Cavernous Sinus Exenteration & Radical Neck Dissection for Dedifferentiated Adenoid Cystic Carcinoma with Neuroendocrine Features

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Background

- Adenoid cystic carcinoma (ACC) is a rare salivary gland malignancy known for an indolent but relentless course and a high affinity for perineural invasion[1,2]
- The "dumbbell" extension from the parapharyngeal space through the foramen ovale into the cavernous sinus creates a significant anatomical constraint, necessitating a multidisciplinary, dual-compartment surgical strategy[3,4]
- Radical exenteration results in a large-volume defect connecting the neck and intracranial space for maximal tumor resection[4,5]

Case Description

- A 35-year-old woman presented with a high-grade epithelioid malignancy of the right parotid gland with parapharyngeal extension.
- Within one week, she developed rapid cranial neuropathies, including near-complete ophthalmoplegia with complete ptosis and pupil sparing (trace downgaze only, relative CN IV sparing), and trigeminal sensory loss and pain involving V1–V2 with minimal V3 sparing.
- Imaging (Figure 1) demonstrated extensive invasion of the cavernous sinus with extension through the foramen ovale, producing a “dumbbell” tumor configuration.
- Cerebral angiography with right ICA balloon test occlusion was tolerated without neurological deficit, confirming adequate collateral circulation.
- Given the fulminant progression and aggressive skull base invasion, a multidisciplinary radical resection was pursued for local disease control.

Surgical Approach

Multidisciplinary skull base resection combining head and neck surgery and neurosurgery for aggressive extracranial–intracranial disease.

Head and neck exposure:
- Modified Blair and neck dissection incision.
- Radical parotidectomy and circumferential mobilization of the parapharyngeal tumor.
- Carotid artery skeletonization for proximal vascular control.
- Tumor-involved external carotid artery branches sacrificed; facial nerve branches preserved and confirmed intact with stimulation.

Cranial exposure:
- Right extended frontotemporal-orbitozygomatic craniotomy with extradural middle fossa approach.
- Anterior clinoidectomy and subfrontal cisternal release for proximal and distal internal carotid artery control.
- Cavernous sinus exenteration performed due to extensive cranial nerve invasion.

Tumor mobilization and resection:
- Tumor extended through the foramen ovale, creating continuity between intracranial and infratemporal/parapharyngeal compartments.
- Coordinated superior–inferior dissection enabled en bloc tumor delivery through the cervical corridor.

Reconstruction:
- Abdominal fat graft obliteration of the expanded foramen ovale and infratemporal defect.
- Pericranial flap reconstruction of the cavernous sinus and middle fossa floor.

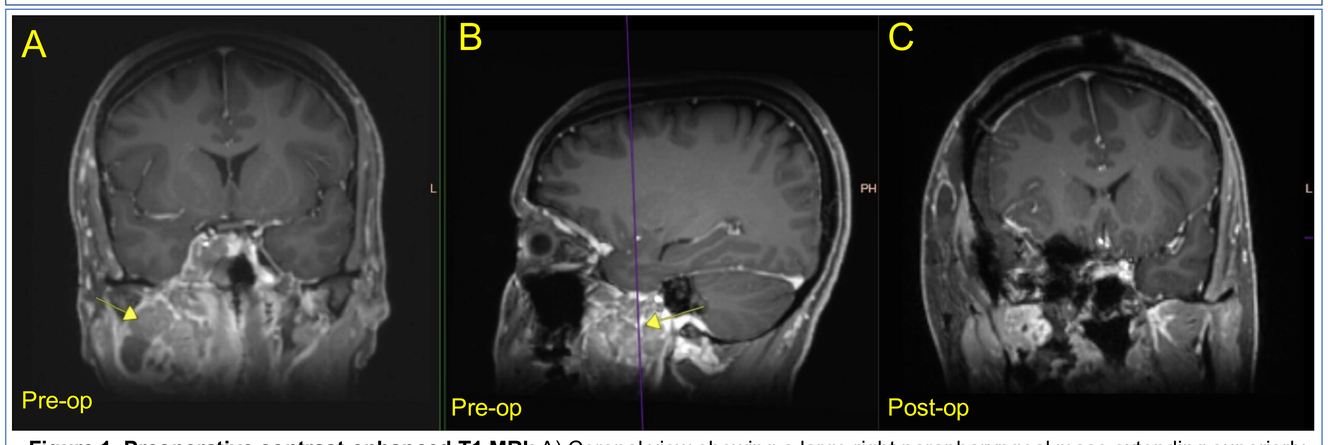

Figure 1. Preoperative contrast-enhanced T1 MRI: A) Coronal view showing a large right parapharyngeal mass extending superiorly to the skull base with medial displacement of the nasopharyngeal pillar, B) Sagittal view demonstrating intracranial extension through the foramen ovale into the cavernous sinus and Meckel’s cave, C) **Post operative** coronal view with complete intracranial tumor resection

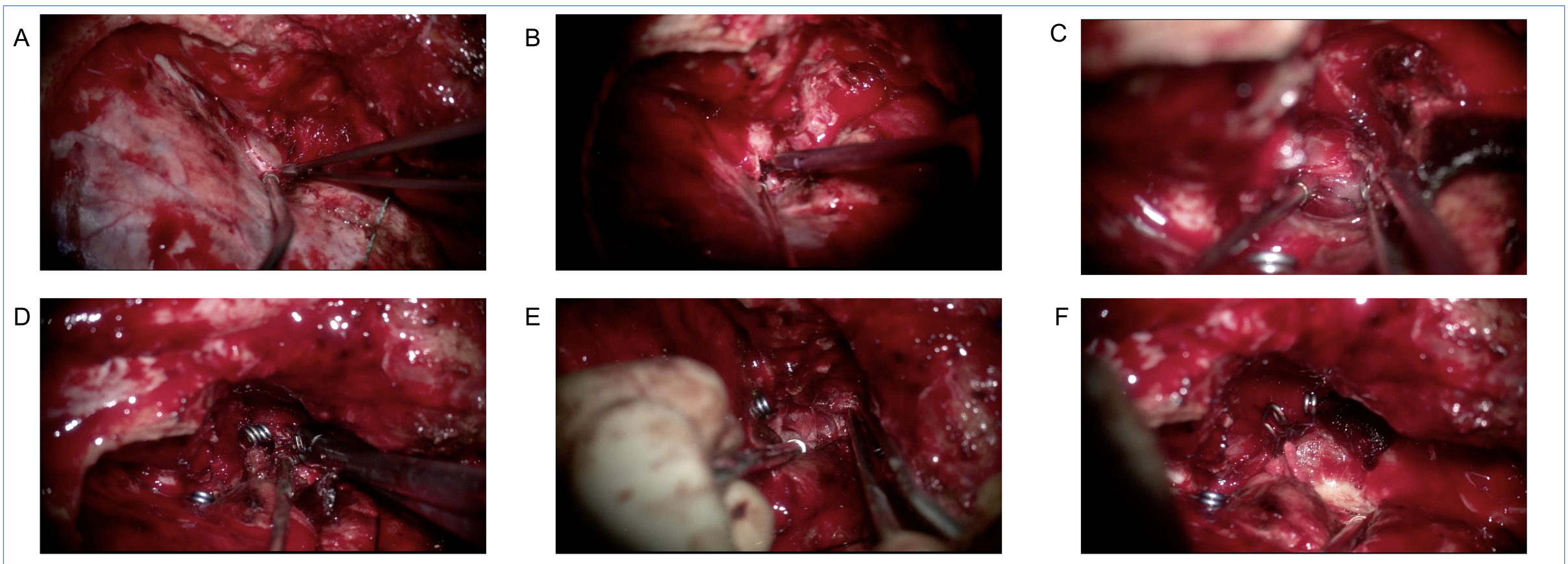

Figure 2. Intraoperative imaging demonstrating**:** A) Mobilization of the meningo-orbital band enabling extradural middle fossa peel , B) Anterior clinoidectomy with temporary supraclinoid ICA clipping for distal vascular control , C) Cavernous sinus tumor resection between CN III and the V1–V2 complex , D) Complete carotid sacrifice with clipping for control at the foramen lacerum given bleeding from cavernous branches, E) Tumor mobilization through the expanded foramen ovale via combined cranial and cervical corridors, F) Final inspection showing a skeletonized cavernous ICA and empty Meckel’s cave.

Clinical Outcome

- Multidisciplinary gross total resection of parapharyngeal and cavernous sinus tumor via combined transcranial and transcervical approach.
- Parent internal carotid artery was sacrificed given complete tumor encasement; involved cavernous branches were sacrificed as required for oncologic resection. Preoperative balloon test occlusion provided an essential safety margin.
- Expected complete right ophthalmoplegia and V1–V3 anesthesia following oncologic cavernous sinus exenteration.
- Facial nerve preserved, with intact stimulation of all branches at case completion, post op exam with trace left droop on activation
- Abdominal fat graft obliteration of the expanded foramen ovale combined with a vascularized pericranial flap; no postoperative CSF leak observed.
- Stable ICU course following complex skull base resection.
- Follow-up: Postoperative imaging confirmed image-complete resection; subsequent recurrence demonstrated favorable response to adjuvant chemoradiation

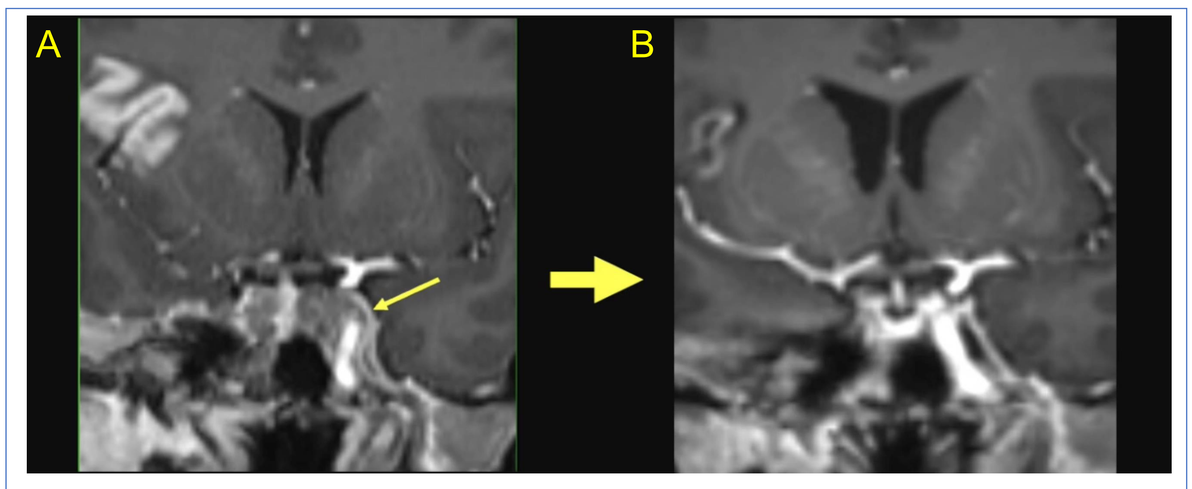

Figure 3. MRI T1 contrast coronal view A) 3-week post op contralateral cavernous sinus syndrome with tumor spread B) no radiographic evidence of disease following overall 6-week course of radiation and chemotherapy with cisplatin and etoposide

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