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ORIGINAL ARTICLE

Endoscopic transnasal resection of optic pathway pilocytic astrocytoma

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Abstract

Purpose Optic pathway gliomas (OPGs) are low-grade neoplasms that primarily affect children. The management of OPGs remains controversial. Reports on the use of the endoscopic endonasal approach (EEA) in OPGs are extremely limited, and no such reports exist on its utility for pediatric OPGs. Here, we report our results and experience with OPGs treated with the EEA.

Methods We retrospectively reviewed the medical records of OPG patients who were treated surgically via the EEA at our institutions from 2015 to 2017. Data on the demographics, clinical presentation, surgical complications, clinical outcomes, radiological imaging, and visual outcomes were recorded for each patient.

Results Four cases were identified, with visual disturbances being the predominant complaint. The mean patient age was 15.5 years. Three cases showed normal preoperative hormonal profiles, but one patient had hypothyroidism. All tumors identified in this study were World Health Organization grade I pilocytic astrocytomas. Surgical complications included hypopituitarism in two patients, meningitis in two patients, cerebrospinal fluid leak in one patient, and transient diabetes insipidus in one patient. No patient experienced worsening neurological or visual symptoms postoperatively.

Conclusions Although our data are preliminary, the EEA provides a direct corridor to OPG with acceptable results in terms of tumor resection and visual outcomes. Hypothalamic-pituitary axis dysfunction remains a limitation of any treatment modality for OPGs and should be considered whenever possible. Definitive conclusions are pending as the learning curve of this approach is steep. Further work is needed to understand patient selection for such an approach.

Keywords Endoscopic transnasal surgery · Endoscopic endonasal approach · Hypothalamic glioma · Optic pathway glioma

Introduction

Optic pathway gliomas (OPGs) are low-grade, pathologically benign neoplasms that primarily affect children [1, 2]. They represent approximately 2–5% of all pediatric intracranial tumors and are found in approximately 15% of patients with neurofibromatosis type I (NF1) [1–3]. Pilocytic astrocytoma is considered to be the most common pathological variant of OPGs [1, 2]. The clinical characteristics of OPGs vary depending on the tumor location, size, and extension, as well

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as patient age at diagnosis; visual disturbances, headaches, and hypothalamic dysfunction are the most common presenting signs and symptoms in patients with OPGs [1, 3].

Given the critical location, the histologically benign nature, and the considerable risk of complications following surgical intervention, OPG is a challenging entity to manage. In cases where surgery is considered, these lesions are usually approached by an anterior or anterolateral transcranial routes [4, 5]. Alternatively, a few recent small series have shown the feasibility of an endoscopic endonasal approach (EEA) for this pathology [5, 6].

Over the past decade, significant advances have been achieved in endoscopic skull base surgery, expanding this approach beyond the limit of pituitary lesions. Currently, the EEA is utilized for intradural lesions, such as meningiomas [7] and craniopharyngiomas [8], and is increasingly utilized for intraparenchymal tumors [5, 6, 9–11]. The minimally invasive endoscopic transnasal route provides a direct visualization to the selected skull base region through multiple corridors, thereby avoiding brain retraction and manipulation. This has been mainly reflected in shorter hospital stays, fewer complications, and better cosmesis [12, 13]. While the EEA has been well described for most common skull base tumors, its utility and limitations for more complex tumors, such as OPGs, remain unclear.

In the present study, the authors report their experience with the EEA in four patients with World Health Organization (WHO) grade I OPGs.

Methods

Patients

We retrospectively reviewed the medical records of OPG patients, who were treated surgically via the EEA at our institutions from 2015 to 2017. Each case was performed by a team composed of neurosurgeons and otolaryngologists, who have experience in endoscopic endonasal skull base surgery. The goals of surgery were primarily to establish the diagnosis, debulk tumor mass, and improve mass effect-related symptoms. Data on demographics, clinical presentation, surgical complications, clinical outcomes, and duration of follow-up were recorded for each patient. All patients underwent a complete preoperative hormonal assessment and were evaluated by endocrinologist, ophthalmologist, and rhinologist. Preoperative magnetic resonance imaging (MRI) was performed for every patient. Patients were seen 2 weeks after discharge from the hospital in neurosurgery and rhinology clinic. Also, the follow-up comprised endocrinological and ophthalmological evaluation. MRI studies were repeated 3 months after surgery and annually thereafter.

Surgical approach

The EEA has been described in details previously [14, 15]. In brief, under general anesthesia via an endotracheal tube, the patient was positioned in the supine position and placed in 3-point head fixation. The neuronavigation system was registered (using a thin cut computed tomography (CT) scan as a reference, merged with a T1-weighted MRI scan with contrast). The approach started with a middle turbinate resection, antrostomy, and an anterior and posterior ethmoidectomy. Next, a nasoseptal flap was elevated and placed into the nasopharynx and a posterior septectomy was performed. The sellar floor, anterior sellar bone, tuberculum sella, and part of the planum sphenoidale were removed, utilizing a 3-mm cutting burr, diamond burr, and kerrison punch. A wide dural opening was utilized to expose the tumor. The resection started with central debulking using an ultrasonic aspirator. The optic nerve, which had better vision, was identified and a small tumor residual was left on the optic nerve to avoid vision loss. Following this, the resection was completed using microscissors, Rhoton dissectors, and the ultrasonic aspirator. Closure was completed in multiple layers (fat, inlay fascia lata, solid reconstruct, nasal septal flap, surgical glue, and a nasal pack (Figs. 1 and 2)).

Results

A total of four patients, three children and one adult, were included in this study. The mean patient age at surgery was 15.5 years (range, 7–32 years), and the ratio of males to females was 1:1. None of the patients were NF1-positive. All patients had different degrees of visual impairment, and other symptoms included headache ($n = 2$), nausea/vomiting ($n = 2$), and cognitive impairment ($n = 1$). Table 1 summarizes the demographic and clinical characteristics of patients (Figs. 3 and 4).

Three patients did not receive any prior treatment for the lesions, but one female patient had undergone multiple prior surgeries for progressive OPGs, including two prior craniotomies for subtotal resection (one pterional craniotomy [complicated by right eye blindness] 7 years ago and one transcallosal craniotomy 4 years ago) and ventriculoperitoneal (VP) shunt placement for hydrocephalus 4 years ago. She underwent fractionated radiation therapy (54 Gy/30 fractions) for tumor progression 1 year after last surgery. Thereafter, she remained in a stable condition for one and a half year. Subsequently, she had tumor progression and was thus referred to our institutions for consideration of endoscopic endonasal surgery.

Subtotal resection was achieved in all cases, and no intraoperative complications were encountered. Histopathological diagnosis determined pilocytic astrocytoma, WHO grade I in all cases. Two patients underwent VP shunt insertion for

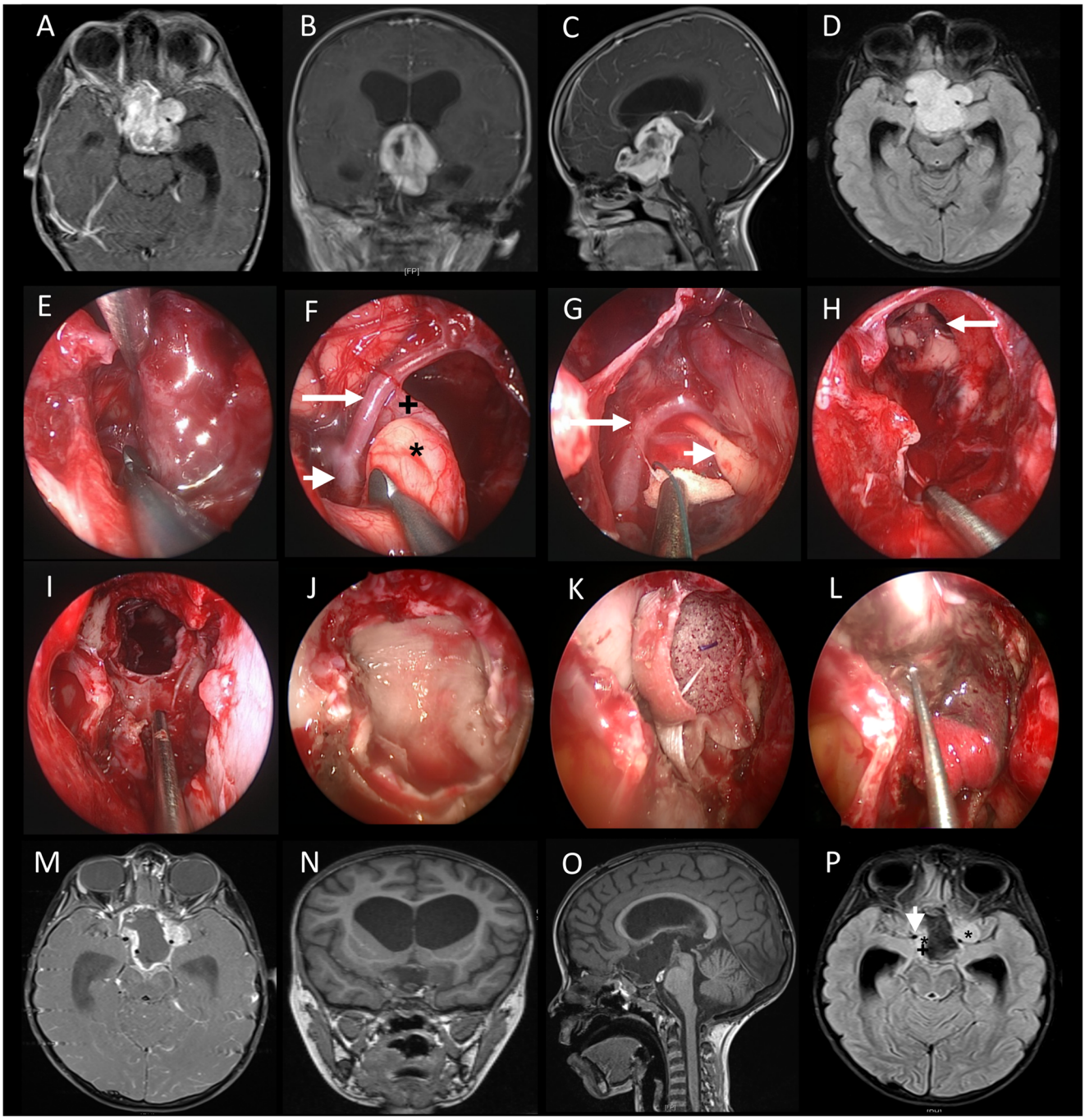

Fig. 1 Illustrative case 1: 7-year-old female who presented with symptoms of gradual vision loss. Physical examination revealed bilateral visual acuity deficits that were worse in the left eye. Pre-operative imaging; **a** axial, **b** coronal, **c** sagittal T1-weighted contrast-enhanced MR imaging, and **d** axial flair MR imaging showing a suprasellar mass involving the third ventricle. **e** After opening the dura, tumor exposure, and central debulking, we proceeded with establishing a plane between the tumor capsule, optic pathway, and ICA (at the side with good vision). **f** Showing the right optic pathway expanded because of tumor infiltration. The ICA (arrowhead), ACA (arrow), optic tract (+), and tumor residual (*) are seen. **g** The basilar artery (arrow) and the third cranial nerve (arrowhead) are clearly seen after completing the resection of the inferior posterior part of the tumor. **h** After tumor resection, the floor of the third ventricle and the foramen of Monro (arrow) are visible. **i** Transnasal view after completing the resection showing the skull base defect. **j–l** Reconstruction started with: **j** inlay fascia lata, **k** medpor solid reconstruct, and **l** septal flap. Post-operative imaging; **m** axial T1-weighted contrast-enhanced, **n** coronal, and **o** sagittal T1-weighted non-contrast MR imaging. **p** Axial flair MR imaging showing residual tumor (*) on the right and left optic nerves. The optic tract (+) and ICA (arrowhead) (correlating with the intraoperative finding shown in figure **f**) are demonstrated. ACA, anterior cerebral artery; ICA, internal carotid artery

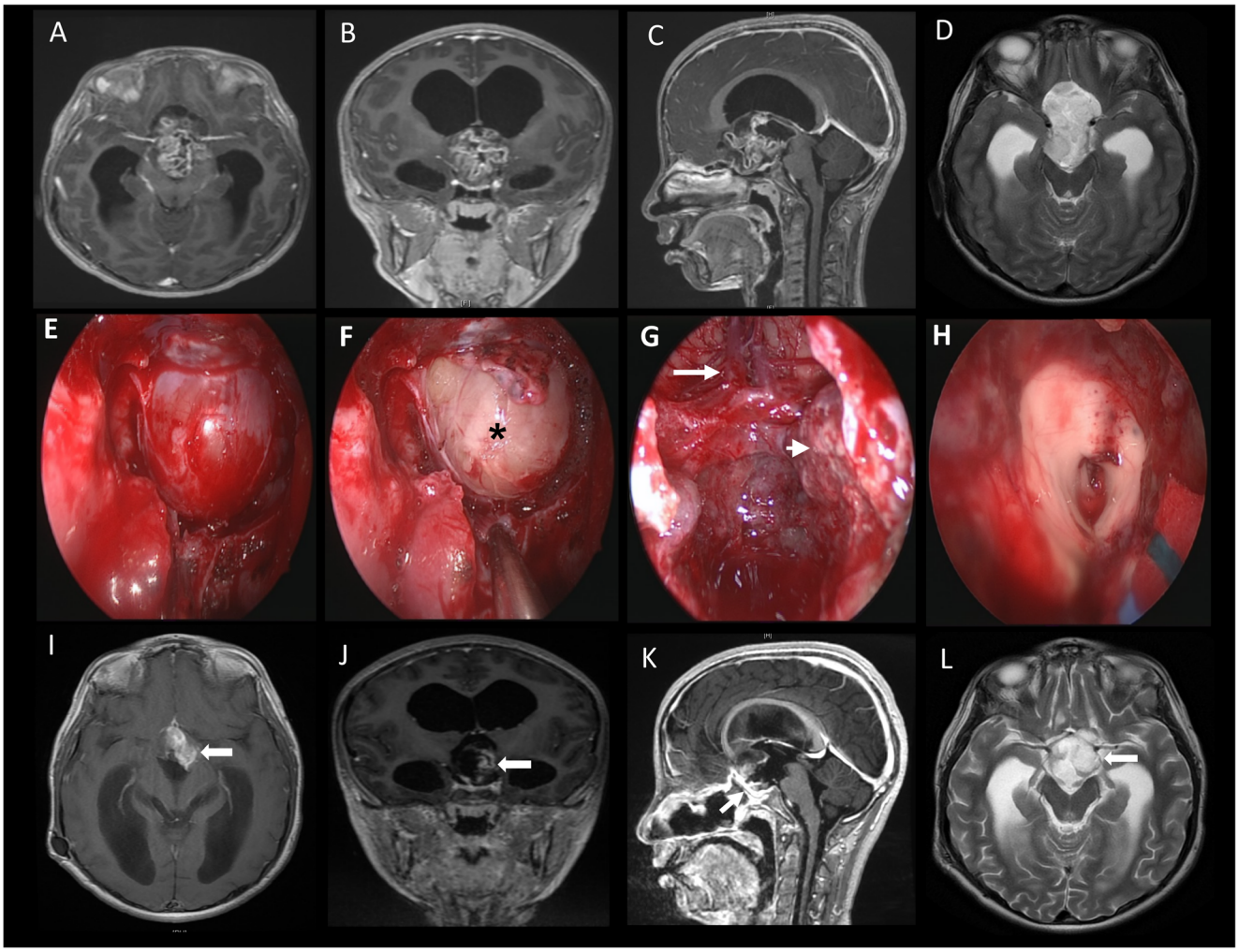

Fig. 2 Illustrative case 2: 12-year-old male who presented with gradually worsening symptoms of vision loss, headaches, nausea, and vomiting. Physical examination determined bilateral visual field and acuity deficits that were worse in the right eye. Pre-operative **a** axial, **b** coronal, **c** sagittal T1-weighted contrast-enhanced, and **d** axial T2-weighted MR imaging, showing a large heterogenous suprasellar mass involving the third ventricle. **e** Intra-operative exposure with the sphenoid and posterior ethmoid sinuses opened. Then, bilateral superior turbinate and right middle turbinate removal and posterior septectomy were done. The dura covering the planum sphenoidale, suprasellar space, and tuberculum sella is exposed. **f** The dura is opened, with the tumor (*) visible. **g** Tumor debulking is completed (arrowhead pointing to the tumor capsule attached to the optic nerve; arrow pointing to the ACA). **h** The floor of the third ventricle after tumor resection is seen. Post-operative **i** axial, **j** coronal T1-weighted contrast-enhanced, and **l** axial T2-weighted MR imaging showing residual tumor (arrow) left on the left optic nerve. **k** Sagittal T1-weighted contrast-enhanced MR imaging demonstrates the septal flap (arrow) in place. ACA, anterior cerebral artery

hydrocephalus preoperatively. Other two patients had intraoperative external ventricular drains, which were replaced with VP shunts 2 weeks and 4 weeks post-operatively, respectively. Three patients continued to have shunt-dependent hydrocephalus at latest follow-up.

One patient had abnormalities in hypothalamic-pituitary axis function at time of surgery which remained unchanged post-operatively. Additionally, the same patient developed transient diabetes insipidus (DI). Of other three patients who had normal pituitary function at time of surgery, two patients had new-onset hypopituitarism (hypothyroidism, permanent DI, and secondary adrenal insufficiency); one patient was off steroids, and another patient was off desmopressin at the latest follow-up visit.

Other complications included tension pneumocephalus and cerebrospinal fluid (CSF) leak, complicated by meningitis in one patient as well as isolated meningitis that occurred in another patient. The postoperative CSF leak was successfully treated with endoscopic endonasal re-exploration. There was no worsening of vision postoperatively in any patient. Opthalmological testing showed stable vision in all cases.

One patient had disease progression after a mean follow-up of 15 months (range, 6–28 months). Other three patients continued to be in stable conditions, which required no adjuvant

Table 1 Patient characteristics and clinical presentation

Case	Age (years)	Sex	Clinical presentation	Previous surgery
1	7	Female	Visual disturbance	No
2	12	Male	Headache, visual disturbance, nausea, and vomiting	No
3	32	Female	Hallucinations, nausea, and vomiting	Yes
4	11	Male	Headache and visual disturbance	No

treatment. Table 2 summarizes the complications and clinical and endocrinological outcomes of patients.

Discussion

OPGs can be classified anatomically into tumors that arise in the anterior visual pathway and those that arise in the posterior visual pathway (optic chiasm, hypothalamus, and third ventricle) [16]. OPGs with NF1 usually arise in the anterior visual pathway and have the potential to remain asymptomatic. Additionally, anterior pathway tumors predominantly present with proptosis or vision loss and are limited to one optic nerve [17], as observed in case 4 in this report. Furthermore, posterior pathway OPGs present with bilateral loss of visual fields as well as visual acuity in addition to abnormalities in the hypothalamic-pituitary axis and hydrocephalus [17].

Although OPGs are considered to be benign, the natural history of OPGs is unforeseeable. OPGs have the potential to rapidly progress, remain stable, or regress spontaneously [18]. Thus, the management of OPGs continues to be a subject of controversy. A significant number of cases, such as patients with NF1, may be asymptomatic and have a stable disease condition. Therefore, observation and conservative management are pursued [4]. Chemotherapy has been the recommended first-line treatment strategy for symptomatic young patients [4, 19, 20]. Primary surgery for tumor resection is not the standard care because of the significant risk of complications. However, surgery can be performed with acceptable results in selected patients, including those with large exophytic or cystic tumors [21]. Therefore, endoscopic endonasal surgery was used for tumor resection in the present report.

The EEA to the skull base has been described in detail for more common skull base tumors, such as pituitary adenomas [22], meningiomas [7], and craniopharyngiomas [8]. While the role of EEA for extra-axial skull base tumors is well-established, the role of EEA in approaching intra-axial tumors remains less clear. Few such reports are available in the literature, and most published reports are in the form of single-case reports or small-case series. Reports on the use of the EEA in resection of intra-axial lesions include cases of pontine ependymoma [9], pontine cavernoma [10], hypothalamic glioma [5], and germ-cell tumor [11].

Prior reports on the use of EEA in OPGs are limited [5], and to the best of our knowledge, no such reports exist on its utility for pediatric OPGs. The majority of cases are included in larger mixed case series discussing the use of EEA for various suprasellar lesions (Table 3) [6, 14, 23–25]. Zoli et al. [5] reported a specific experience with the EEA for

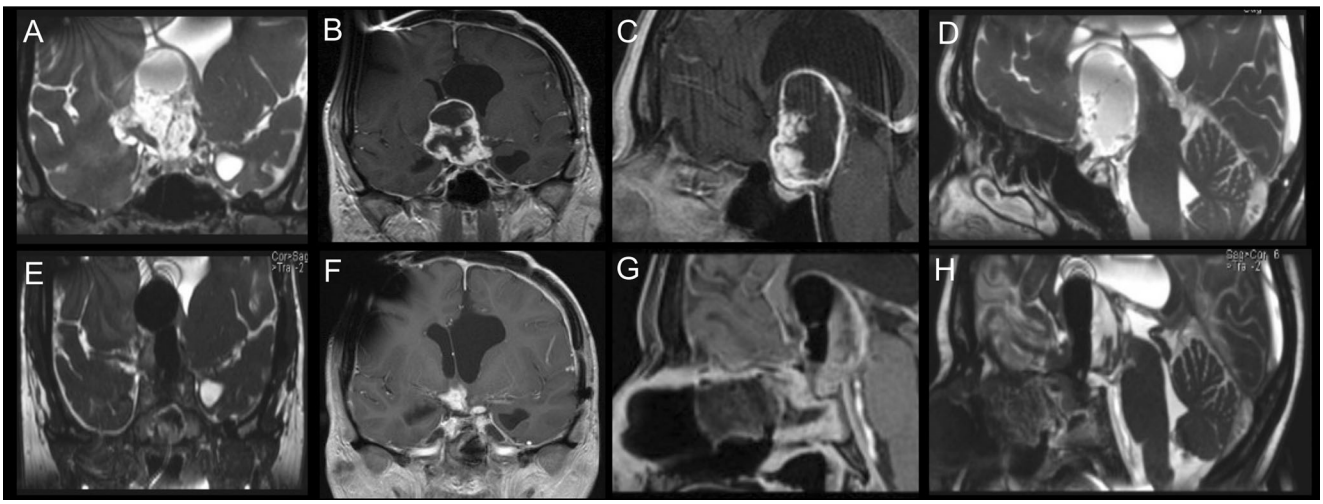

Fig. 3 Case 3. Pre-operative imaging; **a** coronal T2-weighted, **b** coronal T1-weighted contrast-enhanced, **c** sagittal T1-weighted contrast-enhanced, and **d** sagittal T2-weighted MR imaging showing a recurrent suprasellar mass in an adult patient (case 3). Post-operative imaging; **e** coronal T2-weighted, **f** coronal T1-weighted contrast-enhanced, **g** sagittal T1-weighted contrast-enhanced, and **h** sagittal T2-weighted MR imaging demonstrate subtotal resection of the tumor

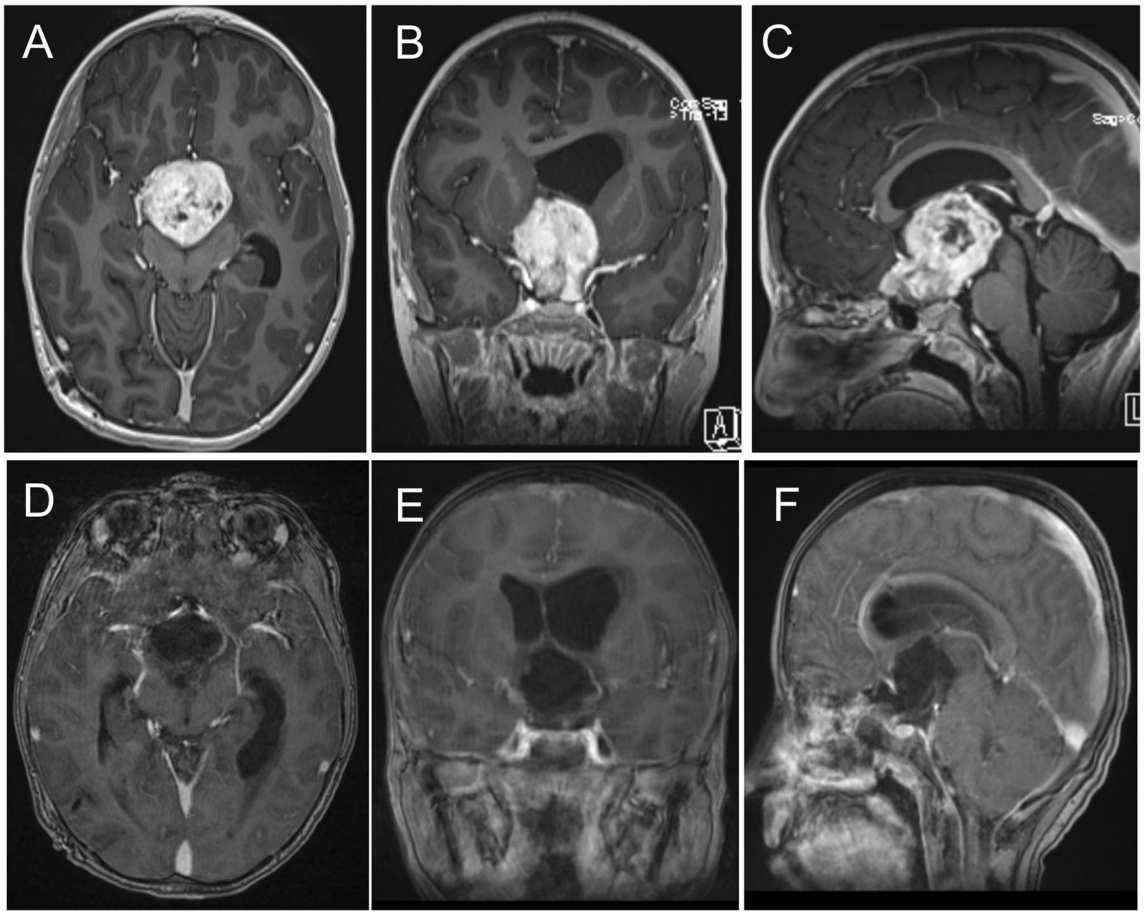

Fig. 4 Case 4. Pre-operative imaging; **a** axial, **b** coronal, and **c** sagittal T1-weighted contrast-enhanced MR imaging, showing a large suprasellar tumor. Post-operative imaging; **d** axial, **e** coronal, and **f** sagittal T1-weighted contrast-enhanced MR imaging demonstrate subtotal resection of the tumor in an 11-year-old male (case 4)

OPGs. They included five cases with a mean age of 32 years (range, 13–44 years); the most common clinical presentations in these cases were visual impairment and hypothalamic-pituitary axis abnormalities. Four cases were pathologically pilocytic astrocytomas while the fifth case was pilomyxoid astrocytoma.

Although total resection of pilocytic astrocytomas is of prognostic importance in general, complete resection of OPGs may result in a high risk of injury to the optic apparatus, hypothalamic-pituitary axis, and the carotids [26–28]. Despite the fact that it is very unusual to achieve total resection of the tumor without jeopardizing vision, Zoli et al. [5] attempted

Table 2 Surgical outcomes in four patients with optic pathway gliomas

Cases	Duration of follow-up (months)	Endocrine outcomes		Visual outcomes		Complications
		Preoperative	Postoperative	Preoperative	Postoperative	
1	28	Normal	Hypopituitarism	Diminished acuity bilaterally (worse in the left eye)	Same	CSF leak, tension pneumocephalus, and meningitis
2	19	Normal	Hypopituitarism	Diminished acuity bilaterally; bitemporal hemianopia (worse in the right eye)	Same	None
3	7	Hypothyroidism	Hypothyroidism and transient DI	Diminished acuity in the right eye; temporal field cut in the left eye	Same	None
4	6	Normal	Normal	Diminished acuity in the right eye	Same	Meningitis

CSF cerebrospinal fluid, *DI* diabetes insipidus

Table 3 Reported cases of optic pathway gliomas treated via an endoscopic endonasal approach

Author	Age (years)/sex	Entity of removal	Pathology	Endocrine outcomes	Visual outcomes	Complications
de Divitiis et al. 2007 [14]	43/F	Subtotal	PA	Normal	Stable	None
Kassam et al. 2007 [24]	16/M	Biopsy	Not reported	Not reported	Stable	Not reported
Arbolay et al. 2009 [25]	42/M	Biopsy	PA	Not reported	Not reported	Meningitis
Paluzzi et al. 2011 [23]	44/F	Subtotal	PA	Adrenal insufficiency and DI	Not reported	None
Zoli et al. 2014 [5][a]	42/M	Subtotal	PA	Anterior panhypopituitarism and DI	Improved	None
	23/F	Biopsy	PA	DI	Stable	None
	13/M	Biopsy	PA	Normal	Improved	None
	38/M	Total	PA	Anterior panhypopituitarism and DI	Stable	CSF leak
	44/F	Total	PMA	Anterior panhypopituitarism and DI	Improved	None
Somma et al. 2017 [6][b]	NR	Subtotal	PA	Stable	Improved	CSF leak
	NR	Subtotal	PA	Stable	Improved	None
	NR	Subtotal	DA	Stable	Stable	None
	NR	Subtotal	DA	Stable	Stable	None
	NR	Subtotal	DA	Stable	Stable	None
	NR	Subtotal	DA	Stable	Stable	None
The present series	7/F	Subtotal	PA	Hypopituitarism	Stable	CSF leak, tension pneumocephalus, and meningitis
	12/M	Subtotal	PA	Hypopituitarism	Stable	None
	32/F	Subtotal	PA	Hypothyroidism and transient DI	Stable	None
	11/M	Subtotal	PA	Normal	Stable	Meningitis

F female, *M* male, *NR* not reported, *PA* pilocytic astrocytoma, *DI* diabetes insipidus, *CSF* cerebrospinal fluid, *PMA* pilomyxoid astrocytoma, *DA* diffuse astrocytoma

[a] CSF leak occurred in two cases in this study

[b] Cases of OPG in this report are not described in details

total resection using the EEA in two patients with OPGs, and one case showed improved vision. OPGs are usually intrinsic to the fibers of the optic nerve, chiasm, and radiation; and in this scenario visual decline is inevitable [19]. It is possible that the tumor in the aforementioned case originated from the hypothalamus and did not infiltrate the chiasm, although the patient showed no symptoms of hypothalamic involvement at time of presentation. In the present series, we attempted subtotal resection considering the young age of patients included in the study and the morbidity probably following total resection. Deterioration of vision was the main concern when surgery was planned. Therefore, with the good visualizing provided by the EEA, the optic nerve that had better vision was identified, and tumor residual was kept on the optic nerve. It is also important to mention that vision was compromised in one eye more than the other. Thus, given the fact that patients had non-functioning vision at one side, we were more aggressive towards the side with worse vision. We do believe that this strategy was effective in our cases as no deterioration of vision was observed postoperatively. Furthermore, preserving the feeding vessels of the chiasm is of critical importance to achieve good visual outcomes [5].

Dedicated reports on the use of the EEA in OPGs are lacking. As mentioned above, few such studies have been performed in a very few number of cases; therefore, the data might not be comprehensive. Given the differences in characteristics of patients receiving various treatment modalities, a valid comparison between surgical and non-surgical patients cannot be performed. However, in two different studies, which reported the outcomes of OPGs treated with multiple modalities, including surgery and chemotherapy, the rate of visual deterioration varied between 9.8 and 21% [1, 4]. Additionally, the risk of visual deterioration in a surgical series using different surgical techniques reached 46% at the surgical side and 15% at the contralateral side [29]. Therefore, we can conclude that the EEA is associated with acceptable results in terms of visual outcomes.

One important factor to consider while treating patients with OPGs is the morbidity, which is related to hypothalamic-pituitary axis (HPA) dysfunction. HPA dysfunction is common in patients with OPGs. It has no doubts that any surgical manipulation to the sellar region will result in abnormalities in HPA regardless of the surgical approach. In this report, new-onset hypopituitarism occurred in two cases. Zoli et al. [5] reported higher rates of postoperative HPA abnormalities (4 out of 5 patients) in patients with OPGs. This is potentially related to their higher rates of total resections as the degree of HPA injury is usually dependent on the aggressiveness of tumor resection. Given that OPGs may progress rapidly, it is somehow difficult to ascertain whether the deterioration of HPA function is primarily related to surgery or to tumor progression [4]. We do believe that the visualization provided by the EEA prompts better endocrine outcomes as noted in its utility for various pathologies. In spite of that, the risk of postoperative HPA dysfunction appears to be higher in OPGs than in other tumors, as noted in this series and previous reports [5, 23]. However, this is not necessarily accurate for pediatric patients as data on the use of EEA in pediatrics are limited, and the role of EEA in that age group is not well established. Furthermore, the results of endocrine outcomes in OPGs concur with those reported by other group for a different pathology in a similar patient population (endocrine function worsened in 54% of patients following craniopharyngioma resection via the EEA) [30]. Similarly, the rate of postoperative HPA dysfunction reached 72% in patients with OPGs resected using a different traditional surgical approach [29]. The risk of endocrine function deterioration is also high with radiation therapy alone [28]. Thus, radiation therapy should be delayed whenever possible in young patients.

Postoperative meningitis occurred in two patients in this report. The risk of meningitis in this study appears to be higher than those reported in other studies. This is mainly explained as patients in this study were at higher risk of developing meningitis. CSF leak and placement of VP shunt prior to surgery are reported risk factors for meningitis that were both present in our patients. Additionally, the rates of CSF leak after endonasal surgery are generally higher in children than in adults, ranging from 4.7 to 15.7% [24, 31–33]. These two factors need to be considered in future surgeries to achieve better outcomes. However, because this report included only four patients, valuable conclusions cannot be drawn. Furthermore, one patient developed fungal meningitis, which is very unusual, especially in an immunocompetent patient. Fungal meningitis has been reported in three cases treated via the EEA [34]. However, all those cases were immunocompromised patients with multiple comorbidities.

Postoperative CSF leak occurred in one case (case 1, a 7-year-old female) in this report. We expect higher rates of CSF leak postoperatively for OPG compared to other sellar lesions. Young age, preoperative hydrocephalus, high flow fistulae, and the extended bone opening used in the EEA have been documented as factors that can increase the incidence of postoperative CSF leak [5]. In this case, the CSF leak was successfully treated with endoscopic endonasal re-exploration.

The need for CSF diversion using VP shunt after tumor resection in optic pathway glioma is not well documented. In two series discussing the treatment and outcomes of OPG, 23% and 26% of patients needed VP shunts as part of treatment without giving sufficient data whether it was used as an initial treatment or after tumor resection [1, 4]. We postulate that addressing tumors from below may not allow establishing the CSF pathway. Additionally, one patient in our series who needed VP shunt after surgery had postoperative meningitis as a complication which caused communicating hydrocephalus (case 1).

Pilocytic astrocytomas of the optic pathway tend to recur despite aggressive tumor resection. The progression rate in cases where total resection is not achieved ranges from 50 to 80% [35]. In this series, we attempted subtotal resection to minimize the risk of morbidity. In fact, this strategy helped achieving good control for tumor, although recurrence occurred in one case in this report (case 1). However, the follow-up duration in this series is relatively short, and a longer follow-up duration is needed to fully evaluate the efficacy of the EEA in tumor control. Additionally, despite the aggressive treatment modalities provided for case 3 in the past, the patient experienced tumor progression multiple times. These findings highlight the complexity and controversy in the management of OPGs.

The retrospective nature of our study and the small number of cases are major limitations of this report. However, it will be difficult to include a large number of patients considering the rarity of this pathology and the unclear role of surgical intervention. In addition, the follow-up duration in this study is relatively short; therefore, the conclusion on long-term outcomes of this approach cannot be drawn. Moreover, if the EEA is elected, it should be performed in centers that have considerable experience in using such an approach.

Conclusions

Although our data are preliminary, the EEA provides a direct corridor to OPG with acceptable results in terms of tumor resection and visual outcomes. Hypothalamic-pituitary axis dysfunction remains a limitation of any treatment modality for OPGs and should be considered whenever possible. Moreover, CSF leak is a major limitation of the endonasal approach in patients with OPGs. Definitive conclusions are pending as the learning curve of this approach is steep. Further work is needed to understand patient selection for such an approach.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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