Persisting embryonal infundibular recess

The persisting embryonal infundibular recess (PEIR) is a rare anomaly of the floor of the third ventricle with a debated pathogenesis. It can be a cause of misdiagnosis in case of cystic lesions of the sellar and suprasellar area.

In 1975 a X-ray examination of the intracranial ventricles with Dimer X made it possible to make the diagnosis of obstructive hydrocephalus due to a mass lesion of the midbrain and abnormal intrasellar recess of the third ventricle in a 34 year old female patient. Considering the location and ontogenetic and phylogenetic facts one can diagnose the second pathological finding as a persisting intrapituitary recessus infundibuli. A malformation due to inhibition during development is to be assumed [1)].

In 1978 a congenital malformation of the anteroinferior aspect of the third ventricle was described. This anomaly has been found only rarely in anatomical descriptions [2)].

In 1979 a new variety of the “empty sella” consisting of polycystic intrasellar dilatation of the infundibular recess was described. The underlying embryonal developmental anomaly of the pituitary gland and diaphragma sellae and the possible effect of increased intracranial pressure are discussed and the malformation is compared with embryonal stages of ventricular development [3)].

In 1982 a patient with thrombosis of the central retinal artery, plain skull radiographs showed an enlarged sella turcica and computerized tomography revealed ventricular dilatation. Neuroradiological examination demonstrated stenosis of the Sylvian aqueduct and an unusual type of empty sella due to intrasellar persistence of the recessus infundibuli and the presence of an intrasellar arachnoidocele. The former anomaly has been reported in only three cases; in none of them was it associated with the latter abnormality [4)].

In 2009 Steno et al., reported 7 previously described cases invariably associated with hydrocephalus and, in 4 reported cases, with an empty sella. These associated findings led to speculations about the role of increased intraventricular pressure in the development of PEIR.

In the case reported by Steno et al., a 24-year-old man without the presence of hydrocephalus or empty sella. Disorders of pituitary function had been present since childhood. Magnetic resonance imaging revealed a cystic expansion in an enlarged sella turcica. A communication between the third ventricle and the sellar cyst was suspected but not apparent. During transcranial surgery, the connection was confirmed. Later, higher-quality MR imaging investigations clearly showed a communication between the third ventricle and the sellar cyst through a channel in the tubular pituitary stalk. This observation and knowledge about the embryology of this region suggests that PEIR may be a developmental anomaly caused by failure of obliteration of the distal part of primary embryonal diencephalic evagination. Thus, PEIR is an extension of the third ventricular cavity into the sella. Although PEIR is a rare anomaly, it is important to identify when planning a procedure on cystic lesions of the sella. Because attempts at removal using the transsphenoidal approach would lead to a communication between the third ventricle and the nasal cavity, a watertight reconstruction of the sellar floor is necessary [5)].

In 2014 Kuroiwa et al., reported that a 81-year-old woman underwent endoscopic transsphenoidal

surgery for the intra- and supra-sellar cystic lesion. Intraoperatively a hole was confirmed over the sella turcica connecting the sellar cyst and the infundibular recess. Liquorrhea did not occur throughout the procedure. A computed tomography (CT) scan obtained immediately after surgery disclosed accumulation of air in the third and lateral ventricles, in addition to the intra- and supra-sellar region. Air accumulation resolved spontaneously after bed rest for 11 days and she was discharged without neurological deficits. However, she required the second transsphenoidal surgery to repair the sellar floor because of bacterial meningitis caused by liquorrhea on the postoperative day 23. A postoperative 3-tesla magnetic resonance image revealed a deep infundibular recess connecting the sella turcica and the third ventricle, which was considered to be PEIR. This is the first reported case describing the intraoperative findings of PEIR [6].

Belotti et al., evaluated two female patients, aged 34 and 50, referred to the Pituitary Surgery Clinic of the University of Brescia with the diagnosis of a sellar cyst and craniopharyngioma, respectively. Endocrine screening and visual field testing were normal. No signs of hydrocephalus or empty sella, as well as other indirect signs of intracranial hypertension, were visible on MRI scans. After a multidisciplinary re-evaluation, diagnosis of PEIR was made in both cases. Both patients are followed but have not developed any disturbance related to the PEIR in the following 18 months.

PEIR is a rare condition, probably unrecognized and the result of dysembriogenesis, which should be included in the differential diagnosis of cystic sellar lesions. Imaging features (funnel pituitary stalk and cyst in the sella) appear pathognomonic. A normal endocrine evaluation might help in the diagnosis and warrants conservative treatment [7].

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