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Dural ectasia and recurrent spontaneous intracranial hypotension in a patient with Marfan syndrome: a case report

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Introduction
Marfan syndrome is a connective tissue disorder that may present with underrecognized neurological manifestations. In this condition, congenital fragility of the dura mater predisposes patients to dural ectasia and increases the risk of cerebrospinal fluid (CSF) leaks, ultimately leading to spontaneous intracranial hypotension (SIH). SIH typically presents with orthostatic headache; however, recurrent or chronic episodes may alter the headache pattern, mimicking chronic primary headache disorders.

Case presentation
A 31-year-old woman with Marfan syndrome presented with severe holocranial pressure-like headache, which worsened with orthostatism and physical exertion, 20 days after an ophthamological procedure. The episode lasted approximately 10 days, and brain MRI performed during this period was unremarkable. She experienced two more episodes one year and four years after the first one. She ultimately evolved to chronic daily headache and analgesics overuse. Due to Marfan syndrome and positional headache, lumbosacral spine MRI was requested. The scans demonstrated marked dural ectasia with multiple spinal meningeal diverticula. Additionally, epidural T2/STIR hyperintensity suggestive of CSF leakage was identified, consistent with the "dinosaur tail sign". She was diagnosed with intracranial hypotension secondary to Marfan syndrome.

Discussion
This case highlights the association between Marfan syndrome, dural ectasia and spontaneous CSF leaks. The classic orthostatic headache presentation may evolve to chronic, non-positional headache over time, making the diagnosis challenging. Normal cranial imaging may also contribute to diagnostic delay, and therefore investigation of the entire neuroaxis is essential in suspected cases. Untreated intracranial hypotension may lead to headache chronification and medication overuse. Early recognition is essential, as specific therapeutic strategies, including epidural blood patch and neurosurgical intervention, may improve patient outcomes.

Conclusion
Spontaneous intracranial hypotension should be considered in patients presenting with orthostatic headache, particularly in the setting of connective tissue disorders. Spinal imaging is crucial in cases with normal cranial neuroimaging. Early diagnosis may prevent headache chronification and reduce morbidity. This case highlights the importance of evaluating dural integrity in the spine of patients with Marfan syndrome and positional headache, even in the absence of typical intracranial changes.

Keywords:
Marfan syndrome
Dural ectasia
Cerebrospinal fluid leak
Intracranial hypotension
Daily headache

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