

Isosexual Precocious Puberty in a Boy with Meningomyelocele and Cerebral Arachnoid Cyst

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Appearance of secondary sexual characteristics before 9 years of age in boys constitutes precocious puberty(1). There are three major types of sexual precocity: true or central, peripheral and combined peripheral and central. Precocious puberty may be isosexual or heterosexual in nature. In this communication we report a case of meningomyelocele who developed isosexual precocious puberty secondary to a cerebral arachnoid cyst.

Case Report

A three-year-old boy presented with the appearance of secondary sexual characteristics since two and a half years of age. Parents first noticed the appearance of pubic hair around the root of the penis and soon they realized that the penis and the testes were enlarged compared with boys of similar age. The child had a meningomyelocele in the dorso-lumbar region and kyphoscoliotic deformity of the spine. The sac measured 6 cm in diameter and was covered by a thin layer of partially epithe-

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lialized tissue. There was intermittent oozing of CSF from the sac. There was no history of meningitis or head trauma in past. The child was developmentally retarded. There were no features of hirsutism, deepening of voice or increased muscularity. Neurological examination of lower limbs revealed flaccid paralysis of both limbs, flexion contractures of both knees and hips, absent deep tendon reflexes, lack of response to touch and pain, urinary dribbling and relaxed anal sphincter. Upper limbs showed hypertonia and exaggerated tendon reflexes. Cranial nerves were normal. Head circumference was 47 cm and fundi were normal. Examination of genitalia revealed dark, curly pubic hair spread sparsely over the pubic area (Tanner stage 3)(2). The penis and the testes were enlarged (Tanner stage 3). Stretched penile length was 7.5 cm and each testis measured 3.5 cm in the long axis.

Spine X-ray showed widening of the spinal canal in the lower dorsal and lumbar regions, spinal dysraphism and marked kyphotic deformity. Partial agenesis of the sacrum was also observed. Right hip was subluxated. Wrist X-ray demonstrated bone age of 6 yr at chronologic age of 3 yr. Skull X-ray was normal. Hormonal assays were not performed due to lack of facility. Cranial CT revealed an arachnoid cyst in the right temporal lobe extending into the parietal region (*Fig. 1*). There was midline shift and dilatation of third and left lateral ventricles (*Fig. 2*). Right lateral ventricle was compressed. The lesion exhibited no contrast enhancement. The child was not offered any therapy as he was severely handicapped.

Discussion

Isosexual precocity may be central or

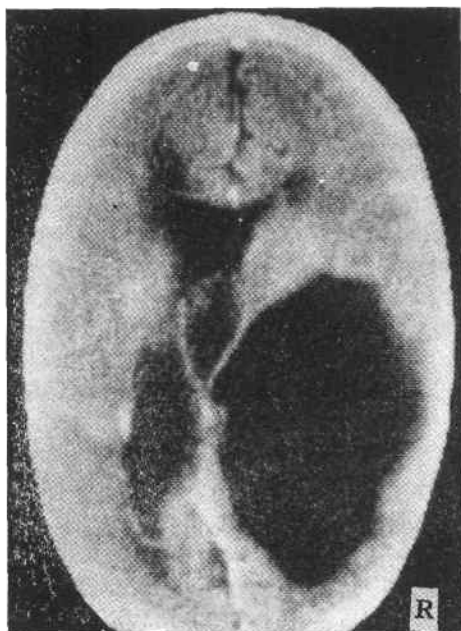


Fig. 2. CT scan showing cyst compressing the right lateral ventricle and causing mid-line shift and dilatation of third and left lateral ventricles.

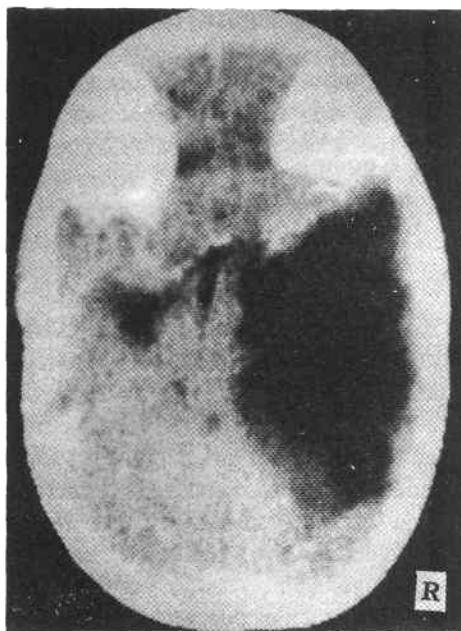


Fig. 1. CT scan showing cystic lesion in the right temporal lobe extending into parietal region.

peripheral in origin. Central or true precocious puberty occurs as a result of the premature release of luteinizing hormone-releasing hormone from the hypothalamus, which stimulates the secretion of pituitary gonadotropins, which in turn stimulate the gonadal sex steroids(3,4). In peripheral precocious puberty, secondary sex characteristics appear without maturation of gonads and there is no activation of hypothalamo-pituitary-gonadal axis(1). Levels of LH, FSH and LH-RH stimulation help differentiate central precocity from peripheral one. In the absence of hormonal investigations it is difficult to tell whether our patient had central or peripheral precocity. However, bilateral testicular enlargement and

the presence of arachnoid cyst in brain point to the central origin of precocious puberty in this boy. Precocious puberty resulting from organic brain lesions is always isosexual(1). Cerebral arachnoid cysts are a rare cause of precocious puberty(5,7,8). In a recent study, none of the 22 children with central precocious puberty had arachnoid cysts as an underlying cause(9). Other CNS lesions resulting in sexual precocity are hypothalamic hamartomas, postencephalitic scars, tubercular meningitis, head trauma, hydrocephalus, tuberous sclerosis, porencephaly, and various brain tumors(1,4-9). Hypothalamic hamartomas are the most common cause of sexual precocity in males and account for 50% of cases(3). All

patients with central precocious puberty should have a CT scan or MRI performed to exclude any underlying brain lesion before making a diagnosis of idiopathic sexual precocity. Cerebral arachnoid cysts are intracranial collections of CSF and account for 1% of all intracranial space occupying lesions(10). They cause neurological dysfunction through expansion that can compress neural tissue and obstruct CSF flow. The association of cerebral arachnoid cyst with meningomyelocele is a rare phenomenon. Zimmerman *et al.*(6) studied various cranial abnormalities in 47 patients with meningomyelocele. Most of the patients had Chiari II malformation and hydrocephalus. Arachnoid cyst was not found in any patient. The mechanism of initiation of puberty in various cerebral disorders remains unknown but hypothalamus may be involved by scarring, invasion, or pressure(1).

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