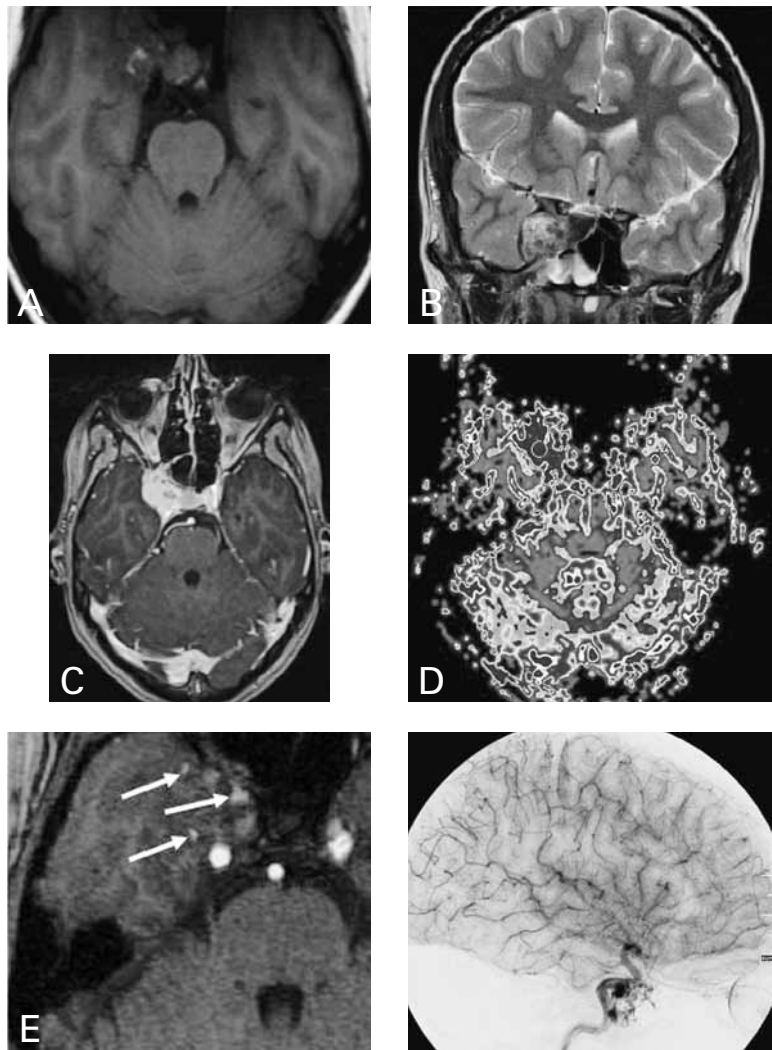


PARAGANGLIOMA OF THE CAVERNOUS SINUS

A. Arain, J. Vandevenne, B. Depeuter, J. Smits, F. Weyns, Y. Palmers¹

Key-word: Paraganglioma

Background: A 15-year-old girl presented in a Dutch hospital with right-sided trigeminal neuralgia. MR-imaging showed a mass lesion in the right cavernous sinus. Differential diagnosis in this hospital was a meningioma or a schwannoma. The patient was referred to the neurosurgery department of our hospital, and resection of the lesion was planned. At surgery, the lesion presented as a subdural bulge surrounded by swollen venous structures. The incision of the dura resulted in profuse hemorrhage of arterial origin, and hemostasis was obtained with difficulty. The tumor showed a fibrillar structure and a strong arterial vascularization which was not concordant with schwannoma. No further exploration of the lesion was performed, and no biopsy was obtained. To clarify the unexpected surgical findings and to reach a diagnosis without biopsy, pre-operative MR-images were reviewed. Digital subtraction angiography (DSA) was performed postoperatively. The lesion did not take up FDG on PET scan. Laboratory results showed increased catecholamines in the urine.



1. Department of Medical Imaging, Campus Sint-Jan, Ziekenhuis Oost-Limburg, Genk, Belgium.

1A	1B
1C	1D
1E	2

Work-up

MRI of the brain (Fig. 1) shows on axial T1-weighted image (A) a mass lesion located in the right cavernous sinus. Coronal T2-weighted image (B) demonstrates a heterogeneous lesion with 'salt and pepper'-appearance, in the right cavernous sinus. Axial contrast-enhanced T1-weighted image (C) visualizes strong contrast enhancement of the lesion. On relative Cerebral Blood Volume (CBV) map of perfusion MRI (D), the relative cerebral blood volume (rCBV) in the lesion is nine-fold increased compared to normal brain parenchyma, suggesting strong angiogenesis, showing nine-fold increased rCBV in the lesion. Axial Time-of-Flight (TOF) image (E) shows presence of arterial flow (white arrows) within the lesion.

Postoperative digital subtraction-angiography (DSA) of the cerebral vessels (Fig. 2) shows strong arterial vascularization of the lesion arising both from internal and external carotid artery branches (recurrent sphenopalatine branch of internal maxillary artery) carotid artery.

Radiological diagnosis

The most likely diagnosis is a large, *hyper-vascular paraganglioma or glomus tumor of the cavernous sinus*. The strong arterial vascularization as seen on TOF MR-images, DSA and surgery, together with the 'salt and pepper' appearance on T2-weighted MR-images, and the presence of increased catecholamine levels in the urine are the arguments in favor of this diagnosis.

Discussion

Paragangliomas are highly vascular neoplasms that arise from paraganglia, which serve as chemoreceptors responsible for monitoring changes in blood pH, carbon dioxide concentration, and rate of blood flow. Approximately 90% of the paragangliomas occur in the adrenal gland (pheochromocytoma), the largest collection of chromaffin cells. The remaining 10% arise from extra-adrenal sites. Most of the extra-adrenal paragangliomas arise in the abdomen (85%), with some in the thorax (12%), and some less commonly in the head and neck area (3%). Paragangliomas of the head and neck (HNP) are rare tumors of neural crest origin, comprising about 0.6% of head and neck tumors and about 0.03% of all tumors. They may occur along the paraganglia's pathway of embryologic migration which extends from the skull base to the pelvic floor. Radiologic evaluation of glomus tumors aids in differentiating them from other neoplastic processes. Paragangliomas usual-

ly show a hyperintense signal on T2-weighted MR-images and a distinct contrast enhancement on T1-weighted images. In larger lesions, On T1-weighted a "salt and pepper" appearance of the tumor matrix on T1- and/or T2-weighted images is characteristic. Prominent arterial vasculature associated with the main lesion may also be seen on TOF sequence of MR-images. DSA is useful in the diagnosis by providing information about vascularity and feeder vessels. CT is useful as it presents a very sensitive imaging procedure for the diagnosis of bony destructions by the paragangliomas often seen in jugular and tympanic paragangliomas. Histological evaluation is the most reliable way to confirm the diagnosis of paraganglioma. Microscopically irrespective of the site, paragangliomas have a common appearance. The tumor contains all three elements normally present in a paraganglion (i.e., type I, chief cells or granular cells; and type II, the supporting or sustentacular cells and numerous capillaries). Paragangliomas are sometimes associated with secretion of several neuropeptide hormones, such as adrenocorticotrophic hormone, serotonin, catecholamine, and dopamine. The incidence of catecholamine secretion is approximately 4%. Frequently, these tumors have a low level of secretion that is not recognized clinically. The treatment options for head and neck paragangliomas include surgical resection, conventional radiation therapy, stereotactic radiosurgery, permanent embolization or a combination of these modalities. The dose commonly recommended for radiation therapy is 45 to 50 Gy; in stereotactic radiosurgery 12 to 18 Gy are usually applied. In this case the surgical resection is not performed to avoid hemorrhagic complications. The patient was referred for radiotherapy.

Bibliography

1. Wasserman P.G., Savargaonkar P.: Paragangliomas: classification, pathology, and differential diagnosis. *Otolaryngol Clin North Am*, 2001, 34: 845-862.
2. Rao A.B., Koeller K.K., Adair C.F.: From the archives of the AFIP. Paragangliomas of the head and neck: radiologic-pathologic correlation. Armed Forces Institute of Pathology. *Radiographics*, 1999, 19: 1605-1632.
3. Lustrin E.S., Palestro C., Vanheesan K.: Radiographic evaluation and assessment of paragangliomas. *Otolaryngol Clin North Am*, 2001, 34: 881-893.
4. Boedeker C.C., Ridder G.J., Schipper J.: Paragangliomas of the head and neck: diagnosis and treatment. *Fam Cancer*, 2005, 4: 55-59.