Management of Large Diaphragma Sellae Meningiomas

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Diaphragma sellae meningiomas (DSM) are rare tumors that represent less than 1% of all intracranial meningiomas. Presenting symptoms include visual deterioration, pituitary insufficiency, and hypothalamic dysfunction.

These tumors typically arise from the retrochiasmatic portion of the diaphragma sellae dura; therefore the optic chiasm is displaced anteriorly by the tumor’s mass, which effectively narrows the transnasal operative corridor via the sphenoidal sinus. Thus, the endoscopic endonasal approach (EEA) provides suboptimal access as the optic nerves would obscure the tumor and increase the risk of iatrogenic injury.

DSMs are further complicated because of their involvement with the pituitary stalk and gland, as well as the hypothalamus. These tumors often encase the small perforating branches and major arteries of the Circle of Willis. Surgical resection requires meticulous planning and expertise.

Because of their location, DSMs may be inadvertently identified as pituitary adenomas, craniopharyngiomas, or tuberculum sellae meningiomas (TSM) on MRI. While these other tumors deviate the chiasm superior and/or posteriorly, DSM have the key feature of displacing the pituitary stalk and optic chiasm anteriorly. Also, DSMs can be differentiated from TSMs because the later cause skull base hyperostosis and frequently involve the optic canals. While EEA is the ideal approach for many suprasellar tumors, a focused craniotomy provides the safest approach when the chiasm is displaced anteriorly and the endoscopic corridor is narrowed.

CASE STUDY

A 54 year-old woman presented with a one year history of progressive bilateral visual problems, somnolence, and fatigue. Visual acuity was 20/100 in the right eye and 20/30 in the left eye. A high-resolution MRI (Figure 1) revealed a 4 x 4 x 3 cm suprasellar meningioma arising from the posterior aspect of the right diaphragma sellae. The tumor displaced the optic chiasm anteriorly and superiorly while deflecting the hypothalamus posteriorly and superiorly. All of the arteries of the Circle of Willis as well as the pituitary stalk were encased by the lesion. The tumor also projected into the sella and displaced the diaphragma dura inferiorly.

Surgical Technique

The tumor was approached through a right focused orbitozygomatic approach (FOZA) developed at the Skull Base Center at the Baylor University Medical Center. This approach allows for extradural access to the optic canal, anterior clinoid process, oculomotor triangle, and the diaphragma sellae (which is the origin of this tumor) while resulting in minimal brain retraction or manipulation. After the optic canal was opened and the clinoid was removed extradurally, feeding vessels to the tumor were coagulated and the tumor was internally debulked. The tumor was then dissected away from the optic apparatus, both the right and left oculomotor (III) cranial nerves, the hypothalamus, and the arteries and perforators of the Circle of Willis.

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The pituitary stalk, which was displaced anteriorly and to the left, was identified in a fold of the tumor. The stalk as well as the hypophyseal arteries were preserved while the tumor was dissected away; a gross total resection (Grade I) was achieved. Histopathology revealed a meningioma with rhabdoid cell changes.

**Post-Operative Course**

The patient did well post-operatively without any new neurological or endocrine deficits. She was discharged to home on post-operative day #4. After one year, MRI (Figure 2) demonstrates complete tumor resection without recurrence. Moreover, there was an absence of temporal and frontal lobe FLAIR or T2 abnormalities, which are typically seen in conventional surgical approaches that traumatize or manipulate the brain tissue.

The patient’s somnolence and fatigue resolved and her vision was markedly improved (OD 20/40, OS 20/20). Her endocrine function was unchanged and there was no evidence of pituitary insufficiency.

**CONCLUSION**

In summary, large diaphragma sellae meningiomas can present significant surgical challenges. Accurate identification and intimate knowledge of the skull base anatomy allows for choosing the ideal surgical approach. Complete resection and favorable outcomes are possible with the advent and implementation of innovative and modern skull base approaches.