

Cavernous Sinus Metastasis of Leiomyosarcoma with Orbital Extension along the Third Nerve, Mimicking Cavernous Sinus Meningioma

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2. Keywords

Leiomyosarcoma; Metastasis;
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1. Abstract

Background: Leiomyosarcoma (LMS) metastasis in the central nervous system is extremely rare. Metastatic LMSs have been described in the orbit, meninges, and skull base, however there are no reports of LMS metastasis into the cavernous sinus with primary origin from lower extremity and long silent disease period of 7 years.

Case presentation: We present a case of a 75-year-old woman with complaints of diplopia for three months. An MRI scan revealed a contrast-enhancing lesion in the cavernous sinus, extending along the third nerve through the left superior orbital fissure. In 2011, the patient was diagnosed with and treated for a leiomyosarcoma of the left lower limb. The lesion mimicked a meningioma in the MRI scan; intraoperatively, it appeared as a schwannoma of the third nerve. The tumor was biopsied and partially resected, and the third nerve was decompressed. Radio surgical treatment was recommended for the remaining tumor. We use an instructional intraoperative video to demonstrate the radiological and intraoperative aspects of LMS metastasis.

Conclusion: LMS can metastasize to the cavernous sinus, leading to varying degrees of ophthalmoplegia. Radio logically, metastatic LMS mimics a meningioma; intraoperatively, it mimics a schwannoma.

3. Introduction

The metastasis of primary lung, breast and skin cancers is most commonly detected in the brain. Other peripheral tumors, including those from the gastrointestinal and urogenital tract, rarely metastasize in the brain. Similarly, leiomyosarcoma (LMS) metastasis in the central nervous system is extremely rare [1-8]. Although metastatic LMSs have been described in the orbit, meninges and skull base [1-14], they have not been reported from the lower extremity in the cavernous sinus. There are several reports of LMSs in the cavernous sinus, however they were not distant metastases – the LMS originated from vascular smooth muscles in the cavernous sinus [8,10,13,15,16]. Here, we report the first case of cavernous sinus LMS metastasis extending through the superior orbital fissure along the third nerve, appearing radiologically as a cavernous sinus meningioma and intraoperatively

as a schwannoma of the third nerve.

4. Case Report

A 75-year-old woman presented with third nerve palsy, experiencing diplopia for three months. Upon examination, the patient was alert and oriented. Visual field was intact, and pupil size was normal and reactive to light. The patient's hearing and facial nerve function were normal. In 2011, the patient received surgical treatment of the leiomyosarcoma on her left lower leg. An MRI scan revealed a contrast-enhancing space-occupying lesion in the left wall of the cavernous sinus, extending through the superior orbital fissure along the third nerve (Figure 1 A-E). No other pathology in the supra or infra-tentorial regions was observed.

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5. Results

Surgical tumor removal and operative findings

Surgery was performed while the patient was in prone position. A lateral supraorbital approach was used, with an OPMI PENTE-RO 900 microscope (Carl-Zeiz, Germany). The optic cistern was opened to release CSF. The tumor was tightly attached to the third nerve and extended through the superior orbital fissure, mimicking a schwannoma of the third nerve (Figure 2 A, B, C and video 1). A biopsy was taken and the tumor was partially removed using standard microsurgical techniques. The third nerve was decompressed (Figure 2D and video 1). The tumor extending to the cavernous sinus was not removed, but treated with stereotactic radiosurgery.

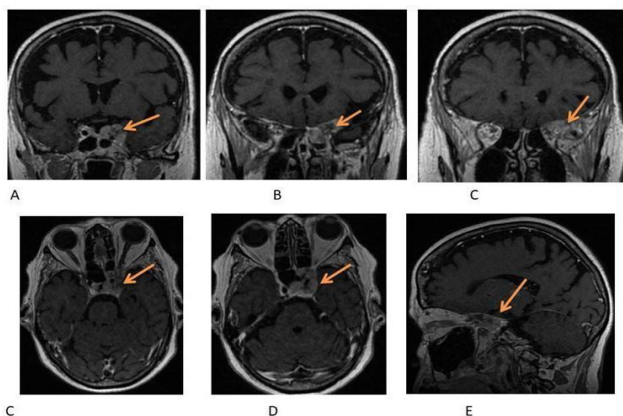


Figure 1: MRI scans showing tumor location and radiological anatomy.

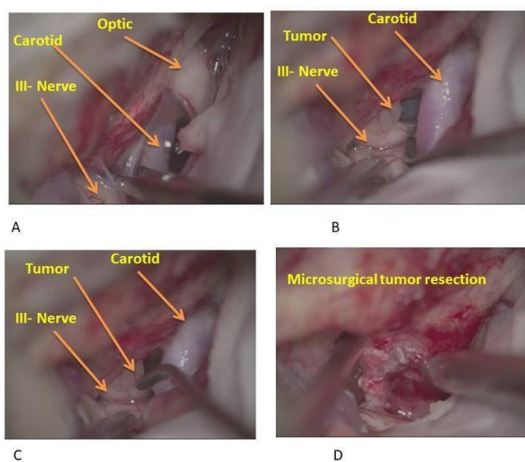


Figure 2: Intraoperative images showing intraoperative surgical anatomy and the steps of surgery, including tumor removal.

Video 1: Demonstration of partial tumor resection, biopsy and third nerve decompression.

Post-operative outcome

The post-operative examination results did not indicate any changes in the patient. No new neurological deficits were observed. A four-week follow-up examination revealed that the patient still experienced ophthalmoplegia of the left eye.

6. Discussion

To the best of our knowledge, this is the first case of LMS originating from the left lower limb that has metastasized to the cavernous sinus after such a long silent period of seven years of initial diagnosis. The LMS extended through the superior orbital fissure along the third nerve, causing unilateral ophthalmoplegia. The metastasis mimicked meningioma on the radiological images, as it showed a homogeneous contrast enhancement and dural enhancement (Figure 1). It is difficult to differentiate an intracranial LMS – either primary or secondary metastasis – from a meningioma with radiological tools[6,10]. Thus, LMS diagnosis is confirmed with histological tools such as immunostaining[11]. We performed surgery with the goal of obtaining a biopsy sample and decompressing the third nerve. Intraoperatively, we found that the lesion was tightly attached to the third nerve, which is where it appeared to originate from. The tumor mimicked a schwannoma of the oculomotor nerve (video 1). The biopsy results confirmed this was metastatic LMS, opposed to meningioma or schwannoma. There is no standard evidence-based treatment of metastatic LMS. Complete resection is the recommended/preferred treatment in brain metastases, but extensive resection of tumors in the cavernous sinus and orbital apex is highly risky as it can increase morbidity. Hence, considering the age and other comorbidities of the patient, we opted to biopsy the tumor and decompress the third nerve along the superior orbital fissure. Radiosurgery was planned for the remaining tumor, as this treatment has been shown to be the best option for partially resected tumors. The prognosis of both primary and secondary LMS is poor. The median survival of patients with metastases in the cavernous sinus is reportedly 2-12 months[4]. However, data on the prognosis of LMS metastasis in the cavernous sinus is lacking due to the rarity of this condition.

7. Conclusion

LMS can metastasize to the cavernous sinus, leading to ophthalmoplegia of varying degrees. Metastatic LMS appears radiologically as a meningioma and intraoperatively as a schwannoma.

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