You are seeing a 12-year-old boy who has missed his health supervision visits for 2 years. His parents say he has been well and they could not afford to take time off from work to bring him in for his yearly checkups, but they are worried about him now because he has had headaches that have caused him to miss several days of school for the past month. The headaches have awakened him at night once or twice in the past 2 weeks. When you evaluate his visual fields, he appears to have trouble with vision on the upper outer third of his visual fields that is worse on the right.

Of the following, the additional physical finding that is MOST contributory to your suspected diagnosis is

A. axillary hair without other signs of puberty
B. decreased linear growth rate
C. decreased weight gain
D. gynecomastia
E. unilateral testicular enlargement
The boy described in the vignette has evidence of a mass pressing on his optic chiasm and causing a visual field disturbance. Masses in this area in children are often craniopharyngiomas arising from the hypothalamus or pituitary, pituitary tumors, or, more rarely, other hypothalamic-pituitary lesions. Tumors in this area are commonly diagnosed because of headache and visual field disturbance, but many are identified because of changes in growth velocity leading to the diagnosis of hypopituitarism. Changes in body composition, including increased fat, decreased muscle mass, and infantile body proportions are more subtle and often missed. Decreased linear growth rate is an important sign of hypopituitarism in the growing child. It may represent growth hormone deficiency, thyroid-stimulating hormone deficiency leading to secondary or tertiary hypothyroidism, or a combination of the two. It is reported in 7% to 93% of children who have craniopharyngioma. Some children who have damage to the hypothalamus or pituitary because of a craniopharyngioma may develop diabetes insipidus, but this is rare before there is other evidence of tumor such as growth attenuation, headache, or visual field abnormalities.

Axillary hair without other signs of puberty is not uncommon in children who have adrenarche or adrenal puberty. Such findings also may be seen after exposure to other, nontesticular sources of androgen. Decreased weight gain rarely is a manifestation of tumors affecting the central nervous system. Both growth hormone deficiency and hypothyroidism usually are associated with somewhat increased fat mass. Craniopharyngiomas may extend to the appetite centers of the hypothalamus and increase hunger, leading to weight gain. Only very rare tumors affecting the appetite center decrease food intake, leading to weight loss (eg, diencephalic syndrome). Gynecomastia is a response to low concentrations of circulating estrogen, as may be seen when androgen of either adrenal or testicular origin is converted to estrogen in the peripheral fat or if there is exposure to another exogenous or endogenous source of estrogen. It is not a sign of pituitary hypofunction. Unilateral testicular enlargement should increase concern for a local testicular tumor but is not expected in hypopituitarism.

Suggested reading:


