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Case Report

Diffuse Dural Ectasia and Basilar Impression in a Child with Marfan Syndrome: A Case Report

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Abstract

Marfan syndrome is a systemic connective tissue disorder caused predominantly by pathogenic variants in the FBN1 gene. Dural ectasia is a well-recognized spinal manifestation, typically involving the lumbosacral region, and although frequently asymptomatic, its recognition is important because progressive dural enlargement may lead to secondary osseous remodeling, neurological manifestations, or cerebrospinal fluid leakage. We report the case of a 9-year-old girl with known Marfan syndrome who underwent spinal magnetic resonance imaging (MRI) because of difficulty maintaining head control and for assessment of associated spinal abnormalities. MRI demonstrated diffuse enlargement of the dural sac, predominantly involving the lumbar and lumbosacral regions, without significant posterior vertebral scalloping, osseous remodeling, or dural diverticula. Quantitative assessment was performed using the dural sac ratio (DSR) from L1 to S1. Evaluation of the craniovertebral junction additionally revealed basilar impression. In the absence of complications attributable to the dural ectasia, conservative management was considered appropriate. This case highlights the importance of recognizing dural ectasia in children with Marfan syndrome, as MRI allows both morphological and quantitative assessment, detection of associated complications, and establishment of a baseline for subsequent evaluation during skeletal growth.

Keywords: *Marfan Syndrome; Dural Ectasia; Dural Sac Ratio; Magnetic Resonance Imaging; Pediatric Imaging; Basilar Impression*

Introduction

Marfan syndrome (MFS) is an autosomal dominant systemic connective tissue disorder, predominantly caused by pathogenic variants in the FBN1 gene encoding fibrillin-1. It is characterized by marked phenotypic variability and primarily involves the cardiovascular, ocular, and musculoskeletal systems [1]. Cardiovascular manifestations, particularly aortic root dilatation and its complications, remain the major determinants of morbidity and mortality, whereas skeletal manifestations include disproportionate long-bone overgrowth, joint laxity, pectus deformities, scoliosis, and other abnormalities that may become increasingly apparent during childhood and periods of rapid growth [1].

The revised Ghent nosology emphasizes cardiovascular and ocular manifestations, molecular findings, family history, and a systemic score incorporating several extracardiac manifestations [2]. Dural ectasia is one of these recognized systemic features and represents an important manifestation of connective tissue involvement in MFS [2].

Dural ectasia is characterized by enlargement of the dural sac, typically predominating in the lumbosacral region. It may be associated with posterior vertebral scalloping, neural foraminal widening, nerve-root sleeve enlargement, dural diverticula, and anterior sacral meningoceles [3]. Magnetic resonance imaging (MRI) is the imaging modality of choice for its detection, morphological characterization, and assessment of potential complications.

We report the case of a 9-year-old girl with known Marfan syndrome in whom spinal MRI demonstrated diffuse dural ectasia, predominantly involving the lumbar and lumbosacral regions, associated with an abnormality of the craniovertebral junction.

Case Presentation

A 9-year-old girl with a known diagnosis of Marfan syndrome was referred for spinal MRI because of difficulty maintaining head control. MRI of the entire spine was performed to assess potential spinal manifestations of her underlying connective tissue disorder, with particular attention to the cervical spine and craniovertebral junction.

Medullar MRI (Figure 1) demonstrated diffuse enlargement of the dural sac along the spinal canal, with clear predominance in the lumbar and lumbosacral regions, consistent with dural ectasia. No associated dural diverticula or posterior vertebral wall defects were identified. The adjacent vertebral bodies showed no significant posterior scalloping or osseous remodeling.

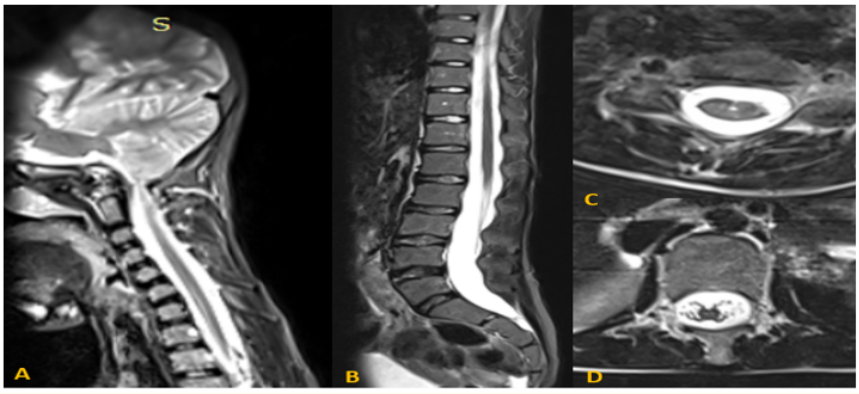

Figure 1: *Spinal MRI findings in a 9-year-old girl with Marfan syndrome. (A) Sagittal T2-weighted image of the cervical spine showing basilar impression at the craniovertebral junction. (B) Sagittal T2-weighted image of the lumbar spine demonstrating diffuse enlargement of the dural sac, predominantly involving the lower lumbar and lumbosacral levels, consistent with dural ectasia, without significant posterior vertebral scalloping. (C, D) Axial T2-weighted images at different spinal levels showing enlargement of the dural sac and increased perimedullary/periradicular cerebrospinal fluid spaces, without associated dural diverticula.*

Quantitative assessment was performed by measuring the anteroposterior diameter of the dural sac and that of the corresponding vertebral body from L1 to S1. The dural sac ratio (DSR), defined as the ratio of the anteroposterior diameter of the dural sac to that of the corresponding vertebral body, was calculated at each vertebral level according to previously described quantitative methods (Figure 2).

Evaluation of the cervical spine and craniovertebral junction additionally demonstrated a craniovertebral junction abnormality characterized by basilar impression.

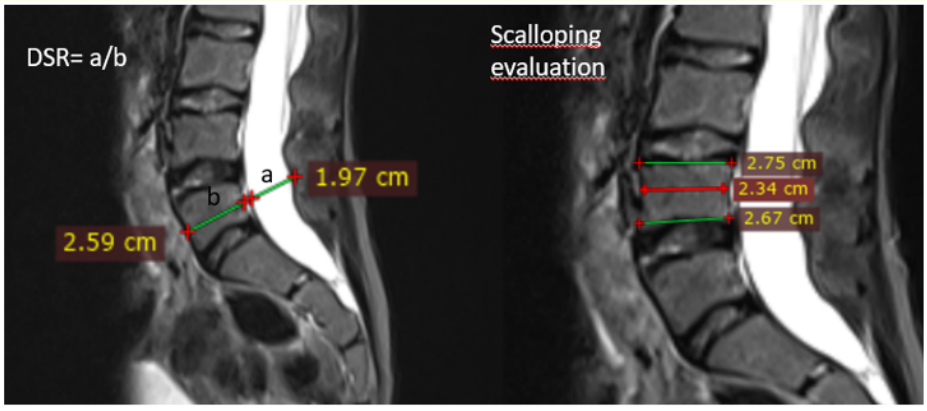

Figure 2: *Quantitative MRI assessment of dural ectasia and vertebral scalloping. Sagittal T2-weighted MR images illustrating measurement of the dural sac ratio (DSR), calculated as the ratio between the anteroposterior diameter of the dural sac (a) and that of the corresponding vertebral body (b) (DSR = a/b). In the illustrated measurement, the DSR was 0.76. Additional anteroposterior vertebral body measurements were obtained to assess posterior vertebral scalloping, with no significant vertebral remodeling identified.*

The overall MRI findings were therefore consistent with diffuse dural ectasia, predominantly involving the lumbosacral region, associated with basilar impression in a child with Marfan syndrome.

Discussion

Marfan syndrome is a multisystem connective tissue disorder with considerable phenotypic variability. Although cardiovascular involvement remains the major determinant of prognosis, systematic assessment should also consider ocular, skeletal, pulmonary, integumentary, and neurological manifestations [1,2]. Skeletal abnormalities may become progressively more apparent during childhood and periods of rapid growth, emphasizing the importance of appropriate longitudinal assessment in pediatric patients.

Dural ectasia is a characteristic spinal manifestation of Marfan syndrome. It is characterized by widening or ballooning of the dural sac and typically predominates in the lower lumbar and sacral regions [3,4]. The underlying connective tissue weakness is thought to reduce the ability of the dura and surrounding structures to withstand chronic cerebrospinal fluid pulsations, favoring progressive dural expansion.

The diagnostic importance of this finding is also well established. Dural ectasia contributes to the systemic score of the revised Ghent nosology [2]. Previous studies have demonstrated that identification of dural ectasia can provide additional diagnostic information in patients with suspected Marfan syndrome, particularly when other manifestations are incomplete or equivocal [5].

MRI is the preferred imaging modality for evaluating dural ectasia because it provides excellent visualization of the dural sac and associated spinal abnormalities. Imaging assessment should not be restricted to dural enlargement alone. The radiologist should systematically search for posterior vertebral scalloping, osseous remodeling or erosion, widening of the neural foramina, enlargement of the nerve-root sleeves, dural diverticula, and anterior sacral meningoceles [3].

Several morphological and quantitative criteria have been proposed to improve diagnostic reproducibility. Ahn., *et al*. proposed criteria incorporating distal dural sac enlargement, anterior sacral meningocele, nerve-root sleeve enlargement, and sacral scalloping

[3]. Oosterhof., *et al*. subsequently developed a quantitative approach based on the dural sac ratio (DSR), comparing the anteroposterior diameter of the dural sac with that of the corresponding vertebral body from L1 to S1 [4].

However, criteria established predominantly in adults should be interpreted cautiously in pediatric patients. Habermann., *et al*. specifically evaluated MRI criteria in children, adolescents, and young adults with Marfan syndrome and demonstrated that the DSR at L5 and S1, together with a sagittal dural sac diameter at S1 greater than that at L4, were particularly useful parameters for detecting dural ectasia in younger patients [6]. Therefore, in children, morphological findings and quantitative measurements should be considered together rather than relying exclusively on adult thresholds.

Dural ectasia may already be prominent during childhood. Pediatric studies have demonstrated a high frequency of dural ectasia among children with Marfan syndrome undergoing spinal MRI, supporting the concept that dural involvement is not exclusively a late manifestation of the disease [7]. This observation is particularly relevant in our 9-year-old patient and emphasizes the value of recognizing dural abnormalities early in the course of the disease.

Clinical significance and potential progression

The clinical relevance of identifying dural ectasia extends beyond its contribution to the diagnosis of Marfan syndrome. Although it may remain asymptomatic for many years, dural ectasia can evolve over time. Long-term imaging studies have demonstrated progression of some morphological and quantitative features of dural ectasia, including increasing dural sac dimensions and development or progression of associated abnormalities [8,9].

Progressive enlargement may lead to secondary remodeling of adjacent vertebral structures, posterior vertebral scalloping, osseous erosion, and neural foraminal widening [3,8]. In more advanced forms, nerve-root involvement may occur, potentially resulting in low-back pain, radicular pain, sensory disturbances, or weakness.

Dural abnormalities associated with connective tissue weakness may also predispose to cerebrospinal fluid leakage. This represents an important, although uncommon, complication because persistent CSF leakage may result in spontaneous intracranial hypotension, typically presenting with orthostatic headache. This association has also been reported in pediatric patients with Marfan syndrome [10].

Therefore, identification of dural ectasia is important not simply because the dural sac is enlarged, but because MRI can detect the structural consequences and potential complications of progressive dural involvement.

This consideration is particularly important in children. Recognition of dural ectasia at an early age provides documentation of the initial extent of dural involvement and establishes an imaging baseline for subsequent comparison when clinically indicated. Given the remaining skeletal growth in pediatric patients, documenting the morphology of the dural sac before the development of significant secondary osseous abnormalities may be particularly valuable [6-9].

Importantly, dural ectasia should not be considered an inevitably progressive or dangerous neurological condition. Many affected patients remain asymptomatic, and progression is variable. Its recognition is primarily important for identifying associated complications and for establishing an appropriate baseline in patients in whom future clinical or imaging reassessment may be required.

Management

Management of dural ectasia depends mainly on clinical manifestations and associated complications rather than on the degree of dural enlargement alone. In asymptomatic patients without significant vertebral remodeling, neurological impairment, or evidence of CSF leakage, treatment is generally conservative [1].

There is currently no established medical treatment capable of reversing the underlying dural ectasia. Consequently, uncomplicated cases are usually managed with clinical observation and symptomatic treatment when required. The indication and timing of follow-up imaging should be individualized according to symptoms, associated spinal abnormalities, and clinical evolution.

Interventional treatment is mainly reserved for severely symptomatic or complicated forms. Patients presenting with significant neurological manifestations related to nerve-root or cauda equina involvement may require specialized neurosurgical evaluation. Surgical approaches have been reported in selected severe cases, but treatment must be individualized because of the intrinsic fragility of connective tissues in Marfan syndrome.

When dural abnormalities are complicated by a symptomatic CSF leak and intracranial hypotension, treatment is directed toward the leak itself. Conservative measures may initially be attempted, whereas persistent symptomatic leaks may require targeted treatment such as an epidural blood patch. Successful treatment with an autologous epidural blood patch has been reported in a pediatric patient with Marfan syndrome and intracranial hypotension secondary to dural ectasia [10]. More invasive procedures are generally reserved for persistent or refractory cases.

In our patient, the absence of significant posterior vertebral scalloping, osseous remodeling, dural diverticula, or neurological manifestations attributable to the lumbosacral ectasia supports a conservative approach. At this stage, the main value of MRI is to document the extent of dural involvement and establish a morphological and quantitative baseline, including DSR measurements from L1 to S1, for subsequent comparison if clinically indicated during growth.

Finally, evaluation of the entire spine rather than the lumbosacral region alone allowed identification of an associated craniovertebral junction abnormality characterized by basilar impression. This finding emphasizes the value of comprehensive spinal assessment when clinically indicated in children with Marfan syndrome.

Conclusion

Dural ectasia is an important spinal manifestation of Marfan syndrome that may already be evident during childhood. MRI is the imaging modality of choice, allowing both morphological and quantitative assessment and systematic evaluation for associated vertebral, foraminal, radicular, and meningeal abnormalities.

In pediatric patients, morphological findings should be interpreted together with age-appropriate quantitative parameters, particularly at the L5 and S1 levels. Early recognition is important not because dural ectasia systematically requires treatment, but because it enables identification of structural and neurological complications and provides a baseline for detecting potential changes during skeletal growth.

Most uncomplicated and asymptomatic cases can be managed conservatively. Treatment is reserved for symptomatic or complicated forms and is directed toward the specific complication, particularly neurological involvement or CSF leakage. In our patient, the absence of significant secondary structural complications supports conservative management and clinical follow-up.

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Conflict of Interest

The authors declare that they have no conflicts of interest.

Patient Consent

Written informed consent for publication of this case and the accompanying images was obtained from the patient's legal guardian.

Author Contributions

All authors contributed to the conception of the work, analysis and interpretation of the imaging findings, drafting and critical revision of the manuscript. All authors read and approved the final manuscript.

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