Pineal cyst

General information

PCs are nonneoplastic and may be due to ischemic glial degeneration or due to sequestration of the pineal diverticulum. They have been regarded as benign, but the natural history is not known with certainty.

PCs may contain clear, slightly xanthochromic, or hemorrhagic fluid. In rare cases, they may enlarge, and like other pineal region masses, may become symptomatic by causing hydrocephalus by aqueductal compression, gaze paresis including Parinaud's syndrome, or hypothalamic symptoms. Positional H/As have been attributed to PCs; the theory is that the cyst could intermittently compress the vein of Galen and/or Sylvian aqueduct. This remains unproven since asymptomatic compression of the vein of Galen and the quadrigeminal plate has been demonstrated on MRI.

Epidemiology

Pineal cyst epidemiology.

Etiology

Etiology is obscure, PCs are nonneoplastic, and may be due to ischemic glial degeneration or due to sequestration of the pineal diverticulum.

Natural history

The natural history is not known with certainty [1].

Clinical features

Rarely, they may enlarge, and like other pineal region masses, may become symptomatic by causing hydrocephalus by aqueductal compression [2] , gaze paresis [3] including Parinaud's syndrome, or hypothalamic symptoms.

Positional H/As have been attributed to PCs, the theory is that the cyst could intermittently compress the vein of Galen and/or sylvian aqueduct [4].

This remains unproven since asymptomatic com- pression of the vein of Galen and the quadrigeminal plate has been demonstrated on MRI [5].

Diagnosis

May escape detection on CT because the cyst fluid density is often similar to CSF. MRI T1WI shows round or ovoid abnormality in the region of pineal recess, signal varies with protein content (isointense or slightly hyperintense). T2 weighted image occasionally show increased intensity [6].

Gadolinium occasionally enhances the cyst wall with a maximum thickness of 2 mm; irregularities of the wall with nodular enhancement suggests the lesion is not benign.

Epidermoid-dermoid cysts may also occur in the pineal region, and are larger and have different signal characteristics on MRI.

Usually an incidental finding (i.e. not symptomatic), seen on ≈ 4% of MRIs [7] or on 25–40% of autopsies [8] (many are microscopic).

PCs may contain clear, slightly xanthochromic, or hemorrhagic fluid.

Treatment

see Asymptomatic pineal cyst management.

Symptomatic lesions or those that show changes in MRI. Surgery to relieve symptoms and/or to obtain a diagnosis.

Hydrocephalus. Surgical options include:

1. Cerebrospinal fluid shunt: recommended only for lesions with appearance to typical PC as it does not obtain tissue for pathology. May not relieve gaze disturbance from direct pressure on tectal plate

a) CSF shunt

b) endoscopic third ventriculostomy (ETV). A few cases of regression of PCs after ETV have been reported

2. aspiration only (stereotactic or endoscopic): may not get enough tissue for diagnosis

3. cyst excision (open or endoscopic): relieves symptoms and establishes the diagnosis. Low morbidity.

Two workhorse surgical approaches to the pineal region include the occipital interhemispheric transtentorial and supracerebellar infratentorial approaches. Each approach provides unique benefits and drawbacks and is associated with morbidities. In this patient, we demonstrate the use of a minimally invasive, supine, lateral supracerebellar infratentorial approach to the pineal region (Video 1). The approach makes use of early access to the foramen magnum to release cerebrospinal fluid for cerebellar relaxation, followed by navigation-guided, minimal dissection of the supracerebellar potential space to arrive at the pineal cyst. Opening of the arachnoid membranes ventrolateral to the confluence of the deep cerebral veins allows for direct access to the cyst with minimal manipulation of the venous complex and risk for injury. The cyst is then fenestrated, and the cyst capsule is

removed. It should be noted that surgical fenestration does not guarantee relief of headache symptoms, and patients should be consoled about this preoperatively [9].

Outcome

They have been regarded as benign, but the natural history is not known with certainty [10]

Incidental pineal cysts typically show a benign course. In the adult population, they do not require long-term neurosurgical follow-up, because pineal cysts tend to remain a stable size. In asymptomatic patients, Nevins et al., recommend a single follow-up MRI scan at 12 months to confirm diagnosis. The patient should then be discharged if the cyst remains stable [11].

Starke et al. aimed to study pineal lesion characteristics on MRI to better distinguish benign pineal cysts from other pineal region malignancies as well as to determine which characteristics were predictive of the latter malignancies. They also aimed to study risk factors predictive of hydrocephalus or malignancy in patients harboring these lesions.

Starke et al. performed a retrospective review of a prospectively compiled database documenting the outcomes of patients with suspected pineal cysts on MRI who had presented in the period from 1998 to 2004. Inherent patient and lesion characteristics were assessed in a univariate logistic regression analysis to predict the following dependent variables: development of hydrocephalus, biopsy-confirmed malignancy, and intervention. Possible inherent patient and lesion characteristics included age, sex, T1 and T2 MRI signal pattern, contrast enhancement pattern, presence of cyst, presence of blood, complexity of lesion, presence of calcification, and duration of follow-up. Inherent patient and lesion characteristics that were predictive in the univariate analysis ($p < 0.15$) were included in the multivariable logistic regression analysis.

Of the 79 patients with benign-appearing pineal cysts, 26 (33%) were male and 53 (67%) were female, with a median age of 38 years (range 9-86 years). The median cyst radius was 5 mm (range 1-20 mm). Two patients (2.5%) had evidence of calcifications, 7 (9%) had multicystic lesions, and 25 (32%) had some evidence of contrast enhancement. The median follow-up interval was 3 years (range 0.5-13 years). Seven patients (9%) had an increase in the size of their lesion over time. Eight patients (10%) had a hemorrhage, and 11 patients (14%) developed hydrocephalus. Nine (11%) received ventriculoperitoneal shunts for the development of hydrocephalus, and 12 patients (16%) were found to have malignancies following biopsy or resection. In the multivariate analysis, contrast enhancement on MRI (OR 1.6, 95% CI 2.86-74.74, $p = 0.013$) and hemorrhage (OR 26.9, 95% CI 3.4-212.7, $p = 0.022$) were predictive of hydrocephalus. Increasing lesion size and hydrocephalus were near perfect predictors of malignancy and thus were removed from multivariate analysis. In addition, contrast enhancement on MRI (OR 8.8, 95% CI 2.0-38.6, $p = 0.004$) and hemorrhage (OR 6.8, 95% CI 1.1-40.5, $p = 0.036$) were predictive of malignancy.

Although cystic abnormalities of the pineal gland are often benign lesions, they are frequently monitored over time, as other pineal region pathologies may appear similarly on MRI. Patients with growing lesions, contrast enhancement, and hemorrhage on MRI are more likely to develop hydrocephalus and have malignant pathology on histological examination and should therefore be followed up with serial MRI with a lower threshold for neurosurgical intervention [12].

Complications

These cysts can become symptomatic, and there have been about 75 verified cases reported in the literature so far.

However, their hemorrhagic manifestation is extremely rare, with only 8 such cases being found in the literature.

Moreover, there have been no occurrences of repeat intracystic hemorrhage reported [13].

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