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Case Report
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Pineal Gland Apoplexy, Case Presentation with Review of the Literature: Is There a Preferred Entry Point for Endoscopic Treatment of Pineal Apoplexy?

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Abstract

Introduction: Pineal apoplexy is defined as hemorrhage into the pineal gland that can be accredited to pineal cysts, tumors, or arteriovenous malformations. Although there are several etiologies, they share the same clinical manifestations of headache, gaze paresis, nausea and vomiting, acute obstructive hydrocephalus, and even death. Pineal apoplexy is a neurologic emergency and therefore an immediate surgical intervention to relieve the pressure of the mass effect caused by the hydrocephalus is necessary.

Case Description: We present the case of a 6 years old female patient who presented to us with signs of increased intracranial pressure. Work up including MRI brain showed pineal apoplexy with obstructive hydrocephalus. Surgical intervention was done including ETV (Endoscopic Third Ventriculostomy) and biopsy of the lesion. According to our practice, we utilized a single-entry point that provides access to the ETV site and that also allowed us to access the pineal region for biopsy.

Conclusion: Being a rare pathology, pineal apoplexy requires urgent treatment due to the obstructive hydrocephalus caused by the lesion. In this paper we present the case of a 6-year-old female patient who presented with obstructive hydrocephalus due to pineal apoplexy that was treated with ETV and biopsy of the lesion. In this paper we are the first to suggest new measurements to be taken when treating pineal apoplexy endoscopically in order to have one ideal entry point that can serve as a good trajectory to both the pineal region and the site of ETV.

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Introduction

Pineal apoplexy is defined as hemorrhage into the pineal gland that can be accredited to pineal cysts, tumors, or arteriovenous malformations. Pineal cysts are generally asymptomatic lesions of the pineal gland with an incidence rate between 1% to 10% of all patients but it can reach up to 40% of patients if examined postpartum [1,2,6]. Pineal gland tumors are rare and account for 0.4% to 1% of all intracranial tumors with histopathological diagnosis comprising ganglioglioma, pineocytoma, choriocarcinoma, meningioma, embryonal carcinoma, and metastatic melanoma [4,8]. It has been reported that only 14% of all intracranial tumors present with a microscopic hemorrhage into the tumor with choriocarcinoma and melanoma being the most common tumors [4]. The remaining etiology is AV malformations and they are considered rare. Although there are several etiologies, they share the same clinical manifestations of headache, gaze paresis, nausea and vomiting, acute obstructive hydrocephalus, and even death [1,4,9]. Pineal apoplexy is a neurologic emergency and therefore an immediate surgical intervention to relieve the pressure of the mass effect caused by the hydrocephalus is necessary. We will present a case of a 6-year-old female patient presenting with pineal apoplexy followed by a review of the literature and our recommendation for an entry point, that we consider ideal, which provides a direct access to the pineal region during endoscopic surgical management to both perform endoscopic third ventriculostomy to treat the hydrocephalus and to take a biopsy from the pineal region.

Case Presentation

We present the case of a 6 years old female patient who was born by cesarean delivery, to non-

Table 1: Literature review showing cases of pineal apoplexy with different treatment modalities.

Patient #	Age (year)/sex	PMH	Symptoms	Other findings	Hydrocephalus	Procedure	Post-op course
1 [3]	51/F	None	Severe headache with brief loss of consciousness	Bilateral papilledema with retinal hemorrhage	Present	Right occipital craniotomy	Upward gaze palsy
2 [4]	35/F	None	Collapses with dull right sided headache, mild gait ataxia, right sided blurry vision	-	Present	Right occipital micro-craniotomy	Transient downgaze and upgaze palsy
3-5 [5]	15/F	None	Photophobia, diplopia, intense headache, nausea and vomiting		Present	None	Spontaneous recovery
	12/F	None	Headache, nausea and vomiting	Cyst	Present	ETV only	Complete recovery
	4m/M	None	Macrocrania	Low grade glioneuronal tumor	Present	ETV	Complete recovery
6 [6]	12/F	None	Headache, nausea, vomiting, syncope	Pineal cyst	Present	EVD followed by ETV + infratentorial supracerebellar approach	Complete recovery
7 [7]	5/M	None	Headache, vomiting, clouding of consciousness	Bacterial meningitis in the past 10 days	Present	ETV	Complete recovery
8 [8]	11/F	None	Nausea and vomiting, lethargy, limitation of conjugate upward gaze, left abducens nerve palsy, bilateral tinnitus, and right hemihypesthesia of the face.	Cavernous angioma	Present	Occipital transtentorial approach + Ommaya reservoir + CSF drainage	Uneventful
9 [9]	31/F	None	Headache, vomiting, photophobia, upward gaze paresis	Pineal parenchymal tumor of intermediate differentiation	Present	VP shunt + infratentorial supracerebellar approach	Complete recovery

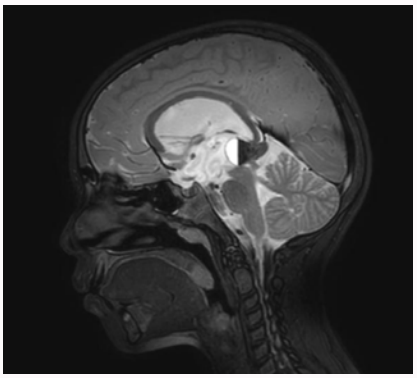

Figure 1: Sagittal T2-weighted image showing a pineal lesion with solid and cystic component containing layering blood products.

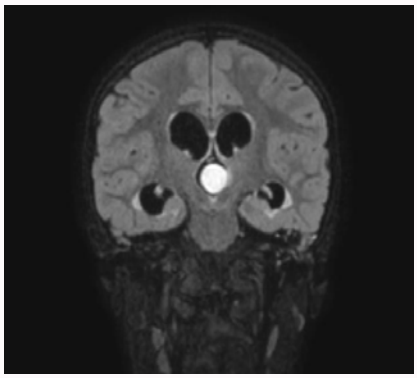

Figure 2: Coronal T2-FLAIR-weighted image showing pineal lesion in the upper part of the aqueduct with resultant moderate to severe acute hydrocephalus, and secondary mild to moderate trans-ependymal CSF seepage.

consanguineous parents who is known to be previously healthy till 2 months prior to presentation when she started to experience retractable headaches awakening her from sleep but mildly resolved on oral pain medication. She then developed blurry vision with vomiting and lethargy. She had normal findings on the neurological physical exam. Lumbar puncture was done as part of the work up that showed normal CSF studies. CT brain showed Pineal region lesion compressing the 4th ventricle and causing ventriculomegaly. MRI brain was then done which showed a heterogeneously enhancing solid lesion in the region of the pineal gland measuring 2.2 × 1.8 × 1.5 (AP × TR × CC) with a cystic component containing layering blood products in the dependent portion (Figure 1). The lesion compresses the tectum and obstructs the upper part of the aqueduct with resultant moderate to severe acute hydrocephalus, and secondary mild to moderate trans-ependymal CSF seepage (Figure 2, 3).

Discussion

While pineal cysts being common lesions, can affect all ages, and arise from many pathologies affecting the pineal gland. Pineal apoplexy is considered a rare etiology behind a pineal cyst [6]. This rare entity is not well understood but can be defined as an acute onset neurological compromise as a result of a hemorrhage of a pineal tumor or a rupture of pineal vascular malformation [8,9]. Many neoplasms have been found to cause pineal apoplexy which sometimes can arise from an ischemic stroke affecting a pineal tumor mainly due tumor mass effect on the supplying vessels or due the tumor being

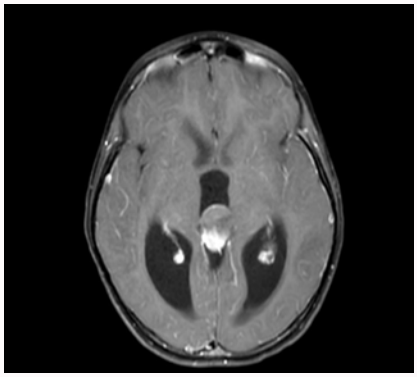

Figure 3: Axial T1-weighted image with gadolinium injection showing a pineal lesion that is heterogeneously enhancing.

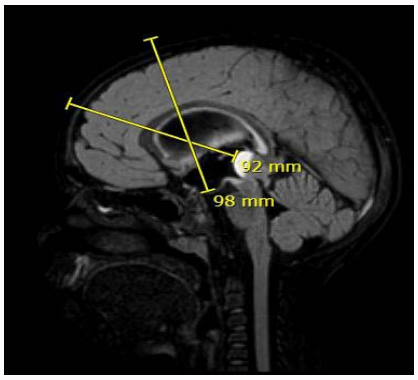

Figure 4: One entry point to target the pineal region and the ETV site.

too big to have adequate blood supply. The symptoms vary and include headache, nausea and vomiting, and visual disturbances which were the most common presenting symptoms in the literature. Hydrocephalus is almost always present on the initial presentation, hence surgical intervention to relieve the obstructive hydrocephalus has to be done in an urgent fashion.

Literature review (Table 1) by Swaroop et al. [8] of patients with pineal apoplexy included patients with average age of 37 years, with no sex preference (1:1 ratio), with the most common presenting symptoms of severe headache with ophthalmologic diseases. Four (4) out of 11 patients had sudden collapse and death as the presenting symptoms. Histopathologic findings include pineal cyst, pineocytoma, and ganglioglioma. Wang et al. [9] showed an average age of presentation of 34 years, with no sex preference (1:1 ratio), with the most common presenting symptom being headache and ophthalmologic symptoms including upward gaze paresis, visual loss, and dilated-non-reactive pupils. Schipmann et al. [7] studied 13 patients with spontaneous regression of the pineal gland lesions showed several hypotheses for regression. They include diagnostic radiation, corticosteroid use, surgical biopsy, treatment of hydrocephalus, lesion apoplexy, and finally the patient's immune response. The reported age range was between 3 months to 73 years with a median of 17 years. The histopathology findings of the lesions include germinoma, meningioma, pilocytic astrocytoma, and Low-grade glioneuronal tumors. Treatment of pineal apoplexy varied in the literature and was divided into either ETV to treat the obstructive hydrocephalus or craniotomy for resection of the lesion with CSF diversion that was either a VP shunt or an ETV.

In our case, the patient presented with sudden neurological deficits and imaging confirmed the presence of hydrocephalus precipitated by a pineal lesion compressing the 4th ventricle.

The decision of surgical intervention was taken. We propose a new entry point that can be used during endoscopic approaches to both take a biopsy from the pineal lesion to confirm our diagnosis and at the same time to treat the obstructive hydrocephalus through performing an ETV. Based on our experience, if we draw a line on a sagittal cut of the MRI image, that crosses the anterior wall of foramen of Monroe lying below the Massa intermedia, with another line that passes through the posterior wall of foramen of Monroe superior to the Massa intermedia we will be able then to have the distance between these 2 lines on the surface of the skin where our entry point can be taken as the midpoint of this line. This has proven to provide an ideal access entry point to perform an ETV and to take a biopsy from the pineal lesion without needing to have 2 different incisions with 2 different entry points. In our case, we started the procedure by marking the right frontal area over the optimal trajectory–which was determined by using the navigation system–for ETV and biopsy at the same time. Burr hole was placed 3.5 cm anterior to the coronal suture and 2.5 cm off midline. Endoscope was introduced and foramen of Monroe was identified on the right side. With the help of a stylet, the floor of 3rd ventricle was opened at the anterior 2/3 of the distance between the mammillary bodies and the infundibulum. We made sure that the Lilliquist membrane was opened and widened as well. Good CSF flow was noted. Then we turned the scope posteriorly and using endoscopic bipolar and grasper tumor was biopsied. The tumor contents were aspirated showing brownish fluid and were sent to cytology. Using the measurements suggested in our cases, access to both the tumor and to the site of ETV was guaranteed without needing to plan different entry points.

Conclusion

Being a rare pathology, pineal apoplexy requires urgent treatment due to the obstructive hydrocephalus caused by the lesion. In this paper we present the case of a 6-year-old female patient who presented with obstructive hydrocephalus due to pineal apoplexy that was treated with ETV and biopsy of the lesion. In this paper we are the first to suggest new measurements to be taken when treating pineal apoplexy endoscopically in order to have one ideal entry point that can serve as a good trajectory to both the pineal region and the site of ETV.

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