Central precocious puberty in girls is defined as the onset of secondary sexual characteristics before the age of 8 years. The majority of cases of central precocious puberty in girls are idiopathic; however, 20 to 30% of cases are organic and are due to a lesion in the central nervous system. The lesions that are the common causes of central precocious puberty are hypothalamic hamartomas, optic nerve gliomas, suprasellar arachnoid cysts, hydrocephalus, germinomas, and other sellar/suprasellar lesions. Central precocious puberty secondary to a cerebellar astrocytoma is extremely rare. The authors report the first case in a girl who presented with several episodes of bleeding per vaginum. There was no clinical or radiological evidence of raised intracranial pressure. (DOI: 10.3171/PED-07/07/066)

KEY WORDS • cerebellar astrocytoma • pediatric neurosurgery • precocious puberty

Case Report

History. This 2-year-old girl presented in January 2006 with a second episode of vaginal bleeding lasting for 2 days. The first episode, which occurred when the girl was 9 months old, was described by her parents as slight spotting lasting less than a day. During the current presentation there were no associated symptoms of fever or pain suggestive of a urinary tract or pelvic infection, and there was no history of foreign body or trauma to her genitalia.

Examination. On examination the child did not have any breast tissue, and there was no evidence of pubic or axillary hair. Her external genitalia appeared normal with no evidence of bruising, injury, or excoriation of the surrounding skin. No café-au-lait spots were observed. Her height was recorded as 88 cm, which was in the 75th percentile for her age, although the mid-parental expectation was less than the 25th percentile. Her weight was noted to be above the 90th percentile. There was no family history of precocious puberty. Examinations of the patient’s blood revealed normal levels of follicle-stimulating hormone, luteinizing hormone, and estradiol, and a pelvic ultrasonography study revealed normal-appearing ovaries and a prepubertal uterus.

She was reexamined in clinic 9 months later, and in the interim had three more episodes of vaginal spotting. Her height had increased to 98 cm (a gain of 10 cm in 9 months), which was in the 90th percentile for her age. Her weight had increased to 16.6 kg, which was above the 95th percentile. She was noted to have breast development with a Stage I left breast and a Stage II right breast. She was also noted to have Stage I pubic hair. Repeated blood investigations revealed that her serum follicle-stimulating hormone and luteinizing hormone were suppressed; however, her serum estradiol had increased to 200 pmol/L (normal range 0–35 pmol/L). A mildly elevated serum prolactin level of 789 mIU/L (normal range 65–455 mIU/L) was also noted.

A repeated pelvic ultrasonography study revealed a peri-pubertal uterine configuration with a relatively large fundus compared with cervical width, implying early maturation. The volume of the left ovary was noted to be in the 95th per-
centile for age and that of the right ovary was above the 95th percentile, also implying advanced maturity. No ovarian mass or cyst was demonstrated.

A computed tomography scan of the brain revealed a large focal lesion within the left cerebellar hemisphere. The lateral and third ventricles were normal in appearance and there was no evidence of a pituitary or suprasellar lesion. Magnetic resonance imaging of the brain confirmed a solid and cystic mass lesion in the left cerebellar hemisphere (Fig. 1 upper), measuring 3.8 × 3.6 × 3.7 cm. The centrally located, somewhat irregular-shaped cystic component was surrounded by a ring of irregular and thickened tissue that homogeneously enhanced and appeared as a high signal intensity on images. In addition, there was a moderate amount of mass effect from the lesion on the midline posterior fossa structures and significant perilesional edema. Supratentorially the ventricles and extraaxial spaces appeared normal.

The amount of myelination was appropriate for a 3-year-old child. There was a normal-appearing sulcal and gyral pattern, hypothalamic and pituitary axes, pineal region, and tectal plate.

Operation. Based on these findings, a diagnosis of isosexual central precocious puberty was made. The patient underwent a posterior fossa craniotomy and complete excision of the lesion (Fig. 1 lower). Intraoperatively the cystic component contained straw-colored fluid typical of a glioma, and the solid component was fleshy with a distinct tumor–brain interface. The histological diagnosis was a pilocytic astrocytoma (World Health Organization Grade I) (Fig. 2), and the immunohistochemical staining was negative for estrogen.

Postoperative Course. The serum estrogen levels returned to normal within 48 hours of the operation. No further episodes of vaginal bleeding have been reported.

Discussion

Central precocious puberty in girls tends to be idiopathic in most cases. However, in about 20 to 30% of cases there is an intracranial mass lesion that causes stimulation of the hypothalamic-pituitary-gonadal axis either by direct invasion of the hypothalamus or by pressure on the hypothalamus secondary to hydrocephalus. In such cases the elevated levels of GnRH produce precocious puberty. This is thought to be due to the impairment of the inhibitory pathways to the GnRH pulse generator. Hypothalamic hamartomas, which consist of congenital heterotopic neural tissue, may contain GnRH-secreting neurons. In many cases treating the raised ICP will result in normalization of the GnRH and a return to the prepubertal state. Sometimes the hypothalamic stimulation becomes autonomous and the raised levels of GnRH persist despite treatment of the raised ICP.

To our knowledge, precocious puberty as a presenting feature of cerebellar astrocytoma has not been previously reported in a girl. We found only three case reports in the literature in which a cerebellar astrocytoma was reported in association with precocious puberty; however, all cases were reported in boys. One of these cases was associated with obstructive hydrocephalus. In another case the patient had symptoms and signs of raised ICP, and he developed precocious puberty after tumor excision. The patient in the final case had a coincidental congenital adrenal hyperplasia with testicular adrenal cell rests.

We report the first case of a cerebellar astrocytoma causing isosexual central precocity in a girl. Interestingly there was no evidence of associated hydrocephalus or any clinical features of raised ICP. The serum estrogen levels returned to normal within 48 hours of removing the lesion. Immunohis-
tochemical staining of the tumor was negative for estrogen, and fluid removed from the tumor also tested negative for estrogen.

The exact mechanism of central precocious puberty in this case is uncertain. We can only speculate that raised ICP was the causative factor even though there were no clinical or imaging features to suggest raised ICP.

References


Address reprint requests to: Vivek Anand Josan, F.R.C.S.(SN), 7 Deeley Close, Birmingham B15 2NR, United Kingdom. email: Josanv@aol.com.