Pineal Cysts

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25.1 Introduction

Pineal cysts (PCs) are better and more often detected with the evolution and wide expansion of magnetic resonance imaging (MRI) machines [1, 2]. Patients present with nonspecific symptoms, with a significant part of them being purely asymptomatic [1, 3]. The natural history of PCs is not yet fully understood, and controversies exist regarding the most appropriate management [1].

25.2 Prevalence and Incidence

The prevalence is between 1% and 4.8% in the general population [2]. Young adults and pediatric patients have the highest prevalence [2], with a pediatric peak incidence of PCs at 10–14 years of age [4], and afterwards prevalence decreasing with age [5]. Several studies have also shown that an overall female predominance exists in pediatric as well as in adult populations [2, 6, 7]. Probably, the rate of prevalence is much higher, as autopsy studies have shown the existence of PCs in 25–40% of all cases [8, 9].

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25.3 Imaging Aspects

Computed Tomography (CT): Some cases are discovered fortuitously with a CT scan, frequently performed for head trauma. On the CT imaging, PCs have a round-shaped aspect and hypodense contents [1]. In some of the cases (30%), hyperdense contents or walls, represented by either hemorrhagic contents or cystic calcifications, can be visible [1, 10].

MRI: MRI is the gold standard in the imaging of the pineal region [1], and even if there are inherent differences between examinations because of different machines, protocols, or modalities used [11], there are some imaging aspects that most of the PCs share. They are round or ovoid, smooth edged, and well-circumscribed lesions, which are better visible on sagittal scans, with contents of the cyst usually homogeneous and isointense to cerebrospinal fluid (CSF). PCs can present as irregular nodular enhancement on MRI images, which is secondary to surrounding venous structures, combined with a displaced pineal gland [1, 11, 12] (Fig. 25.1a, b).

The existence of internal septations may be difficult to assess on routine MRI studies [11], but when high-resolution studies are performed, such as three-dimensional (3D) fast imaging employing steady-state acquisition (FIESTA) or brain volume imaging (BRAVO), a large proportion of cysts can be found to have one or multiple internal septations [11, 13].

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Non-peripheral contrast enhancement of a PC is considered atypical and is correlated in one study with a 70% malignancy rate [14]. Unfortunately, there is no 100% clear way to differentiate pineal cysts from other malignancies that originate in this area, such as pineocytomas, pineoblastomas, germinomas, or mature teratomas [12].

25.4 Natural History

The natural history of PCs is still unknown, with literature data suggesting that around 80% are stable for at least 3 years after diagnosis and some even longer [1, 2, 15] (Fig. 25.2a, b). Some

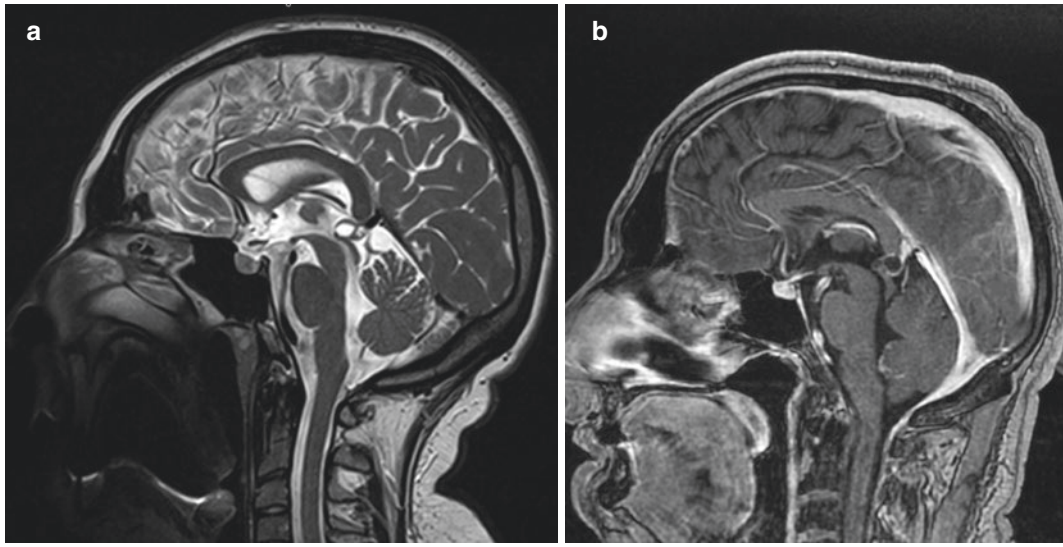

Fig. 25.1 (**a**, **b**) Sagittal T2 and T1+ contrast MRI sequences of a 23-year-old female performed for menses disturbance. Fortuitous discovery of a PC in an asymptomatic patient with no signs of hydrocephalus or visible postcontrast enhancement

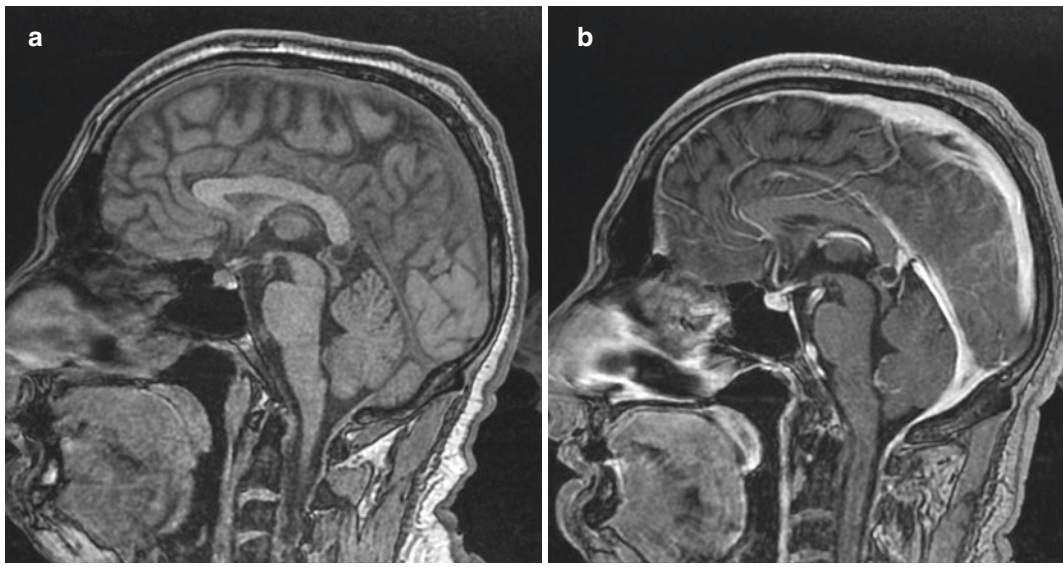

Fig. 25.2 (**a**, **b**) Sagittal T1 and T1+ contrast MRI sequences in a 30-year-old male with nonspecific headache. The cyst presents a peripheral contrast enhancement that is stable in size at 1-year follow-up (**b**).

cysts can even undergo a decrease in size, even to the point of complete involution [15]. A small proportion of pineal cysts can show an enlargement. The mechanisms behind this are poorly understood and are thought to be related to hemorrhage in the pineal cyst [16], the coalescence of smaller cysts [17], or hormonal influences, which can at least, in part, also justify the higher prevalence seen in females [5, 17, 18].

25.5 Clinical Features

The relation between size and symptoms: A few reports have tried to link the size of cysts to neurological signs and symptoms. Diameters larger than 1–1.5 cm appear to correlate more often with symptoms [8, 17, 19]. Nevertheless, other authors have described multiple cases of asymptomatic patients with cysts larger than 1 cm [2, 12, 15]. Up to this point, the size is not considered as a surgery indication in asymptomatic cases [1].

Nonspecific symptoms: Headache is a common symptom affecting half of the adult population [20], therefore it is not uncommon to have patients presenting both PCs and headaches. However, it is difficult to corelate one to each other in the absence of obstructive hydrocephalus [21]. There is data suggesting that melatonin dysfunction can be linked to headaches [22, 23] and it is important to look for other clinical symptoms related to melatonin secretion dysfunction, such as insomnia, delayed sleep phase syndrome, and desynchronosis in patients presenting with PCs [21].

Other non-specific symptoms that can be related to PCs are vertigo, nausea, fatigue, diplopia, tremor, seizures, and paresthesia [3], but thorough examinations should be performed before linking these symptoms to the presence of a PC (Fig. 25.3a, b).

Mass effect and typical neurological signs: Rarely, pineal cysts can grow large enough to cause compression on neighboring anatomical structures with associated typical textbook neurological signs: the quadrigeminal plate–oculomotor disturbances, Parinaud's syndrome, pupillary abnormalities, nystagmus retractorius, Sylvius aqueduct–obstructive triventricular hydrocephalus, Galen venous complex–intracranial hypertension, cerebellum–gait disturbances, ataxia, coordination anomalies, and the fornix–memory disturbances [1].

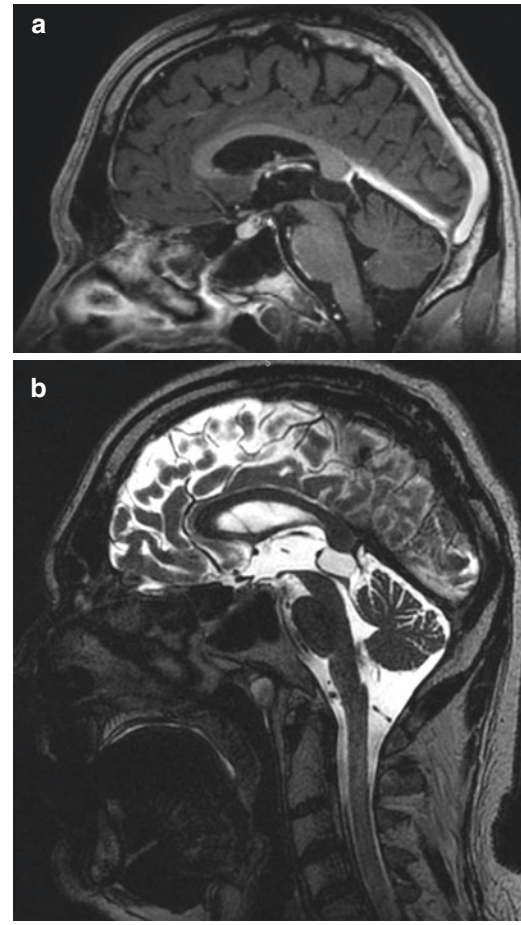

Fig. 25.3 (**a**, **b**) Sagittal T1+ contrast and T2 MRI sequences in a 54-year-old male who presented with morning generalized headache and diplopia. Imaging shows a PC with a diameter of about 2 cm, with the slight distortion of the Sylvius aqueduct. Nevertheless, a thorough neurological examination diagnosed the patient with myasthenia gravis with complete remission of symptoms following medical treatment.

25.6 Role of Surgery

Rationale for surgical treatment: The stable size of PC on long-term follow-up has convinced most authors to recommend that asymptomatic PC should not be surgically treated [1]. Surgery is recommended for symptomatic cases, presenting with clear neurological signs of compression and/or obstructive hydrocephalus [3].

Surgical treatment of cases with nontypical symptoms is controversial, with a recent study showing that venous congestion due to PC can lead to modified MRI biomarkers suggestive of a central venous hypertension syndrome [24]. The same authors report very good results, albeit in a small cohort, following PC removal in these nontypical symptom patients, but the exact mechanism by which these results are achieved is not clear [25].

25.6.1 Surgery Type

Microsurgery: Complete resection of the PC leads to far better results than just cyst fenestration [25]; this can be achieved either by supracerebellar infratentorial (SCIT) or by occipital transtentorial (OTT) approach. Unilateral SCIT in sitting position provides a natural corridor, and with surgical protection of the dominant transverse sinus and sparing of some cerebellar bridging veins, the risk of common complications such as diplopia and venous infarction of the cerebellum can be reduced [3]. The largest cohort for PC operated via sitting SCIT approach reported no postoperative complications [26]. OTT approaches the lesion from above, requires slight retraction on the occipital, and can lead to higher rates of postoperative complications, most common being transient hemianopsia, in 16.1–79% of cases [3] (Fig. 25.4a, b).

Shunting: In the absence of hydrocephalus, a recent study shows poor results with shunting, and the authors do not recommend the use of this surgical technique [25].

Other surgical techniques: Endoscopic fenestration or stereotaxic biopsy can only allow for cyst fenestration, and even if these procedures seem safe without significant complications, there are no clear reports of long-term results [3].

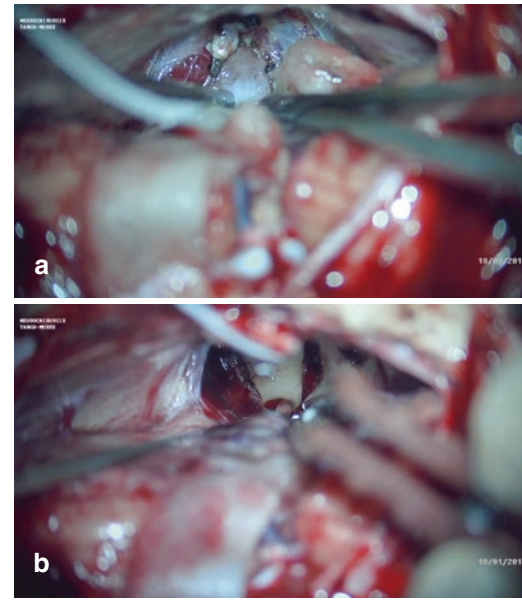

Fig. 25.4 (**a**) Intraoperative aspect showing the operating corridor via an SCIT approach and the thick whitish posterior wall of a pineal cyst. (**b**) Intraoperative aspect after complete removal of the cyst, with the opening of the third ventricle and visible Sylvius aqueduct

25.7 Conclusions

Pineal cysts are still a controversial pathology. Asymptomatic cysts should be monitored regardless of size. Complete resection of the lesions via supracerebellar infratentorial approach seems to provide the best results, even in cases with nonspecific symptoms, but due to limited data in the literature, surgical indications should still be limited and only placed after a thorough evaluation.

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