Spinal Hemangioblastomas in von Hippel Lindau Disease: Clinical and Radiological Characteristics

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Background

- Visceral lesions

- CNS hemangioblastomas
  - Cerebellum (45-50%)
  - Spinal Cord (40-45%)
  - Medulla (5-10%)

Park (2007)

Background

Spinal cord hemangioblastomas

- Hypesthesia
- Weakness
- Hyperreflexia
- Ataxia
- Pain
- Incontinence

- Asymptomatic

Park (2007)

Background

- Intervention spinal hemangioblastoma
  - Surgical resection
  - Embolization
  - Radiotherapy

In general applied for symptomatic lesions

Background

- Intervention spinal hemangioblastoma
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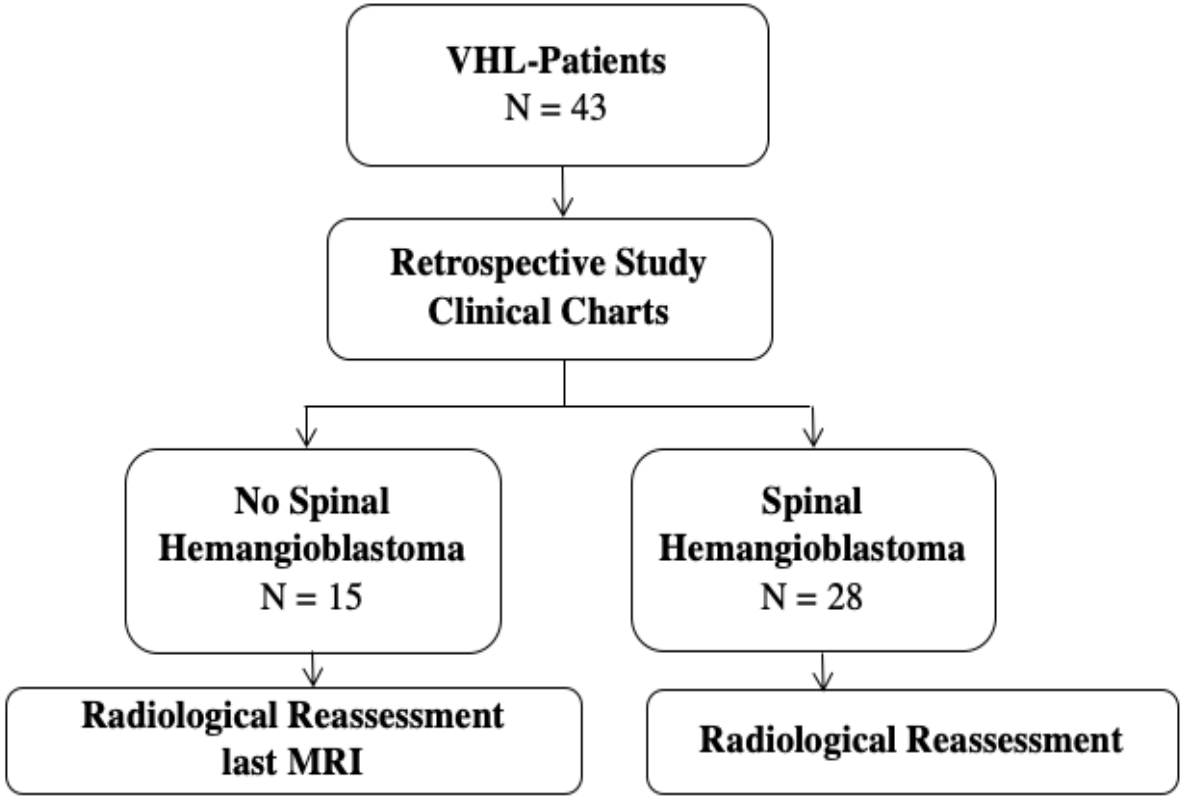
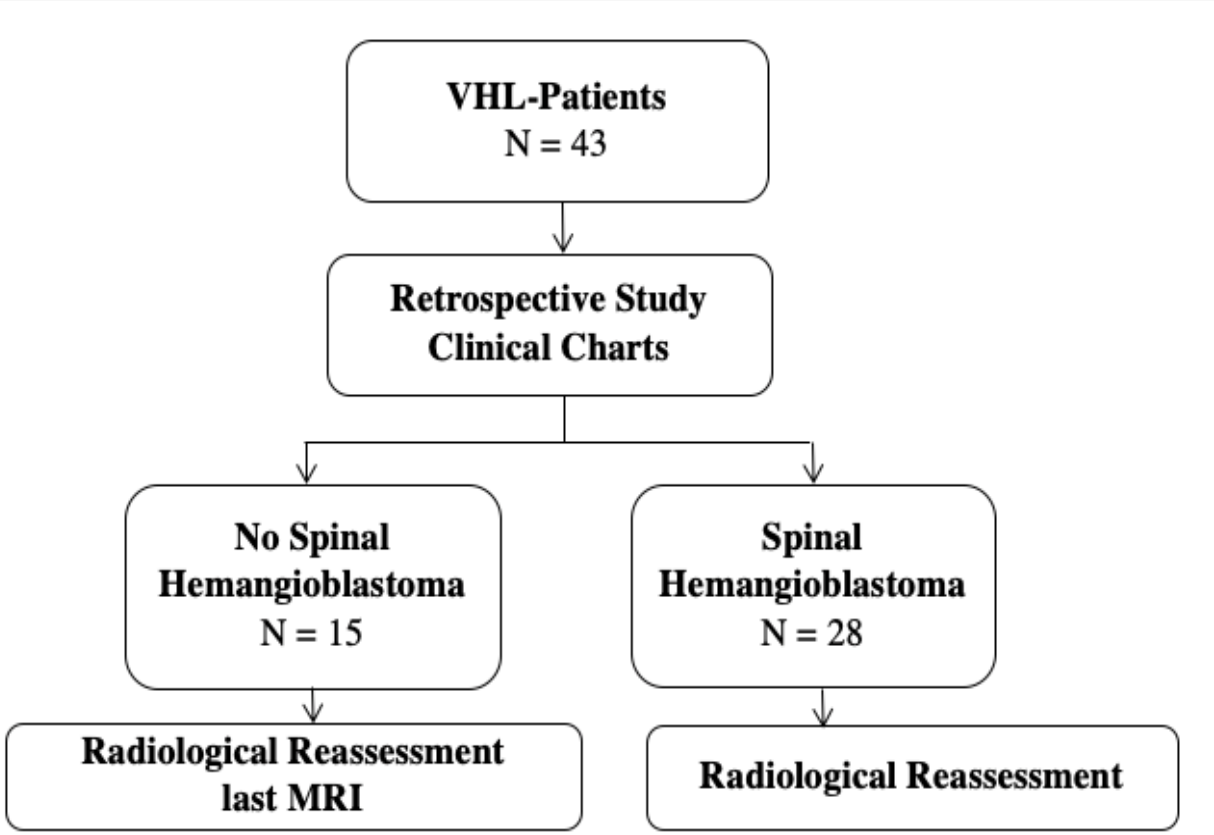
In general applied for symptomatic lesions

- **Aim: To investigate the relation between MRI findings (peritumoural edema and cysts) and symptoms**

Methods


VHL-Patients
N = 43
Retrospective Study
Clinical Charts
No Spinal
Hemangioblastoma
N = 15
Spinal
Hemangioblastoma
N = 28
Radiological Reassessment
last MRI
Radiological Reassessment

Methods



Radiological reassessment

- Localization and volume Hb
- Growth Hb
- Peritumoral edema
- Spinal cysts

Review patient charts

- Hypesthesia
- Weakness
- Hyperreflexia
- Ataxia
- Pain
- Incontinence

Results

- 78 spinal hemangioblastomas
- 10 patients spinal cyst
- 9 peritumoral edema
- 28 patients
  - 22 multiple hemangioblastomas
  - 6 single hemangioblastoma

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Results

MRI

- 78 hemangioblastomas
  - 38 cervical
  - 27 thoracic
  - 13 lumbar

- Peritumoral edema: 1 patients
- Spinal cysts: 2 patients
- Both: 8 patients

Patient charts

- 18 symptomatic

- 10 asymptomatic

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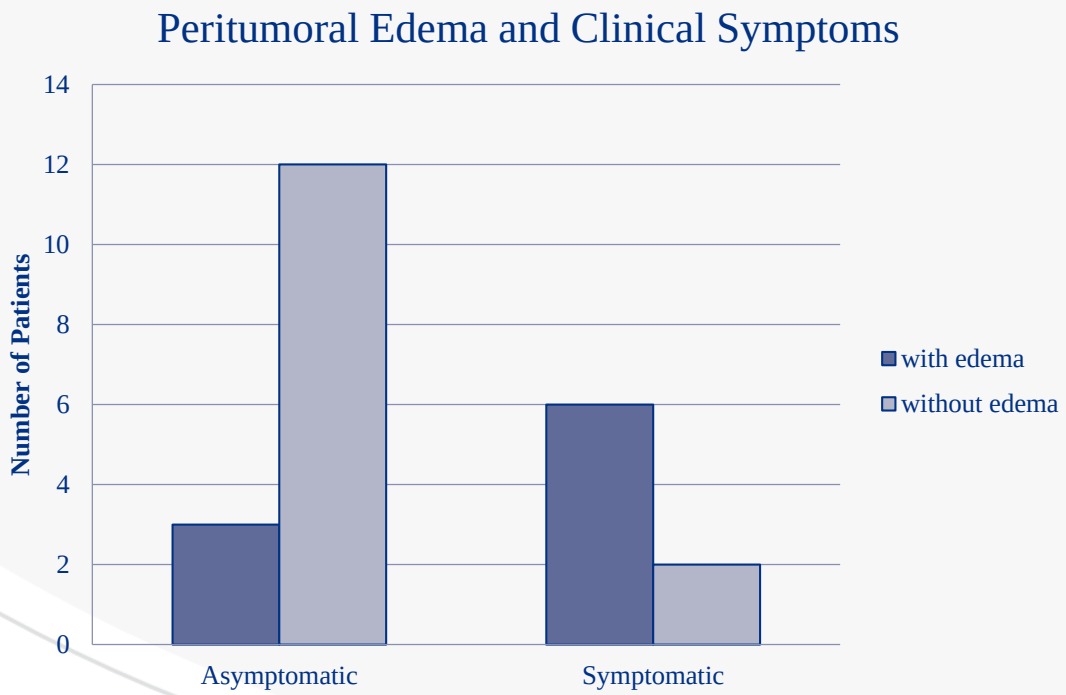
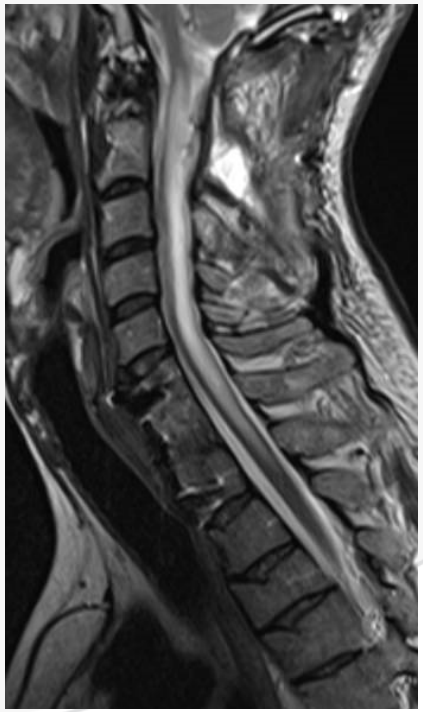
Peritumoral Edema and Cysts





* Association between Peritumoral Edema and Symptoms
(p = 0.023)

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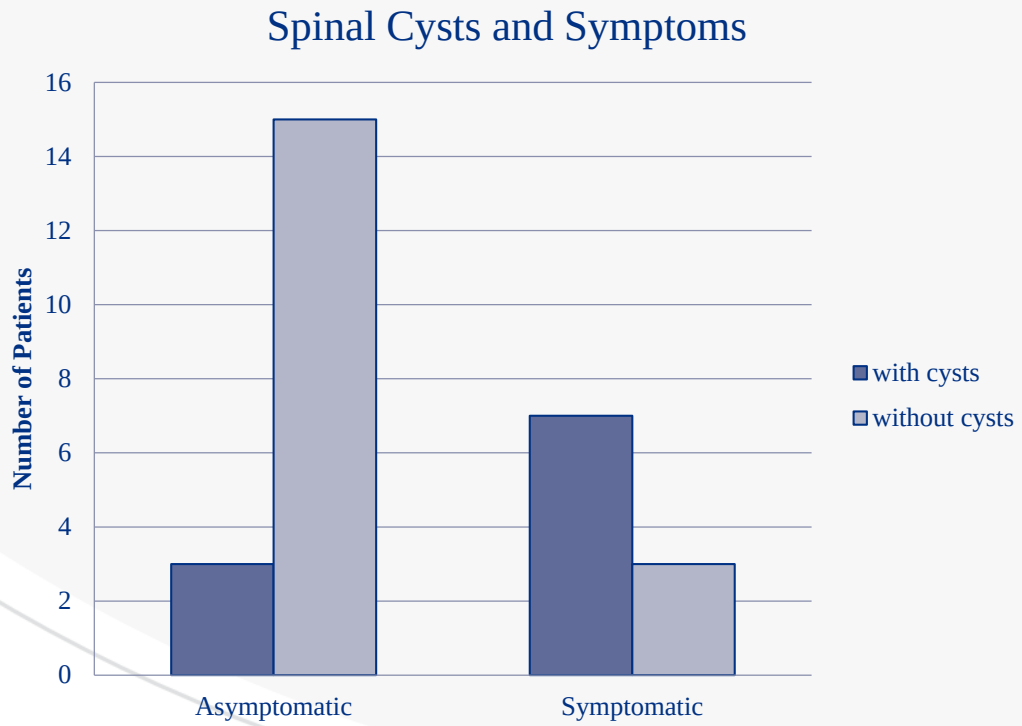
Peritumoral Edema and Cysts



* Association between Spinal Cysts and Symptoms
(p = 0.011)

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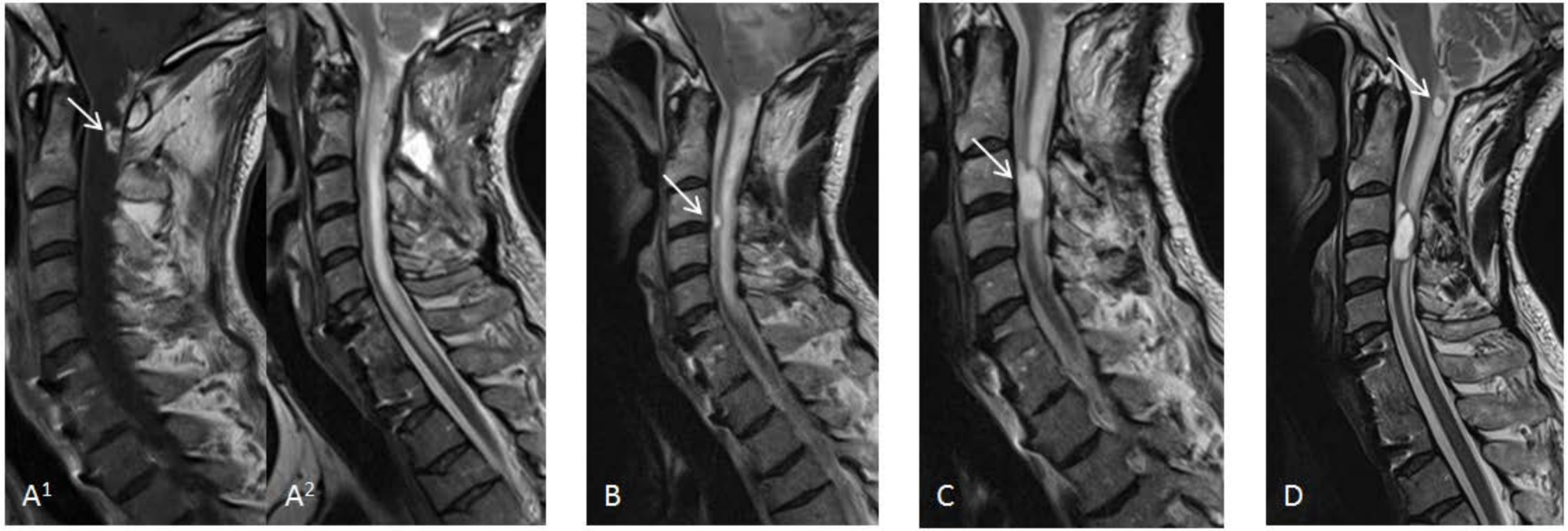
Peritumoral Edema and Cysts



- Observation: development of spinal cyst preceded by the formation of peritumoral edema

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Limitations

- Retrospective
- Study population
- Variance in MRI characteristics

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Conclusion

- Nearly 90% of VHL patients will develop one or more CNS hemangioblastoma(s)
- Hb growth is unpredictable, which makes a regular follow up necessary
- The presence of peritumoral edema and spinal cysts are associated with symptoms
- Further (prospective) studies with larger study populations are needed

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Take home message

The presence of peritumoral edema and/or spinal cysts on MRI in VHL-patients with spinal hemangioblastomas is associated with symptoms and may alert the clinician, to intensify the radiological and neurological follow up in order to prevent irreversible morbidity.

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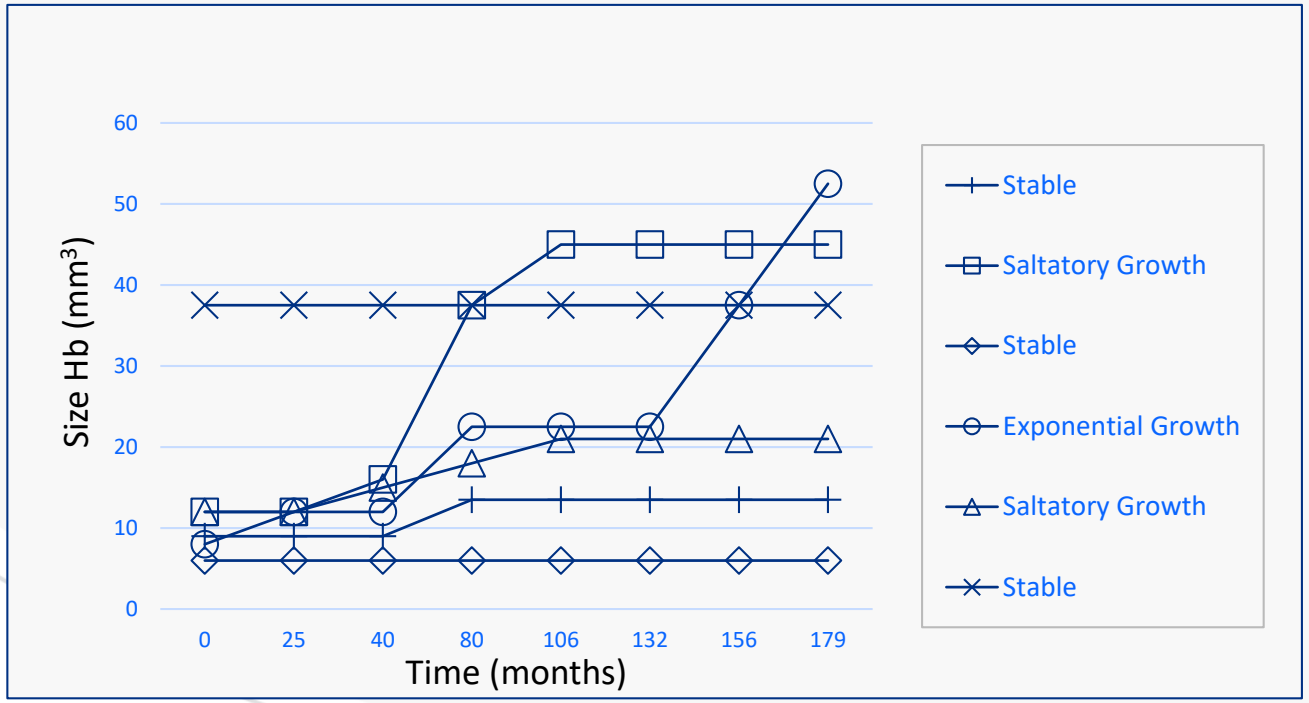
Additional Slides
Results - Growth pattern Hb



- 18 Stable

- 25 Progression
  - 14 Saltatory
  - 4 Linear
  - 7 Exponential

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