Sd. Guillain-Barré atípico

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Sección: Neuropediatría
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Caso clínico

- Niña 24 meses con cojera izquierda de 5 días de evolución
- Cuadro catarral desde hace una semana
- Dificultades para cambio posición (tumbada a sentada)
- Valorada UPED en dos ocasiones: diagnóstico de sinovitis transitoria de cadera
- Tratamiento: ibuprofeno. sin mejoría

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Caso clínico

- AP: sin interés. Desarrollo psicomotor normal.
- AF: sin interés

Caso clínico

Exploración física:

- BEG. Activa y reactiva. Dos manchas café con leche en tórax
- ORL: faringe levemente hiperémica
- ACP: normal
- Abdomen: normal
- Locomotor: no limitación movilidad caderas ni rodillas. Masas musculares normales. No corrección lordosis lumbar ni dolor a la palpación apófisis espinosas. Imposibilidad para cambios posturales de forma espontánea (sentada y bipedestación) por dolor
- NRL: difícil valorar, por poca colaboración. Glasgow 15. PICNR, PPCC normales. Movilidad simétrica de las 4 extremidades. Marcha algo inestable por debilidad y dolor, no atáxica. No cojera. ROTs normales. No dismetría, no temblor

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- AS:
  - Hemograma normal.
  - Iones, transaminasas, CK, PCR normal.

- Ecografía caderas: normal

INGRESO

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Caso clínico

- **Sospecha diagnóstica:**

  Espondilodiscitis

  - Se solicita RMN columna

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Caso clínico

- **Diagnóstico diferencial:**
  - Afectación muscular: miositis
  - Afectación neuromuscular: botulismo, miastenia gravis
  - Afectación nervio periférico: neuropatía tóxica, porfiria, difteria
  - Afectación motoneurona anterior: poliomielitis
  - Afectación médula espinal: compresión medular, mielitis transversa

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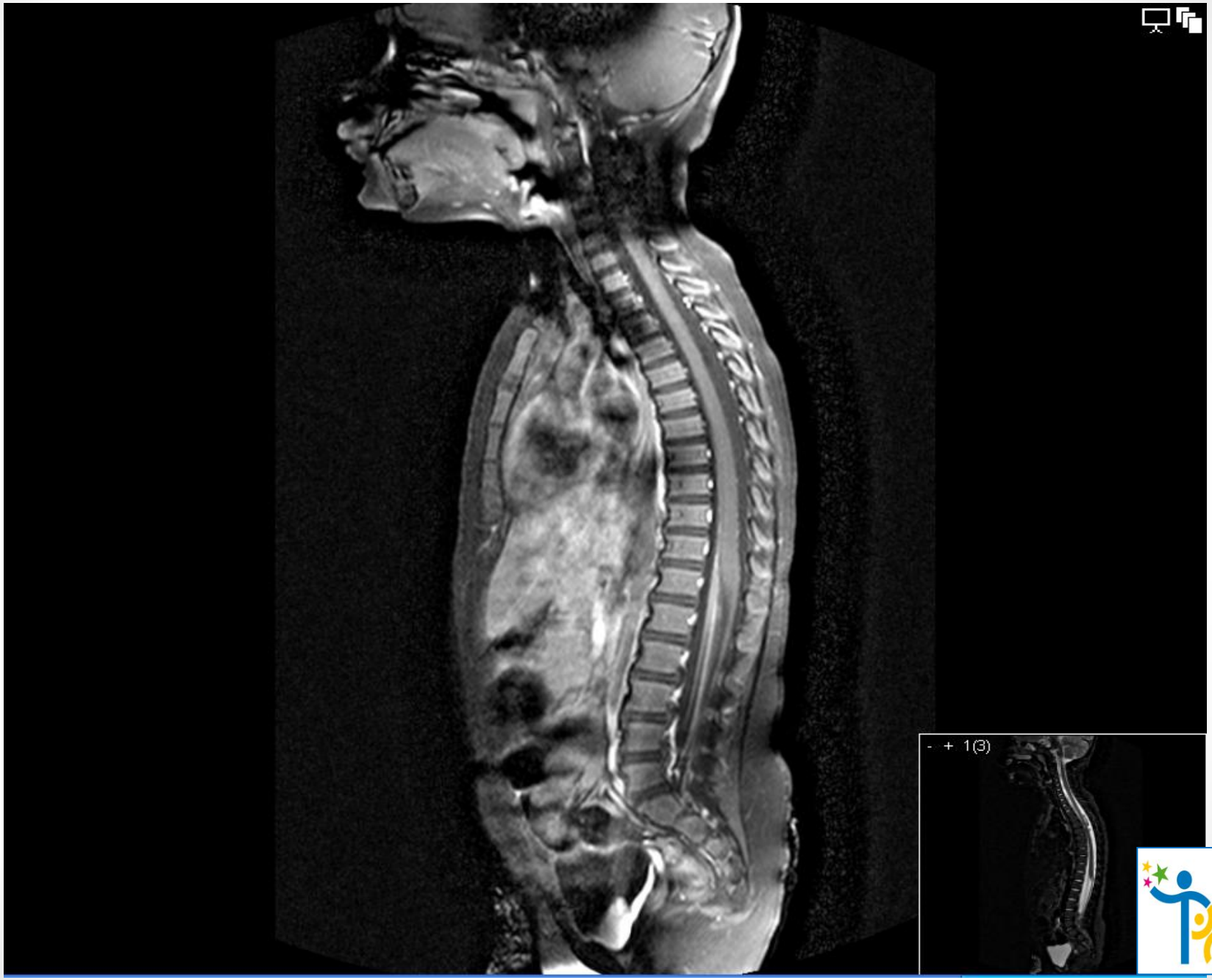
- RMN:
  - Cuerpos vertebrales y discos intervertebrales sin alteraciones
  - Canal raquídeo de calibre normal
  - Médula espinal de morfología e intensidad normal
  - **Captación intensa de la cola de caballo tras la inyección de contraste, compatible con polirradiculitis**



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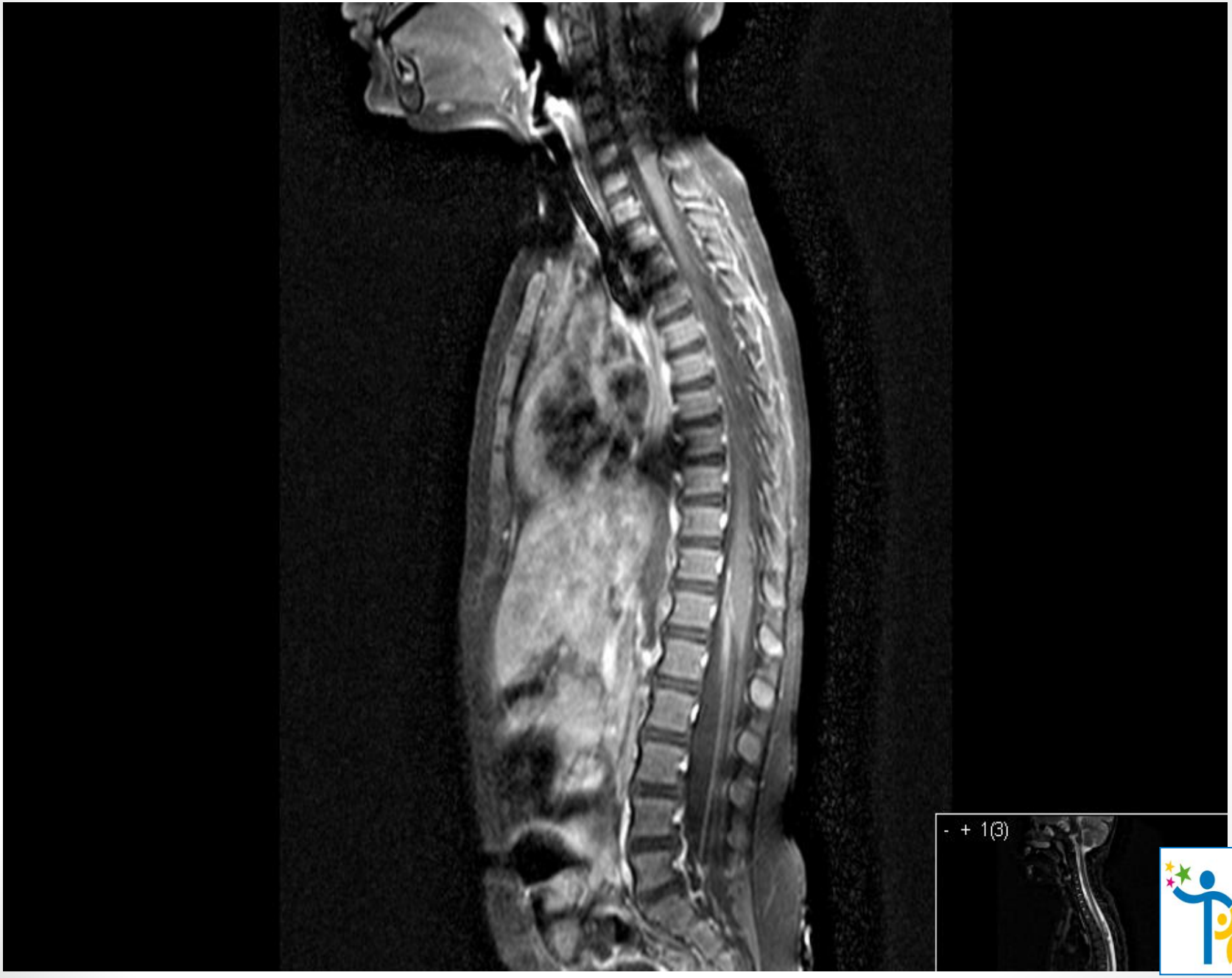

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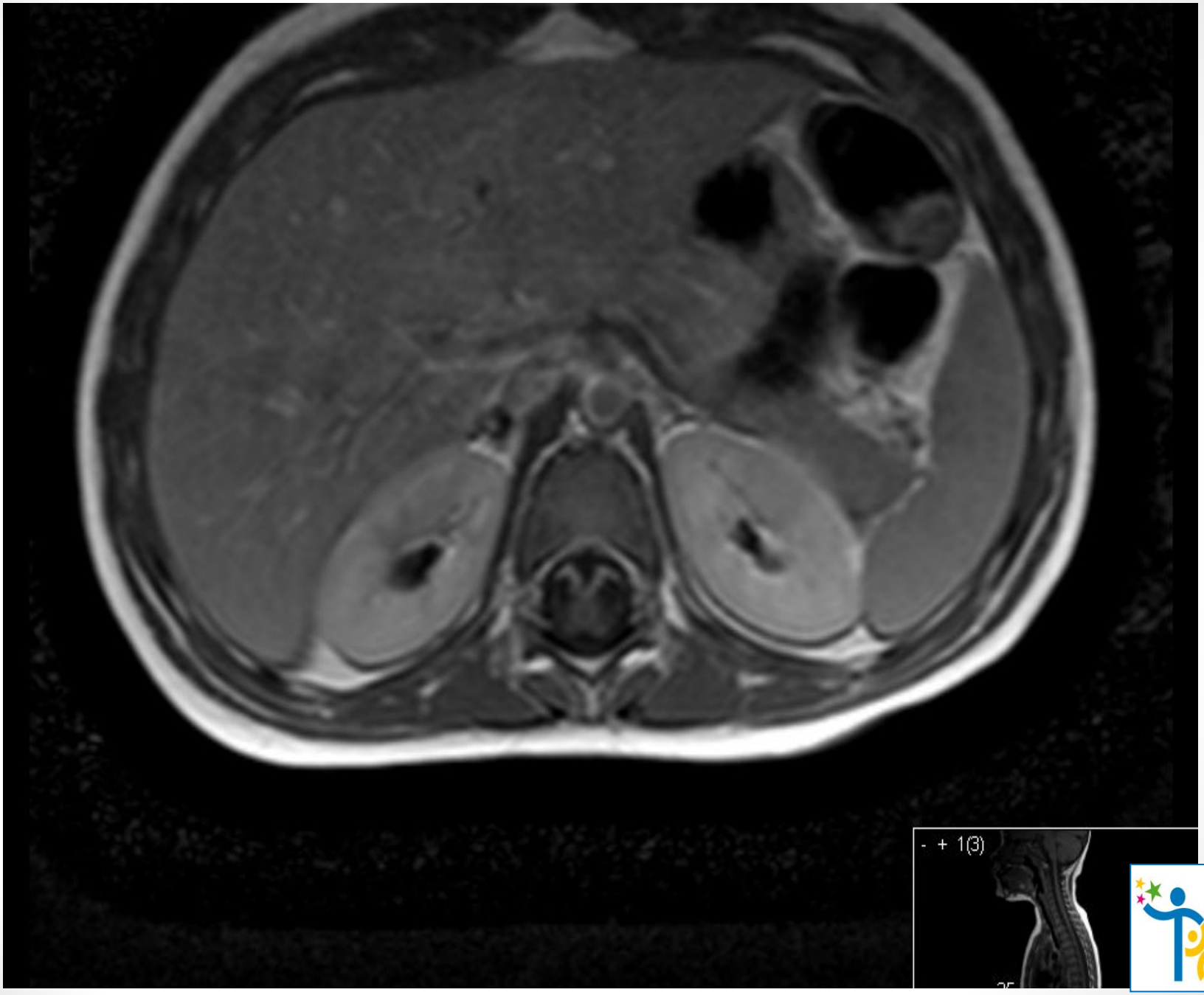

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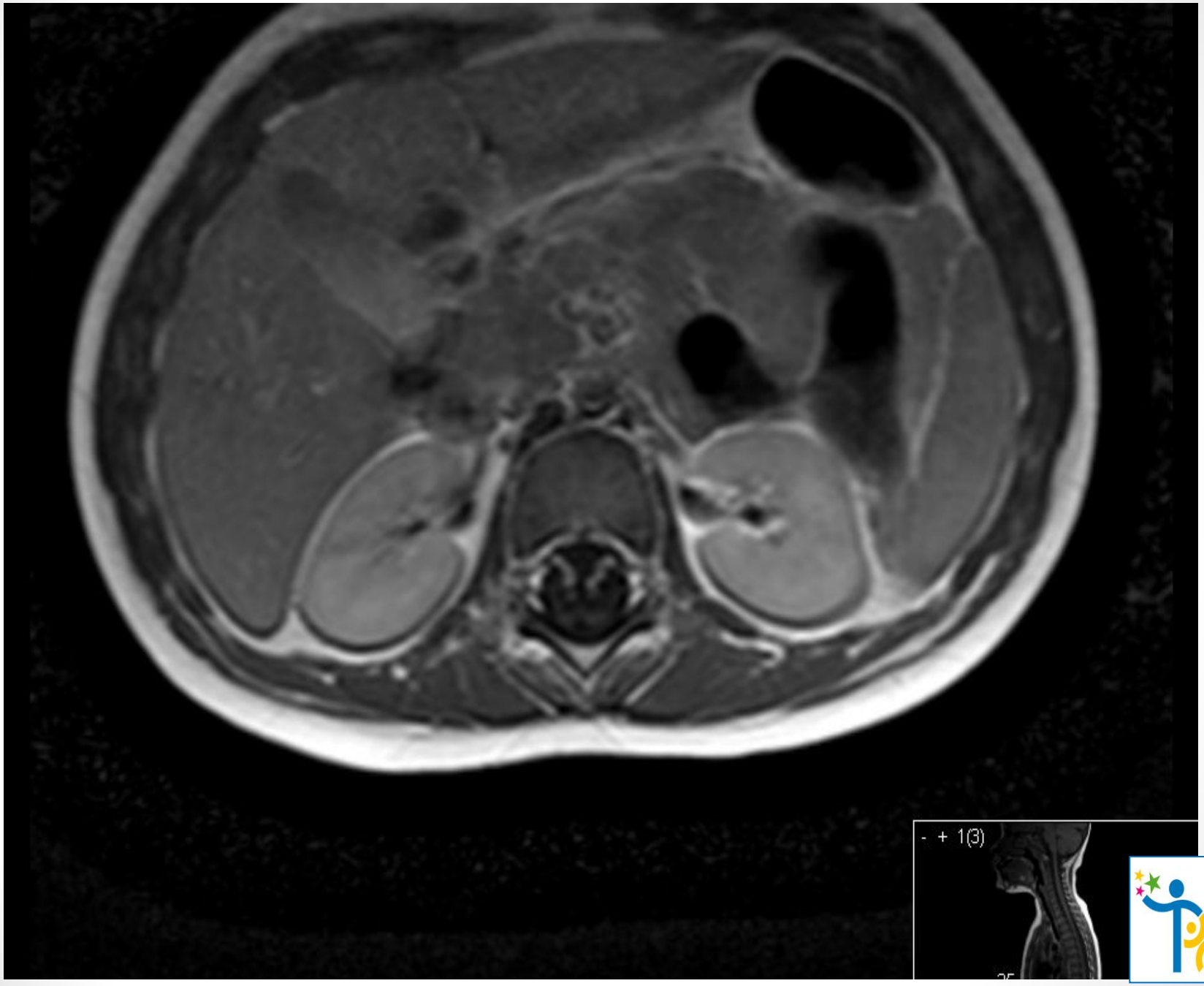

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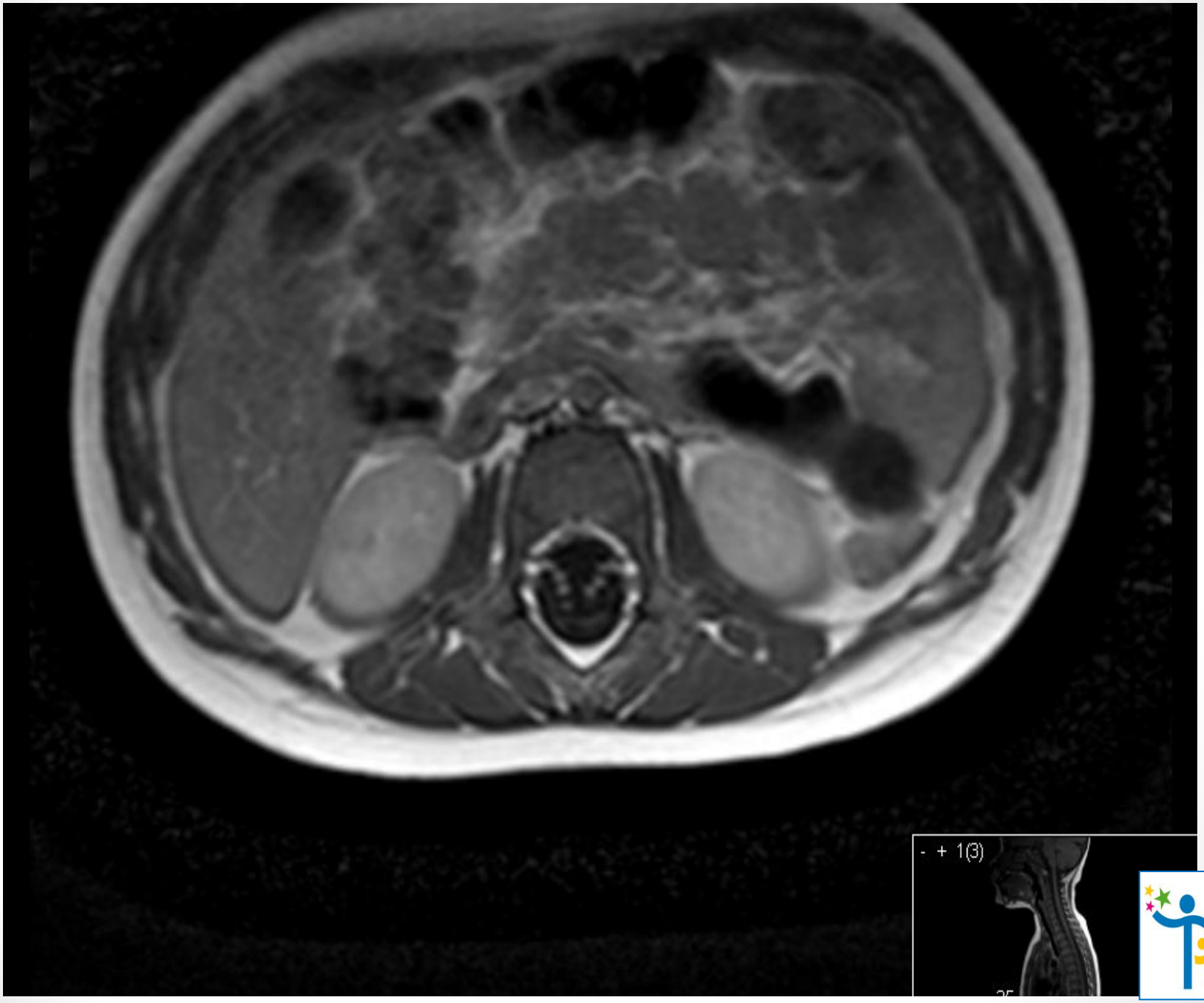

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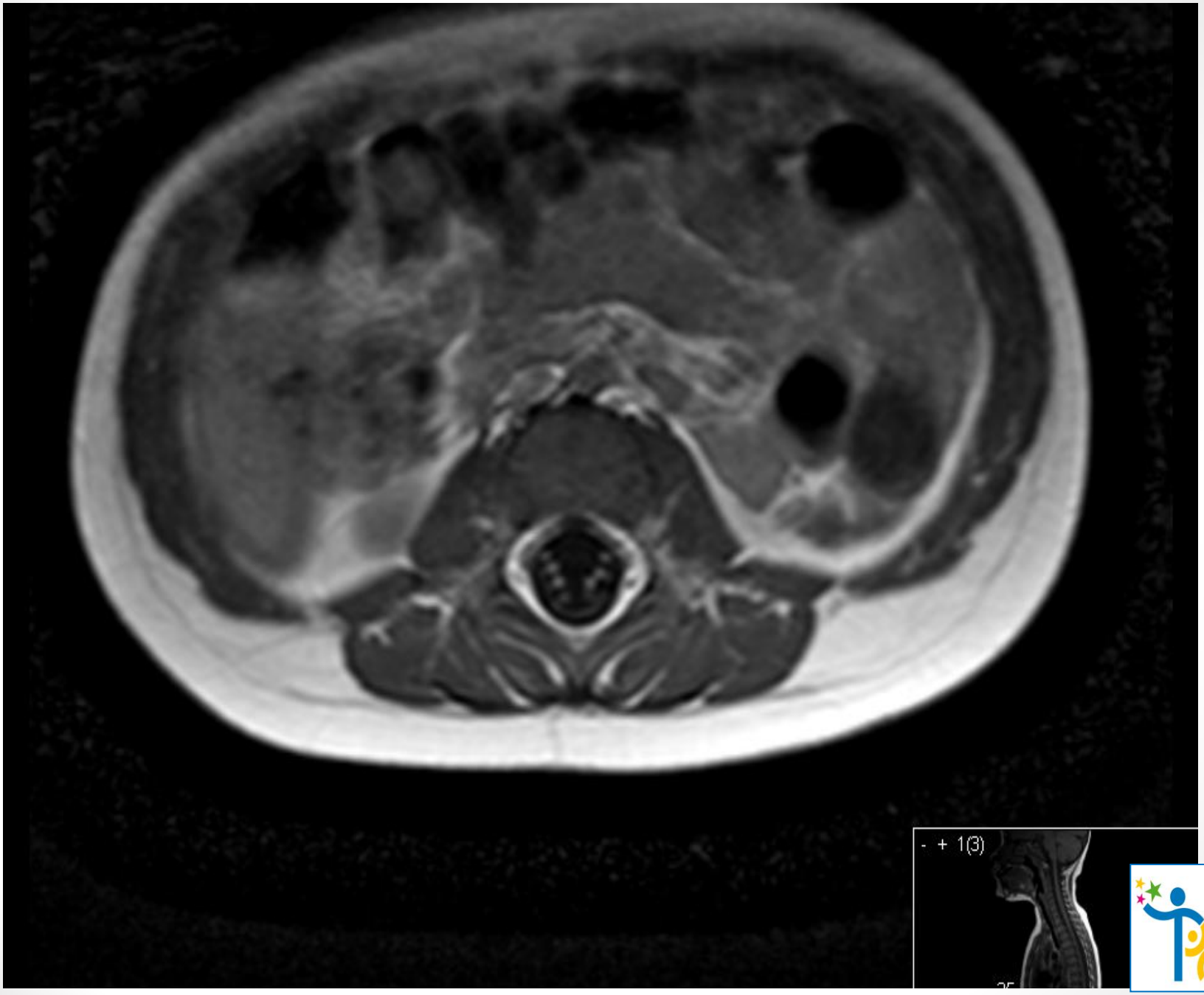

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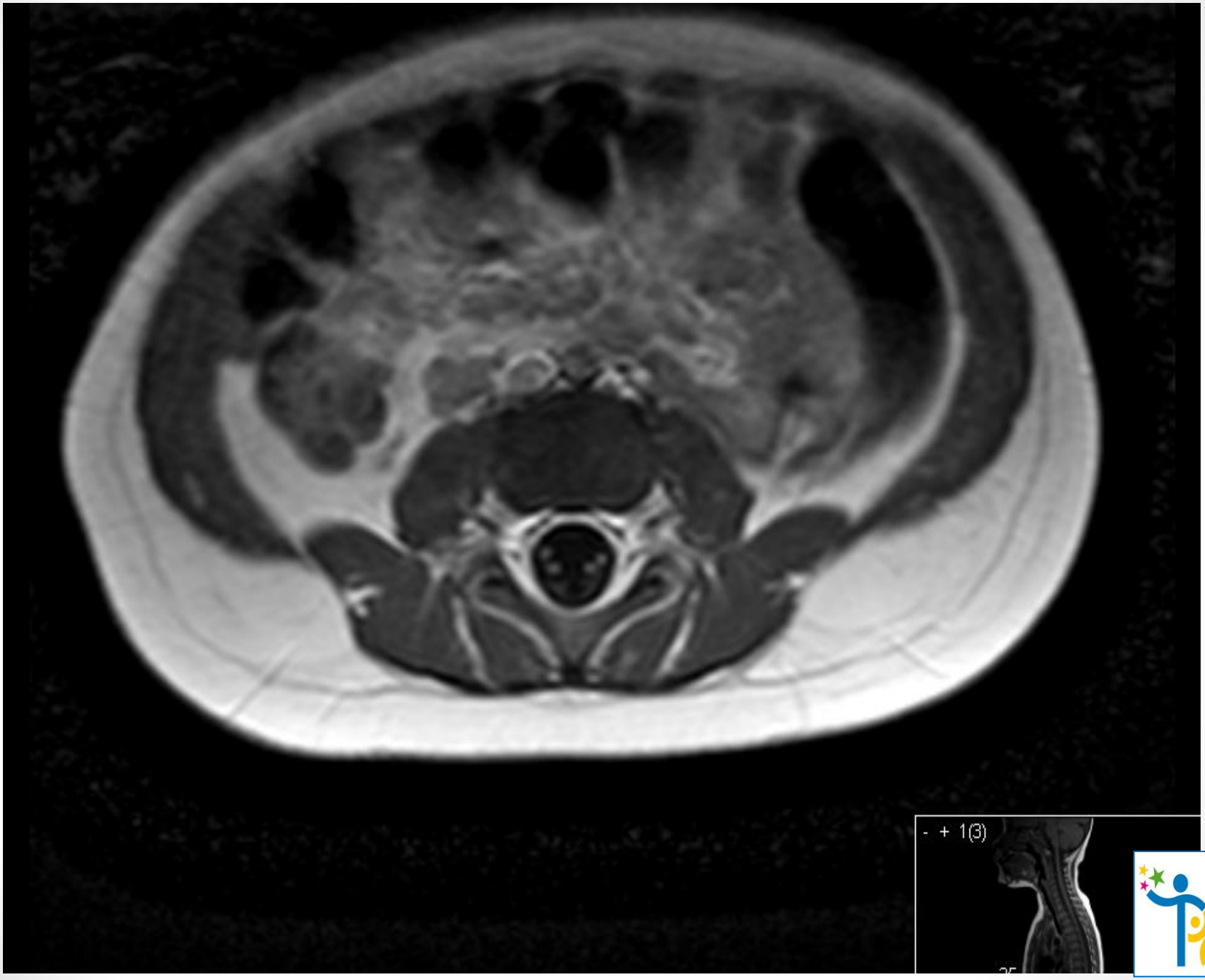

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Caso clínico

- **Evolución:**
  - Empeoramiento progresivo marcha, con incapacidad deambulación sin apoyo y caídas por claudicación (9ºd)
  - Precisa apoyo para mantener bipedestación (10ºd)
  - Hipotonía MMII y cintura escapular
  - ROTs normales
  - No afectación pares craneales

Caso clínico

- Se solicitan:
  - Serologías Campylobacter, Mycoplasma, etc
  - EMG
  - PL

Caso clínico

- EMG:
  - Estudio dentro de límites normales
  - Ausencia potenciales evocados somatosensoriales en músculo tanto MS como MI (que si se obtienen desde columna), aunque no es definitivamente patológico a esta edad
  - Dado los resultados de RMN se recomienda realización PL
- PL: Disociación albúmino-citológica (proteínas 98 mg/dl, ñeucocitos 8/ul)
- Serología: negativa
- Ac. Antigangliósidos: negativos

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Caso clínico

- **Resumen:**
  - Niña 2 años con debilidad MMII progresiva, con incapacidad para la marcha
  - ROTs presentes
  - RMN: Polirradiculitis cola caballo
  - EMG "normal"
  - Disociación albúmino-citológica en LCR

Sd. Guillain-Barré atípico

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- El 10ºdía, se inició tratamiento con Igs, 2g/kg
- Inicia recuperación 3-4 días tras infusión de Igs
- Alta a los 21 días, con marcha independiente mínimamente inestable

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Sd. Guillain-Barré

- Polineuropatía aguda inflamatoria
- Etiología autoinmune
- Incidencia: 0.4-1.3 casos /100.000 niños <15 años
- Forma clásica:
  - Debilidad muscular simétrica progresiva y ascendente
  - Disminución y/o ausencia ROTs
- Afectación sensitivo-autonómica (dolor, parestesias, HTA…)
- Antecedentes infección respiratoria o intestinal semanas previas (Campylobacter jejuni)
- Hasta 10% pueden cursar con ROTs normales o exaltados

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ORIGINAL COMMUNICATION

Guillain–Barré syndrome associated with normal or exaggerated tendon reflexes

Nobuhiro Yuki · Norito Kokubun · Satoshi Kuwabara · Yukari Sekiguchi · Masafumi Ito · Masaaki Odaka · Koichi Hirata · Francesca Notturno · Antonino Uncini

Abstract Areflexia is part one of the clinical criteria required to make a diagnosis of Guillain–Barré syndrome (GBS). The diagnostic criteria were stringently developed to exclude non-GBS cases but there have been reports of patients with GBS following *Campylobacter jejuni* enteritis with normal and exaggerated deep tendon reflexes (DTRs). The aim of this study is to expand the existing diagnostic criteria to preserved DTRs. From the cohort of patients referred for anti-ganglioside antibody testing from hospitals throughout Japan, 48 GBS patients presented with preserved DTR at admission. Thirty-two patients had normal or exaggerated DTR throughout the course of illness whereas in 16 patients the DTR became absent or diminished during the course of the illness. IgG antibodies against GM1, GM1b, GD1a, or GalNAc-GD1a were frequently present in either group (84 vs. 94%), suggesting a close relationship between the two groups. We then investigated the clinical and laboratory findings of 213 GBS patients from three hospital cohorts. In 23 patients, eight presented with normal tendon reflexes throughout the clinical course of the illness. Twelve showed hyperreflexia, with at least one of the jerks experienced even at nadir, and exaggerated reflexes returning to normal at recovery. The other three had hyperreflexia throughout the disease course. Compared to 190 GBS patients with reduced or absent DTR, the 23 DTR-preserved patients more frequently presented with pure motor limb weakness (87 vs. 47%, $p = 0.00026$), could walk 5 m independently at the nadir (70 vs. 33%, $p = 0.0012$), more frequently had antibodies against GM1, GM1b, GD1a, or GalNAc-GD1a (74 vs. 47%, $p = 0.014$) and were more commonly diagnosed with acute motor axonal neuropathy (65 vs. 34%, $p = 0.0075$) than with acute inflammatory demyelinating polyneuropathy (13 vs. 43%, $p = 0.0011$). This study demonstrated that DTRs could be normal or hyperexcitable during the entire clinical course in approximately 10% of GBS patients. This possibility should be added in the diagnostic criteria for GBS to avoid delays in diagnosis and effective treatment to these patients.

Keywords Guillain–Barré syndrome · Acute motor axonal neuropathy · Acute inflammatory demyelinating polyneuropathy · Anti-ganglioside antibody · *Campylobacter jejuni* · Tendon reflex

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Sd. Guillain-Barré

- **Formas clínicas:**
  - *PDIA*: afectación vaina mielina. Más frecuente Europa y EEUU
  - *NAMA*: afectación axonal motora. Más frecuente China y Sudamérica
  - *NASMA*: afectación axonal sensitiva y motora
  - *Miller-Fisher*: oftalmoplejia, ataxia y arreflexia
  - *Otras*: saltatoria, faringo-cervico-braquial, etc.

Sd. Guillain-Barré

- **Diagnóstico:**
  - Criterios clínicos
  - Laboratorio:
    - LCR: disociación albúmino-citológica (criterio indispensable)
    - Ac antigangliósidos
  - EMG: puede ser normal al inicio de la enfermedad
  - ± Imagen: RMN

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Sd. Guillain-Barré

Nerve Root Enhancement on Spinal MRI in Pediatric Guillain-Barré Syndrome

Sarah B. Mulkey, MD*, Charles M. Glasier, MD†, Bassem El-Nabbout, MD‡, William D. Walters, MD, MPH*, Christian Ionita, MD*, Michael H. McCarthy, BA*, Gregory B. Sharp, MD*, and Rolla M. Shbarou, MD*

Guillain-Barré syndrome diagnosis is based on clinical presentation and supportive diagnostic testing. In its early stage, no single, reliable diagnostic test is available. However, a finding of nerve root enhancement on spinal magnetic resonance imaging may be useful. We evaluated the frequency of nerve root enhancement on spinal magnetic resonance imaging in children with Guillain-Barré syndrome. At a single tertiary pediatric center, we conducted a retrospective chart review of children with Guillain-Barré syndrome who had complete spinal or lumbosacral spinal magnetic resonance imaging with gadolinium administration from January 2002-January 2009. Twenty-four consecutive patients were identified. Spinal nerve root enhancement with gadolinium was present in 92% (22/24) of children with Guillain-Barré syndrome on initial spinal magnetic resonance imaging (95% confidence interval, 0.745-0.978). This finding increased to 100% of patients, after two patients underwent repeat spinal magnetic resonance imaging that did reveal nerve root enhancement. Patterns of enhancement were variable, but involved the thoracolumbar nerve roots in all patients. Enhancement of nerve roots with gadolinium on initial spinal magnetic resonance imaging was frequently present in these children with Guillain-Barré syndrome. Spinal magnetic resonance imaging is a sensitive diagnostic test and should be considered an additional diagnostic tool in select cases. Published by Elsevier Inc.

Mulkey SB, Glasier CM, El-Nabbout B, Walters WD, Ion-

Introduction

Guillain-Barré syndrome is an autoantibody-mediated cause of acute motor weakness in children and adults, and is found worldwide [1]. The reported incidence rate is 0.6-4 cases per 100,000 people per year, but this rate may be slightly lower in children [2,3].

A diagnosis of Guillain-Barré syndrome is based on clinical signs, and is supported by testing. Traditional diagnostic tools include lumbar puncture to reveal elevated cerebrospinal fluid protein, and nerve conduction studies to evaluate for conduction blocks or slowing. Cerebrospinal fluid protein typically increases after the first week of illness, but may be at a normal level at the time of presentation [4]. Nerve conduction studies can also be helpful, but like cerebrospinal fluid protein levels, may also indicate normal results during initial presentation [5]. Therefore, neither of these tests is highly sensitive early in the course of the disease. Moreover, pediatric nerve conduction studies are not readily available in some hospitals.

Studies in pediatric and adult patients demonstrate a >90% sensitivity of nerve root enhancement on spinal magnetic resonance imaging in typical cases of Guillain-Barré syndrome. However, information on this finding in children is limi [6-8]. Two previous studies investigated a combined t of 17 pediatric-aged patients [6,7]. Our intent was conduct a retrospective study of the largest consecu series of pediatric patients with Guillain-Barré syndro evaluating the frequency of nerve root enhancement spinal magnetic resonance imaging. We sought

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Conclusiones

- SGB es la causa más frecuente de parálisis flácida aguda en pediatría
- Presencia ROTs es rara, aunque no excluye el diagnóstico
- LCR criterio indispensable para el diagnóstico
- EMG puede ser normal al inicio del cuadro
- RMN puede ser útil en casos dudosos

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Las personas mayores nunca comprenden nada por sí mismas y es muy aburrido para los niños tener que darles una y otra vez explicaciones.

El Principito
Antoine de Saint-Exupéry, 1943

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