

Primary Intracranial Thalamic Leiomyosarcoma: Clinical Report of a Case and Review of the Literature

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Abstract: *Purpose:* The incidence of the primary intracranial leiomyosarcoma is extremely rare, and few cases have been previously reported worldwide to date. This report was to clarify the potential role of radiotherapy in the management of primary intracranial leiomyosarcoma.

Methods and Materials: This report presented a 49-year old man with a 3-month history of a progressively headache and walking unsteadily. The diagnosis was confirmed with thalamic leiomyosarcoma of high-grade malignancy according to the pathologic examination after neurosurgical biopsy. The patient didn't undergo surgical resection because of a high risk death. After biopsy, radiotherapy using 3D-CRT technique to the mass site with 55.8Gy/31f/43d was given accordingly.

Results: The mass didn't reduce much at the end of radiotherapy. The patient refused systemic chemotherapy, he was alive without signs of local relapse and brain side-effects with 6 month's follow-up. After living eleven months and three weeks after radiotherapy, he died of local progression.

Conclusions: Through literature review, the current therapeutic approaches including surgery, radiotherapy as well as chemotherapy appear to have limited effect, but could be beneficial of patients in tumor local control and improvement of the life quality.

Keywords: Leiomyosarcoma, Intracranial, Thalamencephalon, Radiotherapy, Surgery.

INTRODUCTION

Tumors of mesenchymal origin occurring either as primary or secondary lesions of the central nervous system are very rare in children as well in adults. Primary intracranial leiomyosarcoma is exceedingly uncommon, most tumors of this kind in the central nervous system (CNS) represent metastatic diseases from other organs or tissues. Primary cerebral leiomyosarcoma is thought to originate from the mesenchymal cells of cerebral meninges, or arise from the cerebral blood vessels epithelium. Although rare, this type of tumor is often included in the differential diagnosis of dural-based lesions. Histo-pathological stains can help to distinguish these tumors from meningiomas. Clinical managements with surgery, radiation and/or chemotherapy should be considered. However, the prognosis is usually discouraged. Here, we present a case with an intracranial thalamic leiomyosarcoma and the relevant literatures were reviewed.

CASE REPORT

Here presents a 49-year-old man with 3 months history of progressive walking unsteadily, gait disturbance, onset of left frontal headache, and signs of slightly elevated intracranial pressure at original consultation. There was no history of accidental injury was found, optical examination demonstrated no bilateral papilloedema and facial palsy. Right-sided weakness was showed in the physical examination. Biochemical laboratory findings including urea and electrolytes, liver function test were normal. Viral serology on human immune deficiency virus (HIV) was negative. Complete blood counts and metabolic parameters were normal. CT scans of the Chest, abdomen and pelvis were negative. An imaging of brain CT scan showed a homogeneous contrast-enhancing occupying lesion with features of slight displacement of the brain parenchyma, the peritumoural oedema was not obvious; the tumor localized to the left thalamic region and did not reach the atrium of the lateral ventricle. No infiltration in the meninges of the brain could be detected. The patient underwent a stereotactic biopsy, and gross resection of the tumor was not performed as the fact of high risk. The diagnosis was confirmed as an intracranial thalamic leiomyosarcoma by biopsy. After biopsy, the

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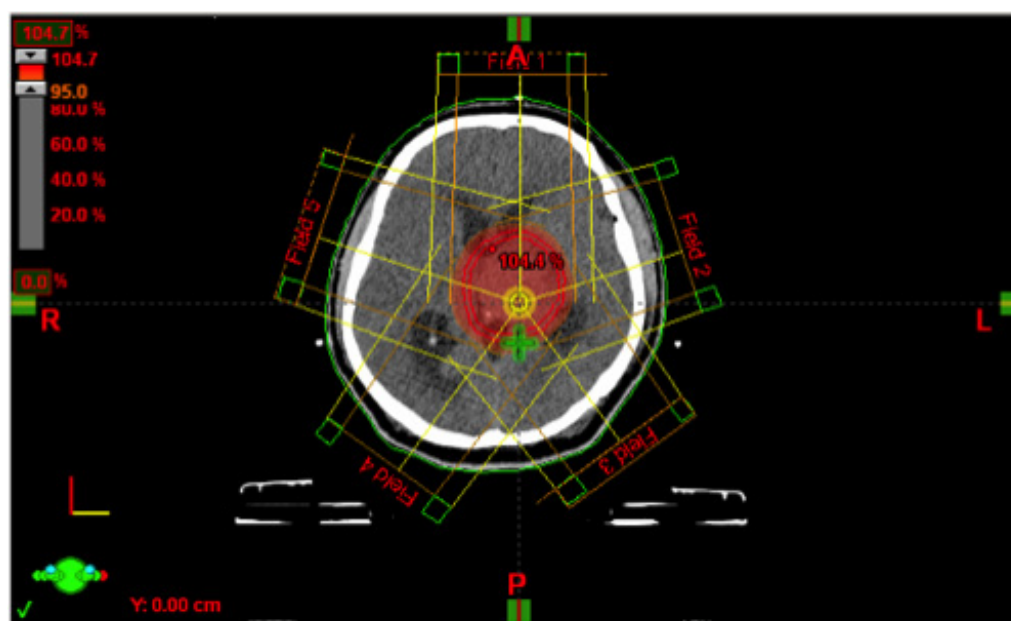


Figure 1: Treatment plan for a leiomyosarcoma in the left thalamic region. The patient received radiotherapy by 3D-CRT technique consisting of 55.8Gy in 1.8Gy per fraction in 31 fractions using Varian Linac 6MV-X rays, the gross tumor volume (GTV) was defined as 2cm from tumor margin, and the 95% plan tumor volume (PTV) isodose line encompassed the target volume.

patient received radiotherapy (RT) using 3D-CRT technique consisting of 55.8Gy in 1.8Gy per fraction in 31 fractions with Varian Linac 6MV-X rays, and the gross tumor volume (GTV) was defined as 2cm from tumor margin (Figure 1). The mass didn't change much in size after radiotherapy. The patient refused systemic chemotherapy thereafter, he was alive without signs of local relapse and brain side-effects after 6 month's follow-up. After living eleven months and three weeks since radiotherapy, he died of local recurrence.

DISCUSSION

Origin of most intracranial soft tissue sarcomas represents metastatic disease of other sites, and the primary intracranial sarcomas are exceedingly rare [1-3], there are around 25 cases reported in the last 30 years up to date (see Table 1). In this case report, the tumor biopsy suggested a cerebral primary thalamencephalon origin.

It was found by review of primary intracranial myomatous tumors in literature that only 1 out of 29 reported cases demonstrated smooth muscle differentiation, other cases might be pure mesenchymal or mixed neural or mesenchymal tumor which shows skeletal muscle differentiation [4]. Paulus W reported 19 primary intracranial sarcomas out of a total of 25,000 brain tumours with biopsies. Of which, there were only three patients with primary intracranial

leiomyosarcoma, two with rhabdomyosarcoma, and other two with angiosarcoma [2]. However, it was demonstrated recent years that slight increased incidence of intracranial leiomyosarcoma has been reported in immunodeficient patients, as occurrence of these leiomyosarcomas associated with EB virus infection and acquired immunodeficiency syndrome (AIDS) [5-10]. But this reported case, however, both HIV and EBV in serology test were negative, suggested a tumor-genesis is not related to the virus infection.

The diagnosis of leiomyosarcoma is usually confirmed by ultrastructural characteristics of smooth muscle cells and immunohistochemistry, S-100 protein staining and epithelial membrane antigen (EMA) could be both negative, and the identification of intracranial leiomyosarcomas with those in other sites relies mainly on immunohistochemical staining and electron microscopy examination.

Regarding clinical management, the best method for the treatment of LMS could not be determined due to the limited number of cases that have been reported. surgical resection of the lesion is usually first recommended, and subsequent adjuvant chemo-and/or radiotherapy should be advocated in the management of the patients. Although multidisciplinary care was given, the prognosis was usually still discouraged with the longest survival reported less than three years [11-13]. Despite limited efficacy,

Table 1: Summary of Patients with Primary Intracranial Leiomyosarcoma in Literature

Ref.	Age/sex	HIV status	EBV	Clinical management	Follow-up
Alijani B <i>et al.</i> [2013]	19/M	negative	negative	total tumor resection	alive after 18 months of follow-up
Takei H <i>et al.</i> [2013]	27/M	negative	positive	gross-total resection	unknown
Kelley BC <i>et al.</i> [2012]	2/M	negative	negative	multiple tumors, large resected	died 3 months after resection
Almubaslat <i>et al.</i> [2011]	47/F	negative	negative	total resection	still alive at 21 months after diagnosis
Sivendran <i>et al.</i> [2011]	43/M	positive	positive	anti-retroviral therapy	no reoccurrence 20 months
Aeddula <i>et al.</i> [2011]	58/M	negative	negative	craniotomy	died 3 wks after craniotomy
Fujimoto <i>et al.</i> [2011]	45/F	negative	negative	subtotal rem + radiotherapy	died 10 months after rem
Gupta <i>et al.</i> [2010]	17/M	positive	positive	radio+ gemcitabine	15 months, then lose followup
Mathieson <i>et al.</i> [2009]	5/M	negative	negative	resection + radio/chemo	alive for 18 months
Hussain <i>et al.</i> [2006]	26/M	negative	negative	surgery; recurrence treated	died 7 months after diagnosis
Suankratay <i>et al.</i> [2005]	--	positive	positive	surgery and radiation	10 months
Lerdlum <i>et al.</i> [2004]	--	positive	negative	surgery and radiation	unknown
Eckhardt <i>et al.</i> [2004]	13/M	negative	negative	resection	died after 15 months
Blumenthal <i>et al.</i> [1999]	--	positive	positive	doxorubicin	24 months
Brown <i>et al.</i> [1999]	--	positive	positive	surgery	n/a
Litofsky <i>et al.</i> [1998]	--	positive	positive	surgery	8 months
Lee <i>et al.</i> [1997]	8/M	negative	negative	resec + Radiother+ Chemo	unknown
Niwa <i>et al.</i> [1996]	--	negative	negative	surgery	died 8 years after diagnosis
Skullerud <i>et al.</i> [1995]	33/M	negative	negative	surgery and radiation	2 years at last followup
Louis <i>et al.</i> [1989]	72/F	negative	negative	surgery	n/a
Asai <i>et al.</i> [1988]	73/M	negative	negative	surgery and radiation/Dox	n/a
Li <i>et al.</i> [1987]	47/M	negative	negative	subtotal resection	died at 1 year after diagnosis
Anderson <i>et al.</i> [1980]	35/M	negative	negative	surgery and radiation	alive after 32months

n/a: not described; rem: removal.

multimodality treatment strategy including optimized surgical resection, postoperative radiotherapy, and chemotherapy should be advocated in most cases to improve patient treatment outcomes and at least life quality.

CONFLICT OF INTEREST

The authors state by present that each of the authors listed below has contributed to, read and approved this manuscript, and there is none of the authors has any conflict of interest, financial or

otherwise on this paper submitted, and we are sure that this manuscript, or any part of it, has not been previously published, and nor is it under consideration elsewhere.

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