.

Conclusion

The presented technique allows direct and safe access to the upper thoracic spine, as it does not involve vital structures including the great vessels or the thoracic duct as classical approaches could do. This is a promising technique with good clinical results, which can be used for the treatment of kyphotic deformities where single posterior approaches have failed, but also instability, spinal cord compression or other pathologies of the vertebral body, as it can achieve a good exposition of them.

KEY WORDS: "kyphoscoliosis", "neurofibromatosis", "infraaxillary approach", "rotary subluxation".

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INTRODUCTION

Neurofibromatosis is **a** disease hereditary that affects the tissues derived from the three embryonic lineages: the neuroectoderm, the endoderm and the mesoderm. Therefore, the clinical manifestations are highly variable, and can affect the skin, nervous system, skeleton, and soft tissues. There are two types of neurofibromatosis. Type **I** is the most common and the criteria for establishing its diagnosis are the presence of "café au lait" skin spots, neurofibromas, optic nerve gliomas, iris hamartomas, and musculoskeletal system abnormalities[1].

Overall, 60% of patients with neurofibromatosis type I have spinal deformities, which may or may not be associated with osseous dystrophy of the vertebrae. This dystrophic component may go accompanied by anomalies of the spinal canal secondary to a malformation medullary. The most common deformities are scoliosis, which affect 2-36% of patients with neurofibromatosis type I (NF1). Almost all of them are short and angular and appear early in childhood. Pure kyphosis is rare, and kyphoscoliosis much more common[2] .

In dorsal kyphoscoliosis there is a clear predominance of kyphosis with respect to the purely scoliotic deformity. The vertebral bodies are often so deformed that they cannot be seen on routine radiographs. The CT allows a complete study of each dystrophic vertebra and the contours of the spinal canal. The three-dimensional reconstructions provide a detailed morphological analysis in all three planes of space. MRI can study the spinal cord and visualize the areas of spinal cord involvement or possible associated spinal cord malformations. The presence of ectasias of the dura mater, which sometimes cause progressive erosion of the vertebral bodies and destabilization of the spine ("scalloping" or "undulation" phenomenon) [2]

Vertebral Rotational Subluxation

The term "rotary subluxation" was coined and described by Duval-Beaupere and Dubousset in 1972, in a study where they analyzed 11 patients with some type of dystrophy due to vertebral to neurofibromatosis and other congenital dysplasias[3] .

They saw that in this type of disease the two dystrophic vertebrae located in the junction of two segments of scoliotic curves (each of them lordotic and opposite rotated) progressively dislocated or sinubluxateadd, dardeospsteinsg a kyphotic and rotational configuration, exerting a shearing effect around the medullary canal. The AP and lateral radiographs demonstrate that the apex of the kyphosis is located exactly at the junction of the two curves scoliotic seen in the AP.

This phenomenon progresses over time. The "hairpin" that forms is responsible for a bayonet deformity of the spinal canal. This progressive deformity is, in turn, responsible for the stretching of the cord in the spinal canal and, therefore, the possible origin of neurological alterations. These deformities also cause respiratory failure due to altered shape and incompetence of the thoracic cage.

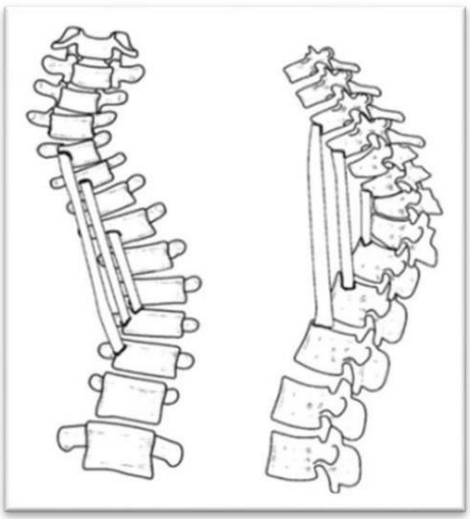
Treatment early: arthrodesis circumferential

It is important to be aware that in dystrophic forms of kyphoscoliosis such as In NF, there is the possibility that progressive rotational subluxation may occur over time, leading to

neurological complications progressive as paraplegia, so stabilization must be early [3. 4. 5].

A prior period of spinal traction is often required to obtain maximum and progressive correction, with possible recovery from neurological abnormalities and better respiratory preparation. Isolated posterior instrumented fusion procedures are often doomed to failure, with recurrence of the deformity[6,7] .

Solid anterior support must be achieved, usually with the aid of autologous fibular **"strut"** grafts [2, 3]. It is preferable to start with the anterior time, in order not to run the risk of completely destabilizing the spine as a consequence of possible bone excisions made previously by a posterior approach, and to maintain the correction obtained with preoperative traction.



After some time the posterior arthrodesis is carried out. The correction of the spinal deformity at this point represents a decompression maneuver of the medullary canal that requires severe torsion of the apex of the kyphosis, therefore it must be performed with special care.

Immobilization, almost always by means of a corset system coupled to a cranial halo, must be prolonged, with a total duration of 6 months to 1 year.

A new approach: the infraaxillary approach

Disorders of the upper thoracic spine

they can lead to severe complications and morbidities, and yet operating on it continues to be a challenge due to the characteristics of the anatomy at this level.

Recently, a new

technique of approaching the upper thoracic spine that up to now has proven to be safe and easily reproducible, the infaaxillary thoracotomy [8], a route normally used in cardiac and thoracic surgery.

This approach route allows us direct access to the vertebral bodies from T2 to T6 inclusive, avoiding the risk of damaging complex anatomical structures that were at risk in the classic anterior approach routes, such as the transsternal or high transthoracic approach, where we find vital structures such as

thoracic duct and great vessels.

Through this anterolateral thoracic approach we can

safely expose the vertebral bodies of the upper thoracic spine and thus treat pathologies of the vertebral body that can cause spinal cord compressions, spinal instability or kyphotic deformities, due to diseases such as metastatic tumors, cold abscesses or progressive deformities such as the case of this article. All this avoiding the

mediastinum and therefore minimizing the risk of damaging structures such as the esophagus, the pleura, the recurrent laryngeal nerve, the vagus nerve or the great

glasses.

MATERIAL AND METHODS

In this article we present the case of a 9-year-old woman with neurofibromatosis type I, who presented

a high dorsal convexity kyphoscoliosis

left: double thoracic curve T1-T5 of 71º left and T5-T10 of 90º right, with an angular kyphosis of 59º. There were no bone malformations on CT and no spinal cord compromise on MRI, showing in the latter a clamping of the discs affected by the scoliosis segment.

On physical examination he had:

• Multiple café au lait stains. • Area of left thoracic fixed rotation. left slightly
• Shoulder
promoted.
• Right pelvis slightly raised. • Erase left fold. • Hamstring contracture.

Peripheral neurological examination was intact and he presented a pattern of respiratory restriction in functional tests.

Image 1: X-rays of the patient prior to the intervention, during admission with halo-traction.

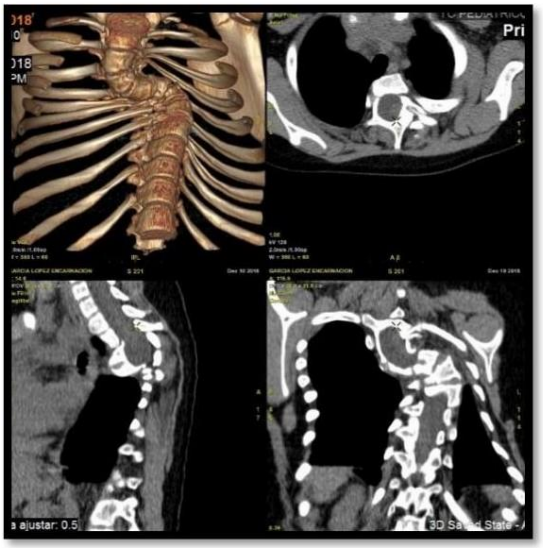


Image 2: CT and 3D reconstructions

First, spinal traction with cranial halo was carried out for a period of one month to achieve a progressive stretching of the kyphosis and thus obtain the maximum possible correction. During this period, the daily neurological examination was normal.

After this preparation, we proceed to perform instarumented posterior arthrodesis for the treatment of the deformity, with the surprise that when the patient is placed in the prone position, motor and sensory potentials in the lower limbs are abolished, forcing the intervention to be suspended.

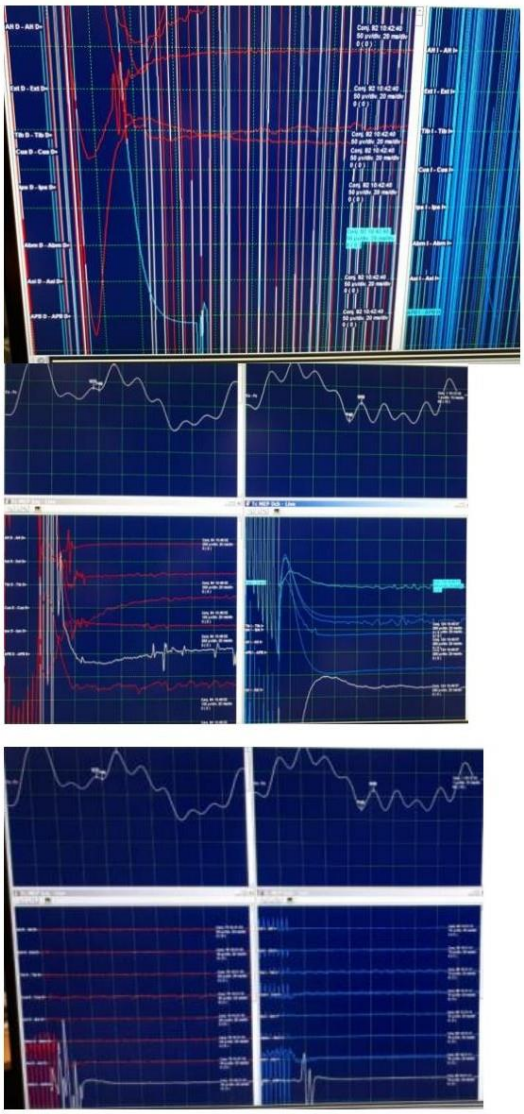


Image 3: intraoperative images of neurophysiological monitoring showing the loss of PESS and PEM in MMII during anesthetic induction.

Conduction was recovered in the supine position. After this eventuality, an update of all the

studies and an extension of them with MRI, angio-CT and 3D reconstructions.

Image 4: MRI slice showing significant kinking of the spinal cord at the apex of the deformity at the T4-T6 level, without identifying intramedullary signal changes suggestive of myelopathy or vertebral anomalies.

RESULTS

Given the suspicion that it could be a rotary subluxation of the upper thoracic spine, it was decided that the most appropriate treatment would be to perform an arthrodesis. circumferential with a double track approach, starting with an **anterior route in the first stage.**

The original technique describes the anterior approach from the concavity of the curve lower scoliotic. However, in our case we decided to approach the curve from its convex part, since due to its anatomy the curve is better exposed and manipulation of the great vessels is avoided.

The patient was placed in the right lateral decubitus. A left anterolateral thoracotomy was performed **at the infraaxillary level,** with rib resection two or three levels above the apex of the deformity.

After careful dissection keeping close to the chest wall, the apex of the deformity is left discovered. To gain access to the apex, the segmental vessels related to these vertebrae were ligated. Decompression and anterior discectomy were performed at the apex level.

Next, a cadaveric tibia graft was obtained, from which two strut-shaped grafts were made. The original technique describes obtaining the graft from the patient's own tibia or fibula, including the use of the resected ribs. In our patient, this option was rejected due to the possible dysplastic characteristics of that bone due to its base NF. Tibia was also chosen for its greater abundance of cancellous bone.

These **two "strut" tibial grafts** were inserted into the concavity of the kyphosis as a palisade or buttress, beginning with a short one at the level of the apex of the kyphosis. In this way, in the concavity, they will support the compression forces. Due to the orientation of the vertebral bodies, the grafts were aligned with gravity in the sagittal plane and more or less oblique in the coronal plane. The gaps between the

Tibial grafts were filled with cancellous bone chips.

Image 5: Intraoperative photographs of the positioning of the patient in right lateral decubitus and the left infraaxillary approach performed.

One month later, after the patient's recovery and her correct clinical and radiological evolution, the **second stage of surgery** was performed using a posterior route, also with intraoperative multimodal monitoring, to complete the circumferential arthrodesis.

In this case, an approach was performed from C7 to T12, Ponte-type osteotomies at the apex level of the deformity, and posterior instrumentation with pedicle screws and costal nails. The potentials remained conserved at all times.

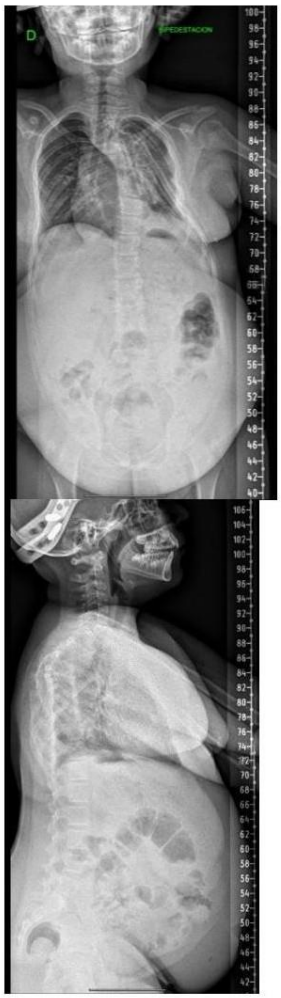


Image 6: Anteroposterior and lateral X-rays of the entire spine in standing position after the first postoperative period.

At follow-up one year later, the patient is asymptomatic, leading a normal life and sports. The evolution has been satisfactory, without neurological complications so far.

Image 7: Intraoperative photographs during the second surgical period with the patient in the prone position, posterior approach route and instrumentation.

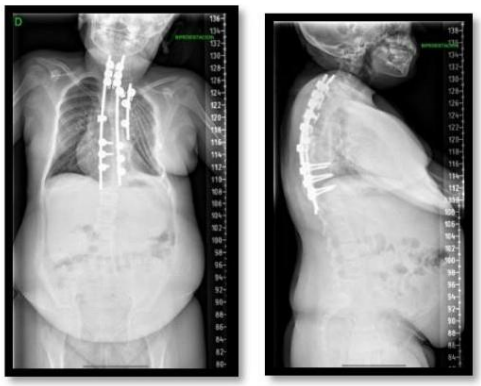


Image 8: Anteroposterior and lateral X-rays of the entire spine in standing position after the second surgical procedure.

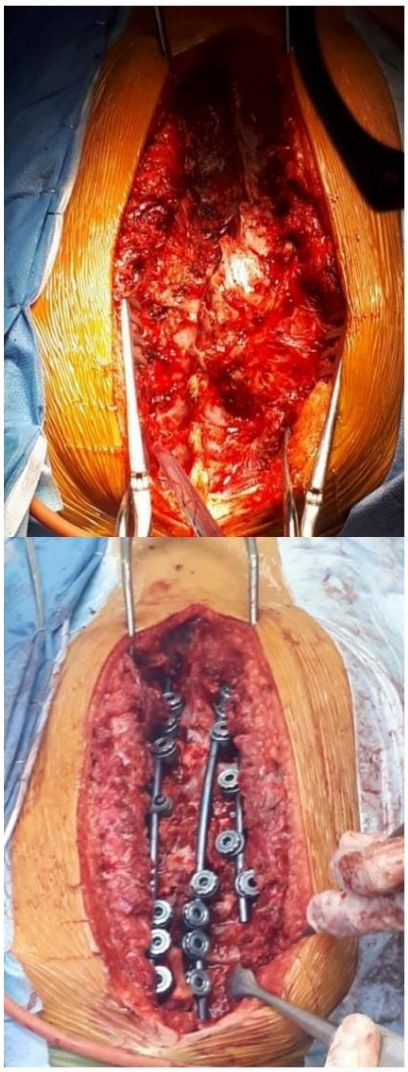


Image 9: Anteroposterior and lateral radiographs of the complete spine in standing position and 3D CT reconstruction slices at follow-up one year after the intervention.

Image 10: Clinical photographs of the patient in consultation at the follow-up one year after the intervention.

CONCLUSIONS

The debut of a rotary subluxation within a kyphoscoliosis is one of the more severe forms of spinal cord deformity, which can have serious neurological consequences.

Its early diagnosis is essential, and is based on the proper identification of the pattern of the curve: the apex of the kyphosis on the lateral radiograph corresponds to the area where two scoliotic curves meet on the anteroposterior radiograph.

This deformity, the angular kyphosis that exists even in the early stages of the disease, progresses rapidly to produce neurological deficits, so early stabilization is essential.

arthrodesis is recommended circumferential with strut grafts through an anterior approach, since it has been seen that in most cases single posterior approaches fail.

To carry out this previous route, recommends the application of the infraaxillary thoracotomy approach, a recent approach that, according to experience to date, is easily reproducible and allows us to have direct access to the vertebral bodies of the upper thoracic spine in a safe manner, with good visualization of the same and adequate space to operate, minimizing the risk of damage to adjacent vital structures.

**All images were obtained and used with the signed consent of both parents and in agreement with the patient. It is declared that there are no conflicts of interest.*

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