Sphenoorbital meningioma

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Spheno-orbital meningiomas represent a delicate subtype of intracranial meningiomas involving the sphenoid wing, orbit and important neurovascular structures such as cavernous sinus, carotid artery or optic nerve. Insidious and aggressive dural, bone and orbital involvement contains several defiances to adequate resection, which provides high rates of recurrence.

Classification

Orbital meningiomas can be classified as primary optic nerve sheath meningiomas, primary intraorbital ectopic (Ob) meningiomas and sphenoorbital meningiomas (Sph-Ob) based on anatomic site.

There are genomic alterations in 68% (13 of 19) of orbital meningiomas. Sph-Ob tumors frequently exhibited monosomy 22/22q loss (70%; 7/10) and deletion of chromosome 1p, 6q and 19p (50% each; 5/10). Among genetic alterations, loss of chromosome 1p and 6q were more frequent in clinically progressive tumors. Chromosome 22q loss also was detected in the majority of Ob meningiomas (75%; 3/4) but was infrequent in ON meningiomas (20%; 1/5). In general, Ob tumors had fewer chromosome alterations than Sph-Ob and ON tumors. Unlike Sph-Ob meningiomas, most of the Ob and ON meningiomas did not progress even after incomplete excision, although follow-up was limited in some cases. A study suggests that ON, Ob and Sph-Ob meningiomas are three molecularly distinct entities. The results also suggest that molecular subclassification may have prognostic implications [1)].

Clinical features

Spheno-orbital meningiomas often present with Visual impairment due to invasion of the optic canal by the tumor.

Differential diagnosis

Intraosseous primary intracranial malignant melanoma

A case of a man with a history of rapidly arising motor slowing associated with urinary incontinence, presenting with mild convergent strabismus caused by paralysis in abduction in the right eye. A brain CT showed a lesion compatible with malignant sphenoorbital meningioma, and the patient underwent gross total resection. Intraoperatively, the blackish lesion infiltrated and eroded the bone; it was placed externally on the dura mater with a mild reaction and without attachment. Histological examination confirmed intraosseous primary intracranial malignant melanoma They report the first intraosseous spheno-orbital PIMM case and present an embryological theory about how this unusual tumour can develop [2].

Treatment

Sphenoidal hyperostosis that results in incomplete resection makes these tumors prone to high rates of recurrence with postoperative morbidity resulting in a nonfunctional globe. High-dose radiation therapy has often been described as the only treatment capable of achieving tumor control, although often at the expense of the patient's progressive visual deterioration.

The complete surgical resection of a spheno-orbital meningioma is sometimes impossible because of the typical involvement of delicate structures of the orbital cone. The use of suitable micro instruments opens the possibility that the complete surgical resection of tumors such as meningioma in this anatomical region be the rule rather than the exception.

Case series

In 4 patients of fourteen, the transorbital endoscopic approach was combined with an endonasal approach. Mean age was 51 and male-to-female ratio was 1:6. In 8 patients (57.1%), an intraorbital involvement was observed, 3 of them (21.4%) showed significant intraconal disease. No patient presented significant cavernous sinus infiltration. Main presenting symptoms were proptosis, diplopia, and visual impairment in 14, 6, and 6 patients, respectively. Mean proptosis improvement was 2 mm (standard deviation 2.3). They observed no major postoperative complications.

This preliminary clinical experience seems to demonstrate that selected spheno-orbital meningiomas can be safely managed by means of an endoscopic transorbital route through a superior eyelid approach. Patients with orbital or cavernous sinus infiltration are at highest risk of persistence [3].

2016

Meningioma-associated proptosis (MAP) can be cosmetically and functionally debilitating for patients with sphenoorbital meningioma and other skull base meningiomas, and there is limited information on the quantitative improvement in proptosis after surgery. Because less extensive removals of tumor involving the orbit fail to reduce proptosis, Bowers et al., has adopted an aggressive surgical approach to the removal of tumor involving the periorbita and orbit. The authors of this study retrospectively reviewed outcomes of this surgical approach.

All surgeries for MAP performed by a single surgeon between January 1, 2002, and May 1, 2015, were reviewed. Age, sex, visual symptoms, number and types of surgical treatments, cavernous sinus

involvement, complications, duration of follow-up, residual tumor, use of adjuvant radiation therapy, and extent of proptosis resolution as measured by the exophthalmos index (EI) pre- and postoperatively and at the final follow-up were recorded.

Thirty-three patients (24 female [73%]) with an average age of 51.6 years were treated for MAP. Of the 22 patients with additional visual symptoms (for example, loss of visual acuity, field cut, or diplopia), 15 had improved vision and 7 had stable vision. No patients had worse proptosis after treatment. The average preoperative EI was 1.39, the average immediate postoperative EI was 1.23, and the average final EI at the most recent follow-up was 1.13. Thus, average overall EI improvement was 0.26, but the average immediate EI reduction was 0.16, demonstrating that proptosis progressively improved during the postoperative period. Residual cavernous sinus involvement was present in 17 of 18 patients who had had preoperative cavernous sinus meningioma involvement. Only 2 patients in the series had recurrent tumor at the orbital region, and their proptosis improved again after reoperation. One case of delayed vasospasm and 2 cases of postoperative trigeminal numbness (V2) were recorded. The average follow-up was 4.5 years (53.8 months).

In this series, all patients experienced proptosis improvement and none had worse visual symptoms at the final follow-up, although proptosis resolution occurred over time. Only 2 patients had tumor recurrence at the orbit that required surgery. Surgical complications were uncommon. Study results suggest that aggressive resection of MAP is well tolerated and offers superior proptosis elimination with infrequent recurrence at the orbit. Importantly, no cases of enophthalmos were noted despite the lack of formal reconstruction of the orbit [4].

2013

A surgical series of 60 spheno-orbital meningiomas is reviewed. The preoperative visual symptoms, the involvement of the optic canal in both neuroradiological studies and surgical descriptions, the different surgical approaches are reviewed. These data are correlated with the postoperative visual outcome.

The 60 spheno-orbital meningiomas were classified in 4 types according to the intraorbital tumor localization: type I, supero-lateral (18 cases); type II, inferomedial (8 cases); type III, orbital apex (22 cases); type IV, diffuse (12 cases). Thirty-six of the 60 patients (60%) had variable decrease of the visual acuity on the tumor side. Forty-three patients (71.6%) had tumor extension into the optic canal on imaging studies. On the whole, 36 patients among 43 with invasion of the optic canal (83.7%) had preoperative visual dysfunction; on the other hand, none among 17 patients without tumor invasion of the optic canal had visual dysfunction. The surgical approaches according to the tumor location were as follows. A supraorbital-pterional approach was used in the 8 inferomedial tumors, in the 22 orbital apex tumors, and in 9/12 diffuse tumors; these last two types had concentric involvement of the optic canal. Three diffuse tumors with significant extension in the infratemporal fossa were operated on via a frontotemporal-orbitozygomatic approach. A wide decompression of the optic canal was performed in all cases, excepting in two inferomedial tumors without optic canal invasion. The 18 patients with lateral tumors were approached via a lateral orbitocranial approach, including removal of the sphenoid wing and lateral orbital wall without bone flap; the resection of the lateral aspect of the optic canal was performed in the 3 cases with canal invasion. Postoperative improvement of the visual function was observed in 18 of 36 cases with visual dysfunction (50%). The rate of visual improvement was significantly higher in cases with lateral involvement (3/3 or 100%) than in those with concentric involvement of the optic canal (11/27 or 40.7%).

The invasion of the optic canal by the tumor is the main reason of visual dysfunction in patients with

spheno-orbital meningiomas. A wide opening of the optic canal must be performed routinely in patients with orbital apex and diffuse orbital tumors, where there is concentric invasion of the optic canal wall. In these cases the supraorbital-pterional approach is the technique of choice. In selected cases with lateral intraorbital tumors and invasion of the lateral aspect of the optic canal the complete tumor resection coupled with good decompression of the optic nerve may be achieved via a less invasive lateral orbitocranial approach without craniotomy [5)].

2010

30 patients, were surgically treated for spheno-orbital meningiomas performing a fronto-pterional approach by or under the supervision of the senior author (J. Meixensberger) between May 2001 and February 2006. There were 22 woman and eight men with a mean age of 54.4 years. The follow-up period ranged from 3 to 75 months (mean: 33.7 months).

The majority of patients presented with a clinical triad of visual impairment (74%), progressive proptosis (55%) and visual field defects (40%). Total microscopic tumor resection was achieved in ten patients (33%). Visual acuity improved in 65% of the patients, and 40% of these returned to normal vision. Pre-existing cranial nerve deficits remained unchanged in the majority of patients (88%) and improved in 12%. Temporary new cranial nerve deficits occurred in three patients. The rate of permanent non-visual morbidity was 10% (three of 30 patients). Eight patients (27%) received post-operative radiotherapy with an overall tumor growth control rate of 63%. The overall recurrence rate was 27% (eight of 30 patients).

Sufficient tumor control can be achieved with minimal morbidity and satisfying functional results [6)].

2005

This series consisted of 25 patients who were retrospectively analyzed over a 12-year period. Visual function was evaluated pre- and postoperatively in all patients. A standardized surgical approach to a frontotemporal craniotomy and orbitozygomatic osteotomy with intra- and extradural drilling of the optic canal and all the hyperostotic bone was performed. Orbital and cranial reconstruction was performed in all patients. The follow-up period was 6 months to 12 years (average 5 years). The patients presented with the classic triad of SOM: proptosis (86%), visual impairment (78%), and ocular paresis (20%). A gross-total resection was achieved in 70% of patients with surgery limited by the superior orbital fissure and the cavernous sinus. Proptosis improved in 96% of patients with 87% improvement in visual function. Ocular paresis improved in 68%, although 20% of patients experienced a temporary ocular paresis postoperatively. There were no perioperative deaths or morbidity related to the surgical approach or reconstruction. Ninety-five percent of patients reported an improved functional orbit. There was tumor recurrence in 8% of patients; in one case recurrence was delayed for longer than 11 years.

Sphenoorbital meningiomas are a distinct category of tumors complicated by potentially extensive hyperostosis of the skull base. Successful resection requires extensive intra- and extradural surgery, necessitating drilling of the optic canal and an orbital osteotomy within anatomical limitations. The bone resection requires reconstruction with autograft, allografts, or alloplast for improved orbital function. All aspects of the clinical triad improved. A radical resection can be achieved with low morbidity, providing a significantly improved clinical outcome in the long-term period [7)].

1)

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