Prolonged survival in a patient with sinonasal teratocarcinosarcoma with cranial extension

Case report

SHUNSUKE TERASAKA, M.D., MAX B. MEDARY, M.D., DONALD M. WHITING, M.D., TAKANORI FUKUSHIMA, M.D., EVALYNNE J. ESPEJO, M.D., AND GIRJA NATHAN, M.D.

Departments of Neurosurgery and Surgical Pathology, Allegheny University of the Health Sciences, Pittsburgh, Pennsylvania

Sinonasal teratocarcinosarcoma is a rare malignant neoplasm characterized by the combined histological features of carcinosarcoma and teratoma. The primary symptoms of this tumor are usually nasal obstruction and epistaxis, and a nasal cavity mass is the most common clinical finding. The authors describe an exceptionally rare case in which the patient presented with massive intracranial extension and exhibited confusion as an initial symptom. He subsequently underwent combined radical surgery and radiation therapy and has remained free of disease for 31 months. The surgical approach to the lesion, histological features, and clinical course are detailed.

KEY WORDS • anterior skull base • cranial extension • transbasal approach • radical surgery • sinonasal teratocarcinosarcoma

History. This 66-year-old man presented with a history of hypertension, having suffered a stroke in 1991 with no residual symptoms. The patient’s wife related a slowly progressive history of increasing somnolence, left frontal headache, and apathy that had worsened over a period of 6 weeks. The patient was admitted after acute deterioration in his mental status.

Neuroimaging and Neurological Examination. Computed tomography scans of the brain revealed a large left frontal mass with acute hemorrhage. The patient was oriented to person and place but his affect was flat and his concentration was diminished. His speech was clear and fluent and his memory was intact. The patient was anosmic, and mild papilledema was observed bilaterally. Visual fields and pupillary responses were normal, and the remainder of his neurological examination was unremarkable.

Radiographic Examination. Magnetic resonance (MR) imaging demonstrated an extensive lobulated mass lesion engulfing much of the left frontal region. This mass also involved the superior aspect of the left nasal cavity and the region of the sphenoid sinus. On the precontrast sequence, there were areas of hyperintensity at the periphery of the lesion, indicating a hemorrhagic component (Fig. 1 left). There was a marked, although somewhat inhomogeneous enhancement of the tumor after intravenous infusion of contrast material (Fig. 1 center and right). No extension of the lesion into the left orbit was seen. We observed marked compression of the ventricular system accompanied by effacement of the left frontal horn. A carcinoma originating from the nasal cavity or esthesioneuroblastoma were considered to be the most likely diagnoses.

Operation. An extended transbasal approach was used for extirpation of this large tumor. After a bicoronal incision was made, a subgaleal flap was turned toward the orbital ridge. Vascularized pericranium was preserved and reflected anteriorly to be used for reconstruction of the anterior skull base. The frontal region was widely ex-
posed, including both orbital rims. A bifrontal craniotomy was performed and the supraorbital bar was removed. Using microsurgical techniques, the subfrontal dura was dissected from the orbital roofs revealing destruction of the floor of the anterior fossa. The tumor occupied the left ethmoid and the sphenoid sinuses. Intraoperative pathological evaluation revealed a carcinoma with multiple mitoses. Therefore, the superior turbinate was removed on both sides and the paranasal mucosae and bones were all radically excised. The tumor was debulked internally, a 10-mm dural margin was removed, and a gross-total resection was performed.

A fascia graft of rectus muscle from the abdomen was used to repair the dural defect. The anterior skull base was reconstructed with split calvaria and secured with titanium microplates; it was then covered with vascularized pericranium.

**Histological Findings.** Microscopic examination revealed a heterogeneous tumor consisting of a mixture of immature teratoid or teratoma-like tissue, benign and malignant epithelial or carcinoma tissue, and benign and malignant mesenchymal or sarcoma tissue.

The teratoma-like tissue consisted of abundant nests of immature nonkeratinizing squamous cells with clear cytoplasm, small nuclei, and no intercellular bridges, resembling fetal oral cavity epithelium (Fig. 2a).

The benign epithelial tissue consisted of glands lined by various types of epithelium, including mucinous columnar, ciliated respiratory, and the previously described clear cell squamous tissue. Transitions of one type of epithelium to another were seen (Fig. 2b). The malignant epithelial tissue or carcinoma consisted of poorly differentiated epithelial tissue with glandular and neural rosettes (Fig. 2c). Some neural rosettes were positive for the neuronal markers neuron-specific enolase, chromogranin, and synaptophysin; these findings were consistent with poorly differentiated carcinoma featuring neuroepithelial differentiation (Fig. 2c).

The benign mesenchymal elements consisted of chondroid elements, including cartilage and chondromyxoid areas (Fig. 2f). Malignant mesenchymal or sarcoma cells consisted of spindle cells with cross-striations (Fig. 2g) that were positive for desmin on immunoperoxidase staining (Fig. 2h); these findings were consistent with rhabdomyosarcoma.

The combination of teratoid or teratoma-like elements with carcinoma and sarcoma are findings of teratocarcinosarcoma, a unique heterogeneous tumor with a diverse histological appearance. The tumor described in this report invaded cerebral tissue.

**Postoperative Course.** The patient tolerated the procedure well and his postoperative course was unremarkable. He received a total dose of 5280 cGy of radiation administered to the head and neck in 24 fractions over 12 weeks. Posttreatment MR images demonstrated no tumor recurrence at 27 months (Fig. 3).

**Discussion**

The term teratocarcinosarcoma was coined by Heffner and Hyams in 1984. They identified the clinical and histological characteristics of this rare tumor and described treatment and survival rates based on their series of 20 cases. To date, another 20 cases have contributed to our knowledge of this rare entity.

The initial symptoms in all of the previously reported patients were related to the paranasal sinuses. The tumor shows a variegated histological pattern with a combination of epithelial and mesenchymal components. Thus, small samples, such as those obtained from biopsy procedures, may lead to a misdiagnosis of squamous cell carcinoma, adenocarcinoma, malignant craniopharyngioma, or esthesioneuroblastoma, resulting in confusion regarding the true incidence of this tumor and its potential treatment modalities.
Fig. 2. Photomicrographs of histological preparations showing the heterogeneous composition of the tumor. a: Immature nonkeratinizing squamous cells with clear cytoplasm, resembling fetal oral cavity tissue. H & E, original magnification × 100. b: Benign mucin-containing glands lined by ciliated respiratory-type epithelium showing transition to clear cell squamous cells. H & E, original magnification × 400. c: Carcinoma cells with neural rosettes. H & E, original magnification × 100. d: Glands and neural rosettes positive for synaptophysin on immunoperoxidase staining. Original magnification × 250. e: Poorly differentiated carcinoma cells showing neuroepithelial differentiation as seen on brown cytoplasmic immunoperoxidase staining to neuron-specific enolase. Original magnification × 250. f: Chondroid areas including cartilage and chondromyxoid tissue. H & E, original magnification × 100. g: Rhabdomyosarcoma consisting of malignant spindle cells with cross-striations. H & E, original magnification × 400. h: Rhabdoid differentiation seen on brown cytoplasmic immunoperoxidase staining to desmin. Original magnification × 300.
To our knowledge, this is the first case in which the patient presented with symptoms related to the central nervous system. Furthermore, the MR images demonstrated minimal nasal cavity involvement.

The treatment of this lesion continues to be challenging because of the tumor’s high rate of malignancy and the low survival times for patients (averaging less than 2 years). Previous reports have shown that for a patient to have any chance of a disease-free survival, sinonasal teratocarcinosarcoma requires an aggressive initial therapeutic approach with a combination of radical surgical resection and radiation therapy. However, tumor response to this treatment, particularly in regard to the effectiveness of radiation therapy, remains unpredictable. The local recurrence rate, in 17 (65%) of 26 patients, is consistent with the mortality rate at 3 years posttreatment.

Only two patients underwent chemotherapy in addition to surgery and radiation therapy. The role of chemotherapy in the management of this disease awaits clarification. Shah, et al., reviewed 71 patients with malignant carcinoma involving the anterior skull base and observed that survival was directly related to evidence of dural or parenchymal involvement. Morita, et al., treated 49 cases of esthesioneuroblasotoma and concluded that the best initial procedure was to accomplish gross-total removal of the tumor. These results support radical resection of this tumor, as was performed in our case.

We used the extended transbasal approach in this case to provide exposure of the subfrontal region, paranasal sinuses, clivus, and anteroinferior region of the cavernous sinus. Such an approach allows radical resection of this rare tumor.

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References


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Address reprint requests to: Takanori Fukushima, M.D., Department of Neurosurgery, Allegheny University of the Health Sciences, 420 East North Avenue, Suite 302, Pittsburgh, Pennsylvania 15212–4746.