Case Report

Extracranial Meningiomas in the Head-and-Neck Region: A 15 Years' Experience

D Krasic, M Stojanovic, V Petrovic[1], Z Pešic

Department for Maxillofacial Surgery, Clinic for Stomatology, Niš, [1]Department for Histology and Embryology, Faculty of Medicine, University of Niš, Serbia

Abstract

Meningiomas constitute a large group of tumors of the central nervous system, with the prevalence of 18%–20%. Extracranial localization of meningeomas could appear due to their extracranial origin or due to the spreading of intracranial meningeomas. In this paper, we present our 15 years' experience in the diagnosis and treatment of extra- and intracranial meningiomas invading the head-and-neck region. In the period from 2001 to 2016 at our clinic, there were five patients (three men and two women), with meningiomas of extracranial origin or meningiomas of intracranial origin, spreading into the head-and-neck region. All patients were surgically treated, with the addition of adjuvant radiation therapy in some of the cases. Postoperative and postirradiation complications were described as well.

Keywords: *Extracranial, head and neck tumors, meningioma*

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Introduction

Meningiomas constitute a large group of tumors of the central nervous system, with the prevalence of 18%–20% of all intracranial neoplasms.[1] Intracranial meningiomas, anatomically situated in the anterior and middle cranial fossa, most frequently spread (similar to skull base tumors) to the orbit, temporal and frontal bone, temporomandibular joint, and paranasal cavities and rarely to the nasopharynx.[2,3] Extracranial meningiomas are rather rare, being described in the literature in about 2% of the cases, most commonly involving the head-and-neck region, especially the orbits and nasal floor.[4-6]

We present here our 15 years' experience in the diagnosis and treatment of extra- and intracranial meningiomas invading the head-and-neck region.

Patients and Results

In the period from 2001 to 2016 at our clinic, 27 patients with meningiomas were followed up. This study involves five patients (three men and two women) with meningiomas of extracranial origin or intracranial meningiomas spreading into the head-and-neck region. The patients were aged 30–65 years (mean, 47.5 years). One patient had a tumor of extracranial origin from the orbit, two had tumors with extracranial spread into the infratemporal and temporal fossa, and two patients presented with the tumor spreading in orbit and nasopharynx.

Detailed clinical examination of each patient was performed, and multislice computed tomography (CT) or nuclear magnetic resonance (NMR) imaging of the head and neck was performed. Preoperative incision biopsy was performed for the purpose of differential diagnosis, and definitive diagnosis was made by histopathological analysis postoperatively [Figure 1a and b]. All patients were surgically treated, with the addition of adjuvant radiation therapy in some of the cases, in accordance with the protocols of the European Society for Medical Oncology. Postoperatively, the patients were followed up for at least 2 years, monthly at first, and later at intervals of 3 and 6 months.

All the patients were divided into two groups by topographical localization of their tumors.

The first group consisted of three patients (two men and one woman), aged 40.5 years on the average. Two of

Address for correspondence: Dr. M Stojanovic, Bulevar dr Zorana Đinđića 52, 18000 Niš, Serbia.
E-mail: stojcha79@gmail.com

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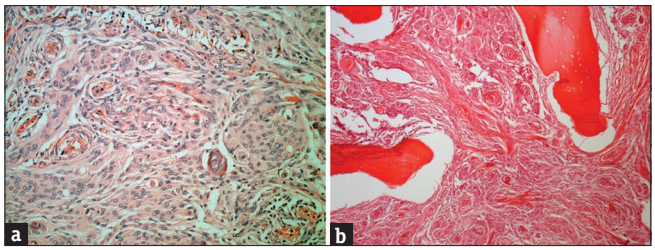

Figure 1: (a) Meningothelial meningioma, (H and E, ×20). Tumor has a lobular structure, with cells arranged as a syncytium. Cells have eosinophilic cytoplasm, round, and uniform nuclei. Furthermore, a psammoma body can be seen on the figure, (b) Meningothelial meningioma, H and E, ×10. Tumor infiltrates osseous tissue

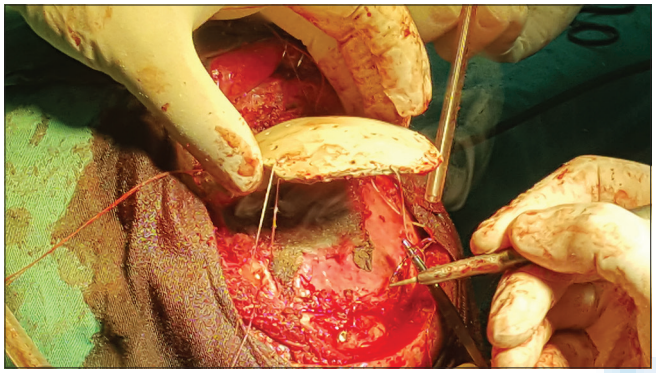

Figure 3: Intraoperative picture: A patient with reconstruction-polyetheretherketone optima prosthesis

them had meningiomas of intracranial origin extending to the orbit and nasopharynx, while one had meningioma of extracranial origin from the orbit. They were treated with anterior craniotomy and orbitotomy, with temporal maxillectomy. Primary reconstruction with local flaps was done in all of the cases. Reconstruction of the defect in the frontoorbital region with a polyetheretherketone implant prosthesis was done as a second-stage procedure in one patient [Figures 2 and 3]. Local flaps for the reconstruction of dura involved the fascia lata, temporal fascial, and pericranial flaps. Bone defects were reconstructed using a titanium mesh and soft tissues with local flaps. Postoperative complications in the form of reduced vision and bulbomotor function were seen in one patient. One patient relapsed after 3 years and was surgically treated.

The second group consisted of two patients (one man and one woman) with the disease spreading extracranially into the temporal and infratemporal space. The surgical treatment involved anterior craniotomy with temporal maxillectomy and resection of the zygomatic arch, and primary reconstruction was performed using local flaps. One patient was postoperatively irradiated due to incomplete resection. There were no relapses in the postoperative period. Complications occurred in both patients in the form of limited mouth opening and paresis of the facial nerve. Postoperative physical

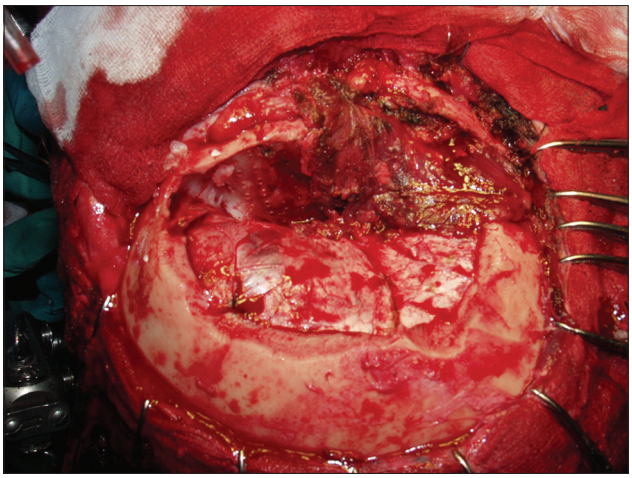

Figure 2: Intraoperative picture of a patient with meningioma with extracranial spreading to orbit and nasopharynx

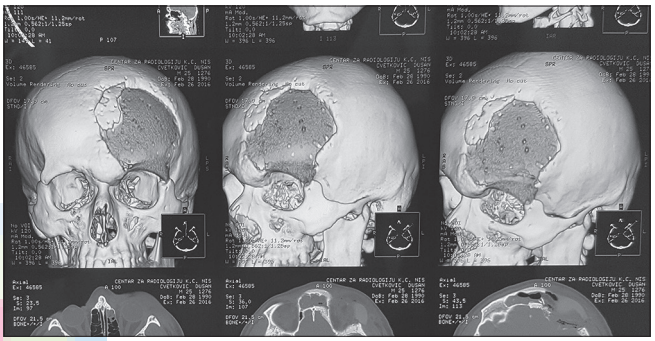

Figure 4: Postoperative computed tomography scan: A patient with reconstruction-polyetheretherketone optima prosthesis

therapy was administered, with symptoms receding by about 60%–70% in both cases.

Control CT scans were done postoperatively to assess the success of surgical procedure [Figure 4]. In all five cases, the pathohistological analysis showed Stage I meningioma.

Discussion

Etiopathogenesis of meningioma has not yet been completely elucidated, some describe their origin from arachnoid cells, while others suggest that they originate from undifferentiated mesenchymal cells and perineural cells.[7-9] As the pioneers in the classification of meningiomas, Hoye *et al*. classified extracranial meningiomas into four groups: Type I – extracranial propagation of meningiomas of intracranial origin; Type II – extracranial propagation of meningiomas with skull base origin; Type III – ectopic meningiomas without any contact with the base, cranial nerves, or endocranium; and Type IV – extracranial metastases with intracranial lesion.[10] Lang *et al*. modified the Hoye's classification adding the subgroups describing calvarial involvement. Furthermore, there is the classification

described by Lopez *et al.* as well, combining different etiological criteria for these tumors.[11] It is beyond any doubt that the origin of meningiomas is related to their clinical characteristics and association with the dura and thus determines further therapeutic management of these tumors.[4] In our study, four patients corresponded to Hoye's Type I and one patient corresponded to Hoye's Type II disease.

CT imaging has been universally accepted in the diagnosis of extracranial meningiomas, where they are seen as a thin, heterogeneous shadow compared to the adjacent tissue, especially with contrast-enhanced scanning.[12,13] NMR imaging can be used to facilitate the diagnosis because it produces better visualization related to adjacent soft tissues.[9] Although some studies suggest fine-needle aspiration as a method of choice in the diagnosis of meningioma, the definitive diagnosis of meningioma is made based on histopathological analysis.[13,14] The World Health Organization (WHO) has classified them into three histological stages: the first, characterized by concentric cellular spirals with calcified bodies; the second, demonstrating a higher mitotic activity with smaller cells and nonconcentric distribution and with the appearance of necrotic areas; and the third type, comprising malignant meningiomas.[13] The WHO protocol indicates adjuvant radiation therapy for Stage I disease with incomplete resection and for Stage II and Stage III anyway.[15]

The treatment of meningiomas is primarily surgical. Throughout the literature, mortality rates for benign lesions are about 5% and for malignant ones around 30%.[4,15] In case of extracranial meningiomas, a timely and precise diagnosis is essential and successful treatment of meningiomas requires multidisciplinary work, as the only way to assure therapeutic success and to avoid possible complications.

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Conflicts of interest

There are no conflicts of interest.

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