Scalloping Spine in a Patient With Type 1 Neurofibromatosis

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A 46-year-old lady known with type 1 neurofibromatosis (NF1) reported a history of mild chronic pain involving her back and limbs. She had multiple neurocutaneous stigmata of NF1 including neurofibromas. (Figure A) She exhibited myelopathic changes in her lower limbs, with increased tone and hyperreflexia noted bilaterally and symmetrically. MRI of her spine showed diffuse dural ectasia from C4-T3, causing severe scalloping of the posterior endplates. (Figure B) Dural ectasia (defined as focal dilation of the dural sac) has an unconfirmed pathogenesis. (1) NF1-associated dural ectasia is rarely reported, occurs most frequently in the thoracic region and treatment remains ill-defined. (2) Considering the extent of her radiological pathology, our patient had mild symptomatology and was managed conservatively. One should be aware of the association of NF1 and dural ectasia, and that patients may present with a subtle and non-specific clinical picture.

References


Figure 1: Panel A: The largest neurofibroma present, which was on the posterior aspect of the patient’s right arm and forearm. Panel B: A sagittal T2-FLAIR- sequence MRI spine, showing diffuse dural ectasia from C4-T3 causing scalloping of the end plates. There is an associated hyper-intense longitudinal segment at C6, suggestive of a syrinx.
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