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Not only endoscopic third ventriculostomy – management of aqueductal stenosis associated with pathologies of the pineal region

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ABSTRACT
Pathologies of the pineal gland region are frequently encountered in daily neurosurgical practice. It is important to remember that, besides usually asymptomatic pineal gland cysts, neoplasms of a different origin may also occur in this region. The rarity of the most dangerous lesions – pineal gland apoplexy and pineal tumors – might cause problems with proper diagnosis and treatment. Their occurrence is commonly associated with symptomatic hydrocephalus caused by aqueductal stenosis. Currently, in such cases, endoscopic procedures, including endoscopic third ventriculostomy (ETV), are commonly and widely chosen as treatment options. Endoscopic procedures not only enable decompression of hydrocephalus but also further diagnosis of its cause. Herein, we present 3 case reports of endoscopic treatment with ETV performed in patients with hydrocephalus related to pineal region pathologies.
Keywords: pineal gland; pineal gland apoplexy; pineal gland tumor; endoscopic third ventriculostomy.

INTRODUCTION

Pathologies of the pineal gland region are among the most common incidental findings in neuroimaging, with pineal cysts being predominant. Whilst data show that such lesions may occur in 1–2% of patients undergoing magnetic resonance imaging (MRI), autopsies reveal its presence in more than 25% of cases, and the prevalence is significantly higher in female patients [1, 2]. In contrast, neoplasms of the pineal gland constitute less than 1% of all central nervous system tumors and occur mainly in male patients. Although pineal cysts and pineal gland tumors have diametrically different origins and dynamics of change, they can both mimic each other, thereby hindering the proper diagnosis. Pineal region pathologies, both pineal cysts and pineal gland tumors, may cause nonspecific symptoms mainly connected to the mass effect and aqueductal stenosis – a direct cause of hydrocephalus [3, 4]. Endoscopic surgery, like third ventriculostomy, is a commonly proposed treatment option as relatively safe and well-established in such cases [5, 6]. Herein, we present 3 case reports of endoscopic treatment with endoscopic third ventriculostomy (ETV) performed in patients with hydrocephalus related to pineal region pathologies.

CLINICAL PRESENTATION

Case 1

A 50-year-old male presented in January 2022 with walking disturbances. He reported a 10-year history of slowly worsening balance, particularly an altered, irregular walking gait, characterized by a wide stance. Upon admission, the patient exhibited good overall health, was conscious, fully cognizant, and presented no additional focal deficits. The MRI revealed obstructive active hydrocephalus caused by a tumor in the pineal gland region (Fig. 1). Laboratory tests returned normal results. In the neurosurgery operating room, a biportal endoscopic operation was performed, starting with an ETV and then a biopsy of the tumor. After the surgical procedure, temporary weakness of the right upper limb was observed, alongside significant improvement in gait. No adverse events were reported during the hospital stay, and the patient was discharged from the hospital in good health, demonstrating more confident walking abilities. The surgical wound healed completely without complications. The histopathological examination revealed a Grade I subependymoma tumor. The patient remains under observation, showing continued improvement in motor abilities.

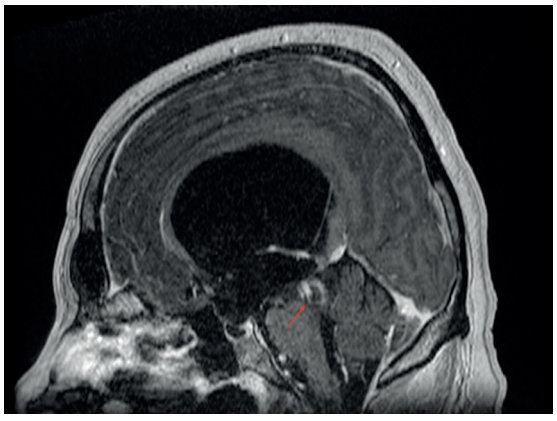

FIGURE 1. Diagnostic magnetic resonance imaging before the operation; the arrow marks the location of the tumor

Case 2

The second patient was an 80-year-old female, who was admitted to the hospital in December 2020. Prior to admission, she presented with gait and balance disorders, as well as memory

impairment. The patient's general condition was good, consciousness and cognition were normal, and she did not exhibit any further neurological manifestations. The MRI images showed a tumor mass in the pineal region, which was responsible for obstructive hydrocephalus (Fig. 2). Laboratory tests did not reveal any abnormalities, and the neurosurgeons performed an ETV with endoscopic resection of the tumor. Upon the patient's discharge, the wound had healed and the neurological condition had slightly improved. The postoperative pathology manifested an Ependymoma Grade II classification. Control MRI scans after 2 months and 1 year confirmed complete resection of the tumor without recurrence (Fig. 3). The course of hospitalization was uneventful. Due to the total resection and the benign character of the tumor, no adjuvant treatment was necessarry. In the almost 2-year follow-up, the patient remains in good general condition with controllable underlying disease and satisfying life quality.

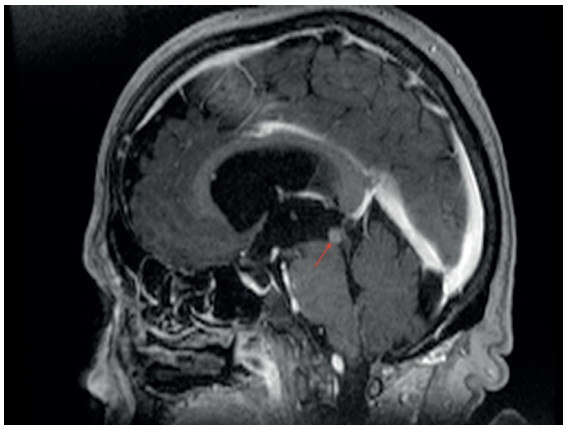

FIGURE 2. Diagnostic magnetic resonance imaging before the operation; the arrow marks the location of the tumor

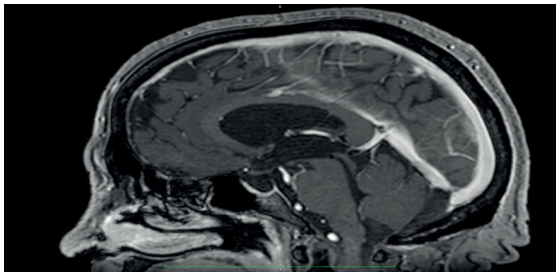

FIGURE 3. Diagnostic magnetic resonance imaging image after the operation; complete resection of the tumor

Case 3

A 21-year-old male presented to the hospital in April 2022 with a 1-month history of headaches, which exacerbation occurring on the day of hospital admission. A computed tomography (CT) scan in the Emergency Room revealed intracystic hemorrhage in a pineal cyst, which was later confirmed by MRI (Fig. 4). The neurological examination revealed a setting-sun sign, indicating upward gaze paralysis. Laboratory tests were normal. The patient was admitted to the Neurosurgical Unit and ETV with marsupialization and subtotal resection of the pineal cyst was performed utilizing a biportal approach (Fig. 5). The post-operative CT scan showed a significant reduction in cyst size (Fig. 6). After the operation, gradual improvement was observed; the wound was healing well, and the patient was discharged from the clinic on the fourth day after the operation. The histopathological examination revealed a pineal cyst with hemorrhage.

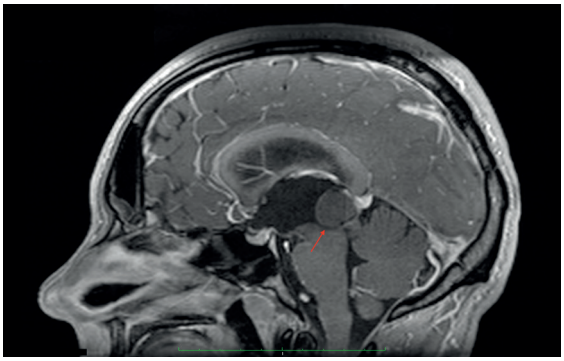

FIGURE 4. Diagnostic magnetic resonance imaging before the operation; the arrow marks the location of the cyst

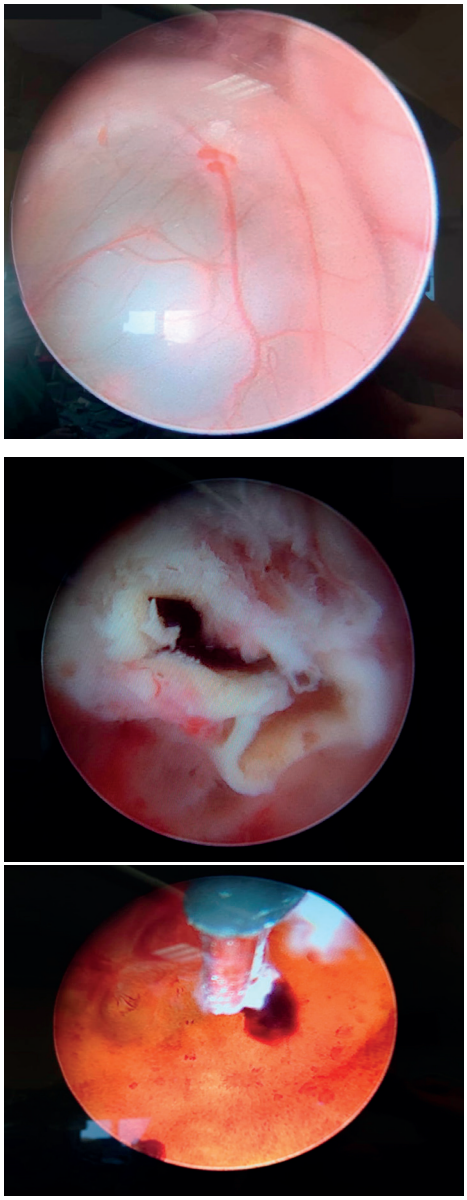

FIGURE 5. Intraoperative image: a) third ventricle floor; b) endoscopic third ventriculostomy being performed; c) completed endoscopic third ventriculostomy – enabled proper cerebrospinal fluid circulation

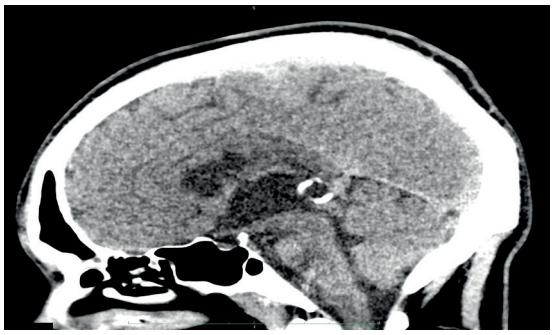

FIGURE 6. Diagnostic computed tomography scan after the operation; a significant cyst size reduction

DISCUSSION

Pineal gland pathologies are frequently encountered conditions in neurosurgical units and although their origins might vary, the treatments are very similar. Pineal region pathologies, such as pineal cysts and pineal gland tumors, are commonly associated with aqueductal stenosis – a direct cause of hydrocephalus – which may cause nonspecific neurological symptoms [3]. The most frequently observed manifestations are headaches and nausea, difficulties focusing sight, unsteady gait accompanied by sudden falls, or leg weakness. Irritability and drowsiness may also occur [7].

While pineal cyst and pineal gland tumors can mimic each other, hindering the proper diagnosis, their origin and dynamics of change are profoundly different. Pineal gland cyst prevalence is established at 1–2% of the population. They are mainly incidental findings on brain CT or MRI. The cysts consist of 3 layers: the glial layer, pineal layer, and Rosenthal fiber layer, containing cerebrospinal fluid [2]. Its incidence is higher in females and adults, and the size is not related to the patient's age or gender [1]. Apoplexy of the pineal gland cyst is an extremally rare diagnosis, and there are only a few case reports of such findings. Magnetic resonance imaging may confirm the intracystic hemorrhage diagnosis based on the characteristic fluid-fluid interface [8].

The causes of intracystic hemorrhages are diverse. The most common risk factor is antiplatelet or anticoagulant therapy [9, 10]. It is often accompanied by hypertension, which is commonly related to hemorrhages in different organs [11]. Conversely, there are reports in worldwide literature about anatomical abnormalities in pineal cysts that may be linked to hemorrhages [12, 13].

In comparison to uncomplicated pineal cysts, tumors of the pineal region commonly require surgical intervention or MRI follow-up [14]. This type of tumor is rare, established in only less than 1% of all brain oncological patients. Most of them occur in pediatric populations [3, 4]. Despite their rarity, pineal gland tumors can be classified as a wide range of different neoplasms. As reported by Regis et al. in their study on 370 patients aged 3–73 years, more than 50% of all pineal gland tumors are germinomas and astrocytomas, less than 25% are pineoblastomas and pinocytomas, and the rest consist of other rare findings are: ependymomas, teratomas, ganglioglioneuromas, lymphomas, meningiomas, and metastases [15].

Endoscopic third ventriculostomy is a commonly and widely chosen treatment option for hydrocephalus and is considered the treatment of choice for obstructive hydrocephalus. It is established as a considerably safe procedure enabling immediate clinical improvement of the patient and avoiding the placement of intraventricular shunt, which is regarded as a possible source of infection [16, 17].

The history of ventriculoscopy dates back to the early 1900s when the father of modern neurosurgery, Walter E. Dandy, performed choroid plexectomy in cases of communicating hydrocephalus using a simple endoscope. Unfortunately, the mortality rate proved to be high, which led to the abandonment of this treatment for the following years. Further attempts at endoscopic treatment of hydrocephalus are inextricably connected to early 20th-century urologists. Victor L'Espinasse, a Chicago urologist, was the first to use a cystoscope to cauterize the choroid plexus. The procedure was not reported by him in literature, dismissed as merely "an intern's stunt". It was not until 1923 that Wiliam Mixter revived the idea of using the urethroscope in the treatment of hydrocephalus, executing the first successful endoscopic ventriculostomy of the third ventricle. The following years brought many modifications such as stereotactic frames, better optic resolution, and upgrades to endoscopic equipment with deflectable tips and working ports [16, 18].

Importantly, endoscopic procedures not only enable the decompression of hydrocephalus but also further diagnostics of its cause. Modern neuroendoscopes allow for highly detailed visualization and documentation of ventricular anatomy. Moreover, this percutaneous technique is extremely valuable in its ability to identify and potentially eliminate the primary cause of hydrocephalus, e.g., a pineal cyst or a tumor of the pineal region. As total resection is often not possible, endoscopic procedures allow for proper diagnosis by providing biopsy material for histopathological examination. It is crucial to select cases properly and provide post-operative care, including monitoring of intracranial pressure and assessing the need for external ventricular drain, to increase the success rate and reduce complications [16].

CONCLUSION

Pathologies of the pineal gland region, though common, may cause difficulties in proper diagnosis. This is because the most dangerous ones – pineal gland apoplexy and pineal tumors – might be misdiagnosed due to their rarity. As a relatively fast and safe treatment, endoscopic procedures, often accompanied by ETV, are frequently used in both cases. These procedures enable a profound diagnostic process and complex therapeutic management, providing clear answers not only for patients but also for neurosurgeons. Endoscopic third ventriculostomy is a short, relatively safe procedure that facilitates complex and multidirectional diagnosis and treatment of aqueductal stenosis associated with pathologies of the pineal region.

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