Craniospinal dissemination in teratocarcinosarcoma

Case report

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There have been fewer than 60 cases of malignant teratocarcinosarcoma (TCS) described in the literature, usually arising in the nose and paranasal sinuses. The authors report on a patient who presented with neurological symptoms caused by a frontal lobe TCS, and in whom widespread spinal tumor dissemination developed. In rare cases, TCSs can occur with a predominantly cranial and neurological presentation and spread to the spinal canal. (DOI: 10.3171/JNS/2008/109/8/0321)

KEY WORDS • magnetic resonance imaging • neoplasm • spinal dissemination • teratocarcinosarcoma

TERATOCARCINOSARCOMAS are aggressive malignant neoplasms characterized by combined histological features of carcinosarcoma and teratoma.1,6–12 Teratocarcinosarcomas are rare tumors that typically arise as a mass in the nasal cavity. Because patients typically present with complaints of epistaxis and nasal obstruction, neurosurgeons seldom encounter these lesions. We describe the case of a patient with a frontal lobe TCS and widespread spinal tumor dissemination. A postmortem examination was not performed in this patient.

Case Report

This 32-year-old man presented with a 2-week history of confusion and abnormal behavior, having become withdrawn and somnolent. On examination he was found to be neurologically intact, but his left pupil was nonreactive. Emergency MR imaging revealed a large 9-cm-diameter mass occupying the frontal lobes with minimal involvement of the ethmoid air spaces and nasal cavity (Fig. 1). The tumor showed inhomogeneous contrast enhancement, with areas of hemorrhage with multiple blood fluid levels. The patient underwent an emergency craniotomy during which a large and extremely hemorrhagic intraparenchymal tumor was partially resected. Histological examination revealed the hemorrhagic tumor consisted of irregular epithelial structures within an atypical stromal background (Fig. 2). Fibrovascular papillary cores were covered with florid epithelial proliferation and glandular formations, some lined by multilayered primitive cells with teratomatous or blastemic areas. The intervening stroma contained numerous pleomorphic spindle cells. Both epithelial and mesenchymal components had a high mitotic rate and a high MIB-1 (Ki 67) labelling index. The epithelial component also showed strong positivity for epithelial markers (Cam 5.2 and AE1/3), and focal staining for S100 protein; there was no reactivity in either component for synaptophysin or glial fibrillary acidic protein. The overall histological appearance was consistent with a high-grade, malignant TCS.

An ear, nose, and throat examination revealed a small hemorrhagic mass between the left middle turbinate and nasal septum. The histological features of the nasal mass were similar to the intracranial TCS. Whole-body dual-modality 18F-fluorodeoxyglucose–PET/CT examination
demonstrated increased uptake consistent with residual nasal and intracranial tumor (Fig. 3). A second focus of faint uptake was noted in the pelvis. However, metastasis from the TCS to the pelvic lymph nodes was excluded on biopsy sampling, which also revealed an unrelated benign schwannoma. The patient underwent a second craniofacial resection 10 weeks after diagnosis. At surgery, the left lateral ventricle was found to be studded with tumor, and whole-brain radiotherapy was started for residual intracranial tumor.

Seven months after the initial diagnosis, the patient presented with back pain, leg weakness, and urinary incontinence. Magnetic resonance images showed stable intracranial tumor but ependymal deposits, extensive nodular enhancement encasing the spinal cord from C-6 to the conus, and infiltration of the lumbosacral thecal sac (Fig. 4). The patient’s condition continued to deteriorate, and he died 1 month later.

Discussion

Teratocarcinosarcoma is a very rare neoplasm. The diagnosis is based on the presence of malignant epithelial elements and ≥ 2 malignant mesenchymal components, such as fibroblasts, cartilage, bone, and smooth muscle. If only a single mesenchymal component is present, the lesion is termed carcinosarcoma. The key teratoid features necessary for recognizing TCS are “fetal-appearing” clear-cell squamous epithelium and organoid structures such as tubular or glandular formations.1,4 Germ cell components are absent in TCS. The tissue heterogeneity and variegated architecture means that small samples from needle biopsy may not show the true histological range of these tumors. In the past, this diversity of histological patterns has resulted in different designations and classifications, including teratocarcinosarcoma,1,4 malignant teratoma biphasic type,8 blastoma,6 and teratoid carcinoma.9 The term teratocarcinosarcoma has been proposed as a unifying diagnosis,2 and fewer than 60 patients with this rare tumor have been described in the literature, mostly in case reports. The histogenesis of TCS is unknown, but these lesions are postulated to arise from primitive cells sequestered in the sinonasal membrane that have the capacity to differentiate into divergent types of somatic cells.1,4,5 Immunohistochemical analysis and ultrastructural findings suggest that they could be primitive neuroectodermal tumors capable of organized divergent differentiation.7,11

Nearly all TCSs arise within the nasal cavity or the paranasal sinuses, and rarely the nasopharynx, orbit, or oral cavity.4,10,12 A few reports have also described intra-
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Fig. 3. Dual-modality 18F-fluorodeoxyglucose–PET/CT image demonstrating increased uptake in the residual tumor.

Fig. 4. Lumbar spine MR image obtained 7 months after diagnosis showing an extensive enhancing tumor encasing the spinal cord and conus medullaris (arrows).

cranial invasion into the dura mater or the frontal lobe at presentation or progression. However, our patient presented with a large intracranial mass and neurological manifestations, and not the typical symptoms of TCS such as epistaxis or nasal obstruction. On MR imaging, it was not immediately appreciated that there was communication between the intracranial tumor and the nasal cavity, and the patient was initially thought to have an intraaxial malignant glial tumor. There has been only 1 previous report of a predominantly intracranial TCS with minimal nasal cavity involvement, and presenting similarly with frontal lobe signs of confusion and somnolence.

Teratocarcinosarcomas are aggressive, rapidly growing tumors, with a poor prognosis. Most patients succumb to extensive disease within 3 years. Magnetic resonance imaging typically demonstrates an infiltrating mass with a low signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and positive enhancement after contrast injection. The recommended treatment is resection, but extensive local destruction often precludes complete removal. Adjuvant radiation therapy may be helpful, but aggressive chemotherapy is less well established. Metastasis has been reported to the cervical lymph nodes and lung, but not dissemination in the cerebrospinal fluid to the spinal canal. Because widespread craniospinal dissemination may occur in malignant intracranial neoplasms such as glioblastoma, medulloblastoma, and lymphoma, this phenomenon would not be unusual for a malignant tumor such as TCS, especially when the intracranial component has spread along the ependymal surface of the ventricles.

This case illustrates that although TCS is an uncommon sinonasal neoplasm, it can present in rare cases as a predominantly intracranial tumor. Neurosurgeons should be aware of this lesion’s close relationship to the nasal cavity and paranasal sinuses, and its potential for cerebrospinal fluid and spinal dissemination.

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References

References


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