Case Report

Primary intracranial leiomyosarcoma presenting with massive peritumoral edema and mass effect: Case report and literature review

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Abstract

Background: Primary intracranial leiomyosarcomas (LMSs) are unusual tumors of the central nervous system (CNS) affecting all age groups, and are recently, becoming more prevalent in immunosuppressive conditions such as in patients with human immunodeficiency virus (HIV) infection. However, only a few CNS LMS case reports exist in the English literature, on the occurrence of this rare entity in immunocompetent adults. Even, rarer is a purely intraparenchymal occurrence without any dural attachment in afflicted individuals. To the best of our knowledge, only four such cases have been reported in the literature until now. None of these cases were associated with marked peritumoral brain edema (PTBE) and mass effect as seen in our case and falsely suggesting an underlying glioma.

Case Description: A 45-year-old male patient, presented with headache, right-sided weakness and difficulties with speech over 4 months along with a single generalized tonic clonic seizure. Physical examination revealed mild to moderate papilledema, motor aphasia, and right-sided hemiparesis. Radiographic evaluation showed a large left temporo-parietal mass extending into the basal ganglia with intense heterogeneous contrast enhancement. There was marked perilesional edema and mass effect with midline shift. The patient underwent a left temporo-parietal craniotomy for subtotal resection of the tumor. The post-operative period was uneventful. Histopathology revealed a spindle cell tumor, which stained immunopositive for smooth muscle actin, vimentin, and S-100, yielding the diagnosis of LMS.

Conclusion: Primary intracranial LMS can rarely occur in immuno-competent adult patients and should be considered in the differential diagnosis of intraparenchymal lesions presenting with significant PTBE.

Key Words: Adult, immunocompetent, intracranial, leiomyosarcoma, primary
INTRODUCTION

Primary intracranial leiomyosarcomas (LMS) is a malignant neoplasm of smooth muscle cell origin and can affect any organ of the body due to the ubiquitous presence of smooth muscle cells. However, involvement of CNS is seen in few cases only. Most reported cases of intracranial LMS represent metastases from other primary sites such as the gut, uterus, retro-peritoneum, or from subcutaneous tissue. Primary intracranial LMS is known to occasionally occur in immunocompromised patients whereas occurrence in adult immunocompetent patients is exceptionally rare.

A thorough literature review revealed only 15 cases of primary intracranial LMSs in immuno-competent adult patients [Flowchart 1]. Most of the cases described thus far showed dural-based lesions, mimicking meningiomas. Purely intraparenchymal involvement has very rarely been seen. Herein, we report an additional case of primary intracranial LMS in an adult immunocompetent male, who presented with massive peritumoral brain edema and mass effect and we will discuss the observations made in the context of the current pertinent literature.

CASE REPORT

A 45-year-old man presented with a history of progressively increasing headaches, right-sided weakness, difficulties with speech for 4 months as well as a single generalized tonic clonic seizure. Clinical examination identified some papilledema, expressive aphasia, and right-sided hemiparesis (MRC grade 4/5). There was no history of intravenous drug use or radiation exposure.

Routine laboratory studies returned as within normal limits and serology was negative for HIV, Hepatitis - B Virus (HBV), Hepatitis-C Virus (HCV), and Epstein Barr virus (EBV). No other comorbidities were noted. Head CT (native + contrast enhanced) demonstrated a heterogeneously enhancing lesion located in the left tempo-parietal areas with extension to the basal ganglia. Extensive perilesional edema and mass effect with midline shift towards the contralateral side was visible [Figure 1].

Brain MRI [Figure 2] again demonstrated a large heterogeneous mass with both solid and cystic components $5.9 \times 4.4 \times 5.2$ cm in size. The lesion imposed hypo-intense on T1W images and was heterogeneously hyper-intense on T2W sequences. After IV gadolinium administration, heterogeneous contrast enhancement of the solid part was seen. There was massive perilesional edema resulting in profound mass effect. Pre-surgical chest X-ray and ultrasound abdomen were normal. Differential diagnosis favored high grade glioma or atypical meningioma.

Patient underwent a left tempo-parietal craniotomy and subtotal excision of the tumor, under image guidance. Intraoperatively, solid and cystic components (containing xanthochromic fluid) were encountered. The solid tumor component was greyish-white in appearance, showed firm consistency that required excision rather than removal by suction-irrigation. This part of the lesion was highly vascular leading to massive blood loss (approximately 2–2.5 liters). Since the tumor displayed poorly defined dissection planes only a subtotal excision could be achieved. Intraoperatively, he received 3 units of blood transfusion. The immediate post-operative period was uneventful and there was no deterioration in her neurological status. Post-operative CT scan showed some residual tumor without tumor cavity hematoma or bleed. Repeat CT scan 3 days after surgery showed resolving mid-line shift and edema. He received steroids in tapering doses for 7 days and was discharged home 10 days after surgery.

Pathology

Microscopic examination [Figure 3] of the resected tissue specimen revealed a spindle cell tumor with marked nuclear atypia and very high mitotic index (17–20/10 HPF). No glial tissue was identified on Glial Fibrillary Acidic Protein (GFAP) staining and areas of hemorrhage and necrosis were evident on histological examination. Immunohistochemical staining was positive for smooth muscle actin (SMA), S-100, and vimentin and negative for desmin, pan-cytokeratin, MIC-2, CD34, glial fibrillary acidic protein, progesterone receptor, and epithelial membrane antigen. Ki-67 positivity was 35–40%. Pathological diagnosis was made of a malignant, mesenchymal, non-meningothelial tumor favoring...
Figure 1: Computed tomography (CT) images showing tumor in left frontotemporal and basal ganglia region with mass effect and midline shift with perilesional edema (a–d) noncontrast images; (e–h) contrast images.

Figure 2: Magnetic resonance imaging (MRI) of tumor tissue showing a solid (predominant) and cystic lesion in the left frontotemporal and basal ganglia region with mass effect and midline shift with perilesional edema (a–d) T1W axial images; (e–h) T2W axial images; (i–l) T1 with gadolinium contrast axial images; (m, n) coronal contrast images; and (o, p) sagittal contrast images.
leiomyosarcoma. Given the diagnosis, a CT scan of the chest, abdomen, and pelvis were done, to evaluate for other potential sites of involvement, but results were negative for systemic disease. His Karnofsky Performance Scale Index was 80. Post-operatively, the patient received adjuvant radiotherapy (60 Gy in 30 fractions). The patient had a final neurological exam that showed improved motor weakness (MRC grade +4/5), but was lost in follow up one year after the surgery.

DISCUSSION

Primary intracranial LMS is an infrequent tumor of CNS arising from the mesenchymal cells of the cerebral blood vessels or duramater. Most reported cases were of dural-based lesions, mimicking meningiomas. Purely intraparenchymal involvement has only been seen in 4 patients identified by our literature review. None of the reported patients presented with marked PTBE and mass effect as seen in our case.

Zhang et al. reported immuno-suppression and radiation induced tumorigenesis as key causative factors for the development of primary intracranial LMSs in their review article summarizing findings from 37 cases (10 children and 27 adults). A major cause of immuno-suppression identified by the authors was HIV infection (15 cases), other etiologies included Common Variable Immunodeficiency Syndrome (CVID), preceding lung transplantation and chemotherapy for brain tumor. Furthermore, it was hypothesized that EBV associated with immnosuppression could be related to the incidence of primary intracranial LMS. However, the mechanism by which EBV-transformed, infected smooth muscle cells contribute to the development of LMS remains largely unknown. Similarly, radiotherapy-induced leiomyosarcomatous transformation of vasculature associated cells may also lead to the formation of LMSs.

After a comprehensive literature review [Flowchart 1], we found only 15 cases of primary intracranial LMS which occurred in immuno-competent adults. Out of 15 patients described, 3 patients had developed LMS after prior surgery (without any adjuvant chemotherapy or radiotherapy). Out of these three, one developed LMS nine years after surgery for a neurofibroma located in the CPA (here the authors hypothesized that heat/mechanical stimulation of the previous surgery may be an etiological factor for tumor development). Another case of intra-ventricular LMS, reported by Louis et al., also underwent surgery for basal cell carcinoma and intra-ductal breast carcinoma before the development of LMS; however, the role of preceding surgery in the pathogenesis of a later occurring LMS remains unclear. None of the above-mentioned etiological factors were present in our case.

No specific symptoms have been identified that are associated with primary intracranial LMS, and symptoms vary largely based on tumor location.

It has been hypothesized that intraparenchymal LMSs originate from smooth muscle cells lining the intracranial blood vessels. Interestingly, Mathieson et al. reported a case of frontal intraparenchymal LMS that seemed associated with a synchronous chronic subdural hematoma in a 5-year-old immune-competent child, which suggested chronic inflammation of the smooth muscle cells in hematoma membrane, responsible for genesis of LMS.

Although the characteristic histopathological features of this rare entity have been highlighted and reviewed by other authors, there is a paucity of radiological data and description thereof in the literature. Table 2 describes the radiographic details of cases of primary intracranial LMSs retrieved in our search in immuno-competent adults.

Smith et al. in their investigation on dural-based mass lesions other than meningiomas, described intracranial LMSs to be typically hypo- to iso-intense on T1-weighted imaging and iso- to hyper-intense on T2-weighted imaging. Avid enhancement after administration of intravenous contrast is typically seen and they share numerous imaging characteristics with meningiomas. Although radiological detail is lacking in literature, they appeared as hyper-attenuating lesions on plain CT, which will show homogeneous contrast enhancement. They also pointed out that these lesions may contain calcifications on CT.

Our purely intraparenchymal case and with solid and cystic components had massive perilesional edema and...
Table 1: Clinical data of primary intracranial leiomyosarcomas in adult immunocompetent patients as described in the literature

| Author, year (ref. no)        | Age/sex | Presentation                                                                 | Tumor size | Comorbid conditions | Location and dural involvement | Treatment                                                                        | Follow-up                |
|------------------------------|---------|------------------------------------------------------------------------------|------------|---------------------|---------------------------------|---------------------------------------------------------------------------------|--------------------------|
| Anderson et al., 1980[4]     | 35/M    | Visual symptoms and headache                                                 | NA         | None                | Sella and suprasellar region    | Partial resection + Radiotherapy                                                | Alive after 2 years and 8 months |
| Asai et al., 1988[5]         | 73/M    | Rapid growth of subcutaneous mass in right temporal region                   | 3 cm diameter subcutaneous mass | None                | Right temporal region           | Complete resection + Radiotherapy                                               | NA                       |
| Louis et al., 1989[17]       | 72/F    | Altered mental status, memory deficits, headache                             | 5 × 4 × 4 cm | Suffered angina     | Left Lateral ventricle          | Complete resection + Radiotherapy                                               | No evidence of disease at 6 months |
| Skullerud et al., 1995[26]   | 33/M    | Diplopia, headache, unsteadiness of gait, Parinaud’s syndrome               | 3.5 cm in diameter | None                | Pineal region                   | Complete resection + Radiotherapy                                               | No evidence of disease at 2 years |
| Oliveria et al., 2002[20]    | 58/F    | NA                                                                           | NA         | None                | Left temporal region            | Complete resection + Radiotherapy                                               | Alive with no evidence of disease at 2.5 years |
| Hussain et al., 2006[11]     | 26/M    | Progressively increasing subcutaneous parieto-occipital mass                 | 5 cm diameter subcutaneous mass | Multiple lung lesions and a liver lesion | Parieto-occipital region Bone invasion present | Complete resection + Radiotherapy + Chemo-therapy                              | Dead of disease after 7 months |
| Fujimuto et al., 2011[18]    | 45/M    | Facial palsy, loss of hearing and partial abducens nerve palsy (Previously operated CPA neurofibroma) | NA         | None                | CPA Dural involvement present   | Partial Resection + Radiotherapy                                                | Dead of disease after 10 months |
| Aeddula et al., 2011[1]      | 58/M    | Headache, seizures                                                          | 1.6 × 1.3 cm | Chronic smoker + CAD + HT | Left antero-medial temporal lobe. No dural involvement | Complete Resection                                                             | Died after 3 weeks of surgery |
| Almubaslat et al., 2011[3]   | 47/F    | Weakness and speech difficulty                                               | 6.3 × 5.0 × 3.0 cm | HT                  | Left Fronto-parietal lobe Dural involvement present | Complete resection                                                            | Alive at 21 months after diagnosis |
| Zhang et al., 2012[20]       | 26/F    | Headache, diplopia (6th nerve palsy)                                         | NA         | None                | Body of corpus callosum          | Partial Resection + Gamma knife radiosurgery                                    | Died after 3 months |
| Alijani et al., 2012[21]     | 19/M    | Progressively increasing subcutaneous parieto-occipital bump                 | 5 cm diameter subcutaneous mass | None                | Parieto-occipital region Bone invasion present | Complete resection + Radiotherapy                                               | Alive with no evidence of disease at 1.5 years |
| Saito et al., 2014[23]       | 75/F    | Acute onset of left eye pain and ptosis of left eyelid, 6th nerve palsy       | NA         | Cerebral infarction and occlusion of left ICA | Sella and suprasellar region    | Biopsy + Radiosurgery (Stereotactic radiation with cyber knife)                | Local recurrence + Died after 3 years and 8 months |

Contd...
Table 1: Contd...

| Author, year (ref. no) | Age/sex | Presentation | Tumor size | Comorbid conditions | Location and dural involvement | Treatment | Follow-up |
|------------------------|---------|--------------|------------|---------------------|-------------------------------|-----------|-----------|
| Gulwani et al., 2014[11] | 55/F    | Trigeminal neuralgia with right 6th nerve palsy | NA         | None                | Right middle cranial fossa base + cavernous sinus involvement | Partial resection + Radiotherapy | Died after 5 months |
| Maslehaty et al., 2016[18] | 43/M    | Progressively increasing palpable swelling at occipital region (previously operated grade 1 right cerebellar astrocytoma) | NA         | None                | Right occipital region Dural involvement present Bone invasion present | Complete resection + Radiotherapy | Alive with no evidence of disease at 6 months |
| Polewski et al., 2016[20] | 41/F    | Headache, expressive aphasia, and Homonymous hemianopsia | 6 × 5 × 4.6 cm | None                | Extra-axial mass arising from left tentorium | Complete resection + Radiotherapy + chemotherapy | Alive 31 months after initial diagnosis |
| Present case, 2016 | 45/M    | Headache, contralateral hemiparesis and seizures | 5.9 × 4.4 × 5.2 cm | None                | Left Temporo-parietal and basal ganglia region | Partial resection + Radiotherapy | Alive after 1 year |

NA: Not available, M: Male, F: Female, CT: Computed tomography, CPA: Cerebellopontine angle, ICA: Internal carotid artery, CAD: Coronary artery disease, HT: Hypertension

Table 2: Radiological data of primary intracranial leiomyosarcomas in adult immunocompetent patients as described in the literature

| Author, year (ref. no) | Location | HCP | Calcification | Cysts | CT findings Plain | CT findings Plain | MRI findings | MRI findings | MRI findings | MRI findings | Remarks |
|------------------------|----------|-----|---------------|-------|-------------------|-------------------|--------------|--------------|--------------|--------------|---------|
| Anderson et al., 1980[4] | Sellar and suprasellar region | None | None | None | None | NA | NA | NA | NA | Large intrasellar tumor extending into suprasellar region and sphenoid sinus X-ray lateral view skull-erosion of dorsum sella + enlargement of sella turcica. |
| Asai et al., 1988[5] | Right temporal region | None | + + | None | ↑ | + + HM | NA | NA | NA | Subcutaneous calcified mass in the right temporal region |
| Louis et al., 1989[17] | Left lateral ventricle | None | None | None | ↑ | NA | NA | NA | NA | Homogeneous high-attenuation mass, filling and distorting Left lateral ventricle |
| Skullerud et al., 1995[26] | Pineal region | None | NA | NA | NA | + + HM | NA | NA | NA | Intense contrast-enhancing tumor filling the posterior part of third ventricle |
| Oliveria et al., 2002[20] | Left temporal region | NA | NA | NA | NA | NA | NA | NA | NA | Large highly vascularized soft tissue mass involving the meninges + invasion of parietal |
| Hussain et al., 2006[12] | Parieto-occipital region | NA | NA | NA | - | - | NA | NA | NA | |

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showed midline shift, both of which had never been reported in literature. The exact cause of peripherally located tumor cysts is not entirely known, although this may be due to degenerative changes, tumor cells secreting cystic fluid, or the formation of loculated fluid collections from scar tissue within or adjacent to the tumor.

The profound peritumoral edema seen in our case may be due to the large tumor size, creating brain compression causing ischemia and secondary brain edema or could be due to enhanced vascularity, cellularity and mitotic activity of the tumor, since highly vascular tumors tend to have significant perilesional edema.

The diagnosis of LMS is based on demonstration of smooth muscle cells and their characteristic ultra-structural features on microscopy. Histologic evaluation of leiomyosarcomas usually depicts elongated spindle-shaped cells with abundant mitotic activity and arranged in intersecting fascicles.

| Author, year (ref. no) | Location | HCP | Calcification | Cysts | CT findings Plain | CT findings Contrast | MRI findings T1W | MRI findings T2W | MRI findings T1W C+ | Remarks |
|------------------------|----------|-----|---------------|-------|-------------------|---------------------|-----------------|-----------------|------------------|---------|
| Fujimoto et al., 2011[18] | Cerebellopontine angle | None | NA | NA | NA | NA | NA | + + HE | bone and displacement of brain parenchyma Tentative diagnosis - Osteoblastic meningioma |
| Aeddula et al., 2011[1] | Left anteromedial temporal lobe | None | None | None | No abnormality | NA | ↓ | ↑ | Absent | FLAIR-↓ |
| Almubaslat et al., 2011[3] | Fronto-parietal lobe | None | None | None | NA | NA | NA | NA | + + HE | - |
| Zhang et al., 2012[20] | Body of corpus callosum | None | None | None | NA | NA | ↔ | ↑ | + + HE | - |
| Alijani et al., 2012[22] | Right parieto-occipital mass | None | None | None | Heterogeneous mass (soft-tissue ↓) | + + HM | NA | NA | | |
| Saito et al., 2014[23] | Cavernous sinus | None | None | None | NA | NA | ↓ | ↓ | + + HM | Lytic lesion which has destroyed the skull and had extra cranial expansion Infiltration along ICA and MCA present Tentative diagnosis - Metastatic malignant meningioma |
| Gulwani et al., 2014[11] | Right middle cranial fossa base involving cavernous sinus | None | None | None | NA | NA | ↔ | + + HM | Destruction of petrous bone + Dural attachment + |
| Maslehaty et al., 2016[19] | Right occipital region | None | None | None | NA | NA | NA | + + HM | Contrast enhancing dural based extra-axial lesion of right posterior fossa + sparing the brain parenchyma Tentative diagnosis - Meningioma or hemangiopericytoma |
| Polewski et al., 2016[22] | Extra-axial mass arising from left tentorium | None | NA | None | NA | NA | NA | + + HE | |
| Present case, 2016 | Left temporoparietal and basal ganglia region | None | None | + | ↑ | + + HE | ↓ | ↑ | + + HE | Solid-cystic lesion with perilesional edema and mass effect |

↑Hyperintense, ↓Hypointense, ↔Isointense, +: Present, HCP: Hydrocephalus, HE: Heterogeneous enhancement, HM: Homogeneous enhancement, ICA: Internal Carotid Artery, MCA: Middle cerebral artery, CT: Computed tomography, MRI: Magnetic resonance imaging, FLAIR: Fluid attenuated inversion recovery images, NA: Not applicable
Difficulties in obtaining a clear resection margin for intracranial LMS are common, and it is generally accepted that the survival rate is poor. Overall, radiation therapy due to ill-defined tumor borders contributes to difficulties in administering an adequate dose of radiation, as well as difficulties in obtaining negative surgical margins. Complete resection of the tumor was possible in only 62% of the reported cases, as shown in Table 1 and it was associated with improved survival rate. Radiotherapy for improved local control of the tumor has also been used widely as an adjunct to surgical resection. 

Recently, gamma knife radiosurgery and stereotactic robotic cyber knife radiosurgery for intracranial LMS have been reported. There are also some reports about the use of chemotherapeutic agents for intracranial LMS, however, results were inconsistent. The overall prognosis of LMS appears relatively poor and seems to be dependent on tumor size and location as well as mitotic rate observed. 

Management

Since rather few case reports exist for this relatively rare clinical entity, there is no established standard treatment regimen for primary intracranial LMS. Based on our literature review, surgery remained the primary treatment, with the aim of obtaining negative surgical margins. Complete resection of the tumor was possible in majority of the reported cases, as shown in Table 1 and it associated with improved survival rate. Radiotherapy for improved local control of the tumor has also been used widely as an adjunct to surgical resection. 

Recently, gamma knife radiosurgery and stereotactic robotic cyber knife radiosurgery for intracranial LMS have been reported. There are also some reports about the use of chemotherapeutic agents for intracranial LMS, however, results were inconsistent. The overall prognosis of LMS appears relatively poor and seems to be dependent on tumor size and location as well as mitotic rate observed. 

Our recommendation is therefore to pursue aggressive multimodality treatment of this rare pathology until more extensive evidence is available about targeted treatment.

CONCLUSION

Primary intracranial LMS occurs very rarely in adult immune-competent patients, which makes the correct diagnosis in this setting challenging. Amongst lesions located purely intraparenchymally, LMS can rarely present with profound perilesional edema and mass effect as in our case, mimicking other malignant intracranial tumor entities. We hope that our literature review will contribute to a better understanding and increased awareness of this uncommon pathology.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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