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The role of the endoscopic transsphenoidal approach in pediatric neurosurgery

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Abstract The endoscopic endonasal transsphenoidal approach to the sellar region for the removal of pituitary adenomas and of other neoplasms in the same area has proved its reliability and effectiveness for the very wide vision it offers, coupled with minimal surgical trauma. Indications and advantages of such a technique are reported, focused on the treatment of lesions of the sellar and parasellar environment in pediatric age-group patients, and based on a consecutive series of 100 patients, 3 of them adolescents, treated during the last 3 years.

Keywords Endoscopy · Pediatric brain tumors · Pituitary adenoma · Transsphenoidal surgery

Introduction

The history of transsphenoidal surgery for sellar-region lesions [42] begins in the early 1900s when Schoffler, following a study on the cadaver by the Italian, Giordano [4, 19], chief surgeon of the Venice Hospital, performed through a superolateral nasoethmoidal route the first extracranial transsphenoidal removal of a pituitary adenoma. After a few years this procedure was better standardized by Cushing and Hirsch who proposed respectively, an oronasal and an inferior endonasal approach. During that century the transsphenoidal technique more than once was about to fall into disuse in favor of transcranial ones (pterional, subfrontal, subtemporal), more invasive, but then capable of offering a major view of the surgical field and a better surgical management. Later, two significant improvements to the transsphenoidal approach, made by Guiot, with the use of intraoperative fluoroscopic control, and by Hardy, utilizing a surgical microscope, favored a better view of the surgical field and a safer orientation, fixing the definite affirmation of the technique, generally acknowledged as first choice in almost all sellar lesions. Moreover, in the past few years, endoscopic procedures have gained ground in neurosurgery, both as techniques combined with microsurgery (endoscope-assisted microneurosurgery), and as autonomous surgical options (pure endoscopy). The latter, added to the contribution of the otorhinolaryngologists in the surgery of the paranasal sinuses with functional endoscopic sinus surgery (FESS) [39, 40, 49, 59, 60, 61] has encouraged a series of attempts at pituitary endoscopic transsphenoidal adenomectomy, endonasal [24, 25] or transnasal [18, 24, 29, 54, 55, 57, 66], unilateral [24, 25] or bilateral [12, 52], endoscope-assisted [14, 16, 18, 22, 25, 26, 41, 45, 48, 58, 67] or purely endoscopic [25, 29, 54, 57, 66], up to the present adjustment of an endoscopic endonasal unilateral transsphenoidal approach by Jho [11, 32, 33, 34, 35, 37]. Jho has made this procedure simple, less traumatic, very effective, and capable of allowing a more extensive surgical removal, respecting the anatomic structures running through.

Surgical approach and technical considerations

All patients undergo an overall "pituitary investigation" including:

- A preliminary evaluation of the pituitary function in basal and dynamic conditions
- An ophthalmologic assessment with visual field instrumental measurement
- A neuroradiological imaging study by MRI of the sellar region with contrast enhancement (focused on the lesion and its relationships with the parasellar structures such as the optic nerves, the cavernous sinuses and the ICA) and CT scan of the nasal and paranasal cavities (for the choice of the most favorable surgical route, left or right, with respect to the nasal septum, the turbinates, and the sphenoid sinus structure), if necessary completed by tridimensional reconstructions of "virtual endoscopy" [5, 53].

The patient is positioned on the surgical table in a cradle like position, that is supine, in slight Trendelenburg, with the trunk hardly raised and the head fixed to the Mayfield without three-point fixation, slightly hyperextended and tilted 10 deg towards the surgeon.

After accurate disinfection and local contact anesthesia of the nasal mucosa, a preliminary endoscopic exploration of the nasal cavities is performed which, together with the radiographic imaging of the nasal cavity and of the lesion's morphology, permits the choice of nostril suitable for the surgical procedure. A 0° rigid diagnostic endoscope is employed, connected to an irrigation system, held in the nondominant hand in the nasal phase of the operation and secured to an endoscope-holder in the sphenoidal and sellar phases of the procedure [8, 35].

After being inserted into the nostril, the endoscope passes between the inferomedial aspect of the middle turbinate and the nasal septum, until it reaches the anterior wall of the sphenoid sinus, where the sphenoid ostium is located. An anterior sphenoidotomy, comprising the removal of the rostrum and of the septum inside the sphenoid sinus is then carried out, in order for the sellar floor to be seen. If drilling of a nonpneumatized sphenoid sinus, the so-called conchal type, is necessary, it can be performed as usual, but it did not happen in any of our cases. The displacement of the sphenoid mucosa covering the sellar floor, its opening and the incision of the dura mater, allow exposure of the lesion, that can be removed. The gradual progression of the endoscope during the removal of the lesion permits a closer view of the surgical target, "inside the anatomy", and the accurate control of the surgical field, particularly with the aid of angular 'scopes (30°, 45°, 70°). The use of the endoscope in this sort of surgery has brought about a modification of the instruments too; in fact, the conventional bayonet-shaped instruments used in the microsurgical transsphenoidal approach are no longer used by our team. Nowadays, straight instruments are in use, that run tangentially to the endoscope, in the same nostril. Some new instruments have been made [10] and others are in development, to optimize the surgical technique, reducing at least the nasal traumatism and improving the possibility of the removal of the lesion. There is a "learning curve" to acquire the essential handling, that is to say, a specific endoscopic skill, that complements previous experience in transsphenoidal surgery: the latter is certainly useful, if added in the context of a global "pituitary culture".

Indications for surgery: clinical aspects

In children, this minimally invasive procedure can be safely and adequately employed in:

- *Pituitary adenomas:* Almost all of them, just with the same indications [32] as those for the conventional microsurgical transsphenoidal approach [21, 65]. It is well-known that these lesions are much less common in children than in adults [2, 3, 13, 17, 46, 50, 51] and represent less than 2% of all intracranial tumors in the pediatric age-group [20, 27], most of them occurring in the postpubertal child [15, 23], usually girls. Presenting-symptoms must be familiar to the pediatrician and to the general practitioner in order for the lesion responsible to be recognized, i.e., the adenoma, which is usually hormonally active and typically a microadenoma, thus manifesting with an endocrinopathy rather than with a mass effect [51]. They cause different endocrine symptoms than in adults, with pubertal and growth delay [47], except GH sec., and/or primary amenorrhoea [2, 50]. The most common is the prolactinoma, followed by the ACTH sec. [50] and there is a different distribution of the various types before, during and after puberty (ACTH sec. is more common before puberty, PRL sec. more common during and after puberty [50]. The third most common secreting pituitary adenoma is the GH sec., very rarely seen before puberty, but, like the PRL sec., very frequently invasive. This distribution is opposite to that observed in adults, where NF predominate [63], but this might be that they are not recognized in children, because they are asymptomatic.

- *Craniopharyngiomas*, with the accepted indications of the transsphenoidal route [1, 44], i.e., enlargement of the pituitary fossa, preferably cystic infradiaphragmatic lesions and preexisting hypopituitaryism [44] or even with supra- [43] or retrosellar extension [28], if symmetrical and well defined.

- *Germinomas with intra- and parasellar locations*, which constitute almost 20% of germ-cell intracranial tumors [30]: they can be managed by this approach, to debulk the tumor, thus decompressing the optic system, which is usually involved, and to confirm the histologi-

Fig. 1 Coronal T1-weighted image after gadolinium (GDTPA) administration: a large intra and suprasellar mass upwards displacing the A1 segments is present, with a peripheral ring enhancement

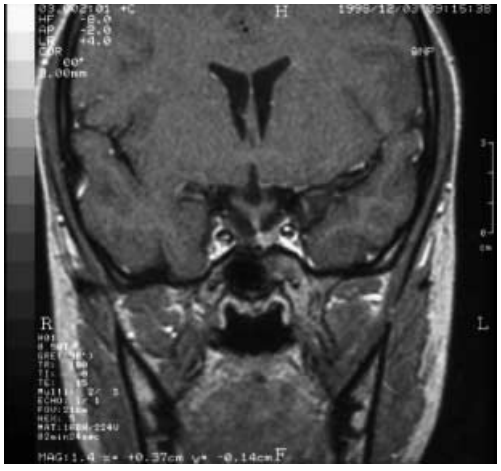

Fig. 2 Coronal T1-weighted image after gadolinium (GDTPA) administration, 1 year after surgery: there is not any evidence of the preexisting tumor mass and the residual pituitary gland lies in the left portion of the sella

cal suspicions, so that the adequate radiation treatment [6] can be planned.

• *Cerebrospinal fluid fistulae* and congenital, iatrogenic or post-traumatic encephalocele [56] of the sphenoethmoidal area have been managed successfully, in recent years, by purely endoscopic transsphenoidal [38] or transethmoidal [62, 64] approaches.

Our group has been the first in Europe [8, 7] to employ the endoscopic endonasal unilateral transsphenoidal approach, achieving a series of 100 consecutive patients (75 macroadenomas, 12 microadenomas, four craniopharyngiomas, three CSF sphenoethmoid leaks, two sphenoid sinusitis, two clival chordomas, one arachnoid cyst, and one residual meningocele), over a range of approximately 3 years (1997–1999); three patients were in the pediatric-adolescent group: a 16 year-old girl with an intrasuprasellar craniopharyngioma, in which a radical removal was possible (see Figs. 1 and 2); an 18 year-old girl with an ACTH silent intrasellar pituitary macroadenoma, totally removed; and a residual nasal meningocele in a 17 year-old boy, who had been operated on through a frontal craniotomy 16 years before because of a frontobasal encephalocele, and in whom a meningeal cyst with a puruloid content was removed through an endonasal endoscopic procedure. There was no difference, compared with the adults, in the technical aspects of the procedure. The follow-up period of the present series is too short to draw any absolute conclusion, even though the results and the pitfalls seem to be at least similar to those of the microsurgical transsphenoidal procedure.

The endoscopic endonasal transsphenoidal approach presents significant advantages over conventional microsurgery [9], because it offers a wider surgical vision of the operating field, with the potential of better and safer tumor-removal; it affords a lower surgical trauma with a shorter hospital stay (an average of 3.3 days in our present series, 41 patients having been discharged on the first or second postoperative day) and a quick recovery; it greatly respects the inner nose, which brings fewer nasal and sinusal postoperative complications, and does not bring any risk of damage to the teeth or tooth buds, which is particularly welcome in the developmental age.

Conclusions and future perspectives

If transsphenoidal surgery is safe and effective in the treatment of pituitary lesions in the pediatric age-group [2, 3, 44, 51], endoscopic endonasal transsphenoidal surgery promises to be even more convenient, thanks to its favorable peculiarities.

The minimal invasiveness of this procedure with respect to both the transcranial and the transsphenoidal microsurgical approach, makes it ideal for the treatment of several pediatric pathological conditions of the sellar region, in which it is essential to preserve the anatomic and functional integrity of the hypothalamic-pituitary axis, in order to assure the correct growth of the child and to maintain a good nasosinusal ventilation.

The pediatric neurosurgeon has to keep up-to-date on the evolution of this topic too, in order to make the right choice at the right moment for the little patient. The surgeon is not asked to have a specific “pituitary knowledge”, nor a background of microsurgical transsphenoidal experience or a specific endoscopic skill, but must be able to select from a wide range of opportunities, the best option for a minimally invasive, maximally effective treatment, which endoscopic endonasal pituitary surgery is showing itself to be.

The use of different angled endoscopes 0, 30, 45 and 70 deg inside the sphenoid sinus, and the wider vision from an optocarotid protuberance to the other, laterally, of the sphenoethmoid planum above, and of the clivus below, point out further surgical approaches to the anterior and posterior skull base, only by removing a restrictive tract of bony covering and after opening the basal dura mater. The first operations already carried out for tuberculum sellae meningiomas [31] or clival chordomas [36] by means of the endoscopic endonasal unilateral transsphenoidal route, show the possibility of minimally invasive procedures targeted on the skull base, but at the same time demand the overcoming of the present technical problems, such as the need for adequate, dedicated instruments, and of the watertight closure of the path crossed, in order to avoid CSF leaks.

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