Clinical Manifestations of Rathke's Cleft Cyst and Natural Progression over Two Years in Children and Adolescents

Jo Eun Jung, MD.¹ ,Mo Kyung Jung, MD.¹ ,Ah-Reum Kwon, MD.¹ ,DukHee Kim, MD.PhD.² ,Hyun WookChae, MD.¹ , and Ho-Seong Kim, MD,PhD¹

Department of Pediatrics, Severance Children's Hospital, Endocrine Research Institute,
Yonsei University College of Medicine, Seoul, Korea
Department of Pediatrics Sowha Children's Hospital, Seoul, Korea

Purpose

Rathke's cleft cyst (RCC), a benign, sellar or suprasellar lesion arising from the embryological remnants of Rathke's pouch, is usually asymptomatic, and found incidentally on autopsy. With increased interest in pediatric endocrinopathies and better imaging modalities available, the prevalence of RCCs is increasing. However, few studies have examined children or adolescents. Therefore, we investigated c linical manifestations, endocrine dysfunction, and radiologic features of RCC in children and adolescents.

Materials and Methods

We retrospectively reviewed medical records of 91 children and adolescents with RCC diagnosed using magnetic resonance imaging (MRI) from January 2006 to December 2015. Results of clinical symptoms and endocrine function tests measuring basal hormones were reviewed, as were initial and follow-up MRI findings.

Results

The two most common clinical symptoms were precocious puberty and headache; presentation varied with age. Preschool-aged children experienced early puberty or headache, while older children experienced a greater variety of symptoms. Most patients had endocrine dysfunction (n=60, 76.9%); cent ral precocious puberty was most common (n=25, 41.7%). There was no significant difference betwee n endocrine dysfunction and MRI findings. However, we found significant differences in radiological f indings according to treatment of choice: patients managed surgically (n=7) had significantly larger c ysts, which were more frequently located in the suprasellar region, than those managed medically. P ost-surgery, endocrine function improved in only one (14.2%) of the patients who underwent surgery . Of 22 patients who had a follow-up MRI performed two years after diagnosis, the size of the RCC h ad increased in about 13.6% (n=3).

Conclusion

Routine follow-up of endocrine function and imaging studies is necessary for patients with RCC, part icularly those requiring surgery.







