ESTHESIONEUROBLASTOMA – CASE REPORT

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Keywords: *esthesioneuroblatoma, multidisciplinary approach*

Abstract: *Esthesioneuroblatoma is a rare tumour arising from the olfactory epithelium of the nasal vault which frequently invades the cranial base, paranasal sinuses and orbits. Objectives: The aim of this study is to analyze the natural history, treatment and prognosis of this pathology. Methods: This paper reports on a 52 year-old male patient, admitted in the Neurosurgical Department of Sibiu, in January 2011 for esthesineuroblasotma with secondary invasion of the frontal lobe, maxillary sinus and orbits. The patient was operated by a multidisciplinary team through a craniofacial approach with gross total tumoral removal and cranial base reconstruction with Titan Micro mesh and periost. Results: The postoperative course was uneventfully and the patient was discharged on the 14th day postoperatively. Conclusions: The very good results are due to the multidisciplinary approach.*

Cuvinte cheie: *estezioneuroblastom, abord multidisciplinar*

Rezumat: *Estezioneuroblastomul este o tumoră rară ce provine din epiteliul olfactiv nazal şi invadează frecvent baza de craniu, sinusurile paranazale şi orbita. Scopul prezentării este analizarea istoricului acestei afecţiuni, a tratamentului şi prognosticului. Metodă: Se prezintă cazul unui pacient în vârstă de 52 de ani, internat în secţia clinică de neurochirurgie Sibiu în ianuarie 2011 pentru estezioneuroblasom cu invazie secundară a lobului frontal, a sinusului maxilar şi a orbitei. Pacientul a fost operat de o echipă multidisciplinară prin abord craniofacial, cu îndepărtarea tumorii şi reconstrucţie de bază de craniu cu placuţă de titan şi periost. Rezultate: Evoluţia intra şi postoperatorie a fost favorabilă. Concluzii: Rezultatele favorabile se datorează abordului multidisciplinar.*

INTRODUCTION

Modern surgery includes orbital surgery in skull base surgery. Orbital surgery is not a separate speciality but it requires a very good interdisciplinary collaboration: ophthalmologist, neurosurgeon, ENT surgeon, OMF surgeon.

Esthesioneuroblatoma is a rare tumour arising from the olfactory epithelium of the nasal vault, which frequently invades the cranial base, paranasal sinuses and orbits. Esthesioneuroblastoma is an aggressive malignant tumour derived from the specialized neuroepithelium of the upper nasal cavity.

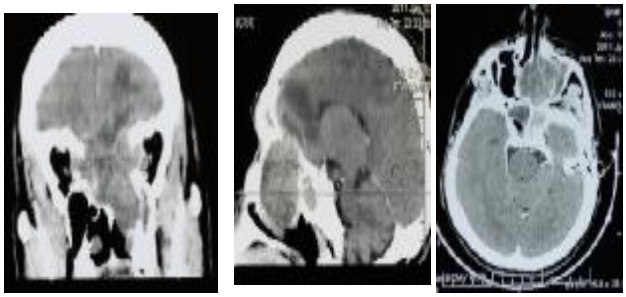
Terminology: olfactory neurocytoma, olfactory neuroepitelioma, neuroendocrine carcinoma, esthesioneurotpithelioma, esthesioneurocytoma, Neuroblastoma, esthesioneuroblastoma

The aim of this study is to analyze the natural history, treatment and prognosis of this case.

CASE REPORT

A 52 year-old male patient, admitted in the Neurosurgical Department of Sibiu in January 2011 for esthesineuroblasotma with secondary invasion of the frontal lobe, maxillary sinus and orbits. The patient was operated by a multidisciplinary team through a craniofacial approach with gross total tumoral removal and cranial base reconstruction with Titan Micro mesh and periost. CT exam (coronal, axial and sagital cuts native, plus contrast) revealed anterior cranial base mass; occupied whole ethmoidal sinus, and upper nasal cavity bilaterally with invasion of maxillary sinus and intracranial invasion of the left frontal lobe; after contrast injection, the tumour presented moderate inhomogenouslly enhancement (figure no. 1).

Figure no. 1. CT exam (coronal, axial and sagital section plus contrast)



Cranial MRI (T1 weighted with gadolinium, coronal, axial and sagital) revealed anterior cranial base tumour with intense inhomogenouslly contrast enhancement with intracranial invasion of the left frontal lobe; invasion of the orbits with mass effect (figure no. 2).

Figure no. 2. Cranial MRI (T1 weighted with gadolinium, coronal, axial and sagital)

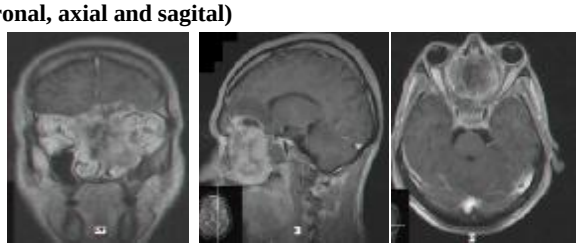


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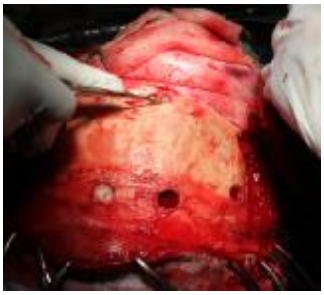
Figure no. 3. Intraoperative aspects. A. Scheme, B. Trepan holes, C. Craniotomy, D. The tumour

Surgical technique

A



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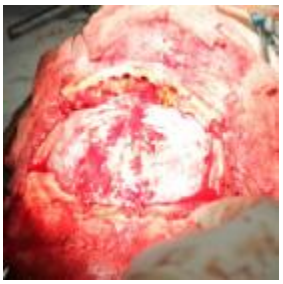

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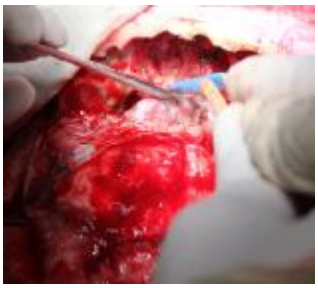

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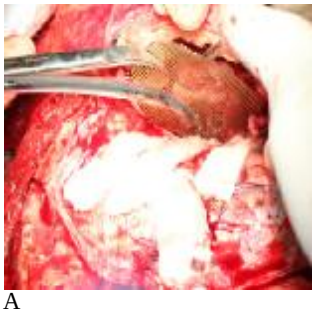
Bicoronal scalp incision, bicoronal scalp flap, bifrontal craniotomy, exenteration of mucosa of frontal sinus (figure no. 3) with cranialisation of frontal sinus, the facial approach of this tumour was performed through (dismantling technique of the facial skeleton) nasal translocation, gross total tumoral removal; en bloc resection, the periorbital membrane was intact, anterior cranial base reconstruction with Titanium Micro Mesh, fascia and periost, watertight suture (figure no.4).

Figure no. 4. Intraoperative aspects, A. Cranial base reconstruction, B. Bone flap fixation, C. Transfacial approach, D. Immediate postoperative aspect



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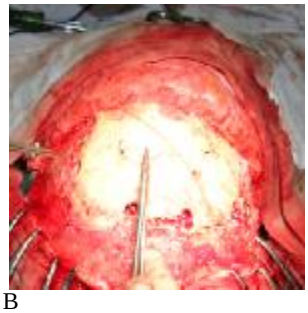

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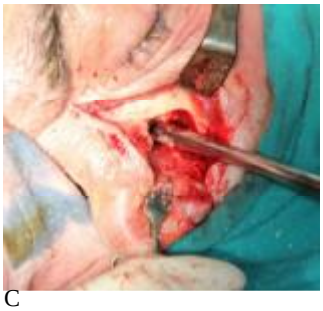

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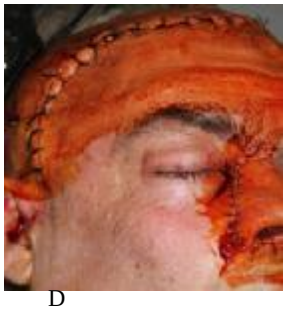


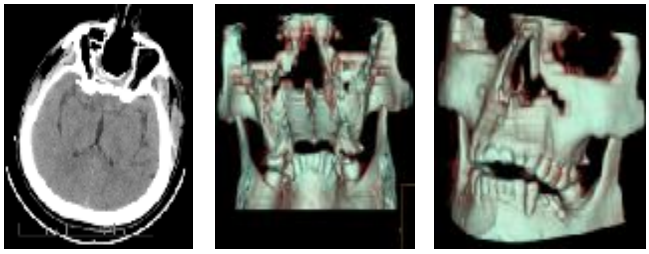
D

RESULTS

The postoperative course was uneventfully and the patient was discharged on the 14th postoperative day. Postoperative CT revealed gross total tumoral removal and no complications (pneumocephalus, hematomas, hemorrhages) (figure no. 5).

Proptosis was reduced and the visual acuity was unchanged. The histopathological exam revealed estesioneuroblastoma. The patient was directed afterwards to the Oncology Clinic where he underwent radiotherapy (50 Gy). The clinic and imagistic control at 6 months did not reveal any relapse.

Figure no. 5. Postoperative CT exam



Conclusions:

Esthesioneuroblastoma is an aggressive malignant tumour derived from the specialized neuroepithelium of the upper nasal cavity. The tumour can spread submucosally in all directions, thereby involving the paranasal sinuses, nasal cavity, and surrounding structures.

Because sinonasal malignant lesions are rare and difficult to distinguish from their benign counterparts, their diagnosis is challenging.

Multimodality treatments are the most frequently advocated interventions, and craniofacial resection is the most common therapy. Craniofacial resection has evolved into the best surgical procedure for achieving safe, en bloc resection of disease.

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