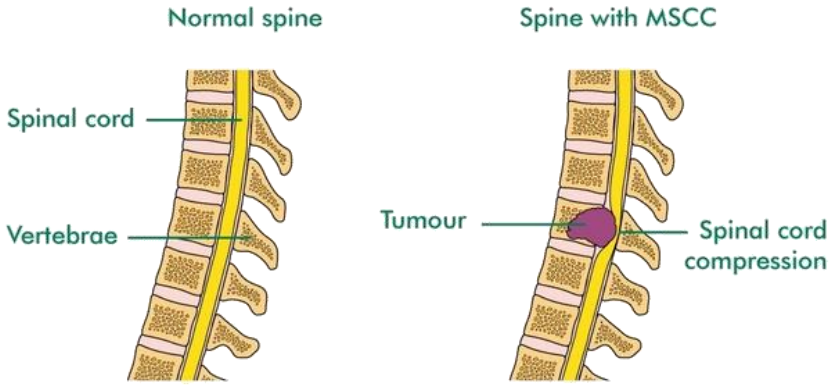
European Reference Network
for rare or low prevalence complex diseases

Network Paediatric Cancer (ERN PaedCan)

SPINAL CORD COMPRESSION: HOW TO TREAT



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ERN PaedCan – Young SIOPE webinar series

"Most challenging cases in paediatric oncology"

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Conflict of interest

Nothing to disclose

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Case 1

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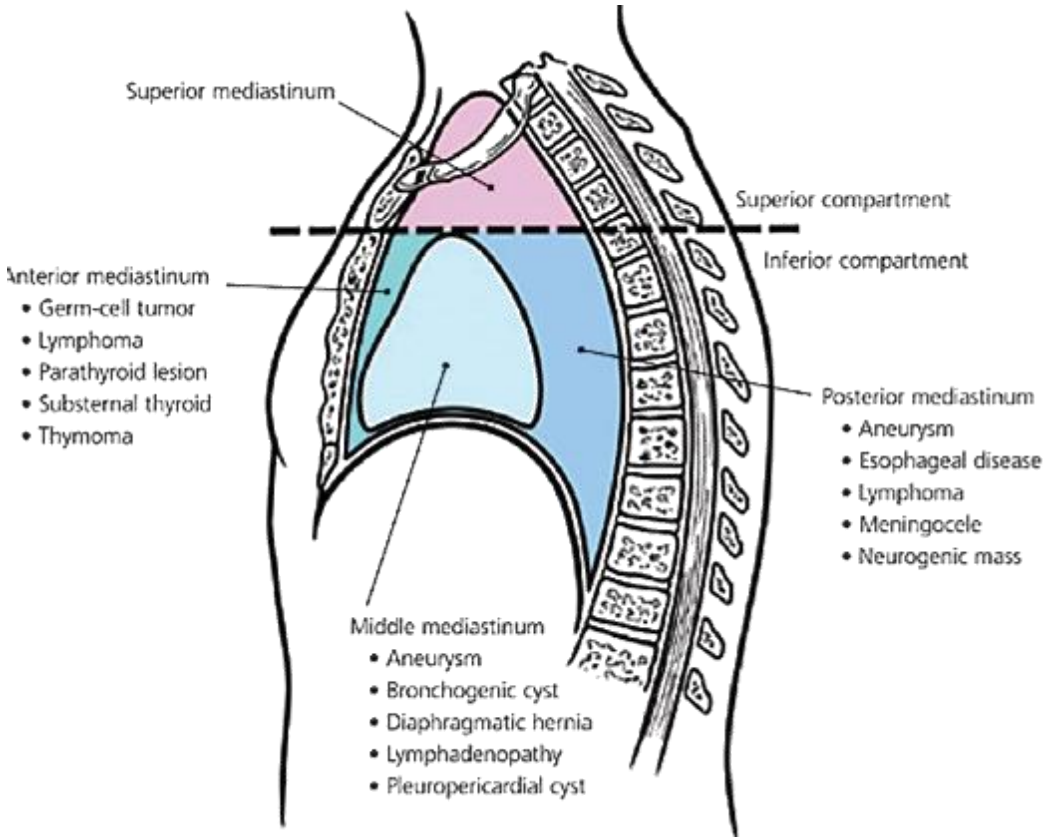
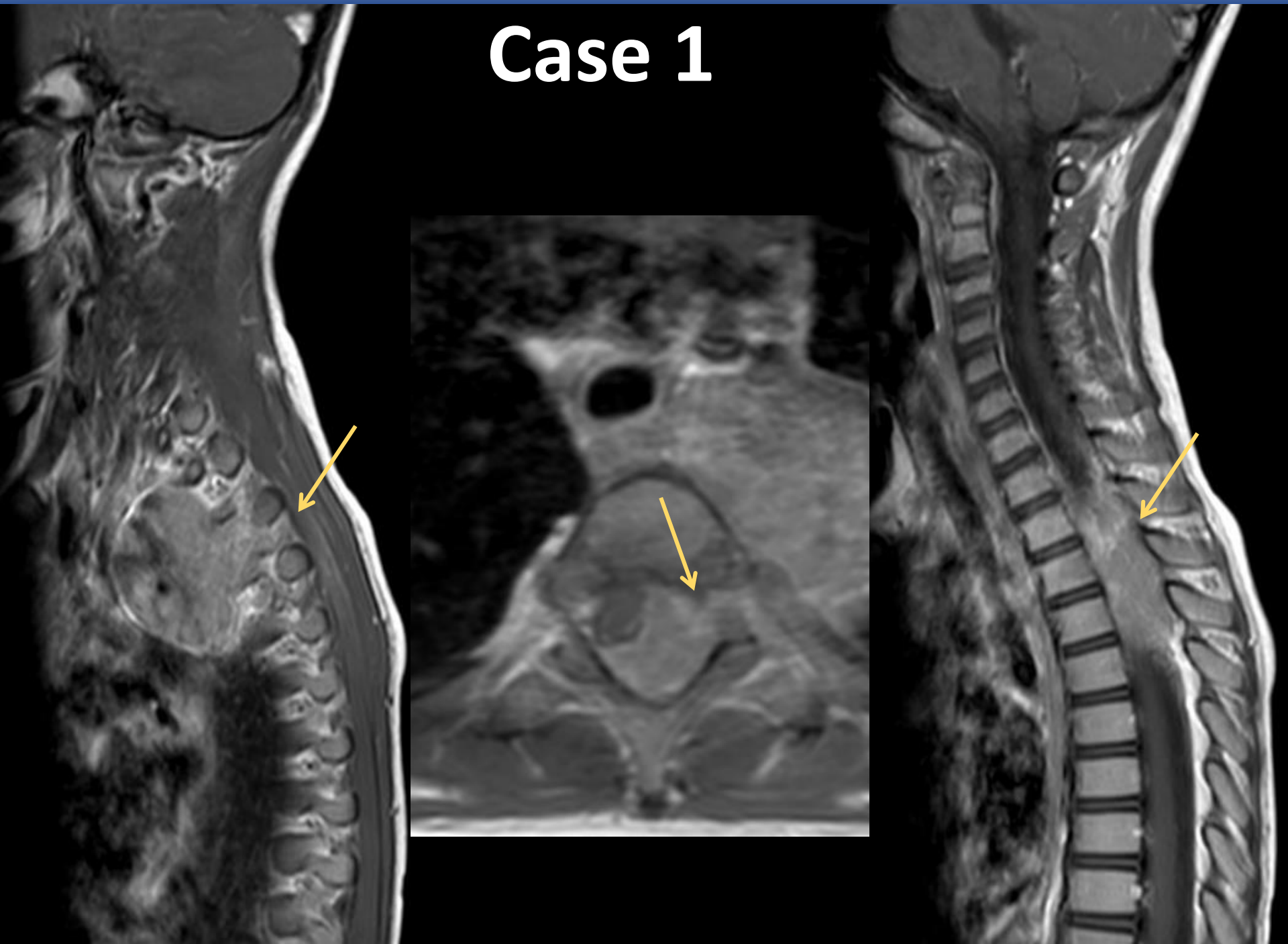
- **4 year-old boy** without any medical history
- 1 month of back **pruritus** + progressive flexion posture
- Initial neurological exam was normal
- Thorax XR and TC showed a **posterior mediastinal mass** with canal invasion from T3 to T5



Case 1



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Case 1

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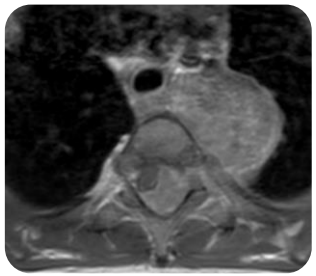

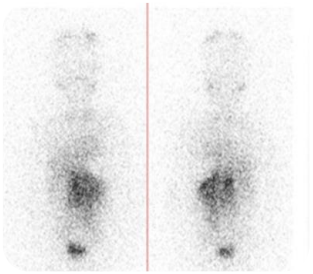

D - 5

- Flexion posture and **pruritus**
- MR > 50% canal invasion

D - 1

- Progressive walking refusal and tiptoe walk
- **Inability to walk, Bilateral Babinski sign and absence of reflexes**
- MIBG injection

D 0

- **Pain, enuresis and weakness (< 24 h) -> T8 paraplegia**

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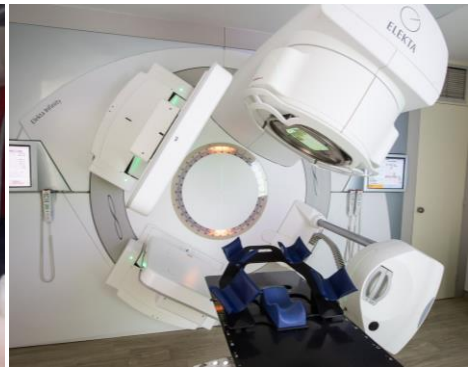
What would you do now?

1. Urgent surgical decompression
2. Tumour sample to decide how to proceed
3. Empiric chemotherapy with cyclophosphamide + vincristine +/- etoposide
4. Urgent radiotherapy



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Case 1

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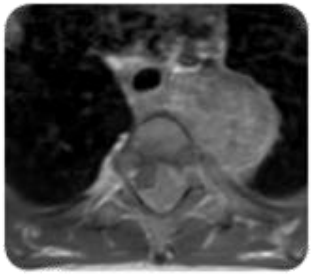

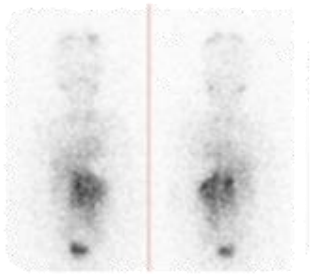

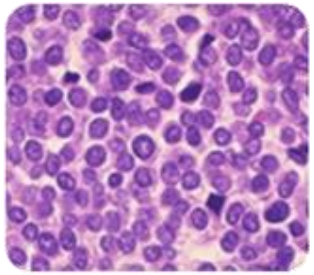

D - 5

- Flexion posture and **pruritus**
- MR > 50% canal invasion

D - 1

- Progressive walking refuse and tiptoe walk
- **Inability to walk, Bilateral Babinski sign and absent of reflexes**
- MIBG injection

D 0

- **Pain, enuresis, paralysis** (< 24 h)
- MIBG: negative
- Biopsy: **small blue round cell tumour**
- **VC**: Vincristine 2 mg/m² + CF 1.2 g/m²

D + 1

- T8 paraplegia
- **EWS-FLI 1** translocation
- **CAV** (D 2): CF 2.1 gr/m² + Doxorubicin 25 mg/m² x 3 d

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Case 1

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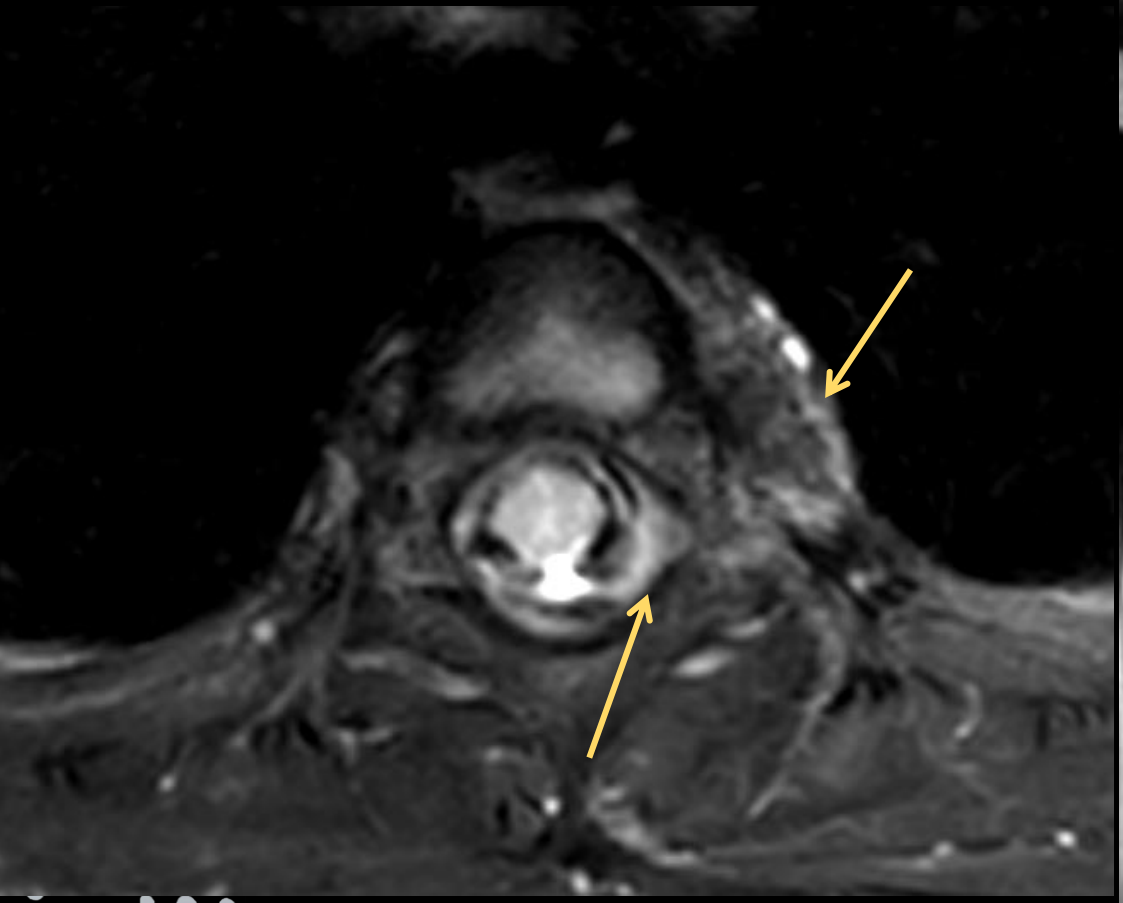
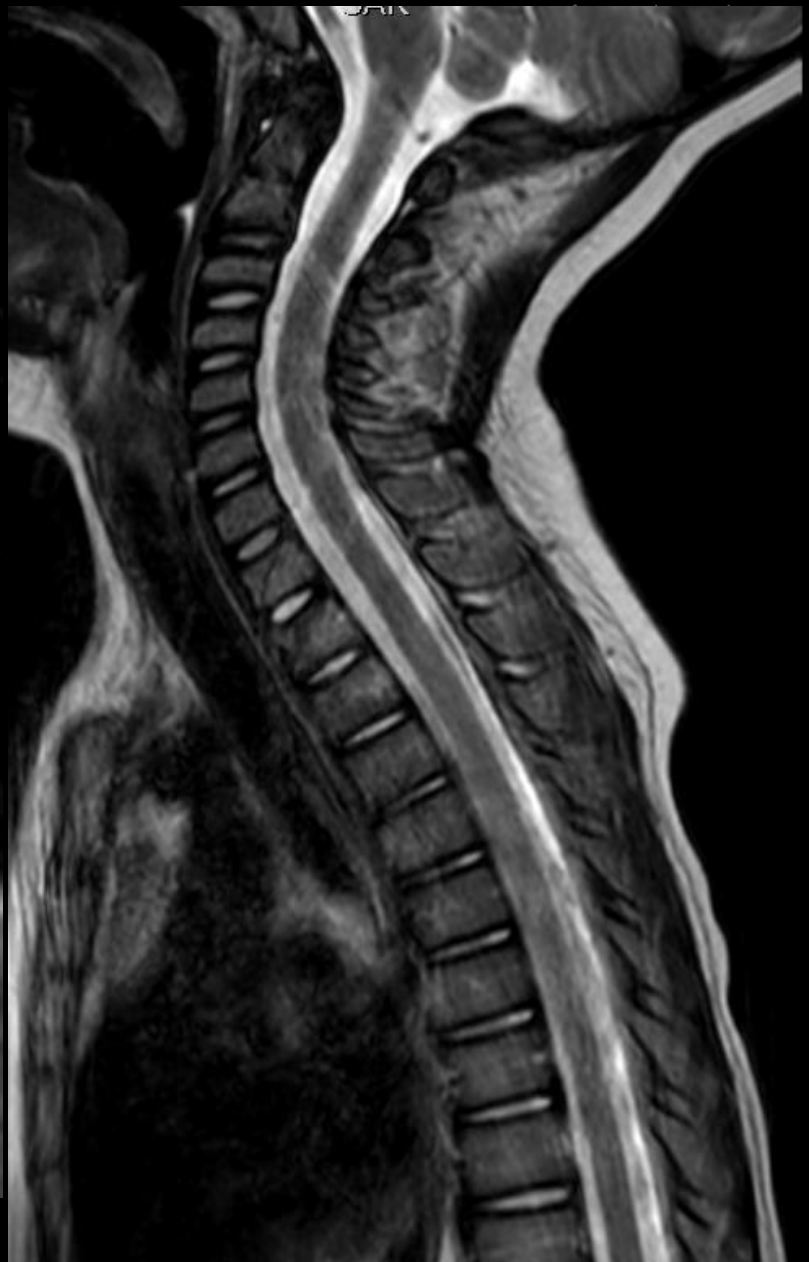
D 0	**T8 paraplegia**: Pain, enuresis and weakness (< 24 h)
D + 3	Initial recovery of low extremities movements
D + 17 C1 (D +2 C2)	Walking with assistance, sphincter control, neutral Babinski.
< 2 month (post C3)	Nearly complete recovery with persistent mild loss of proprioception. MR

Case 1

MR after 3 cycles

Neurological recuperation





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Case 1

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D 0	**T8 paraplegia**: Pain, enuresis and weakness (< 24 h)
D + 3	Start recovery of low extremities movements
D + 17 C1 (D +2 C2)	Walking with assistance, sphincter control, neutral Babinsky.
< 2 month (post C3)	Nearly complete recovery with persistent mild loss of proprioception. MR
Complete **GEIS 21-SR** protocol	5 cycles (CAV - CAV - IE - CAV – IE) + RT EOT evaluation (6 m later): residual mass (2 x 0.5 x 2.5 cm)
2 y and 11 month	CR, no daily life limitations Mild loss of proprioception

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Case 2

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8 year-old girl with **relapsed Ewing sarcoma (upper thoracic)**

First line: **EuroEwing 2012 trial** including right **thoracotomy** (tumour resection + 3rd-4th ribs) + **radiotherapy**

EOT: **local recurrence** -> **surgery** + chemotherapy in **rEEcur** trial (IT x 6 cycles) + **HD-melphalan/VP** + **RT**

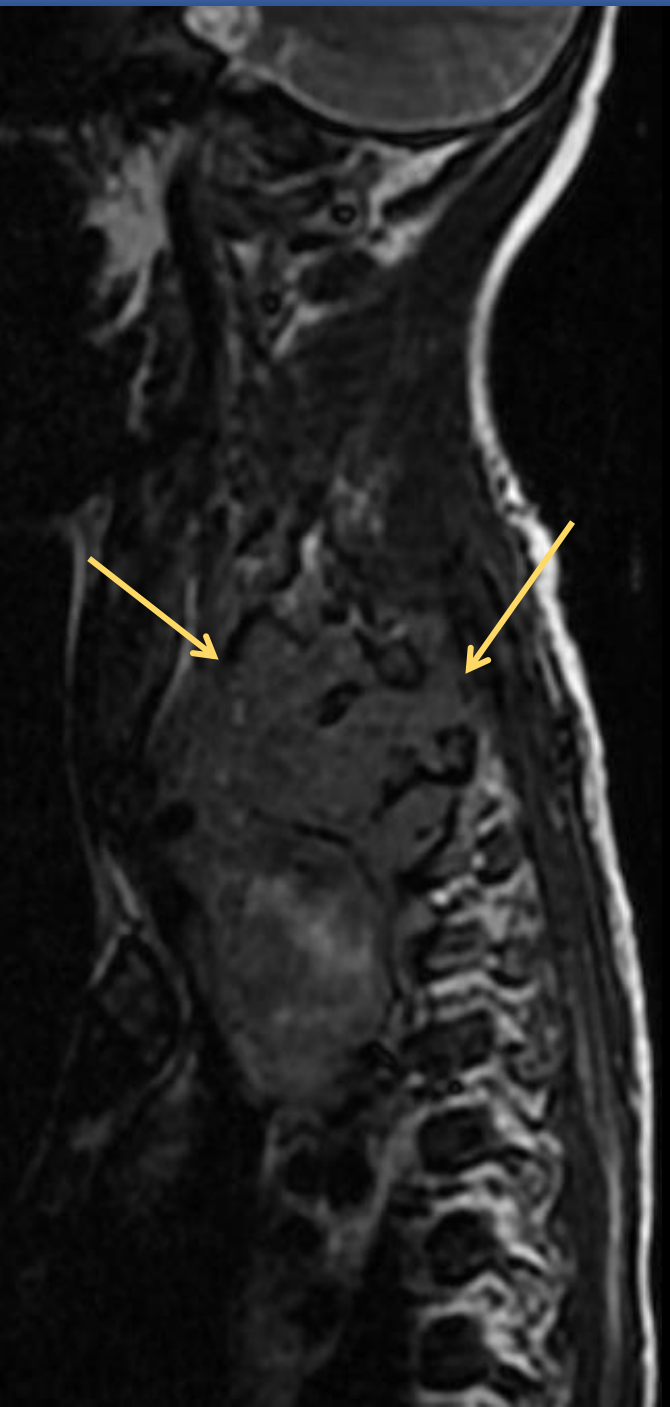
5 m of EOT -> **Local relapse: Cyclo-Topo every 4 weeks**

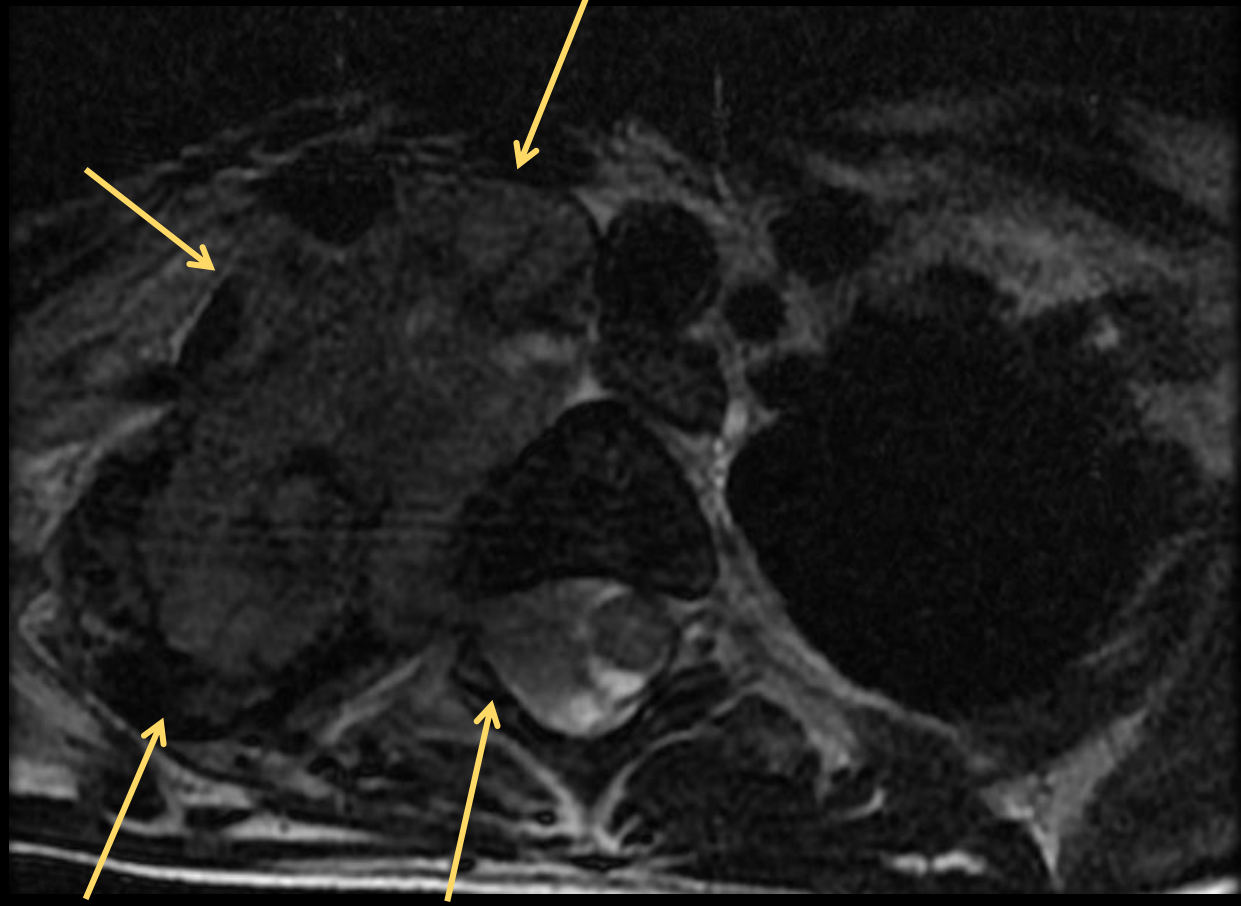
Prolonged thrombocytopenia delayed 2nd cycle

Presented with:

low extremities weakness and inability to walk (< 24 h)

Case 2





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What would be your preferred treatment?

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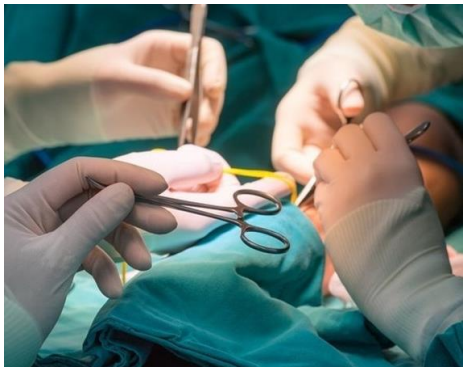
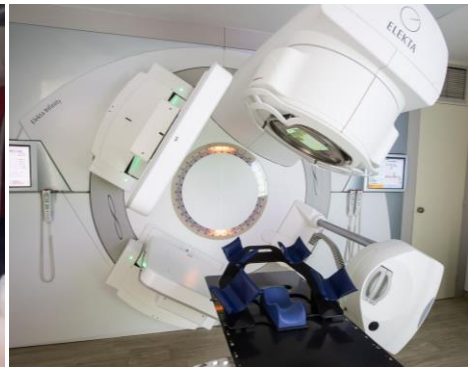
1. Palliative symptomatic treatment
2. Laminectomy
3. Chemotherapy
4. Radiotherapy



Case 2
After neurosurgery

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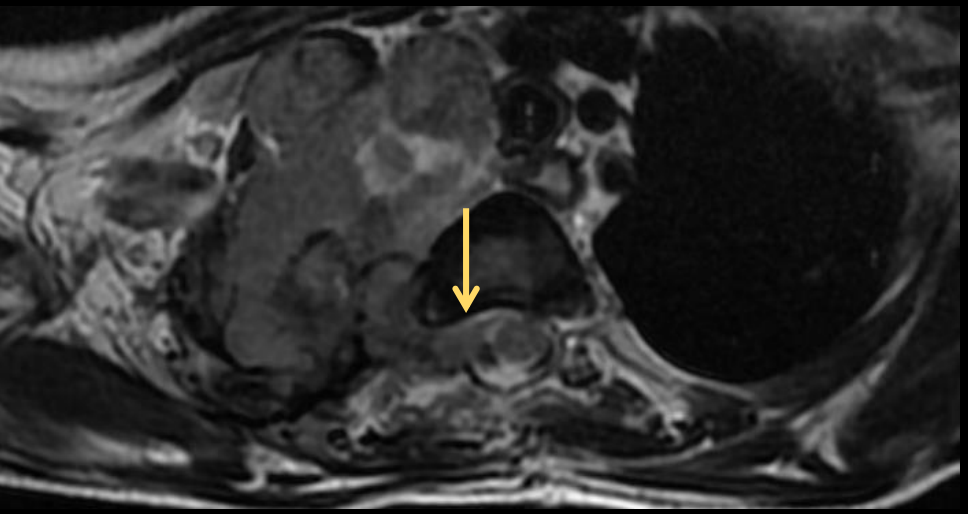

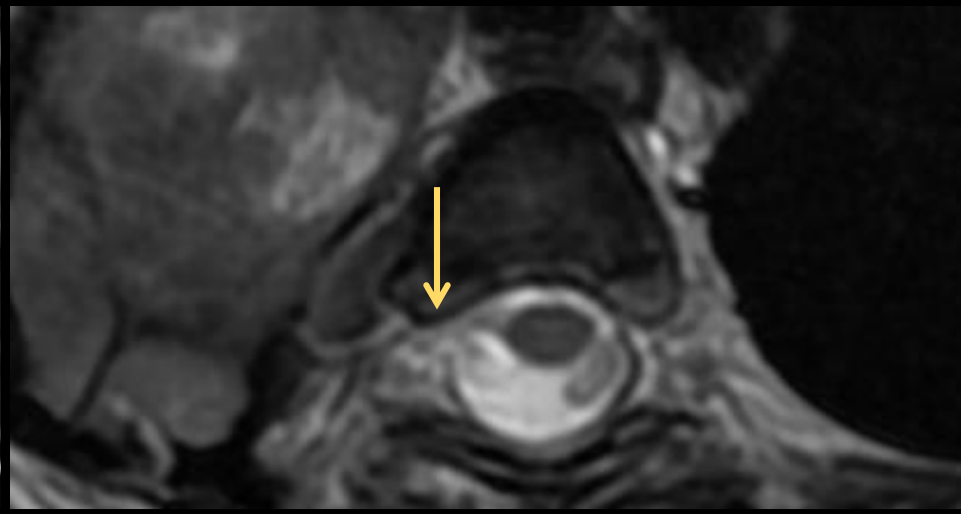

C7-D2 laminectomy
Discharged on D +7, progressive recovery of walking ability

Local radiation
Trabectedin + Irinotecan

+ 5 m: **Lung progression** -> Cabozantinib + palliative support

Died from disease **8 months later**.
No spinal cord compression symptoms again

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Discussion

- When and why is it worth to wait until tumour diagnosis
- How the tumour diagnosis influences treatment
- Surgery, radiation and chemotherapy: pros and cons

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Take home message

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- Spinal cord compression is a **complex emergent situation**
- In order to decide how to treat:
  - It is worth having a **tumour sample** as some tumours respond speedily to chemo (Ewing, NBL, Lymphomas).
  - Severity of neurological symptoms do not implies necessity of surgical decompression.
  - **Balance** between risk and benefits:
    - Children -> **growth expectancy**
  - **Resources availability** always plays a role.