

Case Report

The “extended” endoscopic endonasal approach for the removal of a mixed intrasuprasellar germinoma: Technical case report

Alberto Di Somma, Carolina Bronzoni, Elia Guadagno¹, Domenico Solari, Giovanni Orabona Dell’Aversana², Marialaura Del Basso De Caro B. S.¹, Paolo Cappabianca

Department of Neurosciences and Reproductive and Odontostomatological Sciences, Division of Neurosurgery, ¹Department of Advanced Biomedical Sciences, Division of Pathology, ²Department of Neurosciences and Reproductive and Odontostomatological Sciences, Division of Maxillo-Facial Surgery, Università degli Studi di Napoli Federico II, Naples, Italy

E-mail: Alberto Di Somma - albertodisomma@hotmail.it; Carolina Bronzoni - Kerol.bronz@libero.it; Elia Guadagno - eliaguadagno84@gmail.com; *Domenico Solari - domenico.solari@unina.it; Giovanni Orabona Dell’Aversana - gdoorabon@yahoo.it; Marialaura Del Basso De Caro B. S. - marialaura.delbassodecaro@unina.it; Paolo Cappabianca - paolo.cappabianca@unina.it
*Corresponding author

Received: 02 August 13 Accepted: 20 September 13 Published: 29 January 14

This article may be cited as:

Somma AD, Bronzoni C, Guadagno E, Solari D, Dell’Aversana GO, De Caro BM, et al. The “extended” endoscopic endonasal approach for the removal of a mixed intrasuprasellar germinoma: Technical case report. *Surg Neurol Int* 2014;5:14.

Available FREE in open access from: <http://www.surgicalneurologyint.com/text.asp?2014/5/1/14/126043>

Copyright: © 2014 Di Somma A. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Background: Intracranial germ cell tumors (GCTs) represent less than 5% of pediatric brain tumors. Neurosurgical treatment remains essential in the management of patients with intracranial nongerminomatous GCT.

Case Description: A 12-year-old girl presented with clinical features of neurohypophyseal dysfunction and rapidly progressive visual worsening. Magnetic resonance imaging (MRI) showed a lesion arising from the sella with a significant suprasellar component, compressing the optic chiasm and extending into the third ventricle. The tumor was removed via an endoscopic endonasal transtuberulum-transplanum approach and the histology revealed a mixed germinoma. In the postoperative course, a conspicuous improvement of visual function was observed; an early postoperative MRI showed near-total removal of the lesion. The patient was referred to pediatric oncologist for the adjunctive chemotherapy and radiotherapy.

Conclusions: The management of primary intracranial sellar and suprasellar germinomas still remains controversial. With this report we highlighted another possible surgical option among therapeutic strategies for these highly malignant tumors.

Key Words: Endoscopic endonasal surgery, intracranial germinoma, pediatric brain tumor, skull base surgery

Access this article online**Website:**

www.surgicalneurologyint.com

DOI:

10.4103/2152-7806.126043

Quick Response Code:

INTRODUCTION

Intracranial germ cell tumors (GCTs) represent 3-11% of pediatric brain tumors, of which nearly two-thirds are germinoma.^[13] The most common primary sites include the pineal (56%) and the neurohypophyseal regions

(28%) with basal ganglia, thalamus, corpus callosum, cerebellum, and spinal cord being less common.^[19] GCT can be broadly divided into two major histological subtypes: (1) Pure germinomas and (2) nongerminomatous germ cell tumors (NGGCT). This differentiation has clinical significance as the histological subtype is the single

most important factor in predicting the outcome. Indeed, pure germinomas are highly radiosensitive tumors and carry a superior prognosis to NGGCTs, which have 5-year survival rates in the range of 30-50%, with many tumors relapsing within 18 months from diagnosis.^[9,18,24]

Despite recent advances in chemotherapy and radiotherapy, neurosurgical treatment remains fundamental in the care of patients with intracranial NGGCTs, providing tissue sampling, cerebrospinal fluid (CSF) diversion and cyto-reduction.

Historically, surgical management of NGGCTs involving sella and suprasellar area has been gained through different transcranial approaches, including the subfrontal, frontolateral and, above all, pterional routes and microscopic transsphenoidal approaches. Nowadays, the evolution of the endoscopic endonasal transsphenoidal technique has permitted to approach sellar lesions also in pediatric age^[6,16] and has then led to the extension of the indications to a variety of lesions involving all the anatomical area surrounding the sella, that is, craniopharyngiomas, anterior skull base meningiomas and giant macroadenomas.^[3,17,27]

It is interesting to observe that up to date the use of the endoscopic endonasal approach for managing intracranial GCT in adult patients has been rarely reported in the pertinent literature^[26] as well as in pediatric population.^[2,4] We herein report a case of a pediatric patient affected by a large, intra and suprasellar mixed GCT, removed through an extended endoscopic endonasal approach.

To the authors' knowledge, this is the first pediatric case of a successfully surgical treatment of a large intrasuprasellar mixed GCT using a pure "extended" endoscopic endonasal transplanum/transsterculum approach.

CASE REPORT

A 12-year-old girl was admitted to our hospital with a few-month history of polyuria, polydipsia, and secondary amenorrhea. A neurological examination, including visual field exam and visual acuity, showed amaurosis in the left eye and rapidly worsening temporal hemianopsia in the right eye. A complete preoperative endocrinological assessment revealed mild elevation of the serum prolactin level (76 ng/mL, n.v. 2-25 ng/mL), secondary hypothyroidism (TSH: 0.15 mUI/ml, n.v. 0.4-4 and FT4: 0.52 ng/dl, n.v. 0.9-1.7) and secondary hypocortisolism (morning cortisol: 31 ng/ml, n.v. 50-200). Neuroradiological investigation, by means of magnetic resonance imaging (MRI) scans, demonstrated a large mass arising from the sella with a significant suprasellar component, compressing the optic chiasm and extending into the third ventricle, showing nonhomogeneous enhancement after intravenous contrast administration [Figure 1a-c].

Our diagnostic hypothesis were either a craniopharyngioma or a GCT. However, the evidence of diabetes insipidus

as a presenting symptom in a pediatric patient was more suggestive of a germinoma. Unfortunately, it was not possible for technical reasons to perform the serum and/or CSF assay of Alpha-fetoprotein (AFP) and Human chorionic gonadotropin (beta-HCG) before surgery.

After careful consideration of many factors, including patient age, daily progressive visual impairment, lesion size and midline location without any parasellar extension, it was decided to treat the tumor by using an extended endoscopic endonasal transsterculum/transplanum approach,^[1,5,17,19,20,23,25] few days after the admittance.

The postoperative course was marked by a conspicuous improvement of temporal hemianopsia in the right eye and light perception was reported in the left eye. No postoperative infection or CSF leak was detected. The patient was discharged without any neurological defect of new onset. The postoperative serum level of AFP was 103.9 UI (n.v. 0-5.5 UI) while level of beta-HCG was 165.7 mUI (n.v. <3.5 mUI). Afterwards, the patient was referred to the pediatric oncologist for the adjunctive chemotherapy and radiation therapy treatment. One month postoperative MRI confirmed near-total tumor removal [Figure 1d-f]. Three months after surgery, the girl was still being medicated with desmopressin acetate for the diabetes insipidus, but her condition was otherwise normal.

SURGICAL PROCEDURE

After the placement of a lumbar drainage in order to allow spinal fluid diversion to prevent postoperative CSF leakage, an extended endoscopic endonasal approach to the suprasellar area was performed according to the principles already described in the literature.^[1,7,14,15]

A binostiril approach was used. A left middle turbinectomy with the resection of the posterior nasal septum and a wide sphenoidotomy was performed. Tuberculum sellae and posterior portion of the planum sphenoidale and sellar floor were removed.

Upon dural opening, a greenish-yellow, bleeding mass came into view, splitting the pituitary gland apart. Initial intracapsular debulking of the tumor was performed with the aid of the cavitron ultrasonic aspirator (CUSA) that allowed the removal of the intrasellar component, appearing elastic and fleshy. Afterwards, fine extracapsular dissection of the right side of the tumor wall was performed and the suprasellar component carefully removed.

The dissection and the removal of the lesion in the suprasellar area followed the same principles of microsurgery, with the use of dedicated instruments.

The last step consisted on the fragmentation and peeling of the tumor capsule, which was found to compress and

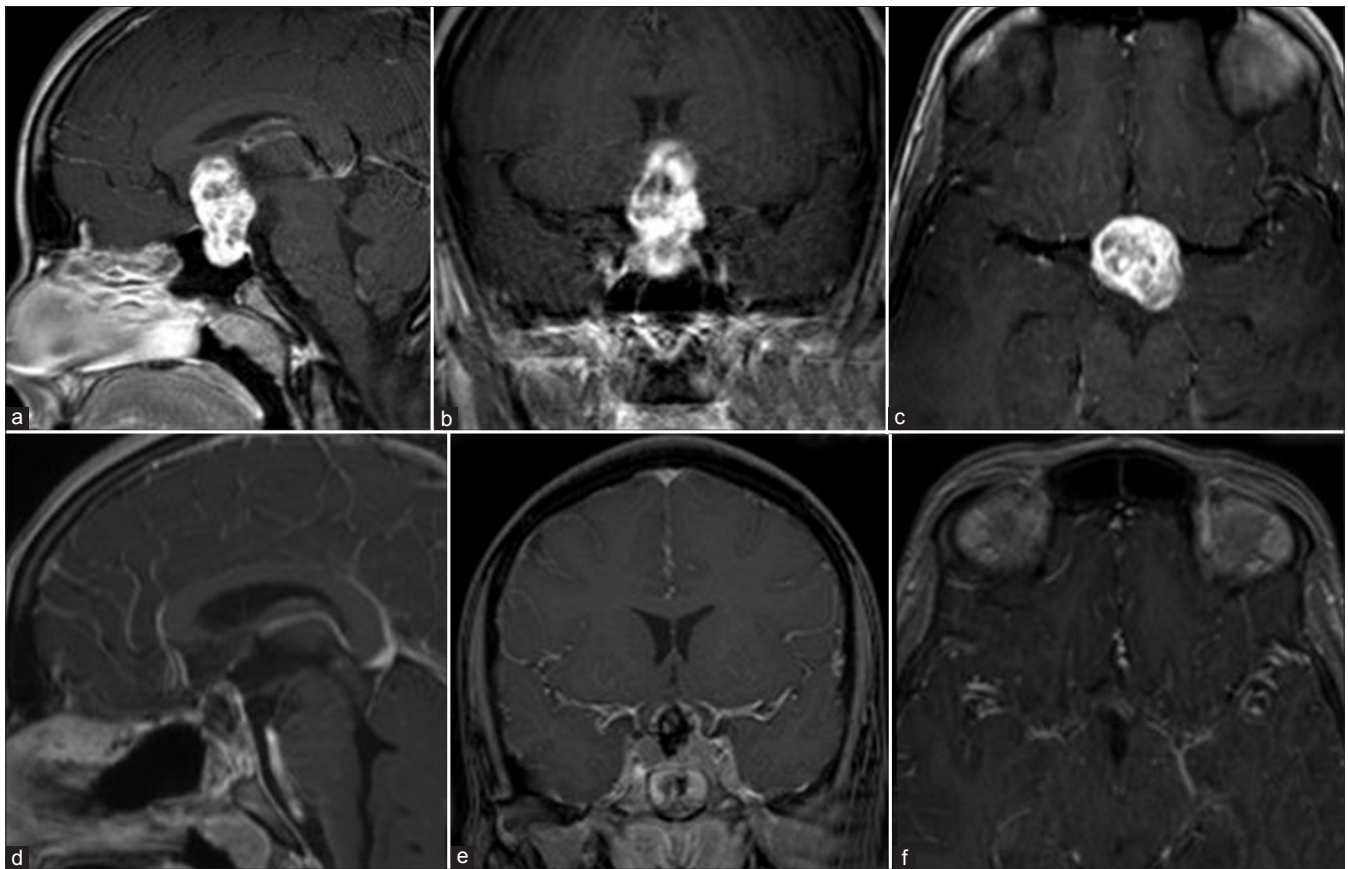


Figure 1: Preoperative axial (a) coronal (b) and sagittal (c) MRI scans showing this case of a large intra and suprasellar mass compressing the optic chiasm and extending toward the diencephalon; postoperative axial (d) coronal (e) and sagittal (f) MRI scans demonstrating the near total removal of the lesion. The autologous fat (hyperintensity), used for reconstruction, fills the surgical cavity

obstruct the infundibular recess of the third ventricle. At the end of the procedure, the pituitary stalk was identified and a little tumor fragment surrounding it and the infundibulum was intentionally left in place. The floor of the third ventricle, as well as mammillary bodies and tuber cinereum along the floor, were carefully explored and no evidence of tumor infiltration was recognized. Multiple fragments of tissue from different areas of the lesion were taken during operation in order to obtain an accurate histopathological diagnosis [Figure 2].

The reconstruction of the osteodural defect was achieved through the overlapping of different materials, placed both in the extradural and the intradural spaces, in a multilayer technique. A sort of “sandwich” composed by multiple layers of dural substitute was created outside the nasal cavity, oversizing the osteo-dural defect and containing a thin stratum of periumbelical fat and a fragment of bone. All these materials were kept together by 8.0 prolene suture. This multilayer “sandwich” was placed in the intradural space with the external coat fixed in the extradural surface. Fibrin glue (Tisseel®, Baxter, Vienna, Austria) was used to hold the material in place and to fill the sphenoid sinus.

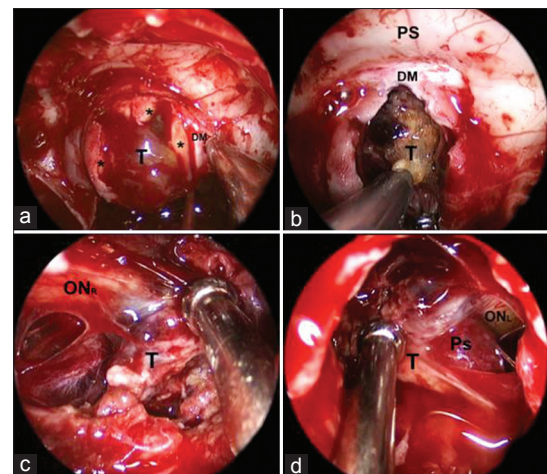


Figure 2: (a) after opening the dura, the lesion comes into view splitting the pituitary gland in several parts; (b) initial intracapsular debulking of the tumor with the aid of the cavitron ultrasonic aspirator (CUSA); (c) fine dissection of the right side of the tumor capsule until the right optic nerve comes into view; (d) left optic nerve exposition T: tumor; PS: planum sphenoidale; DM: dura mater; ONr: right optic nerve; ONl: left optic nerve; Ps: pituitary stalk *: pituitary gland

Finally, a Foley catheter balloon was positioned to keep in place the reconstruction materials [Figure 3].

Pathology report

This lesion consisted of immature mesenchymal tissue, comprehensive of myxoid, microcystic, and hemorrhagic areas, among which there were groups of large atypical individual cells with a round nucleus, a conspicuous nucleolus and abundant clear cytoplasm, often in apoptosis, surrounded or infiltrated by small sized lymphocytes; in other areas, cohesive cells showed a cord-like and glandular pattern, strong anaplasia and a big eosinophilic nucleolus; sometimes neoplastic cells became thinner and lined microcystic structures within a myxoid and hemorrhagic stroma.

All these aspects belong to a mixed GCT

The immunohistochemical profile confirmed this diagnostic hypothesis: The large cohesive cells showed positivity for PLAP and CD117 (germinoma component), while anaplastic cohesive cells were positive for CK7, CD30 and PLAP (embryonal carcinoma-like); there was also a focal signal for AFP in some microcystic structures, suggestive of a well differentiated yolk sac tumor-like component; HCG, GFAP, CgA, synaptophysin, neurofilament, and CK20 were negative.

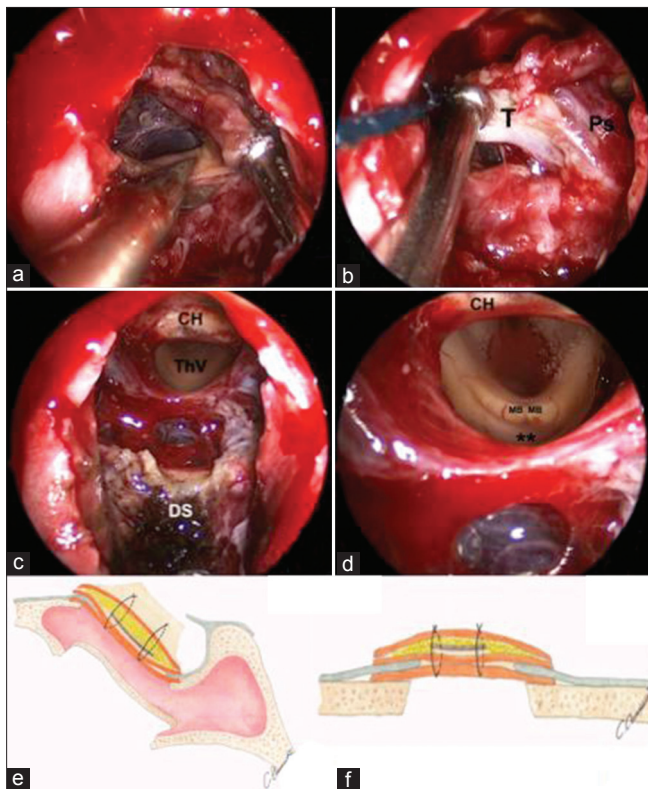


Figure 3: (a) Removal of the suprasellar part of the tumor; (b) identification of the pituitary stalk leaving intentionally in place a little tumor fragment surrounding it and the infundibulum; (c) panoramic view; (d) close-up view of the floor of the third ventricle; (e) schematic picture showing the reconstruction technique used after the approach; (f) particular of the multilayer “sandwich” placed in the intradural space with the external coat fixed in the extradural surface T: tumor; Ps: pituitary stalk; CH: chiasm; ThV: third ventricle; MB: mammillary bodies; **: tuber cinereum

The histological aspects and the immunoprofile lay for a mixed GCT [Figure 4].

DISCUSSION

Intracranial GCTs are malignant neoplasms arising from remnants of primitive germ cells that have failed to migrate to the genital crest during embryonic life.^[10] They show a striking predilection for sites along the midline, namely the pineal region, the third ventricle, and the suprasellar-hypothalamic area.

Radiotherapy with/without chemotherapy is now accepted as the gold standard for the treatment of germinomas.^[12] However, although radiotherapy remains the mainstay of current therapy for intracranial germinoma, a debate has

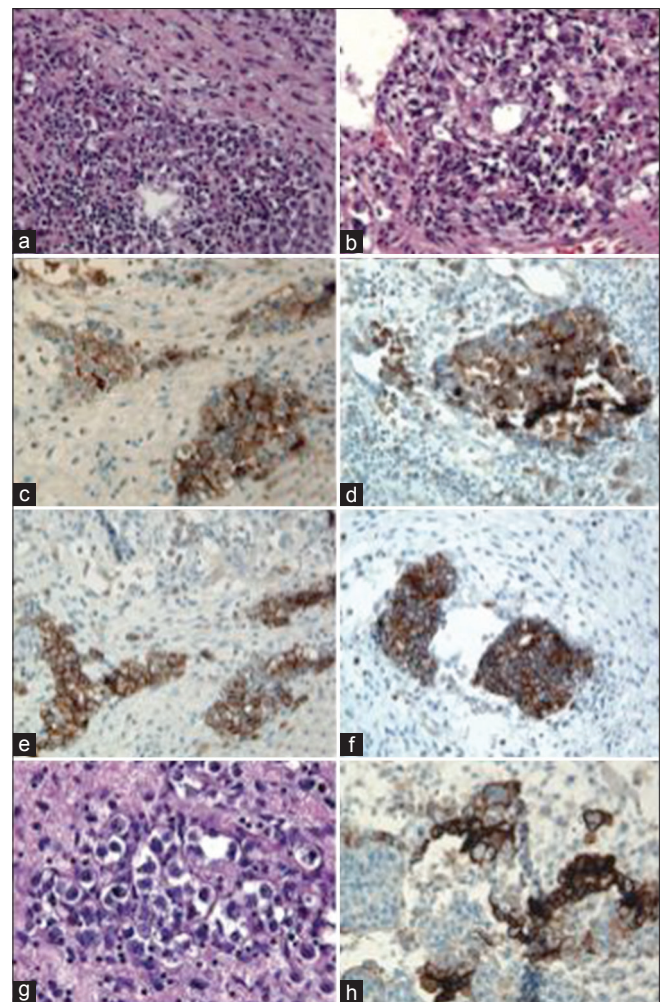


Figure 4: Mixed germ cell tumor: (a) germinoma pattern: large individual cells with abundant clear cytoplasm and a round nucleus, surrounded by small lymphocytes (H and E, ×10) (g) high power view (H and E, ×40); (b) embryonal carcinoma pattern: cohesive cells with a glandular pattern, showing a strong anaplasia and a big eosinophilic nucleolus (H and E, ×10); positivity for PLAP (c) and c-Kit (e) in the germinoma component and for PLAP (d), CD30 (f) and CK7 (h) in the embryonal carcinoma component

emerged concerning the ideal management of NGGCT. For this kind of lesion, radiation therapy has produced less satisfactory results, so that it has highlighted the possible role of the surgical procedure. Aside from possibility of achieving direct tissue sampling, surgery could grant mass debulking thus reducing the need of radio and chemo therapies^[19,20,23] and moreover of their side effects.

The rapid worsening of vision presented by the patient in our report has given us the opportunity to perform an endoscopic endonasal transtuberulum transplanum approach to the suprasellar area, in order to safely remove the tumor. Histology revealed a NGGCT.

The endoscopic endonasal approach extended to the suprasellar area has been successfully used in the management of different suprasellar lesion such as craniopharyngiomas, Rathke cleft cysts, tuberculum sellae meningiomas, suprasellar pituitary adenomas, etc.^[8,11,14,17]

The main advantage of the extended endonasal transsphenoidal approach consists in providing a direct view of the inferior aspect of the suprasellar neuro-vascular structures without any brain retraction and nevertheless ensuring, a multiangled, close-up view of surgical landmarks in the sellar-suprasellar area. Moreover, as shown in this case, an early devascularization of the tumor can be achieved reducing the intraoperative blood loss, which is a very important factor especially in pediatric patients.

Particularly in this case, this route minimizes the risk of postoperative visual loss, which is strictly related to the integrity of the vascularization of the optic chiasm. In fact, most of the blood supply to the optic system comes from the superior surface, from the branches of the anterior cerebral and anterior communicating arteries, thus rendering the inferior approaches potentially less dangerous.^[21,22]

Another possible advantage of the surgical approach, as a first option in the treatment of some of this kind of suprasellar tumors, consists in the fact that the subsequent radio-chemotherapy will be directed toward a neoplastic target significantly reduced in volume. However, there are also disadvantages performing surgery before a radio-induced remission of the tumor, above all the increased bleeding risk approaching a consistently larger and more vascularized lesion.

With this report, we aim to show another possible surgical option in the challenging therapeutic management of these highly malignant lesions. We think that an extensive mass reduction with an extended endoscopic endonasal approach allows a worthwhile starting point for subsequent treatment procedures that can act on a more affordable target.

ACKNOWLEDGMENTS

The authors would like to thank Dr. Carmela Chiamonte for the precious contribution in creating the artworks.

REFERENCES

- Cappabianca P, Cavallo LM, Esposito F, De Divitiis O, Messina A, De Divitiis E. Extended endoscopic endonasal approach to the midline skull base: The evolving role of transsphenoidal surgery. *Adv Tech Stand Neurosurg* 2008;33:151-99.
- Carlos Chung KH, Owler BK, Dexter M, Chaseling R. Paediatric germ cell tumours of the central nervous system: Results and experience from a tertiary-referral paediatric institution in Australia. *J Clin Neurosci* 2013;20:514-9.
- Cavallo LM, Solari D, Esposito F, Cappabianca P. The endoscopic endonasal approach for the management of craniopharyngiomas involving the third ventricle. *Neurosurg Rev* 2013;36:27-37.
- Ceylan S, Koc K, Anik I. Extended endoscopic approaches for midline skull-base lesions. *Neurosurg Rev* 2009;32:309-19.
- Couldwell WT, Weiss MH, Rabb C, Liu JK, Apfelbaum RI, Fukushima T. Variations on the standard transsphenoidal approach to the sellar region, with emphasis on the extended approaches and parasellar approaches: Surgical experience in 105 cases. *Neurosurgery* 2004;55:539-47.
- de Divitiis E, Cappabianca P, Gangemi M, Cavallo LM. The role of the endoscopic transsphenoidal approach in pediatric neurosurgery. *Childs Nerv Syst* 2000;16:692-6.
- de Divitiis E, Cavallo LM, Cappabianca P, Esposito F. Extended endoscopic endonasal transsphenoidal approach for the removal of suprasellar tumors: Part 2. *Neurosurgery* 2007;60:46-58.
- de Divitiis E, Esposito F, Cappabianca P, Cavallo LM, de Divitiis O. Tuberculum sellae meningiomas: High route or low route? A series of 51 consecutive cases. *Neurosurgery* 2008;62:556-63.
- Echevarria ME, Fangusaro J, Goldman S. Pediatric central nervous system germ cell tumors: A review. *Oncologist* 2008;13:690-9.
- Endo T, Kumabe T, Ikeda H, Shirane R, Yoshimoto T. Neurohypophyseal germinoma histologically misidentified as granulomatous hypophysitis. *Acta Neurochir (Wien)* 2002;144:1233-7.
- Gardner PA, Prevedello DM, Kassam AB, Snyderman CH, Carrau RL, Mintz AH. The evolution of the endonasal approach for craniopharyngiomas. *J Neurosurg* 2008;108:1043-7.
- Hardenbergh PH, Golden J, Billet A, Scott RM, Shrieve DC, Silver B, et al. Intracranial germinoma: The case for lower dose radiation therapy. *Int J Radiat Oncol Biol Phys* 1997;39:419-26.
- Hoffman HJ, Otsubo H, Hendrick EB, Humphreys RP, Drake JM, Becker LE, et al. Intracranial germ-cell tumors in children. *J Neurosurg* 1991;74:545-51.
- Jane JA Jr., Prevedello DM, Alden TD, Laws ER Jr. The transsphenoidal resection of pediatric craniopharyngiomas: A case series. *J Neurosurg Pediatr* 2010;5:49-60.
- Kassam A, Snyderman CH, Mintz A, Gardner P, Carrau RL. Expanded endonasal approach: The rostrocaudal axis. Part I. Crista galli to the sella turcica. *Neurosurg Focus* 2005;19:E3.
- Kassam A, Thomas AJ, Snyderman C, Carrau R, Gardner P, Mintz A, et al. Fully endoscopic expanded endonasal approach treating skull base lesions in pediatric patients. *J Neurosurg* 2007;106 (2 Suppl):75-86.
- Kassam AB, Prevedello DM, Carrau RL, Snyderman CH, Thomas A, Gardner P, et al. Endoscopic endonasal skull base surgery: Analysis of complications in the authors' initial 800 patients. *J Neurosurg* 2011;114:1544-68.
- Maity A, Shu HK, Janss A, Belasco JB, Rorke L, Phillips PC, et al. Craniospinal radiation in the treatment of biopsy-proven intracranial germinomas: Twenty-five years' experience in a single center. *Int J Radiat Oncol Biol Phys* 2004;58:1165-70.
- Matsutani M, Sano K, Takakura K, Fujimaki T, Nakamura O, Funata N, et al. Primary intracranial germ cell tumors: A clinical analysis of 153 histologically verified cases. *J Neurosurg* 1997;86:446-55.
- Oka H, Kawano N, Tanaka T, Utsuki S, Kobayashi I, Maezawa H, et al. Long-term functional outcome of suprasellar germinomas: Usefulness and limitations of radiotherapy. *J Neurooncol* 1998;40:185-90.
- Rhoton AJ. Microsurgical anatomy of the sellar region. In: Landolt A, Vance M, Reilly P, editors. *Pituitary Adenomas*. New York: Churchill Livingstone; 1996. p. 241-8.
- Rhoton AJ, Natori Y. The Orbit and the Sellar Region: Microsurgical Anatomy and Operative Approaches. New York: Thieme; 1996. p. 311.

23. Saeki N, Murai H, Kubota M, Fujimoto N, Yamaura A. Long-term Karnofsky performance status and neurological outcome in patients with neurohypophyseal germinomas. *Br J Neurosurg* 2001;15:402-8.
24. Schild SE, Scheithauer BW, Haddock MG, Wong W, Lyons MK, Marks LB, et al. Histologically confirmed pineal tumors and other germ cell tumors of the brain. *Cancer* 1996;78:2564-71.
25. Schwartz TH, Anand VK. The endoscopic endonasal transsphenoidal approach to the suprasellar cistern. *Clin Neurosurg* 2007;54:226-35.
26. Terasaka S, Kawabori M, Kobayashi H, Murata J, Kanno H, Tanaka S, et al. Neurohypophyseal germinoma with abundant fibrous tissue. *Brain Tumor Pathol* 2012;29:58-62.
27. Zada G, Lin N, Laws ER Jr. Patterns of extrasellar extension in growth hormone-secreting and nonfunctional pituitary macroadenomas. *Neurosurg Focus* 2010;29:E4.