Case Report

A rare case of intracranial teratocarcinosarcoma: Case report and review of literature

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Abstract

Background: Teratocarcinosarcoma (TCS) is a rare malignant neoplasm with epithelial and mesenchymal components such as fibroblasts, cartilage, bone and smooth muscle. With less than 100 total reported cases, this malignant neoplasm is rarely encountered by neurosurgeons because it primarily involves the nasal cavity and paranasal sinuses.

Case Description: A 55-year-old male with chronic frontal headaches was found to have a frontal mass with involvement of nasal sinus and right ethmoid sinus. The patient underwent preoperative embolization of tumor followed by bilateral frontal craniotomy for near total resection of the tumor. Patient did well postoperatively without new neurological deficits.

Conclusion: Although cases with intracranial involvement are scarce, treatment with surgical resection with or without adjuvant treatments of chemotherapy and radiation therapy is the most widely accepted with goal for gross total resection. In our case, we achieved near total resection and the patient continues to do well without any gross neurological deficits.

Key Words: Brain tumor, neuro-oncology, neurosurgery, oncology, teratocarcinosarcoma

INTRODUCTION

Teratocarcinosarcoma (TCS) is a rare malignant neoplasm with epithelial and mesenchymal components, such as fibroblasts, cartilage, bone, and smooth muscle, with less than 100 cases described in the literature.[1] The term was first coined in 1984 by Heffner and Hyams, the key features to properly diagnose TCS are the presence of “fetal-appearing” clear-cell squamous epithelium and organoid structures such as tubular or glandular formations.[2,5] Germ cell components are absent in TCS.[16,18] Benign and malignant epithelial, mesenchymal, and neural elements are present, which include immature tissue with teratoid features.[4,5]

Nearly all reported cases of TCS arise within the nasal cavity or paranasal sinuses, with a few reports having described intracranial invasion into the dura mater or the frontal lobe at presentation or progression, as...
in our case.[5,15,16] Misra et al. reviewed 86 reported cases of sinonasal teratocarcinosarcoma (SNTCS) and found a strong male predilection (87%), mean age of 54.5 years; 18/86 (20.9%) cases described intracranial extension, cribriform plate, and anterior cranial fossa involvement.[10] Mean survival has been reported at 1.7 years with a 60% mortality rate within 3 years.[4] One case series of 10 patients (100% male) had no cases with intracranial and dural/or dural extension.[14] Given the rarity of intracranial involvement of this neoplasm, this tumor is rarely encountered by the neurosurgeon. We report a case of TCS and describe the surgical approach and postoperative care.

**CASE REPORT**

**History and presentation**

A 55-year-old male presented with frontal headaches 3 times a week with progressive memory problems, word-finding difficulties, and new-onset seizures. Magnetic resonance imaging (MRI) of the brain revealed a 7.0 × 6.2 × 4.4 cm enhancing mass extending across the cribriform plate and into the right anterior cranial fossa and right nasal cavity [Figure 1]. The patient was initially referred to ENT for biopsy which revealed TCS; he was then referred to the neurosurgical service for further evaluation.

**Operative intervention**

Given the preoperative diagnosis via transnasal biopsy and presumed hypervascularity of the neoplasm, the patient underwent preoperative embolization followed by bilateral frontal craniotomy via a bicoronal incision with pericranial flap preservation. ENT assisted with exposure of the tumor using the transglabellar subcranial approach. Tumor was found to be pale and hypervascular with a dural breach in the low frontal plane with involvement of the superior ethmoid sinus [Figure 2]. Endonasally, the right middle turbinate and remaining ethmoid sinuses were resected. Gross total resection was achieved with significant decrease in blood loss once the tumor was completely removed. The inferior right anterior pole of the dura just above the superior ethmoids appeared to have tumor invasion. This area of dura was resected and reconstructed using dural repair graft. Then, tisseal was placed within each intervening layer of dural graft and duragen, which was placed over the dural closure. The pericranial flap was placed in the nasaethmoidal defect and an additional 10-cc tisseal was applied. The frontal sinuses were fully cranialized bilaterally. Excess pericranium flap on the left was replaced over the frontal bone.

**Histological staining**

Staining was positive for CK-PanAE1-3CAM52, vimentin, EMA, S100, CK 5/6, P63, synaptophysin, chromogranin, and GFAP. Staining negative for CD99 and neurofilament [Figure 3].

**Postoperative course**

The postoperative course was significant for pulmonary embolism and was otherwise unremarkable. Twenty-four months postoperatively the patient is and doing well without any seizure-like activity with return to baseline neurological status. MRI showed no evidence of recurrence with expected postoperative changes [Figure 1]. He completed adjuvant chemotherapy with three rounds of cisplatin therapy and radiation therapy consisting of 60 Gy in 30 fractions to the nasal cavity, frontal sinuses, and partial brain. He is back to work and is currently being weaned from antiepileptics.

**DISCUSSION**

TCS is a rare malignant neoplasm with epithelial and mesenchymal components. The most common presenting

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**Figure 1:** Postcontrast MRI post-contrast with AP (a) and sagittal (b) images showing enhancing lesion extending from nasal sinus to anterior cranial fossa with frontal lobe invasion. Post-operative postcontrast MRI post-contrast shows Axial axial (c) and AP (d) views with near total resection of mass.

**Figure 2:** Intra-operative images delineating our designated craniotomy (a). The dysplastic appearing dura visualized once craniotomy performed (b).
signs and symptoms are nasal obstruction and epistaxis, found in ~75% to 90% of documented cases.\(^4,10,13,14\) Given the primarily intracranial involvement in our case, the presenting symptoms were neurological in nature, which is rare for this type of neoplasm. To our knowledge, this is the only documented case with seizure as a presenting symptom.

A review of literature revealed 11 cases of TCS with intracranial invasion [Table 1].\(^3,6-9,16,17,19-21\) Although the documented cases with predominant intracranial involvement are scarce, treatment with surgical resection with or without adjuvant treatments of chemotherapy and radiation therapy is the most widely accepted with goal for gross total resection. Despite few intracranial cases described in the literature, patients undergoing gross total resection fare better than those with subtotal resection. Further areas of research should focus on whether neo-adjuvant treatment with chemotherapy/radiation therapy can

| Age, sex | Presentation | Location | Surgery/treatment | Outcome |
|----------|--------------|----------|-------------------|---------|
| Terasaka 1998  | 66, male | Somnolence, apathy, HA, AMS, anosmia | FL, NC, SS, ES | Craniotomy for GTR followed by RT | No recurrence at 27 months |
| Kleinschmidt-DeMasters 2000 | 59, male | Hyponatremia SIADH | FL, FS, ES, SS | Combined endoscopic and craniotomy approach with NGTR | Recurrent disease in right orbit 2 month postoperative underwent re-operation and died 6 months later |
| Endo 2001 | 71, male | EP with tumor in NC | FL, NC, orbit | NACT and surgery for nasal lesion with local recurrence 4 years later and then underwent resection and RT. Recurrence in FL 1 year later and underwent subtotal resection twice | Died from local extension of tumor 7 years from initial onset |
| Wellman 2002 | 60, male | EP | ES, FL | Combine craniofacial approach for resection and subsequent RT | Developed radiation necrosis 3 years postoperatively and recurrent disease localized to NC 4 years postoperatively, then underwent total laser excision. Disease-free 5 years postoperatively 7 months after diagnosis patient presented with extensive nodular enhancement encasing spinal cord and died 1 month later |
| Tchoyoson Lim 2008 | 32, male | AMS, somnolence | FL, ES, NC | Craniotomy for partial resection followed by 2nd craniofacial resection 10 weeks later (tumor found to extent to left LV) |  |
| Vijay 2010 | 55, male | HA, blurry vision, AMS | FL, NC, corpus callosum | Right frontal craniotomy for NGTR followed by RT | Lost to follow up for CTx and died 6 month later |
| Kim 2011 | 46, male | Nasal obstruction | NC, ES, FL | Endoscopic sinus surgery for resection but with remnant in FL. Underwent 2nd surgery craniofacial approach for GTR and subsequent RT. | 7 week postoperative imaging revealed multiple recurrent lesions in FL. Underwent craniotomy for GTR and CTx. Disease free for 29 months since initial diagnosis |
| Weinberg 2014 | 22, female | 16 weeks gestation with HA, N, V, weight loss, AMS | RF, NC | Combined transnasal and cranial approach for NGTR. RT delayed for 5 months secondary to pregnancy | 27 months after RT had elevated BHCG and recurrent FR mass, and subsequently, underwent RT. Resolution of RF lesion 1 month after RT but with multiple new lesions in PF. Died 1 month later |
| Joshi 2014 | 44, male | EP | FL, NC, ES, NP | Unknown extent of resection followed by RT | Recurrent extensive tumor 5 months postoperatively. Started on CTx with significant decrease in tumor size |
| Joshi 2015 | 38, male | EP, HA, V, diplopia | FL, FS, ES SS, NC | NACT followed by combined endoscopic and craniotomy for GTR. Postoperative CTx and RT | Disease-free at 2-month follow-up after completion of CTx and RT |
| Joshi 2015 | 48, male | EP | NC, FL, ES | NACT followed by endoscopic GTR of residual from ES. Postoperative CTx and RT | Disease-free at 3-month follow-up after CTx and RT |

TCS: Teratocarcinosarcoma, HA: Headache, EP: Epistaxis, N: Nausea, V: Vomiting, AMS: Altered mental status, FL: Frontal lobes, NC: Nasal cavity, FS: frontal sinus, SS: Sphenoid sinus, ES: Ethmoid sinus, NP: Nasopharynx, LV: Lateral ventricle, GTR: Gross total resection, NGTR: Near gross total resection, RT: Radiation therapy, CTx: Chemotherapy, NACT: Neo-adjuvant chemotherapy, PF: Posterior fossa, SIADH: Syndrome of inappropriate anti-diuretic hormone secretion, BHCG: Beta-human chorionic gonadotropin
In our case, the benefit of surgical neurology is imperative to obtain a water-tight closure to prevent pseudomeningocele. When dural invasion is suspected, cranialization, exenteration, and obliteration of frontal and gross total resection. Goal of gross total surgical resection is standard with or without chemotherapy and/or radiation therapy. Combined ENT and neurosurgical approaches can limit the number of surgical procedures required for gross total resection. Metastasis is uncommon but has been reported in cases with postoperative residual mass.

**CONCLUSIONS**

TCS is a rare neoplasm rarely encountered by the neurosurgeon given its predilection for sinonasal involvement with majority of cases presenting with sinonasal obstruction and/or epistaxis. Neurological signs and symptoms are very rare with this neoplasm. To our knowledge, this is the first reported case with seizure as a presenting sign. Diagnosis requires ample tissue sample. Once diagnosis is established, the surgeon can plan for the optimal surgical approach to facilitate gross total resection. Goal of gross total surgical resection is standard with or without chemotherapy and/or radiation therapy. Combined ENT and neurosurgical approaches can limit the number of surgical procedures required for gross total resection.

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**Conflicts of interest**

There are no conflicts of interest.

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