Management of Central Nervous System Hemangioblastomas in VHL

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• No conflicts of interest.

• Goals:
  • Why is hemangioblastoma a problem?
  • What is done to address the problem?
  • Discuss new findings about hemangioblastomas.
Structure

• Introduction

• Case discussions

• Future directions: update on drug trial
What is a hemangioblastoma?

Central nervous system
Brainstem
Retina
Cerebellum
Endolympathic sac
Spinal cord
So, why is it a problem in the brain?

- Limited space
  - Tumor growth
  - Swelling
  - Cyst

www.trauma.org
Case #1: Tumor Growth
Tumor Growth
Tumor Growth
Effect of Surgery
Case #2: Cyst
Cyst
Cyst
Cyst
Case #3: Swelling
Swelling
Swelling
Swelling
What about other locations?

- Brainstem: Tumor growth, cyst, swelling, syrinx
- Spinal cord: tumor growth, cyst, swelling, syrinx
Case #4
Brainstem Hemangioblastoma
Case #5
Spinal Cord Hemangioblastoma
Vorinostat Trial
The New Genetics: Man Into Superman
TIME
SPECIAL REPORT
COUNTDOWN TO WAR
SOLVING THE MYSTERIES OF DNA
The 50th Anniversary: Reliving Watson and Crick's historic discovery
How gene science has changed our lives
Visions of the future
Want to Know My Future?

New genetic tests can point to risks—but not always a cure

BY BONNIE ROCHMAN
Personalized Medicine in Oncology

- Cancer genetics
- BRCA1 and BRCA2 for risk stratification.
- Gleevec (imitanib) to inhibit ABL in CML.
- Herceptin (trastuzumab) to interfere with HER2/Neu receptors.
- IL13-PE infusion in DIPG.
• Mutation detection rate up to 100% if VHL is definite.

• 10% large deletion

• 30% partial deletions

• 60% point mutations

• 38% missense mutation
Proteostasis Modulators Prolong Missense VHL Protein Activity and Halt Tumor Progression

Chunzhang Yang,1,2 Kristin Huntoon,1,2 Alexander Ksendzovsky,1 Zhengping Zhuang,1 and Russell R. Lonser1,3,*
Patient with missense mutation VHL disease AND symptomatic CNS hemangioblastoma.

Consent

Admission to inpatient unit. Day 0

Vorinostat Days 1 - 7

Surgery and tissue procurement Day 8
Questions and Comments