Unusual Cranial-Orbital Metastasis of a Malignant Solitary Fibrous Tumor

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Abstract

Background: Malignant solitary fibrous tumors (MSFT) are uncommon spindle cell neoplasms thought to arise from cells of mesenchymal lineages. We describe an unusual MSFT, originating within the orbital compartment and metastasizing to the calvarium and liver.

Results: A 59-year-old man had a history of MSFT of the left eye status post enucleation in 2008, complicated by hepatic metastases, status post stereotactic body radiation therapy, surgical resection, and intraoperative radiofrequency ablation in 2014. One year later, the patient presented with progressive left-sided weakness in the setting of a subacute growing calvarial mass the patient had attributed to unresolved sequelae of a minor head trauma. MRI of the brain demonstrated a right parietal transcalvarial mass with intraaxial extension. The patient subsequently underwent a biparietal craniotomy for tumor resection.

Conclusion: This is the first reported case of a primary orbital MSFT with metastasizing hepatic and cranial metastases. This case illustrates the capacity for significant tumor investment within the central nervous system and its vasculature at the tumor interface, which may place patients at increased risk of postoperative complications.

Case Report

The patient is a 59 year-old male with a remote history of testicular seminoma treated with surgical resection and adjuvant radiotherapy who initially presented with occasional blurry vision. On ophthalmologic evaluation he was found to have a left retinal mass, thought to represent a vascular malformation (Fig. 1). He was treated with multiple intracranial injections of Avastin, however the mass did not respond and was in fact noted to increase in size. Due to the increase in lesion size combined with progressive visual disturbances, the patient underwent left eye enucleation. Pathology demonstrated a spindle cell neoplasm consistent with a malignant solitary fibrous tumor (MSFT). No extracranial extension was seen, thus he did not receive adjuvant therapy.

Later years the patient underwent an abdominal MRI that incidentally demonstrated multiple hepatic masses suspicious for metastases (Fig. 2). An ultrasound biopsy confirmed metastatic MSFT. He was treated with stereotactic body radiation therapy (SBRT) with initial response, however on follow-up imaging the masses were noted to have enlarged. He then underwent wedge resection to treat the larger lesions combined with radiofrequency ablation of the smaller ones.

For the recurrent brain lesion the patient was treated with stereotactic body radiosurgery. He was also initiated on daily Sunitinib, a protein kinase inhibitor, a treatment which is currently ongoing.

Figure 1: 1A) MRI of the orbit with contrast-enhancing lesion within the left retina (arrow). B) Fundal photograph. (C) Fundal angio gram.

Primary Orbital MSFT

Figure 2: Gadolinium-enhanced MRI of the abdomen demonstrating three discrete hepatic metastases (arrows).

Hepatic Metastases

Figure 3: A) Gadolinium-enhanced T1-weighted coronal image demonstrating the predominantly right-sided lesion centered in the right parietal bone with both intra- and extra-axial extension. B) Magnetic resonance venography demonstrating occlusion of the middle portion of the superior sagittal sinus with present distal flow. C) Post-operative MRI demonstrates extensive resection of the mass throughout the extra- and intra-cranial compartments, with sacrifice of the middle superior sagittal sinus.

Cranial Metastasis

Figure 4: A) Intraoperative photograph demonstrating the extra-axial extension of the tumor. The tumor easily dissociated from the overlying galea with blunt dissection. B) Intraoperative photograph after biparietal craniotomy.

Figure 5: A) Pathologic examination revealed dense sheets of epithelioid tumor cells arranged in a spindled growth pattern with high nucleus-to-cytoplasm ratio, abundant mitotic activity, and focal areas of necrosis, consistent with a high-grade neoplasm. B) Tumor cells demonstrated strong immunostaining for STAT6, a known oncogene frequently overexpressed in mesenchymal tumors.

References