

CASE REPORT

Primary intracranial leiomyoma - A case report and literature review

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Abstract

Primary intracranial leiomyoma is a rare tumour of mesenchymal origin, with less than 30 cases reported in literature including two cases from the Indian subcontinent. In this article, we describe a case of primary intracranial leiomyoma in an immunocompromised patient with a brief review of literature.

Key words: Epstein-Barr virus; human immunodeficiency virus; immunocompromised; intracranial; leiomyoma; mesenchymal; primary

Introduction

Leiomyomas are benign tumours of mesenchymal origin (ideally non-metastasising) predominantly seen in the genitourinary system. However, metastasis to distant organs like spinal cord, breast, pleura, brain, rib and vertebrae, appendix, parametrium, heart, vessels, bone, muscle, soft tissue, lymph node, and retroperitoneum have been reported.^[1] Our search of literature revealed around 30 reported cases of primary intracranial leiomyoma. In this article, we describe a case of primary intracranial leiomyoma in an immunocompromised adult with a brief review of available literature.

Case History

A 46-year old male presented with the chief complaint of giddiness for an hour associated with an episode of aphasia following which he had difficulty in speaking. He complained of intermittent headaches for the past 6 years. The headache began as shooting type, then progressed to

dull type, localised to left temporal region. There were no exaggerating or relieving factors, no history of trauma, no bowel and bladder disturbances.

He was a known case of HIV and Hepatitis B virus infection on antiretroviral therapy.

On Examination, the patient was afebrile with normal vital signs. Examination of the central nervous system was unremarkable.

Investigations revealed that the patient had mild leukopenia 3.9×10^6 cells/ μ L, ESR-47 mm/hr, He was seropositive for both HIV and Hepatitis B surface antigen (HBsAg).

Contrast enhanced Computed Tomography (CT) scan of the head was performed which showed a well-defined heterogeneously hyper dense lesion [Figure 1] in the left middle cranial fossa measuring $4.1 \times 3.5 \times 3.2$ cm with

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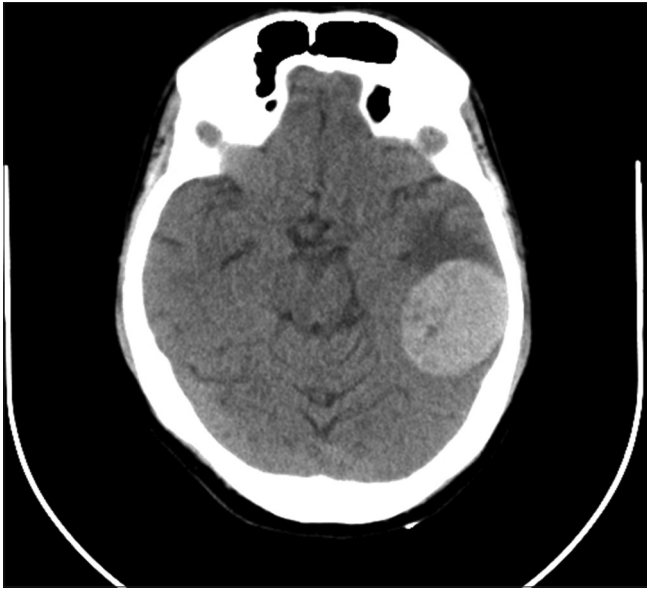


Figure 1: Axial non contrast CT of the brain showing a hyperdense lesion in the left temporal region with perifocal edema



Figure 2: Axial post contrast CT brain showing enhancing lesion in the left temporal fossa with perifocal edema

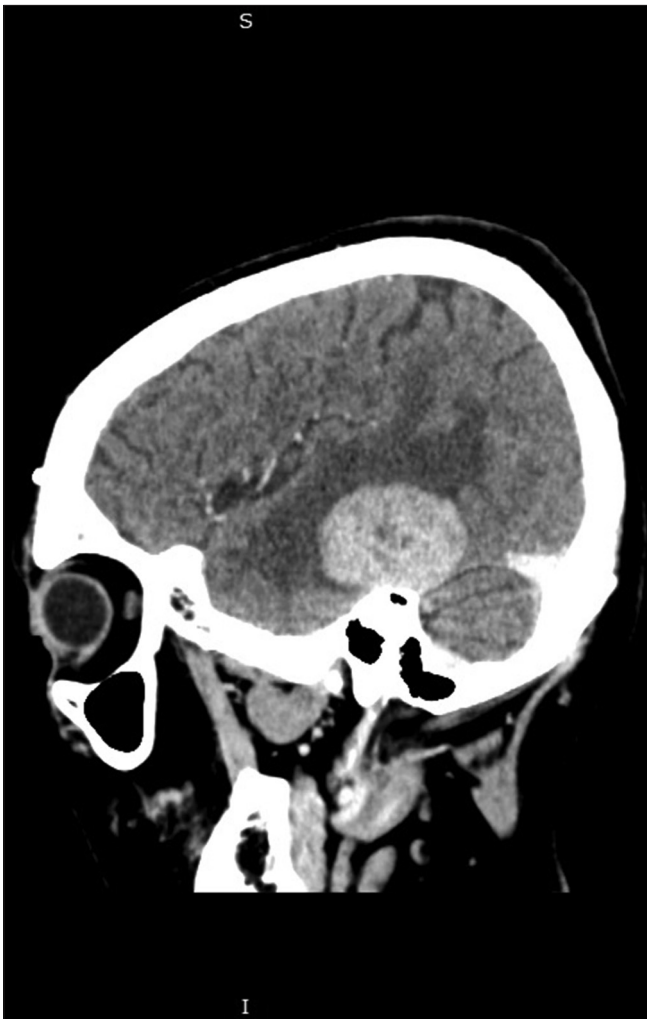


Figure 3: Sagittal reformat of post contrast CT brain showing the same lesion

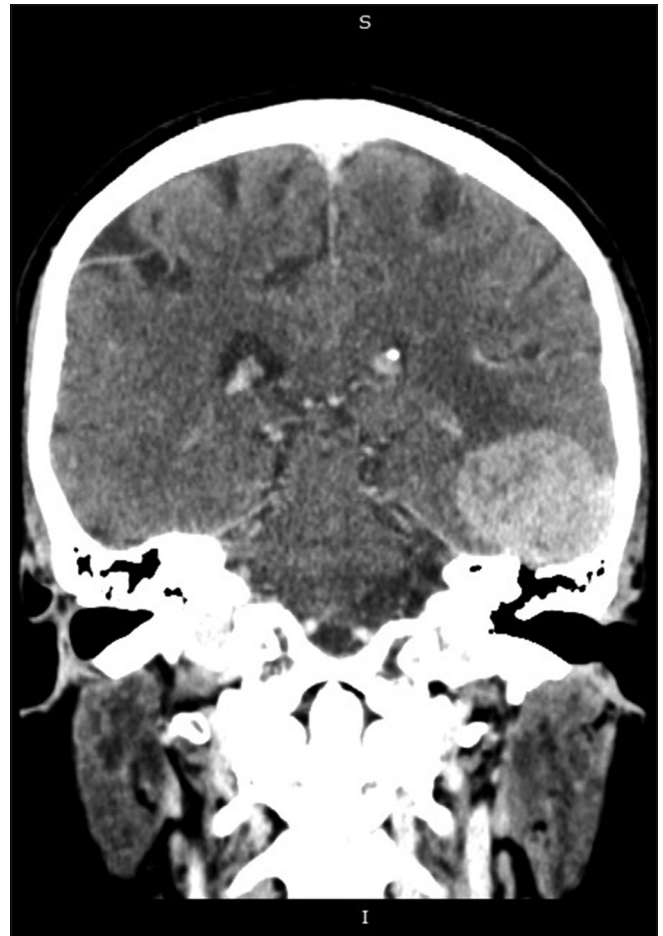


Figure 4: Coronal reformat of post contrast CT brain showing the dural based location of the lesion

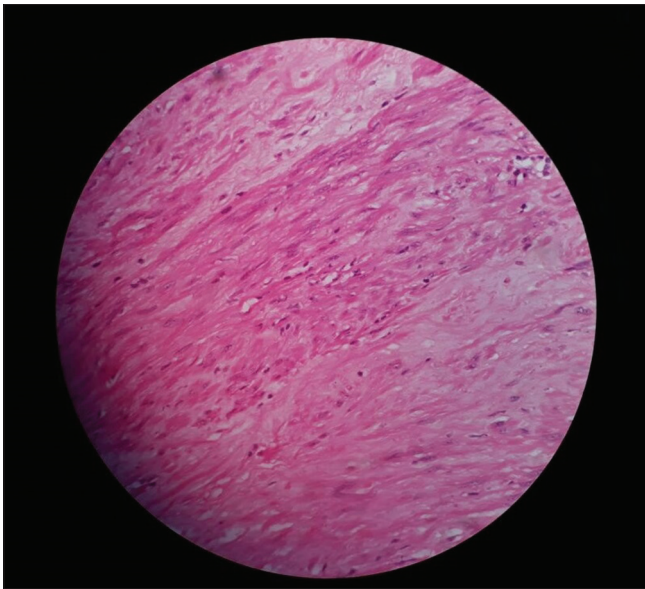


Figure 5: Microscopic images showing spindle cells arranged in broad interlacing fascicles with elongated, slender nuclei

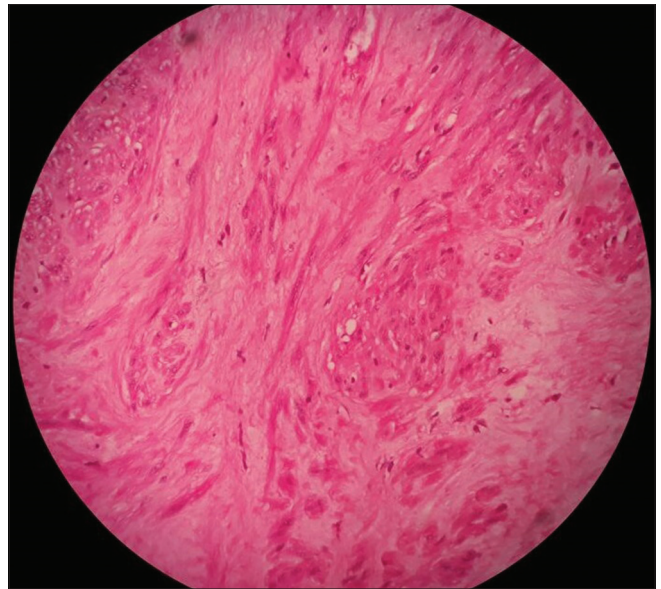


Figure 6: Microscopic images showing spindle cells arranged in broad interlacing fascicles with elongated, slender nuclei

Table 1: Review of literature

Year Author	Locaion of the lesion	Immune status	Gender	Age	HIV	EBV	SIZE in cm	Follow up and Recurrence	Others
1968 Kroe <i>et al.</i>	Sellar	NA	F	68	NA	NA	2.5-3.0	9 m, NR	-
1978 Thierauf &Weiland	Suprasellar	NA	F	4	NA	NA	4.0-5.0	NA	-
1992 Kazumoto and Matsumoto	R Temporal	NA	F	38	NA	NA	3.5	NA	-
1996 Lin <i>et al.</i> /Lai PH	L Temporal	NA	F	20	NA	NA	3 × 2 × 1 cm	24 m, NR	Calcifications
1997 Wang <i>et al.</i>	R Temporal	NA	M	12	NA	NA	NA	56 m, NR	-
1998 Kleinschmidt <i>et al.</i>	Intra and supra sellar	NA	F	56	NA	(-)	3.2	6 m, Residual tumour	-
	RCavernous sinus	Immunocompromised	F	34	(+)	(+)	1.2	4.5 m, Increase in size	-
1999 Kim <i>et al.</i>	R Temporoparietal	NA	F	40	NA	NA	4 × 4.5 × 7	NA	Calcifications
1999 Karpinski <i>et al.</i>	L Sphenoid ridge	Immunocompromised	M	26	(+)	(+)	3	NA	Synchronous intradura extramedullary at C2-3 level
1999 Bargiela <i>et al.</i>	L Occipital lobe	Immunocompromised	F	32	(+)	NA	5	Patient died due to other complication of HIV	Central necrosis
2000 Citow and Kranzler	R Clivus, Upper lobe of right lung	Immunocompromised	F	31	(+)	(-)	3	NA	Low MIB, denovo at both sites
2004 Kumar <i>et al.</i>	R Basal ganglia	Immunocompromised	M	10	(+)	(+)	4	6 m, NR	-
2006 Ali <i>et al.</i>	R Frontal floor	NA	F	29	NA	(-)	1.2	NA	-
2009 Zevgaridis	L Temporal pole	Immunocompromised	F	45	NA	(+)	4×4	7 yr, NR Patient died due to unrelaed causes	Transplant

enhancement post contrast [Figures 2-4]. Oedema noted in the left temporal and parietal lobe causing a midline shift

of 5 mm to the contralateral side -suggestive of a left middle cranial fossa meningioma.

Under general anaesthesia, left frontotemporal craniotomy with excision of the tumour was done. The tumour was extra axial in location with clear demarcation with the adjacent structures.

Histopathology revealed spindle cell neoplasm composed of cells arranged in broad interlacing fascicles. Cells showed elongated, slender nuclei and indistinct cytoplasmic border. Hyalinised stroma with 2-3 mitotic figures/hpf [Figures 5 and 6]. On immunohistochemistry, the tumour cells were positive for smooth muscle actin (SMA), Vimentin and Desmin and negative for epithelial membrane antigen (EMA). This confirmed the diagnosis of leiomyoma.

Discussion

Leiomyoma is a type of smooth muscle tumour considered to be a benign entity although may occasionally metastasise resulting in lesions in distal sites like the brain. Intracranial leiomyomas are rare, with the primary variety being even rarer. The first report of primary intracranial leiomyoma was done by kroe *et al.* in 1968. They suggested that the origin of the tumour to be the vascular smooth muscle^[2] of the brain. Several cases were reported after kroe *et al.* - Most of which were in immunocompromised patients. There is a significant rise in these immunosuppression related

smooth muscle tumours in the recent decade due to the advances in antiretroviral therapy and increase in number of organ transplants and associated immunosuppressive therapies. These smooth muscle tumours may be either benign -leiomyomas or malignant - leiomyosarcoma.

Our search of literature revealed about 30 cases^[3,4] of primary intracranial leiomyoma [Tables 1 and 2] reported, of which only 2 cases have been reported from India. The peak age of incidence was between 25-50 years with the youngest and oldest patients of age 4 and 68 years respectively. There was a slight female preponderance with 17 of the 30 cases being females.

Site: The commonest reported site of occurrence was the middle cranial fossa/temporal lobe followed by the sellar/parasellar/cavernous region. Other reported sites include basal ganglia, frontal region and intraventricular in the lateral ventricle.

Size: The tumours were usually small at presentation with the mean size around 2-4 cm. The largest on record measured 8 cm.

Immune status: Intracranial leiomyomas are commonly associated with immunocompromised states, however

Table 2: Review of literature, continued

Year Author	Locaion of the lesion	Immune status	Gender	Age	HIV	EBV	SIZE in cm	Follow up and Recurrence	Others
2009 Hua <i>et al.</i>	L Middle fossa	Immunocompetent	M	45	(-)	NA	1.5 × 2 cm	14 m, NR	Calcifications
	R Middle fossa	Immunocompetent	M	35	(-)	NA	2 × 3	18 m, NR	-
2010 Dorwal P	L Infratemporal fossa	Immunocompetent	M	17	(-)	(-)	5.4 × 5 × 6 cm	-	Calcifications Bone erosion
2011 Ibebuike	R Frontal lobe	Immunocompromised	M	37	(+)	(+)	4.8 × 4.6 × 5.2 cm	20 wks, NR	Invasion of SSS
2013 Linden E Fornoff	L Middle fossa	Immunocompetent	M	20	(-)	(-)	6 × 8 cm	Multifocal recurrence after 9 years, repeat surgery done, 135 mnths from first surgery -NR	Bone invasion
2013 Andrew Ko	R Sellar	Immunocompetent	F	24	NA	NA	0.7 × 1 × 1.7	NA	-
2013 Andrew Ko	Sellar	Immunocompetent	M	50	NA	NA	3 × 2.5 × 2.5	NA	complex cyst
2013 Lohan <i>et al.</i>	L Sylvian fissure	Immunocompromised	F	55	NA	(+)	NA	NA	Transplant
2014 Issarachaikul	NA	Immunocompromised	F	34	(+)	(+)	NA	NA	-
	NA	Immunocompromised	M	39	(+)	(+)	NA	NA	-
	NA	Immunocompromised	F	32	(+)	(+)	NA	NA	-
	NA	Immunocompromised	F	29	(+)	(+)	NA	NA	-
2016 Garces	L Lateral ventricle	Immunocompetent	M	30	(-)	(-)	1 × 1.2 × 1.9	2.5 yr-NR	-
2017 Upasana	L Frontal	Immunocompromised	M	43	(-)	(-)	2.73 × 1.20 cm	10 m, NR	Renal Transplant
2017 Raheja	L Cavernous sinus	Immunocompromised	F	65	(-)	(+)	0.6 × 0.8 × 1.2 cm	3 mnths, Stable	ALL with allogenic peripheral blood stem cell transplantation
2018 Current case	L Temporal	Immunocompromised	M	46	(+)	NA	4 × 3.5 × 3.2 cm	-	-

seven of the reported cases were in patients with no known immunodeficiency. Of the remaining cases 11 were positive for HIV, four were post-transplant recipients and immune status was unavailable in eight of the patients.

Epstein-Barr virus (EBV) has been implicated in many of these smooth muscle tumours particularly in association with HIV infection. EBV positivity was noted in 12 cases. Data on EBV status was unavailable in these 12 cases. Eight cases were both HIV and EBV positive. Extensive literature is available on the smooth muscle tumours in immunocompromised patients particularly in association with HIV and EBV infections. However, whether the association is significant or just a confounding finding given the immunocompromised state of several of these patients remains unknown. Our patient was positive for both HIV and HBsAg.

Imaging features: Imaging feature of primary intracranial leiomyoma closely resembles that of meningioma with hemangiopericytoma, schwannoma, solitary fibrous tumour as the differentials. The imaging features may resemble lymphoma in the setting of immunosuppression. Other differentials to be considered include glioblastoma multiforme, intracranial angioleiomyoma, meningeal sarcoma and metastasis.

Most tumours appeared as hyper dense solid extra axial masses with perifocal oedema with only one case presenting as a complex cyst. Calcification was reported in four cases, bone erosion/invasion was reported in two cases and invasion of the superior sagittal sinus in one case. Invasion into the adjacent pituitary was noted in two cases. Multifocality was noted in two of the cases, but were still included under primary intracranial leiomyomas because of low MIB (proliferation index). Limited literature is available on the MR imaging of these tumours. T1 hypointensity and T2 hyperintensity was seen with homogeneous enhancement post contrast. However,

absence of diffusion is a feature that differentiates it from tumours like lymphoma. Therefore in an immunocompromised patient with an intracranial extra axial lesion, leiomyoma should be considered in addition to the usual differentials.^[5]

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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Conflicts of interest

There are no conflicts of interest.

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