



Review Article

Hemiparkinsonism secondary to an epidermoid cyst with complete recovery after surgical resection: Case report and review of the literature[☆]



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ABSTRACT

Hemiparkinsonism is an exceptionally rare form of secondary Parkinsonism that manifests with unilateral symptoms and is often accompanied by ipsilateral body atrophy. Epidermoid cysts are benign, slow-growing tumors that can present with varying neurological symptoms. We report a case of a 30-year-old male who exhibited symptoms of Hemiparkinsonism secondary to a right-sided epidermoid cyst. The patient presented with three months of progressive resting left arm tremor and two weeks of left hand weakness. The patient exhibited a central left facial droop and left arm weakness with rigidity and bradykinesia. Unified Parkinson Disease Rating Scale (UPDRS) Part III score was 13. Gross total resection was achieved using a right fronto-temporal craniotomy via a preauricular, subtemporal approach. Complete reversal of symptoms occurred after surgical resection.

1. Introduction

Parkinsonism, although commonly associated with Parkinson's disease, consists of a neurological spectrum of different movement disorders. The respective association of these symptoms with primary brain tumors was first described in 1923 [1]. Since then, several cases of tumor-induced Parkinsonism have been reported, mostly due to primary gliomas and meningiomas [1–7]; however, few cases have ever been reported on the development of hemiparkinsonism secondary to these lesions [8–16]. Hemiparkinsonism, described by Klawans [17] in 1981, is an exceptionally rare form of secondary Parkinsonism that manifests with unilateral symptoms and is often accompanied by ipsilateral body atrophy. The clinical manifestations of the condition are heterogeneous and are often associated with variable levels of action-induced unilateral dystonia, magnetic resonance imaging (MRI) findings, and disease progression [18]. The pathogenesis of the disease is still poorly understood; it is hypothesized that an association between genetic deficiencies in dopaminergic neuron expression and environmental insults that result in subsequent nigrostriatal degeneration may

be the underlying cause [19]. Epidermoid cysts are benign, slow-growing tumors that can present with varying neurological symptoms [20]. As is true for other brain tumors, their presentation is dependent on anatomic location. We present the first reported case of a patient who exhibited symptoms of hemiparkinsonism secondary to an epidermoid cyst, with complete reversal of symptoms after surgical resection. This outcome may shed further light onto the neurological pathogenesis of the disease process.

2. Case report

2.1. History and examination

A 30-year-old otherwise healthy, left-handed male presented with chief complaint of three months of progressive left arm tremor and two weeks of left hand weakness. The tremor was characterized as being worse at rest and improving with movement. The patient also reported a mild left leg tremor that was present only upon elevation. He denied any gait disturbances, falls, constipation, olfactory loss, symptoms of

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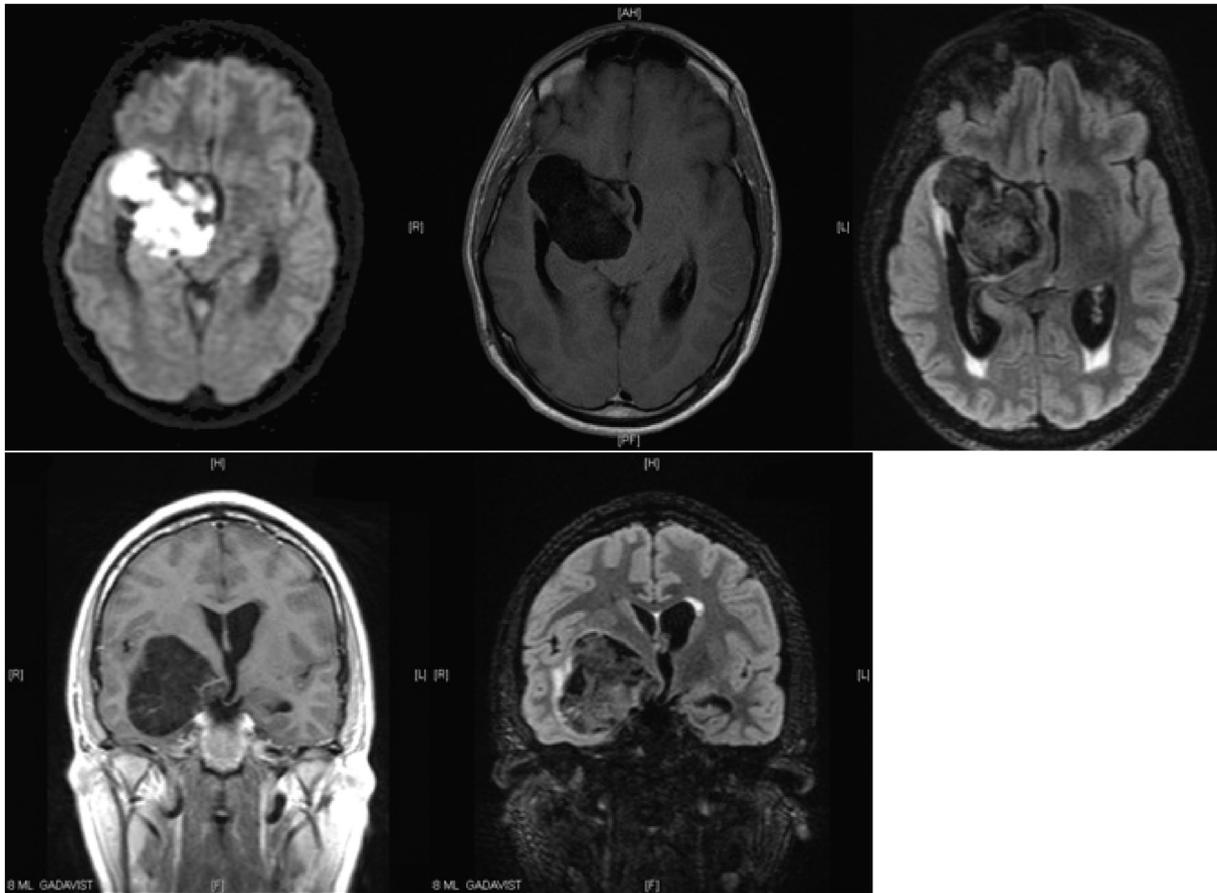


Fig. 1. Pre-operative imaging.

Top row shows axial DWI, axial T1 contrast, axial T2 FLAIR.

Bottom row shows coronal T1 contrast, coronal T2 FLAIR.

The lesion measured approximately 6.5 (anteroposterior) by 3.0 (transverse) by 5.0 (craniocaudal) centimeters. There was significant mass effect and edema with enlargement of the lateral ventricles with transependymal flow. There was a very small area of enhancement.

orthostasis, or memory loss. His family history was significant for a mother, maternal grandfather, and brother who had essential tremor.

On neurological exam, the patient exhibited a central left facial droop, left pronator drift, 4/5 strength of the extensor muscles of the left arm, and left-sided hyperreflexia. He also demonstrated rigidity and bradykinesia disproportionate to the weakness of the left arm, as well as a high frequency, proximally predominant resting tremor of the left arm, which re-emerged with postural changes. His arm tremor had a mild action component when performing finger-to-nose testing, and there was a postural, low frequency tremor of the left leg. He had decreased left arm swing with prominent tremor while walking. His Unified Parkinson Disease Rating Scale (UPDRS) Part III score was 13.

MRI of the brain demonstrated a $6.5 \times 3.0 \times 5.0$ cm lobulated, right temporal lesion that was T1 hypointense and T2 hyperintense, with an increased diffusion-weighted imaging (DWI) signal suggestive of epidermoid cyst (Fig. 1). Secondary to the lesion, the resulting hydrocephalus, periventricular edema, and compression of the right, lateral, and third ventricles, as well as the ventral midbrain resulted in a 6 mm right-to-left midline shift (Fig. 5a–c).

2.2. Operation

Urgent neurosurgical resection was indicated given the respective size of the tumor and subsequent mass effect on the brainstem. Based on the deep location of the lesion, an extradural modified, middle cranial fossa approach was employed (Fig. 6a). The patient was placed in a Mayfield head holder at a 40 degree rotation to the left, and a right

frontotemporal craniotomy was performed using a preauricular, subtemporal approach. A large portion of the sphenoid ridge was drilled out anteriorly and was continued down to the superior orbital fissure. A portion of the root of the zygoma was drilled down to maximize exposure to the floor of the middle cranial fossa (Fig. 6b). The bone of the temporal squamous coming down to the air cells was also drilled to identify the floor of the middle cranial fossa.

Using BrainLab (Munich, Germany) navigation, the middle and inferior temporal gyri were identified and a large corticectomy was performed with bipolar cautery. Since this was the non-dominant right side, we resected the temporal tip to 3 cm to expose the mass, and a larger linear corticectomy was performed on the inferior temporal gyrus to allow us direct access to the tumor (Fig. 6c). The lesion itself appeared shiny and pearly white, consistent with an epidermoid tumor. The sphenoparietal sinus was dissected out and coagulated to allow for easier mobility of the Sylvian fissure, and a Cavitron Ultrasonic Surgical Aspirator (CUSA) was used to debulk the lesion (Fig. 6d). Gross total resection of the tumor was achieved. We were able to preserve the crossing middle cerebral artery branches and the arachnoid around this area, as well as perforators to the brainstem.

2.3. Pathological findings

A $5.1 \times 4.6 \times 0.3$ cm specimen of white-tan soft tissue was sent to surgical pathology. Under microscopic inspection, the specimen revealed abundant keratin and fragments of a cyst wall lined by stratified squamous epithelium, which was consistent with an epidermoid cyst

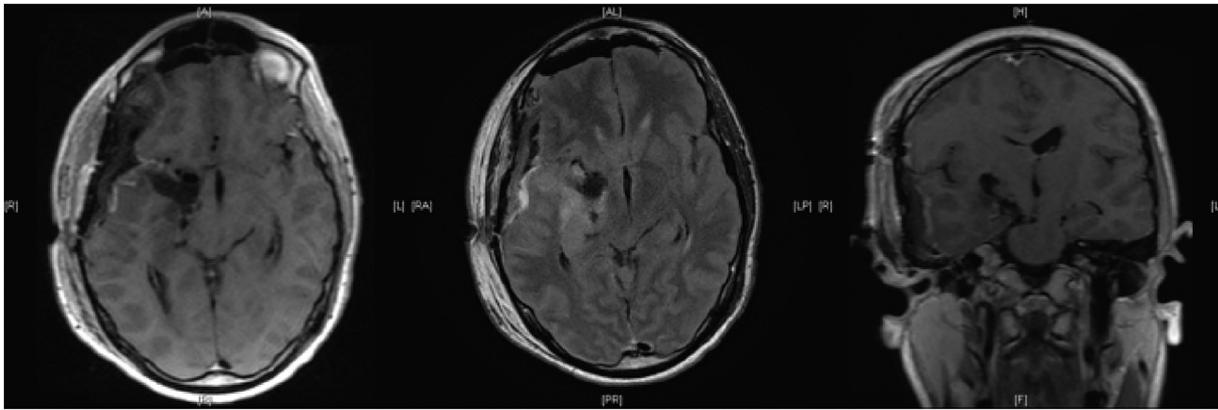


Fig. 2. Post-operative imaging.

Axial T1 contrast, axial T2 FLAIR, and coronal T1 contrast: Post-surgical changes are noted in the area of the tumor resection, documenting gross total resection and no secondary hydrocephalus.

(Fig. 3).

2.4. Postoperative course

Postoperatively, the patient remained neurologically stable and received phenytoin and levetiracetam for increased epileptiform activity of the right temporal lobe that was seen during surgery. Continuous video electroencephalogram (EEG) was performed, which demonstrated epileptiform discharges and right temporal slowing (Fig. 4). Follow up examination two weeks after surgery showed resolution of the left arm tremor, normal strength, and absence of pronator drift. His postoperative UPDRS Part III score was 1.5. Postoperative MRI showed gross total resection of the epidermoid cyst along the medial aspect of the right temporal lobe with decreased right-to-left midline shift (Fig. 2). A repeat EEG six weeks postsurgery showed no epileptiform discharges. Unfortunately, the patient was lost to follow up six months postoperatively, despite multiple attempts to contact him. At the time

of last visit, the patient reported complete resolution of symptoms.

3. Discussion

This is the first reported case of an epidermoid cyst causing Hemiparkinsonism. An earlier study discussed an isolated tremor caused by an epidermoid tumor, though the patient did not exhibit signs of bradykinesia, rigidity, or postural instability [3]. Epidermoid cysts are slowly growing, benign lesions, which account for 0.2–1.8% of all brain tumors and can present with varying neurological symptoms [20]. Although they are considered to be extra-axial lesions arising from the basal surface of the skull, studies have shown that they may also be intra-axial, as demonstrated by a retrospective review which showed 38 intraparenchymal cases, most commonly in the frontal and temporal lobes [21]. These results suggest that the location of epidermoid cysts is influenced by developmental factors that result secondary to anomalies of epithelial cell migration or retention of

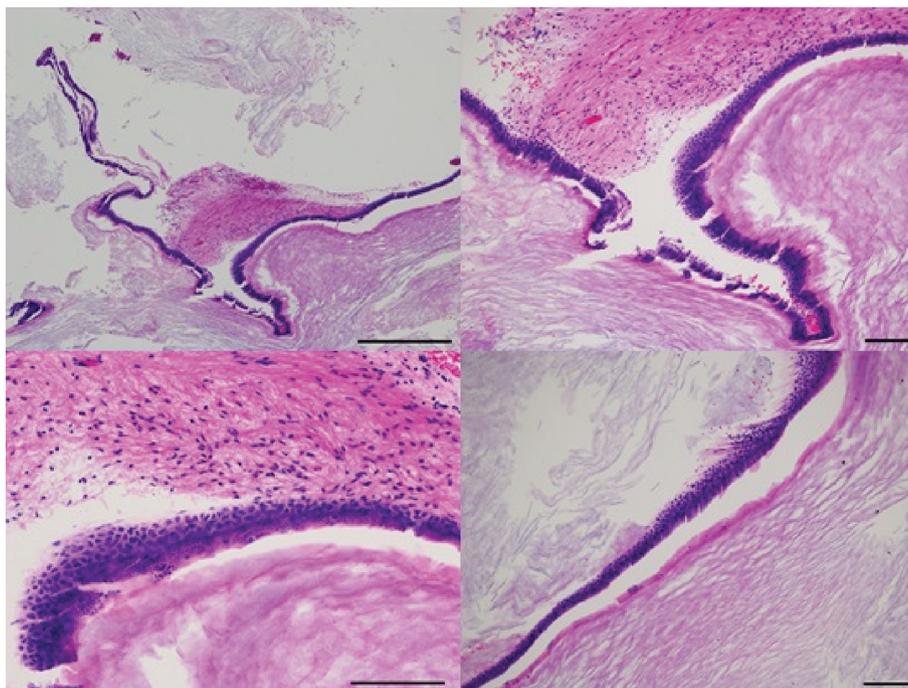


Fig. 3. Histopathology.

Hematoxylin and eosin (H&E) stain revealing layers of keratin and fragments of a cyst wall lined by stratified squamous epithelium, consistent with an epidermoid cyst.

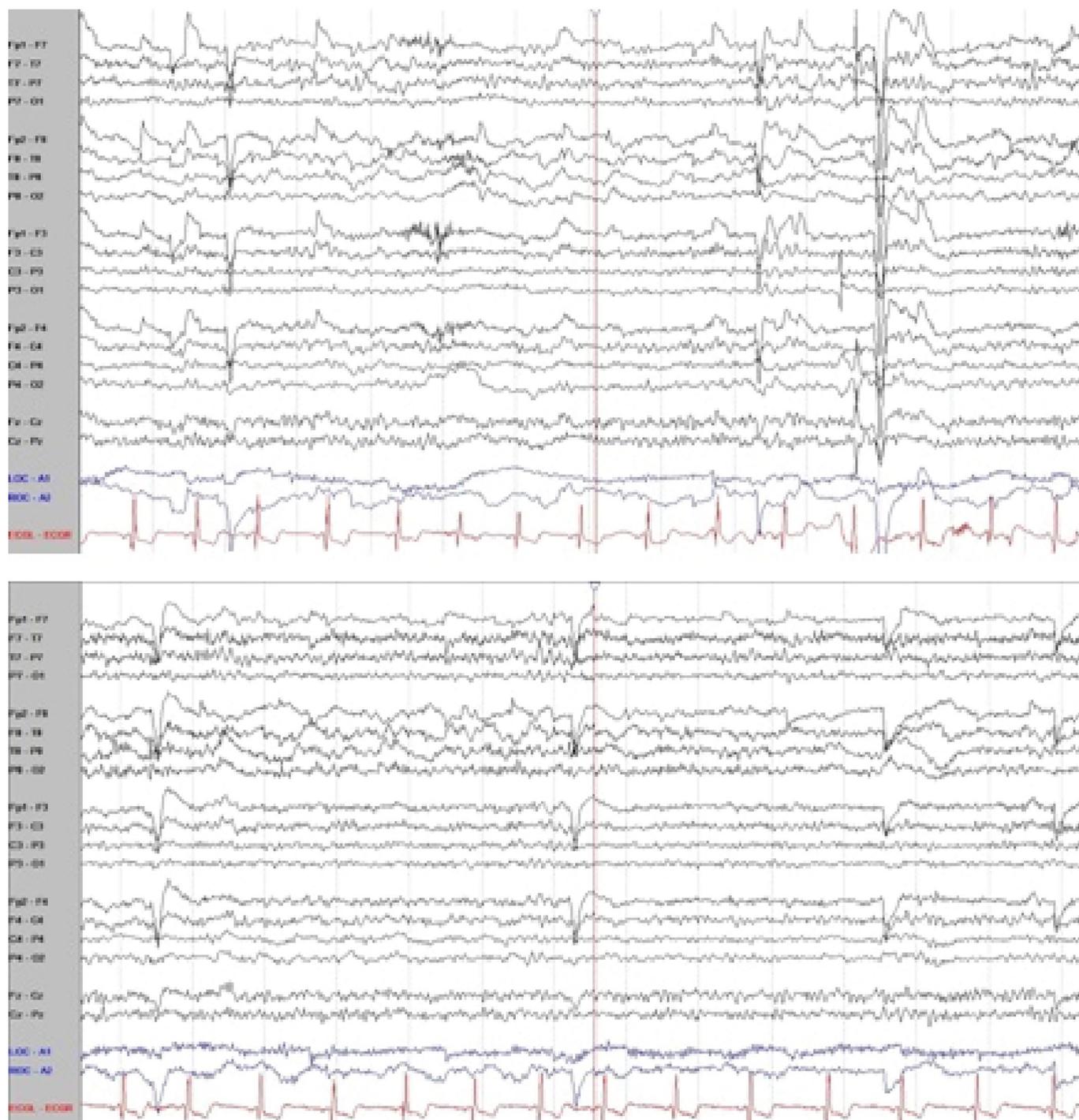


Fig. 4. Intra-operative EEG findings. Continuous EEG tracings demonstrating right temporal slowing.

ectodermal elements [19]. These changes occur when the neural progenitor cells divide from the ectoderm, and remnants of cells are found on the inner or outer surface, or within the neural tube, explaining why these epidermoid tumors can appear intraventricularly, on the surface, or within the brain parenchyma itself [19,22].

A large case series from 1953 analyzing 474 patients with primary brain tumors first demonstrated the occurrence of tumor-induced movement disorders [1]. In this study, ten patients had abnormal movements including dystonia and paresis, and 7 of them had Parkinsonism. This study highlighted the importance of recognizing such lesions as primary causes for these symptoms. The respective locations of

the tumors from this patient population included the frontal, temporal and occipital lobes, as well as the corpus callosum, thalamus, and basal ganglia. The pathological diagnoses included astrocytoma, glioblastoma, and meningioma. Tumor-induced Parkinsonism occurred with an incidence of 0.3%, most commonly from supratentorial masses that compressed and distorted the basal ganglia [1,3]. Intrinsic basal ganglia and thalamic tumors were found to be rare causes of Parkinsonism [2].

Several mechanisms have been proposed to describe the pathogenesis of Hemiparkinsonism. These proposals include compression of the basal ganglia from supratentorial tumors, distortion and stress of the

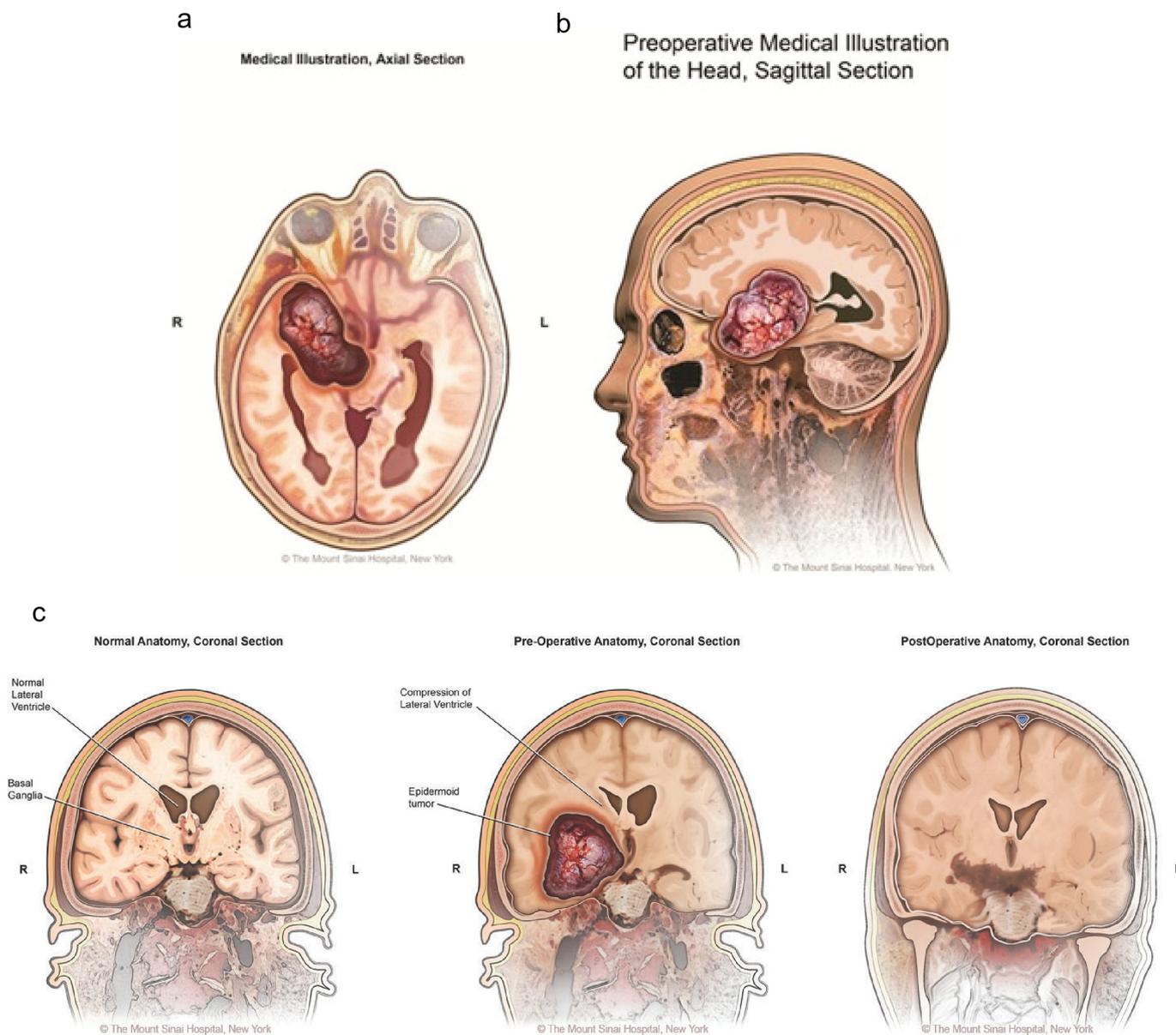


Fig. 5. ab: Cross-sectional illustration of pre-operative lesion with overlying radiographic imaging. Axial (left) and sagittal (right) cross-sections.

c: Medical rendition of disease progression with overlying radiographic imaging.

A three-panel illustration of normal, pre-operative, and post-operative brain anatomy depicting the respective dimensions of the lesion and architectural changes.

midbrain resulting in deficient oxygenation and decreased dopamine production [19], invasion of the substantia nigra with direct involvement of the basal ganglia [11,18], as well as damage to fibers connecting the basal ganglia with higher-order cortical regions [7]. In our case, we believe that the most likely mechanism, in light of the patient's immediate recovery, is transient distortion and compression of the basal ganglia without significant destruction of dopaminergic neurons.

Evaluation for tumors in patients presenting with symptoms of Parkinsonism is recommended in those with young onset, atypical presentations, poor response to levodopa, postural instability without rigidity, and other unexplained neurological symptoms [3]. Treatment and prognosis often depend on the mechanism of disease progression, as well as tumor type and location. With compressive lesions, resection may help to improve symptoms. For epidermoid cysts, regardless of location, the rate of recurrence depends on the amount of tumor resected; subtotal resection rather than total resection has been shown to result in faster rates of recurrence [10].

In our case, we were able to achieve gross total resection by employing an extradural, modified middle cranial fossa approach. This approach minimized brain manipulation and maximized operative corridor [23]. Moreover, the resection of the sphenoid wing and its bony contours allowed for multiple operative trajectories. Advantages of this modified skull base approach include the ability to operate without retractors, as well as providing a flat, wide angle exposure for maximal illumination under the microscope.

4. Conclusion

Hemiparkinsonism can present in a variety of clinical disorders. We report here the first description of an epidermoid cyst causing true neurological Hemiparkinsonism in which the symptoms improved immediately with surgical resection. The deep location and clinical presentation of this tumor may help to elucidate a further understanding of the pathogenesis of Hemiparkinsonism and its disease states. Surgical

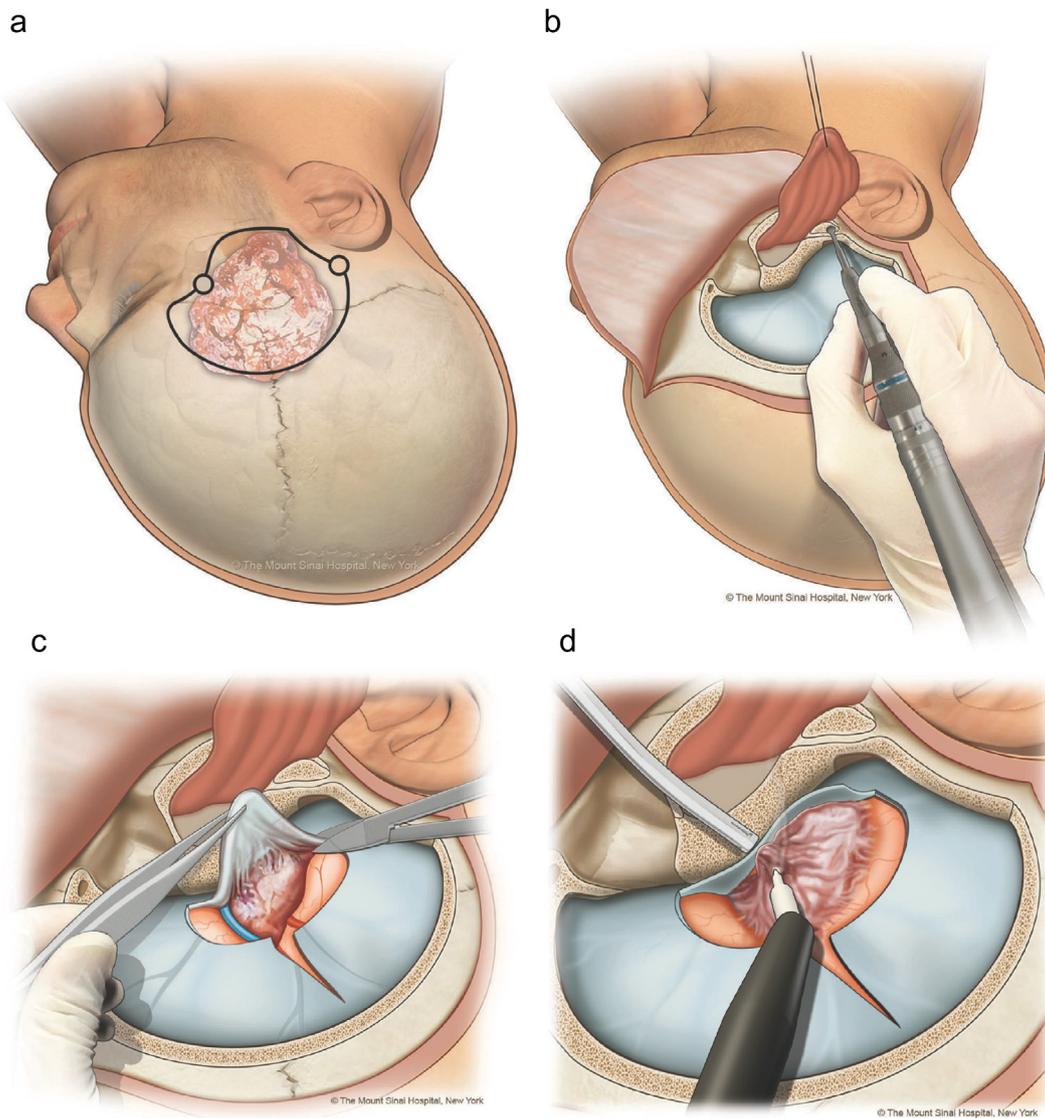


Fig. 6. Surgical procedure illustrations.

- Modified subtemporal approach with ghosted craniotomy.
- Zygoma bone drilling and resection for exposure and trajectory.
- Initial dural opening and tumor exposure.
- Intra-operative debulking with sonic aspirator.

resection of these lesions may prove beneficial for symptoms resulting secondary to compression of brain architecture.

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