Primary extradural leiomyosarcoma involving cavernous sinus in an immunocompetent patient

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ABSTRACT

Intracranial leiomyosarcoma (LMS) are uncommon malignancies and usually encountered after systemic metastases. Limited cases of primary intracranial LMSs have been reported in the literature. It mostly affects immunocompromised individuals in association with Epstein–Barr virus infection. This is the unusual first case being reported of primary LMS in immunocompetent patient with involvement of cavernous sinus.

Key words: Cavernous sinus, immunocompetent, leiomyosarcoma

INTRODUCTION

Leiomyosarcoma (LMS) is a relatively rare soft tissue sarcoma that arises from smooth muscle cells. These tumors are commonly encountered in uterus, stomach, small intestine and retro peritoneum. However, rarely these tumors arise in brain from pluripotent mesenchymal stem cells of the dura mater or cerebral blood vessel epithelium.[1] Dura based lesions may clinically and radiologically mimic meningioma's. We hereby describe a very rare case of primary LMS in immunocompetent patient with involvement of cavernous sinus.

CASE REPORT

A 55-year-old female presented with severe neuralgic pain in the right half of the face in ophthalmic distribution of trigeminal nerve of 3 months duration and right sixth cranial nerve palsy of 15 days duration. There were no features of raised intracranial pressure. There was no history of smoking, intravenous drug abuse or sexual promiscuity. On examination, she had sensory loss in right V1 and V2 distribution, mild wasting of the temporalis and masseter muscles along with right sixth cranial nerve palsy. Routine laboratory studies were within normal limits. Serology was negative for HIV. Her magnetic resonance imaging brain [Figures 1 and 2] revealed an isointense lesion on T2-weighted images with brilliant enhancement on contrast involving right middle cranial fossa base with cavernous sinus involvement and destruction of petrous bone. A possibility of skull base meningioma was considered.

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Operative details

Patient underwent right temporal craniotomy, zygomatic osteotomy, interdural approach and tumor decompression under intra-operative neuronavigation. Intra-operatively, tumor was soft to firm, highly vascular, and extra-axial. The portion extending into the cavernous sinus was left behind due to significant bleeding. Resected tumor was sent for histopathology examination.

Pathology findings

Histological examination revealed a tumor with pushing borders that was adhered to the nonneoplastic nerve bundle. Tumor was composed of spindle shaped cells that were arranged in interweaving fascicles. The tumor cells had cigar shaped nuclei with pointed ends and indistinct cytoplasmic borders [Figure 3a and b]. There was brisk mitotic activity with several bizarre and giant nuclei. The stroma of tumor was richly vascular with areas of hyalinization. On immunohistochemistry (IHC), the tumor cells were positive for desmin, smooth muscle actin and vimentin [Figure 4a]. The tumor cells were negative for S-100, epithelial membrane antigen (EMA), glial fibrillary acidic protein and CD117. Ki-67 staining revealed a high proliferation index of 25-30% [Figure 4b].

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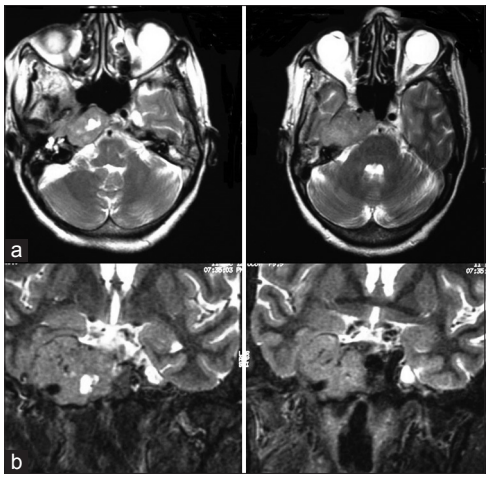

Figure 1: T2-weighted image (a) Axial (b) Coronal shows isointense lesion involving petrous apex, cavernous sinus with encasement of internal carotid artery and extension into middle cranial fossa

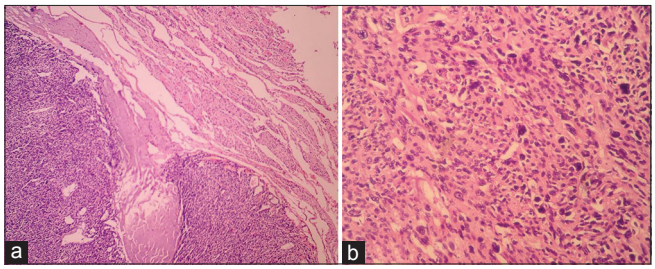

Figure 3: (a) Photomicrograph showing pushing tumor margins with adherent nerve bundles. (b) High power image showing spindled tumor cells arranged in fascicles. There is striking nuclear pleomorphism with tumor giant cells

Based on the histological findings and IHC, diagnosis of LMS involving the cavernous sinus was made.

Postoperatively, patient recovered with no new neurological deficits. Her postoperative computed tomography (CT) scan showed residual tumor in the right cavernous sinus [Figure 5]. Staging CT scan of the chest, abdomen and pelvis and positron emission tomography scan didn't reveal any other site of involvement by tumor. The patient tested serologically negative for HIV, herpes simplex virus and Epstein–Barr virus (EBV) antibodies. The patient was referred to cancer hospital for adjuvant therapy.

DISCUSSION

Intracranial soft tissue sarcomas are rare tumors that account for only 0.1-0.2% of all central nervous system (CNS) tumors.[2] Most intracranial soft tissue sarcomas represent metastases and primary sarcomas arising in brain are quite rare. Among the sarcomas, children are preferentially affected with rhabdomyosarcoma and adult's usually have malignant fibrous histiocytoma and chondrosarcoma.[3] LMS is a rare intracranial malignant

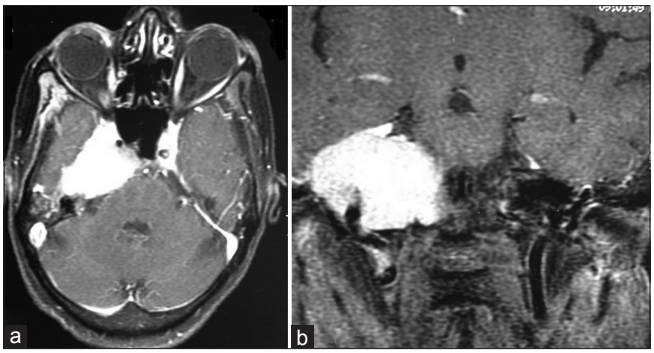

Figure 2: T1-weighted image with contrast (a) Axial (b) Coronal showing brilliantly enhancement of the lesion

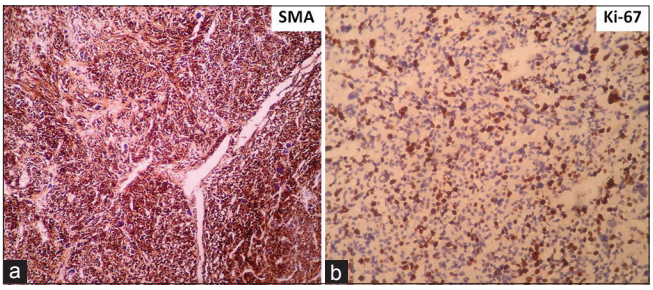

Figure 4: (a) Immunohistochemistry (IHC) stain for SMA demonstrating strong cytoplasmic staining in tumor cells. (b) IHC stain for Ki-67 showing high proliferative index

tumor that mostly arises in the dura and parasellar region. Age of presentation in previous case reports ranged from 2 to 73 years.[4,5] There is usually no sex predilection and the median duration of symptoms before presentation is nearly 4 months.[6]

Intraparenchymal location of the tumor is mostly observed in systemic metastases and primary examples are rare. Systemic LMSs have a predilection for hematogenous spread to CNS and most frequent primary sites include uterus and gastrointestinal tract followed by retroperitoneum, lung and heart.

An interesting association has been observed for occurrence of primary dural CNS LMSs in immunocompromised patients with latent EBV infection.[7] The latter is commonly associated with HIV and organ transplantation. Previous exposure to radiation has also been associated with LMS.

The present case is the first to be reported of primary intracranial LMS in immunocompetent patient involving cavernous sinus. Literature search revealed only 10 cases of primary intracranial LMS in immunocompetent adults [Table 1].

Intracranial LMS need to be distinguished from malignant meningioma, myofibrosarcoma and fibrosarcoma.

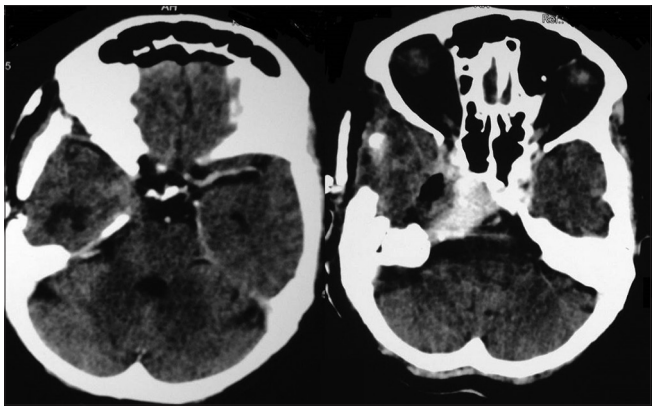

Figure 5: Postoperative computed tomography scan (axial images) with contrast: Adequate decompression of middle cranial fossa portion with residual tumor in cavernous sinus and petrous apex

Table 1: Reports of primary intracranial leiomyosarcoma in immunocompetent adults

Report of patient case	Publication year	Patient age	Sex	Location	Dural involvement
Anderson *et al.*[8]	1980	35	Male	Sella	No
Li[9]	1987	47	Male	Pineal gland	No
Asai *et al.*[5]	1988	73	Male	Right temporal	Yes
Louis *et al.*[10]	1989	72	Female	Left lateral ventricle and choroid plexus	No
Skullerud *et al.*[11]	1985	33	Male	Pineal area	No
Oliveira *et al.*[12]	2002	58	Female	Left temporal	No
Hussain *et al.*[1]	2006	28	Male	Right parieto-occipital	Yes
Aeddula *et al.*[6]	2011	58	Male	Left temporal	No
Almubaslat *et al.*[13]	2011	47	Female	Left frontoparietal	No
Alijani *et al.*[14]	2013	19	Male	Right parieto-occipital	Yes

Diagnosis of LMS is confirmed by immunohistochemical staining. The present patient showed positive staining for smooth muscle actin and desmin and was negative for S-100 and EMA. Malignant meningioma can sometimes show sarcomatous differentiation mimicking LMS. Meningeal cells are usually positive for epithelial markers - EMA and cytokeratin. Fibrosarcoma is tumor of fibroblasts that is positive for vimentin and Type I collagen and on reticulin staining there is characteristic positivity in fibers surrounding each cell.

Surgery, radiotherapy, and chemotherapy are the current choices of treatment for primary intracranial LMS. However, the prognosis of this tumor remains poor with the longest survival reported in literature being 32 months.[6] Patient survival is limited due to several reasons. Gross resection of tumor with adequate surgical margins isn't achievable in most cases and the behavior of tumor is usually aggressive with limited response to chemotherapeutic drugs. Our patient was referred to cancer hospital for adjuvant radiotherapy and chemotherapy. However, she died within 5 months due to systemic complications.

CONCLUSION

Our patient is a rare case of primary extradural LMS involving cavernous sinus in an immunocompetent patient. Although intracranial LMSs are more common in immunodeficient individuals, increasing cases are being encountered in immune competent patients as well. Research studies are needed to identify the etiologic factors responsible for intracranial LMSs in immunocompetent individuals.

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