

Puzzler Page Answers: November 2010

Syndrome or Condition—You Decide

Although McCune-Albright syndrome was strongly considered in the differential diagnosis, the patient was diagnosed as having central precocious puberty. To read the original case, please view page 58 in the November copy either in print or online at www.endo-society.org/endo_news (and click on “Past Issues” to find November).

Below are excerpts from Drs. Maala Daniel and John Buchlis’ **ENDO 2010** poster:

“Central precocious puberty (CPP) results from activation of the hypothalamic-pituitary-gonadal (HPG) axis traditionally before the age of 8 years in girls and 9 years in boys.^{1,2} The specific age has been debated in African American girls. Some suggest this age should be lowered to age 6 or 7.³ In girls the majority of cases are idiopathic and benign, although any intracranial disturbances can cause CPP.⁴ The mechanism is thought to be either related to increased excitatory signals or altered inhibition of gonadotropin releasing hormone (GnRH) secretion.⁴ The classic laboratory criteria for diagnosis is usually a basal LH level over 0.6 IU/L, FSH level over 2 IU/L, and stimulated peak post GnRH agonist LH level over 6.9 IU/L and FSH Level over 5 IU/L.⁴ However, prepubertal girls tend to have an FSH predominance. Therefore, FSH is not as helpful in diagnosing CPP.

“McCune-Albright syndrome (MAS) is a cause of peripheral precocious puberty (PPP) or gonadotropin-independent precocious puberty. It has been described as a triad of café-au-lait spots, precocious puberty (PP), and fibrous dysplasia of the bones. The syndrome is due to mutations of the *GNAS* gene. Genetic testing is available but not routinely used because of its unreliability. The diagnosis is typically made clinically.⁵

“Isolated menses is uncommon. It has been described as vaginal bleeding in the absence of other signs of precocious sexual development. The etiology is often unknown. It has been suggested that increased endometrial sensitivity to estrogen and/or ovarian stimulation may be the cause for this bleeding.⁶ The bleeding is thought to be benign and self-limited in nature.

“Our case most likely represents premature menarche as the initial sign of central precocious puberty.

“McCune-Albright syndrome was strongly considered in our differential diagnosis. Suppressed gonadotropins are expected for the diagnosis. However, long-standing PPP can lead to activation of the HPG axis resulting in secondary CPP.⁵ Since our patient presented with a short duration of symptoms this theory was discounted. Also, GnRH agonists should not have prevented the vaginal bleeding if PPP was the cause.

“Our patient presented with isolated menarche despite not meeting the strict criteria for central precocious puberty. It appears as if the patient does have the diagnosis since

vaginal bleeding has stopped with GnRH agonist therapy. It is possible her underlying undiagnosed seizure disorder was the cause of her precocious puberty.”

References:

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