SAINT JOHN'S HEALTH CENTER BRAIN TUMOR CENTER



Daniel F. Kelly, M.D., Director of the Neuro-Endocrine Tumor Center at the John Wayne Cancer Institute and Saint John's Health Center in Santa Monica, California.

RATHKE'S CLEFT CYSTS (RCCs)

Rathke's Cleft Cysts are not true tumors or neoplasms; instead they are benign cysts. Rathke's pouch forms as part of normal development and eventually forms the anterior lobe, pars intermedia and pars tuberalis, of the pituitary gland. This pouch normally closes in fetal development, but a remnant often persists as a cleft that lies between the anterior and posterior lobes of the pituitary gland. Occasionally, this remnant enlarges to form a cyst, the RCC that can cause pituitary failure, headaches and in some instances, vision loss.

Symptoms

Symptomatic RCCs are relatively common pituitary lesions, accounting for 5-10% of surgically removed pituitary masses. RCCs can be seen at any age, although most are identified in adults. Intrasellar RCCs are usually asymptomatic and are found incidentally at autopsy or on a magnetic resonance imaging (MRI) scan. However, larger RCCs may cause visual disturbances, symptoms of pituitary dysfunction, and headaches.

Diagnosis

The best means of visualizing a RCC is with an MRI or a computer tomography (CT) scan of the pituitary region performed without and with contrast. A complete pituitary hormonal blood evaluation should also be performed. Other possible diagnoses to consider when a cystic mass is seen in the area of the pituitary include a cystic pituitary adenoma, craniopharyngioma or arachnoid cyst.

Treatment

The best treatment for a symptomatic RCC causing pituitary failure, headache or visual loss is surgical removal through an endonasal transsphenoidal approach. The goal of surgery is to completely remove the cyst contents while improving or preserving pituitary function vision and alleviating headache and visual loss if present. Attempts to remove the cyst lining should be avoided because this can result in pituitary gland damage. A complete removal of RCCs is possible in 80-95% of cases although they can recur at a rate of 5-15% over 5 to 10 years.