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SNI: Unique Case Observations

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Case Report

Inflammatory myofibroblastic tumor masquerading as an anterior choroidal artery fusiform aneurysm

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ABSTRACT

Background: Inflammatory myofibroblastic tumor is a rare, poorly understood tumor that has been found to occur in almost every organ tissue. Its location within the central nervous system is uncommon, and patients tend to present with nonspecific symptoms.

Case Description: A female in her eighth decade presented to neurosurgery clinic with complaints of headache and dizziness. Initial imaging was consistent with a low-grade, benign brain lesion in the region of the left choroidal fissure. She was recommended for observation but returned 1 month later with progressive symptoms and doubling of the lesion size. She underwent surgical resection and was found to have an IMT arising from the wall of the left anterior choroidal artery.

Conclusion: Intracranial IMT remains a rare and poorly understood entity. The present case demonstrates a novel presentation of IMT in an adult patient and exemplifies the heterogeneity of the disease presentation.

Keywords: Aneurysm, Anterior choroidal, Inflammatory myofibroblastic tumor, Spindle cell tumor

INTRODUCTION

Inflammatory myofibroblastic tumors (IMTs) comprise a spectrum of enigmatic lesions that occur in almost all organ systems, but rarely involve the central nervous system (CNS).[4,5] It has been termed inflammatory pseudotumor, plasma cell granuloma, and myofibroblastoma. Histologically, these lesions are characterized by myofibroblastic spindle cells mixed with a polyclonal inflammatory infiltrate.[8] Intracranial inflammatory myeofibroblastic tumor is uncommon. Patients tend to present with nonspecific complaints, including headache, visual disturbance, and seizure. The radiologic appearance is nonspecific, and diagnosis rests on surgical pathology.[2,9-11,24] We present an atypical case of CNS-IMT that initially appeared to be an extra-axial tumor on imaging, but with gross appearance of a fusiform appearance at the time of microsurgical resection.

CASE REPORT

History

A female in her eighth decade presented to the neurosurgery clinic complaining of 1 week dizziness and right-sided weakness. Neurologic examination was normal. MRI brain demonstrated a contrast-enhancing lesion centered at the left choroidal fissure between the amygdala and cerebral peduncle [Figure 1a and b]. She was recommended for observation, but returned 1 month later with complaints of progressive headache, confusion, and gait instability. Repeat MRI brain demonstrated that the lesion had nearly doubled in size [Figure 1c and d]. Because of the location of the lesion in the crural cistern and the discrete, spherical appearance on MRI, there was concern that this could represent an aneurysm. CTA did not demonstrate any arterial filling of the lesion [Figure 2]; although the anterior choroidal artery was not seen on that study.

Operation

The patient was placed under general anesthesia in a Mayfield head holder with the head rotated at 45 degree angle to the right with a bump placed under the left shoulder. A left frontotemporal craniotomy was performed in standard fashion. Using frameless stereotactic guidance, we made a corticectomy in the anterior portion of the superior temporal gyrus. Brain tissue was removed along the inferior bank of the Sylvian fissure down to the region of the amygdala and the anterior portion of the temporal horn. We identified a discrete mass medial to the uncus that was firm and somewhat vascular. Initial biopsy demonstrated inflammatory tissue. After circumferential dissection, the mass was found to be clearly arising from the anterior choroidal artery and there was concern that the lesion represented a thrombosed fusiform aneurysm. Vascular neurosurgery was invited to participate in further surgical management of the lesion. Repeat biopsy demonstrated inflammatory and smooth muscle cells, suggesting sampling of the tunica media of a vascular lesion. Micro-Doppler was performed and demonstrated patency of the left posterior communicating artery and no flow within the left anterior choroidal artery. It was felt safe to sacrifice the anterior choroidal artery without causing harm. A straight Yasargil aneurysm clip was applied to the anterior choroidal artery proximal to the lesion. Circumferential microdissection of the lesion was carried out. The artery was then sectioned and the mass was sent for permanent pathology as a single specimen. Hemostasis was then obtained, and the wound was closed in usual sterile fashion.

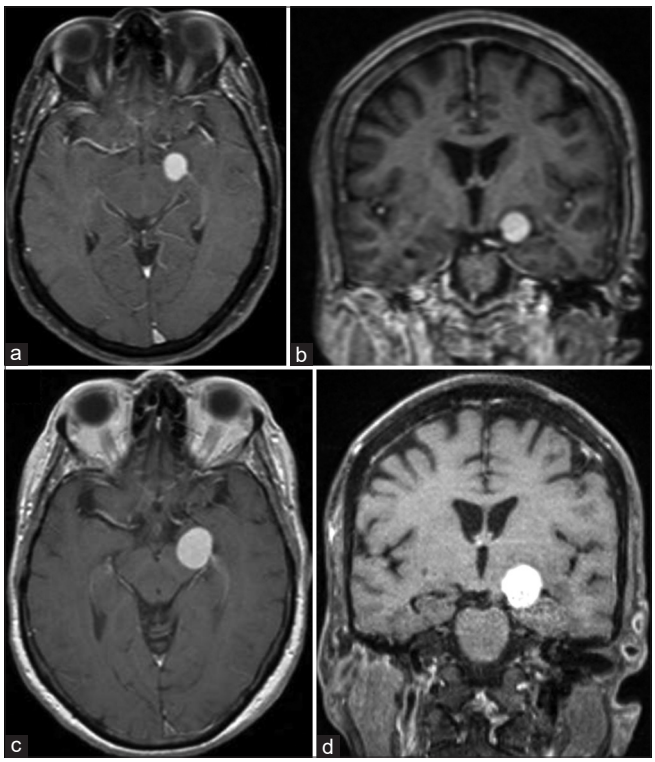

Figure 1: (a) Axial plane contrast-enhanced MRI demonstrating lesion at time of initial presentation. (b) Coronal plane contrast-enhanced MRI demonstrating lesion at time of initial presentation. (c) Axial plane contrast-enhanced MRI demonstrating interval growth 1 month after initial presentation. (d) Coronal plane contrast-enhanced MRI demonstrating interval growth 1 month after initial presentation.

Pathological findings

Gross examination of the tumor revealed a red-brown, ovoid portion of tissue with apparent encapsulation by a thin fibrous layer. Serial sections of the specimen revealed tan-white, partially hemorrhagic soft cut surfaces.

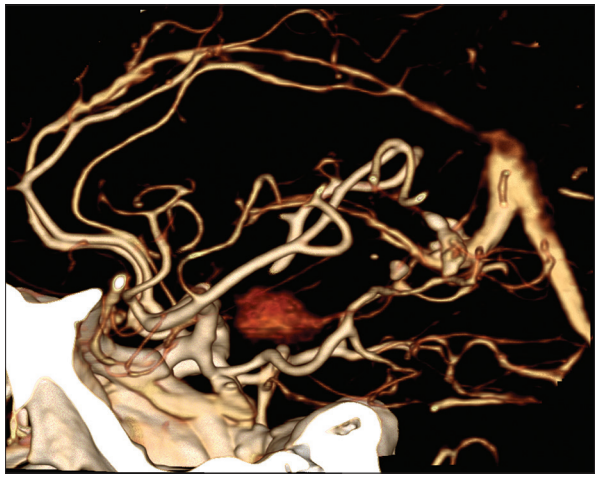

Figure 2: Three-dimensional reconstruction of CT angiography. Lateral projection demonstrates contrast uptake of hypervascular lesion with no discrete arterial connection. The anterior choroidal artery is not visualized.

Microscopic examination of the tumor with hematoxylin and eosin stained sections demonstrated a proliferation of spindle cells arranged in short fascicles, with a prominent background of acute and chronic inflammation [Figure 33]. Immunohistolochemical studies revealed tumor cells to be diffusely and strongly reactive for smooth muscle actin, vimentin, and focally positive for desmin. The background tumor stroma was highly vascular, as elucidated by CD34 and ERG immunoreactivity within vascular endothelial cells; however, tumor cells were negative for these studies. Tumor cells were negative for pancytokeratin, myogenin, and anaplastic lymphoma kinase (ALK)-1. Immunostudy with S100 highlighted background inflammatory infiltrates, but was nonreactive within neoplastic cells.

A representative formalin-fixed, paraffin-embedded tissue block of tumor was macrodissected for RNA extraction, which was directed toward sequence using the Archer® FusionPlex® Sarcoma kit from ArcherDx on the Illumina NextSeq instrument to high uniform depth. Sequence data were processed using the kit manufacturer's analysis pipeline (Archer Analysis v5.1.3) designed to accurately detect gene fusions. Per sequencing study, no pathological fusions were detected.

Postoperative course

Postoperatively, she developed right upper extremity weakness. MRI demonstrated a small infarct of the left posterior limb of the internal capsule consistent with an anterior choroidal artery infarct [Figure 4]. There was no residual tumor remaining. She was ultimately discharged to inpatient rehab and has made significant improvement. Postoperative MRI at 1 year follow-up has not demonstrated any evidence of recurrent disease.

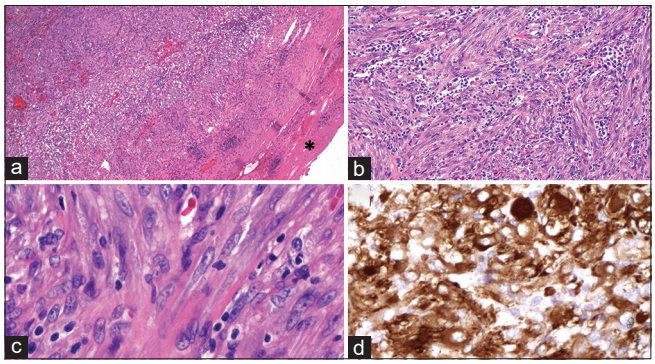

Figure 3: Inflammatory myofibroblastic tumor. Examination of the tumor on low magnification revealed a fairly well-demarcated cellular proliferation, demonstrating an incomplete fibrous pseudocapsule on the periphery (a, asterisk, ×5). The tumor was composed of spindled cells arranged in short fascicles with a background of prominent, multifocal acute and chronic inflammation, as well as frequent blood vessels (b, ×20). Tumor cells showed elongated, monotonous nuclei with small nucleoli (c, ×40) and were diffusely and strongly reactive for smooth muscle actin immunohistochemistry (d, ×40). (a-c) Hematoxylin and eosin. (d) Smooth muscle actin immunohistochemistry.

DISCUSSION

IMTs constitute a rare group of heterogeneous lesions characterized histologically by myofibroblastic spindle cells mixed with a polyclonal infiltrate of plasma cells, lymphocytes, and eosinophils.[5,8] Historically, the variability and heterogeneity of the immune cell infiltrate have made these tumors difficult to classify. The terms plasma cell granuloma, inflammatory myofibrohistiocytic proliferation, and inflammatory pseudotumor have previously been used interchangeably. The present term, IMT, was designated by the World Health Organization in 2002 – formally classifying these lesions as soft-tissue tumors. IMTs most often occur in the lungs, mesentery, and omentum.[8] IMTs are usually benign and have a predilection for children and young adults.[3]

IMT involvement of the CNS (IMT-CNS) is a rare occurrence, with a total number of sporadic IMT-CNS reported of less than 100.[14,22,23] When confined to the intracranial vault, it is classified as intraparenchymatous, meningeal, mixed intraparenchymatous, and meningeal, and extending to the cranial cavity/sphenoid sinus.[11] Meningeal lesions represent the most common subtype, frequently presenting with headache or seizure.[12] Radiologically, IMT may be confused for meningioma, lymphoma, or plasmacytoma. Diagnosis is dependent on histopathologic analysis. Emerging evidence suggests a neoplastic nature, with recurrence rates between 12.5% and 40%.[10] Expression of ALK confers a more aggressive phenotype, with 33% of IMT positive tumors recurring following gross total resection versus 9%

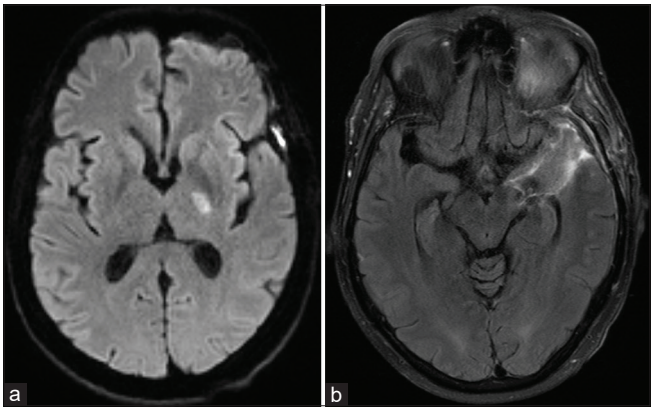

Figure 4: Postoperative MRI brain. (a) Diffusion-weighted imaging demonstrates a small area of restricted diffusion in the left internal capsule. (b) Fluid-attenuated inversion recovery sequence demonstrates postoperative changes of the transgyral approach to the tumor.

in IMT negative tumors.[6,7] Irrespective of ALK expression, gross total resection of IMT-CNS results in improved tumor control with decreased rates of recurrence.[7,19] Crizotinib, an oral chemotherapeutic tyrosine kinase inhibitor, was initially developed for the treatment of ALK positive nonsmall cell lung carcinoma.[20] There have been few reports of its use in treating extra CNS IMT with good response,[1,16] suggesting a possible form of salvage therapy for ALK positive recurrent IMT-CNS.

The present case is an illustrative example of the difficulty in diagnosing and treating IMT-CNS. Although imaging characteristics favored meningioma, rapid growth over short interval was inconsistent with the benign appearance of the WHO Grade I meningioma. Accelerated growth has been reported of pulmonary IMT in the setting of corticosteroid therapy. The precise mechanism is unclear, but has been postulated to be related to cellular proliferation due to immunosuppression.[18] The effect of corticosteroids on IMT remains unclear. Other authors have reported corticosteroid therapy to result in tumor control and a reduction in tumor volume following administration.[2,15] Regardless, the patient had minimal peritumoral edema and was not treated with corticosteroids during the growth period. Moreover, the present case does not fit the current CNS-IMT classification scheme, as it was not associated with dura nor located within parenchyma. The tumor was encountered within the ambient cistern, intimately associated with the anterior choroidal artery. There have been rare reports of intracranial arterial involvement, predominantly occurring in infants and children.[5,13,17,21,25] To the best of the authors' knowledge, this is the first reported case of IMT-CNS associated with an intracranial artery in an adult.

CONCLUSION

Intracranial inflammatory myeofibroblastic tumor remains a rare and poorly understood entity. Radiographic diagnosis is difficult, as the IMT appearance mimics more common CNS pathology. The present case demonstrates a novel presentation of IMT in an adult patient and exemplifies the heterogeneity of the disease presentation.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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