Low back pain in a child: a rare case of intramedullary schwannoma initially resembling juvenile spondyloarthritis

Sirs,

Hip and back pain in children requires a broad and often challenging differential diagnosis, excluding mimickers of different aetiologies (inflammatory, neoplastic, infectious) (1). It is crucial to distinguish between malignancy and juvenile spondyloarthritis (jSpA), the latter being more familiar in our rheumatological setting (2). Intramedullary schwannomas are a rare cause of paediatric back pain, representing 0.3% of intraspinal tumours and 1.1% of intraspinal schwannomas (3).

We describe the interesting case of a 12-year-old Asian female who presented a sudden progressively invalidating inflammatory-like atraumatic afebrile lumbar and right hip pain, showing a partial response to NSAIDs and a completely negative x-ray examination. The pain was mainly present at nighttime and early morning, therefore exhibiting an inflammatory pattern, at least initially. Over two months, the back pain worsened, being present almost anytime, even at rest and during the night. Becoming bedridden, the patient was admitted to our ward with the initial suspicion of jSpA. The child was in an obligated supine position, and her right hip had a normal range of motion. Pain was evoked on palpation of the lumbosacral spinal processes and the sacroiliac joints; Faber test was positive, eliciting back pain, while the Schober test was not done due to her inability to stand. However, the physical examination revealed neurological signs (reduced patellar reflexes, lumbosacral pain elicited by neck flexion, and lower limbs Mingazzini's sign), raising the suspect of another possible aetiology. Routine laboratory tests (including a complete blood count, inflammatory markers, and lactate dehydrogenase) and a pelvic MRI were negative. A targeted contrast lumbosacral spine MRI showed a 3 cm in diameter oval-shaped intramedullary non-aggressive slow-growth neoformation localised at L2 level (Fig. 1). Given this finding, the patient received dexamethasone with an excellent response to the pain, and she was successfully treated with a surgical resection of the lesion, which was histologically proven to be a cellular schwannoma.

This reported case highlights the importance of an accurate medical history with particular attention to the evolution of pain over time and physical examination. Indeed, despite a similar subtle onset of both jSpA and Schwannoma, excruciating pain is uncommon in children with axial inflammatory involvement, as well as becoming bedridden (4). Furthermore, although it is not specific, night pain may represent a critical clue to diagnosing certain conditions. In patients with spondyloarthritis, the production of inflammatory cytokines during the latter part of the night is likely responsible for the low-back pain (4). On the other hand, in patients with schwannomas, intense nighttime pain is thought to be caused by factors that interfere with the dynamics of cerebrospinal fluid (CSF) production and reabsorption during the dark phase (5). A comprehensive approach involving clinical assessment, a combination of imaging techniques, and laboratory tests is essential to reach the correct diagnosis.

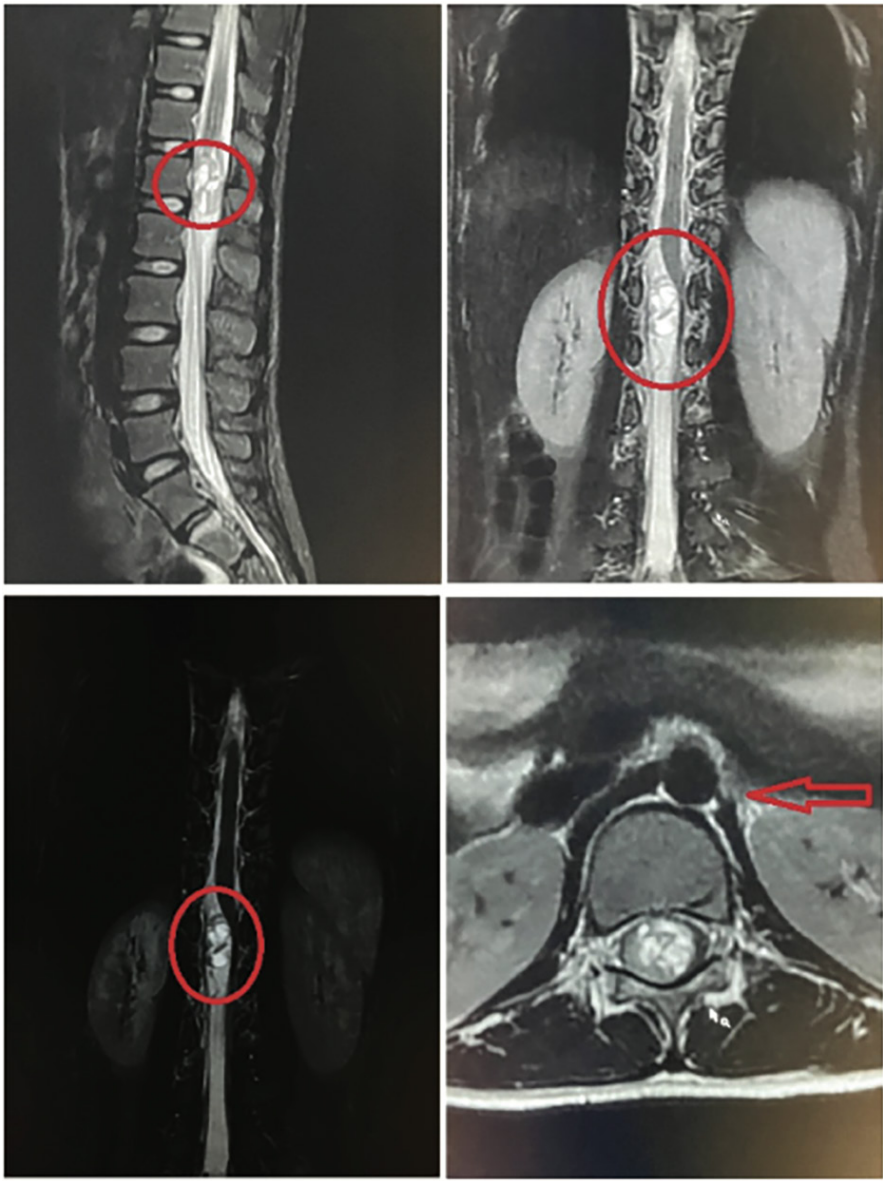

Fig. 1. Lumbosacral spine MRI. Sagittal T2 weighted image (upper left), coronal T2 weighted image (upper right), contrast-enhanced T1 weighted coronal (lower left) and axial (lower right) images.

To date, twelve paediatric cases of intramedullary schwannoma without neurofibromatosis have been reported (7-9). Schwannoma typically presents as a soft tissue mass with non-specific pain and/or focal neurological findings. If multiple Schwannomas are present, they are likely associated with neurofibromatosis type 2. The diagnosis involves imaging modalities (MRI) alongside biopsy for confirmation (9). Red flags for the diagnosis of schwannoma are persistent nighttime pain, abnormal neurological examination, and antalgic obligated supine position (1, 6).

Despite being extremely rare, intramedullary schwannoma should be considered among the differential diagnoses in paediatric low back pain. Although the clinical presentation at the very beginning of this case might have similarities with jSpA, a thorough neurological examination and the evaluation of the pain course allowed us to reach a definite and rather infrequent diagnosis.

L. Giudice[1,2], *MD**
G. Guida[3,4], *MD**
S. Costi[5], *MD*
A. Marino[5], *MD, PhD*
C.B. Chighizola[1,5], *MD, PhD*
R.F. Caporali[1,2], *MD*

**These authors contributed equally.*

[1]*Department of Clinical Sciences and Community Health, Università degli Studi di Milano, Milan,*
[2]*Rheumatology Unit, ASST G. Pini-CTO, Milan,*
[3]*Department of Biomedical and Clinical Science, Università degli Studi di Milano, Milan,*
[4]*Paediatric Department, Vittore Buzzi Children's Hospital, Milan,*
[5]*Paediatric Rheumatology Unit, ASST G. Pini-CTO, Milan, Italy.*

The work should be attributed to: Paediatric Rheumatology Unit, ASST G. Pini-CTO, Milan and the Department of Clinical Sciences and Community Health, Università degli Studi di Milano, Milan, Italy.

Please address correspondence to:
Achille Marino
Clinica Reumatologica Pediatrica,
ASST G. Pini-CTO,
Via Gaetano Pini 9,
20122 Milano, Italy.
E-mail: achillemarino6@gmail.com

Competing interests: none declared.

References

1. LAMB M, BRENNER JS: Back pain in children and adolescents. *Pediatr Rev* 2020; 41(11): 557-69. https://doi.org/10.1542/pir.2019-0051
2. CIVINO A, ALIGHIERI G, PRETE E *et al*.: Musculoskeletal manifestations of childhood cancer and differential diagnosis with juvenile idiopathic arthritis (ONCOREUM): a multicentre, cross-sectional study. *Lancet Rheumatol* 2021; 3(7): e507-e516. https://doi.org/10.1016/S2665-9913(21)00086-2
3. HAYASHI F, SAKAI T, SAIRYO K *et al*.: Intramedullary schwannoma with calcification of the epiconus. *Spine J* 2009; 9(5): e19-23. https://doi.org/10.1016/j.spinee.2008.11.006
4. FISHER C, CIURTIN C, LEANDRO M, SEN D, WEDDERBURN LR: Similarities and differences between juvenile and adult spondyloarthropathies. *Front Med* (Lausanne) 2021; 8: 681621. https://doi.org/10.3389/fmed.2021.681621
5. STEFFENSEN AB, EDELBO BL, BARBUSKAITE D *et al*.: Nocturnal increase in cerebrospinal fluid secretion as a circadian regulator of intracranial pressure. *Fluids Barriers CNS* 2023; 20(1): 49. https://doi.org/10.1186/s12987-023-00451-2
6. WEISS JE, STINSON JN: Pediatric pain syndromes and noninflammatory musculoskeletal pain. *Pediatr Clin North Am* 2018; 65(4): 801-26. https://doi.org/10.1016/j.pcl.2018.04.004
7. LANDI A, GRASSO G, GREGORI F, IACOPINO G, RUGGERI A, DELFINI R: Isolated pediatric intramedullary schwannoma: case report and review of literature. *World Neurosurg* 2018; 115: 417-20. https://doi.org/10.1016/j.wneu.2018.04.220
8. WANG K, ZHAO J, ZHANG Y, SU Y: Pediatric intramedullary schwannoma with syringomyelia: a case report and literature review. *BMC Pediatr* 2018; 18(1): 374. https://doi.org/10.1186/s12887-018-1341-2
9. YAZOL M, DERINKUYU BE, BOYUNAGA O: Three different faces of Schwannoma in pediatric patients. *Curr Med Imaging* 2023 Apr 14. https://doi.org/10.2174/1573405620666230414115555