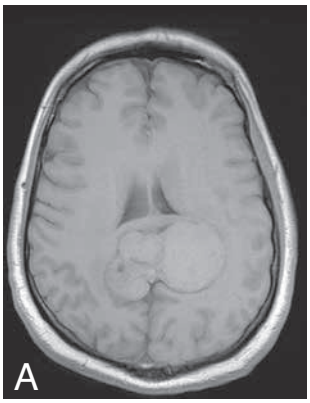
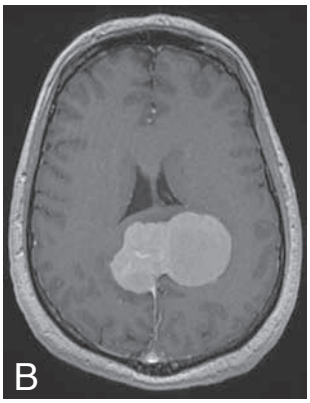
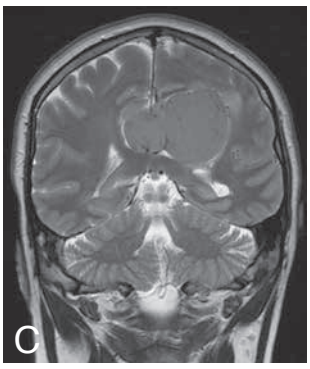
JBR–BTR, 2014, 97: 315.

IMAGES IN CLINICAL RADIOLOGY







Hemangioperiocytoma simulating meningioma in a 41-year-old man

E. Rabaey, K. Desmet, M. De Maeseneer, T. Vanderhasselt, T. Stadnik[1]

A 41-year-old patient was seen in the neurology clinic because of severe memory loss and vision problems. He described his vision problem as "a sail moving in front of his eyes". He also reported loss of sensation and strength in his right arm and leg. On clinical examination loss of sensation on the right side of his face, arm and leg were evident. This was accompanied by loss of strength in his right arm and leg, and right sided asteriognosia.

MR imaging showed a bilobed mass, measuring 3.6 × 6.9 × 4.1 cm, that was in contact with the falx cerebri. There was significant mass effect on adjacent structures in particular the corpus callosum. The diagnosis of meningioma was entertained. On transverse T1 weighted images (A, B) an extra-axial bilobed mass is seen with marked homogenous contrast enhancement. On T2 weighted images (C) flow voids and marked mass effect are present.

At surgery the tumor had a highly vascular appearance. It was seen to extend on both sides of the falx. Profuse bleeding occurred during surgery, and hence made complete resection impossible, only the right portion of the tumor was removed.

Macroscopically the removed fragments of tumor had a brown color. Microscopically the tumor was of high cellularity. Polymorphic cells with large nuclei were seen. The cells were embedded in a delicate reticulin network. No psammoma bodies were seen. Voluminous vascular structures ('Staghorn vessels') were present. On immunohistochemistry the tumor tested positive for vimentin, CD 31, and CD 34, and negative for epithelial membrane antigen and GFAP. These findings were consistent with a hemangioperiocytoma without pathological signs of anaplasia (grade II tumor).

Postoperative MR imaging showed presence of residual tumor. Therefore a second surgical procedure was performed during which residual tumor could be removed.

Comment

Hemangioperiocytoma is a mesenchymal neoplasm that is histologically unrelated to meningioma. Morphologically it is part of a continuum with solitary fibrous tumors. It arises from primitive mesenchymal cells throughout the body and most commonly involves soft tissues of the lower extremities, pelvis and retroperitoneum. Approximately 15% of hemangioperiocytomas occur in the head and neck region. Hemangioperiocytomas are classified as mesenchymal, non-meningiothelial tumors, WHO grade II (generally low grade malignant tumors).

The average age of onset is 42 years and there is a predilection for men. Headache is a common symptom. Other symptoms related to the mass effect of the tumor also occur. There is only an interval of a few months between the onset of symptoms and diagnosis due to the fast growth rate.

Treatment consists of surgery. Chemotherapy has not shown to be effective. There is a 5 year survival rate of 93% following surgery and a disease free survival rate of 89% following surgery.

Macroscopically, hemangioperiocytoma presents as a well circumscribed, encapsulated and firm mass with a dural attachment. It is quite vascular and has a tendency to bleed during surgery. Microscopically, hemangioperiocytoma is highly cellular. It is homogenous and consists of randomly oriented plump cells embedded in a dense network of reticulin. A 'stag horn' vascular pattern is typical and consists of lobules with tumor cells surrounding wide, branching capillaries. On immunohistochemistry hemangioperiocytoma is positive for vimentin, similar to meningioma. Hemangioperiocytoma is negative for epithelial membrane antigen, whereas meningioma is positive for this antigen.

Tumors typically have a supratentorial and occipital location. They involve the falx, tentorium or dural sinuses. Their diameter varies from 2 to 9 cm. The tumor has an extra-axial location with dural attachment. Surrounding edema is common.

On non-enhanced CT, a lobulated hyperdense lesion is seen. Bony erosion may be present. Calcifications and hyperostosis are typically absent in contradistinction to meningioma. After contrast administration heterogeneous enhancement is seen. A dural tail sign may be observed. Low density areas corresponding to cysts or necrosis are common.

Both on T1- and T2-weighted MR images a heterogeneous mass, isointense to gray matter is seen. Areas of flow void may be apparent. Adjacent edema and mass effect are common. After contrast administration marked and heterogeneous enhancement is seen. A dural tail sign is present in 50% of patients. Central areas of necrosis may be seen.

Hemangioperiocytoma and (malignant) meningioma present with similar clinical and MR imaging findings. Hence, differential diagnosis may be difficult. Bone erosion, presence of areas of signal void, and heterogeneous contrast enhancement are more characteristic for hemangioperiocytoma.

Reference

1. Sibtain N.A., Butt S., Connor S.E.: Imaging features of central nervous system haemangiopericytomas. *Eur Radiol*, 2007, 17: 1685-1693.

1. Department of Radiology, Vrije Universiteit Brussel-Universitair Ziekenhuis Brussel, Brussels, Belgium.