New treatment strategy &
New presentation of
Endolymphatic sac lesion in VHL

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Endolymphatic sac

Endolymphatic sac

Lonser et al., J Neurosurg, 2008
Endolymphatic Sac Tumor (ELST)

• The prevalence of ELST in VHL disease:
  - in the literature is up to 16%
  - in the European study is 3.6%
• Bilateral occurrence of ELST is a strong indicator for VHL-associated disease:
  - in the literature is up to 30%
  - in the European study is 19%
• 32% of patients with VHL presented with ELST as the first symptom heralding VHL
• There are no specific genotype-ELST correlations, also any given mutation of the VHL gene can predispose to ELST

Manski et al., JAMA, 1997
Bausch et al., Head Neck, 2016

Endolymphatic Sac Tumor (ELST)

• Once the diagnosis of VHL has been established, contrast-enhanced MRI of the petrous bone and audiology assessment must be part of surveillance with yearly recommended checkups
• Equally important is the possibility of a curative complete tumor resection and preservation of hearing acuity
• An interdisciplinary diagnostic and treatment protocol
• Preoperative arteriography with embolization
• Surgery as early as possible

Bausch et al., Head Neck, 2016
Embolization of the maxillary artery

Pre-embolization

Post-embolization

Embolization of the occipital artery

Pre-embolization
Emboliization of the occipital artery

New treatment strategy for Endolymphatic sac lesion in VHL

Endolymphatic Sac Tumor (ELST)

- ELST are challenging to remove because these tumors are extremely well vascularized and are encased in the temporal bone
- The standard treatment is an extensive endovascular embolization followed as complete a resection as possible
- The endovascular embolization after the preoperative angiography cannot be completed due to risk to embolize the feeding vessels with potential neurological complications (for example branches of internal carotid artery) or because catheterization is not possible (tortuous and/or small arteries)

Endolymphatic Sac Tumor (ELST)

- We developed, in collaboration with the interventional neuroradiologist, a new approach consisting of a 4-steps procedure to treat this tumor
STEP 1

- The neuro-otology surgeon drill into the bone surrounding the tumor and open an access for the radiologist

STEP 2

- The radiologist, under the same general anesthesia, realize a classical endovascular angiography with femoral artery access to analyze the anatomy and to assess the devascularization with control of potential side effects at the end

STEP 3

- The direct puncture technique is used: the lesion is punctured with needle under permanent fluoroscopic guidance using the 3D XperGuide planning software
- This software allows the planning of a virtual needle trajectory defining a precise entry point and a trajectory up to the target while avoid normal brain parenchyma. Then, the superimposition of the virtual trajectory onto the real-time fluoroscopic image allows the guidance of the needle up to the defined depth using two orthogonal positions of the C-arm (entry point view and progression view)
- After puncture, we confirmed the position in the tumor, and direct embolization was performed with mixture of glue made of n-butyl cyanoacrylate and an oily contrast agent
STEP 4

- The neuro-otology surgeon realize the ablation of the tumor 24 to 48 h after the puncture

New presentation of Endolymphatic sac lesion in VHL

Classical aspect
Young man 16 years

New 4-steps procedure

Endolymphatic sac cyst

Surgical view

THANK YOU!

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