

Primary supratentorial intracerebral malignant paraganglioma

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ABSTRACT

Paragangliomas are extra-adrenal neuroendocrine tumors that derive from neural crest. In general, they are benign tumors but few cases had shown a tendency to metastasize. Malignant forms have been reported previously with intracranial metastasis from duodenal origin, but primary intracranial origin represents a rare and unusual location for such tumors. Here, we report a rare case of a 48-year-old lady who presented with symptomatic right-sided insular mass with negative metastatic work up. A complete surgical resection had been done with an unexpected diagnosis of primary gangliocytic paraganglioma with malignant features.

Key words: Brain, chemodectoma, gangliocytic paraganglioma, intracerebral, intracranial

INTRODUCTION

Paragangliomas are rare neuroendocrine tumors. They arise from the paraganglia distributed along the paravertebral sympathetic chains and related ganglia, as well as from the parasympathetic paraganglia such as aortic body, carotid body, and vagal nerve. As a result of their neuroectodermal origin, few paragangliomas can be functional. Hence, autonomic dysfunctional symptoms may occur such as excessive sweating, hypertension, and tachycardia secondary to vasoactive substances release. They are known to be benign tumors with WHO Grade I. However, around 5-10% of them may transform malignant along the course of the disease.^[1]

Paraganglioma may also arise from intracranial or intra-spinal origin. Several explanations were proposed for their existence which include possible growth from ganglionic cells, growth from paraganglia associated with blood vessels or growth from embryonic neuroepithelial rest.^[2] They have a tendency to grow in the sellar/parasellar region or cauda equina region. However, the intraparenchymal growth of paraganglioma is uncommon with seldom reported cases in the literature. An unusual case of primary

intraparenchymal paraganglioma with malignant features was encountered lately at our center. The case is considered to be the first of its kind, and it does reflect the potential risk for malignant primary growth of paraganglioma within the brain parenchyma. A review of the literature for intracranial supratentorial paragangliomas along with the predisposing genetic mutation will be discussed.

CASE REPORT

A 48-year-old healthy female presented to the emergency department with 1-week history of headache and slurred speech. She noticed that her balance had got worse 3 days prior to presentation. On examination, she was noticed to be ataxic with left sided pronator drift and left lower facial asymmetry. The initial computed tomography (CT) scan of her brain showed an iso-dense mass in the right sub-insular area measuring 3.6 cm × 4.1 cm × 3.7 cm, along with perilesional vasogenic edema and mass effect over adjacent structures [Figure 1]. There was no evidence of calcification or hemorrhagic foci within the lesion. It carried slightly low intensity signal in T1-weighted image with heterogenous signal intensity in T2-weighted image. The lesion was well-demarcated and had homogenous gadolinium uptake with restricted diffusion in diffusion-weighted image/apparent diffusion coefficient map [Figure 2].

The patient was admitted to the hospital for a right-sided fronto-parieto-temporal craniotomy for tumor resection guided by neuronavigation. Intraoperatively, the tumor

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10.4103/2347-8659.154431

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was firm in consistency and very vascular in nature but eventually we were able to achieve gross total resection. The early postoperative course was unremarkable for any neurological deficits until patient deteriorated few hours thereafter and had asymmetric blown pupils. Owing to the urgency of the case, the patient was taken for surgery with a provisional diagnosis of postoperative bleeding. Surgical re-exploration was done, and large intracerebral hematoma within the tumor bed was evacuated. The patient, unfortunately, woke up postoperatively with dense left sided hemiplegia [Figure 3].

Microscopic examination of the tumor revealed nests of highly pleomorphic, spindle, epithelioid,

and large atypical cells with prominent nuclei and nucleoli [Figure 4a]. Abnormal mitotic figures and apoptotic nuclei were common. The tight collections of neoplastic cells were surrounded by rich fibrovascular stroma forming the architectural patterns of “Zellballen”. The majority of the neoplastic cells were strongly positive for chromogranin, synaptophysin (SYN), and neuron-specific enolase, and focally for tyrosine hydroxylase [Figure 4b]. Ganglionic cells displayed strong cytoplasmic reaction for SYN and less often tubulin [Figure 4c]. Glial fibrillary acidic protein showed very strong reaction in the cells outlining the edges of the neoplastic congregates, in a pattern seen in sustentacular cells of extracranial paragangliomas [Figure 4d]. The rest of the immunohistochemistry is outlined in Table 1.



Figure 1: Preoperative plain computed tomography head shows a right-sided isodense lesion within the insular region

At this point, an extensive metastatic work up had been done which included CT of her chest, abdomen, and pelvis, as well as mammogram. Due to the fact that this tumor has been frequently originating from the duodenum, a duodenal scope was also performed. The analysis resulted negative for any primary lesions. As some paragangliomas can be functionally active, serum and urine metanephrines, and catecholamines were investigated and resulted also negative. Given these results and the pathological features of the resected tumor, the assumption was made being a primary malignant gangliocytic paraganglioma. The patient was approached in a multidisciplinary team including the radiation oncology and offered local tumor bed radiation. Due to low karnofsky performance

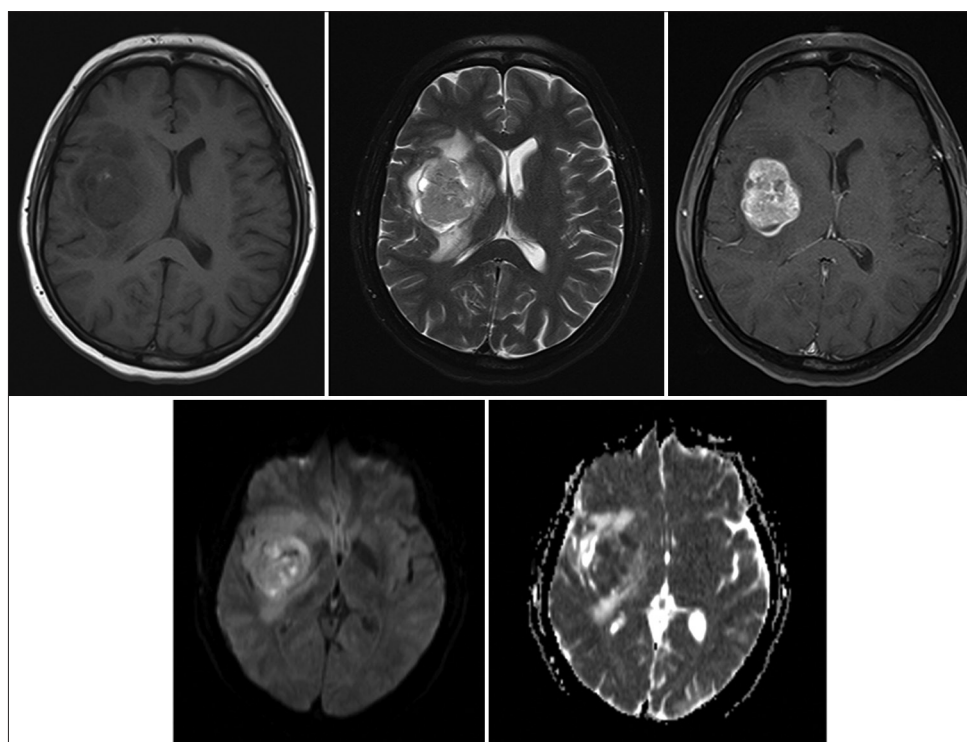


Figure 2: A well-circumscribed tumor that is hypointense in T1-weighted image with heterogeneous texture in T2-weighted image. The lesion was homogeneously enhancing post-gadolinium with restricted diffusion

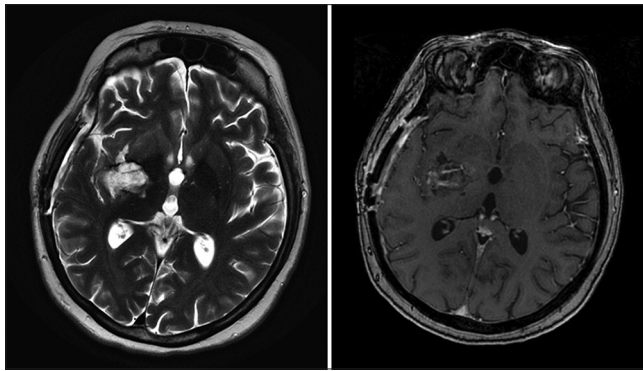


Figure 3: Brain magnetic resonance imaging after 6 weeks from surgery depicts a tumor bed cavity with complete resection. A slight gadolinium uptake was seen within the tumor cavity which represents remnant of fibrillary surgi-cells packing (ethicon) from the second surgery

scale (KPS) of 30, the radiation treatment was deferred until the patient gains more functional strength. She remained stable along with no tumor recurrence on serial follow-up magnetic resonance imaging.

Unfortunately, 12 months postoperatively she presented with a 5 cm × 5 cm ipsilateral enhancing lesion associated with mass effect. Although she was asymptomatic, we opted for surgical resection given the size of the mass that would interfere with radiation therapy. As expected, the pathological work up revealed similar findings to previously resected tumor. Although her surgical resection went uneventful, she continued to decline her functions with lower KPS, which affected the delivery of radiation treatment. She was referred eventually into palliative care unit and died after 2 months from her second surgery.

DISCUSSION

Parangliomas are neural crest-derived tumors that originate in the paraganglia of the autonomic nervous system. Although they are usually considered benign growths, but occasionally malignant forms may occur. They can be functional lesions with active release of catecholamines upon manipulation to result in hemodynamic alterations. The incidence of paraganglioma is frequently reported in combination with pheochromocytoma owing to its rareness along with analogous histological features. The combined annual incidence in United States, for instance, is 500-1600 cases per year with 50% of them present with hypertension.^[3] Parangliomas are usually sporadic but genetic and syndromic associations have been described. In about 27-32% cases of paraganglioma, genetic mutations have been discovered among which the succinate dehydrogenase complex mutation is considered one of the most commonly affected genes.^[4,5] In addition, several recent works have also correlated the SDHB gene mutation and the malignant behavior of paragangliomas.^[6-9]

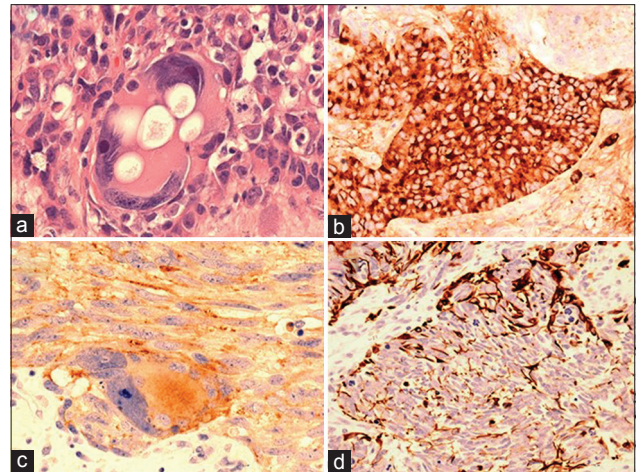


Figure 4: (a) Large neoplastic ganglionic cell with prominent nucleoli and cytoplasmic vacuolation HE; (b) strong chromogranin immunoreactivity in epithelioid tumor cells; (c) cytoplasmic synaptophysin immunoreactivity in a large ganglionic cell and the processes of the adjacent tumor cells; (d) glial fibrillary acidic protein immunoreaction accentuated at the periphery of the tumor nest characteristically seen in sustentacular paraganglioma cells

There have been only 37 reported cases of primary intracranial supratentorial paragangliomas since 1960, and they all harbor benign growth features [Table 2].

In this manuscript, we have excluded from our review all intracranial metastatic paragangliomas and lesions originated in the infratentorial compartment. We found that the mean age of presentation is 47 with 1:1.3 female:male ratio. In around three-quarter of these cases, the sellar/parasellar region was the most common location with symptoms of headache, ophthalmoplegia, and endocrinopathy.^[43] Again, the benign behavior was a character of all of them with few cases required radiation therapy for incomplete resection.

In term of intraparenchymal origin, only three cases had been reported and were harboring benign pattern. Their location within the brain represents a rare condition. Reithmeier *et al.*^[23] had reported the first case of primary paraganglioma in the insular region. This patient was successfully treated with a complete gross total resection, but dense left sided hemiplegia had occurred postoperatively secondary to cerebral vasospasm. In another case, a left temporal melanotic paraganglioma had developed in a patient who was treated previously with chemotherapy and radiotherapy for langerhans cell histiocytosis.^[35] The lesion was partially resected followed by postoperative radiation treatment. The third case had occurred in premotor region, and it was resected completely with the patient remaining stable at short-term follow-up.^[38]

In this report, we presented a unique case of paraganglioma in which primary intraparenchymal growth with malignant features and ganglionic

Table 1: Immunohistochemical evaluation result for primary malignant ganglionic paraganglioma

Antibody	Distributor	Abbreviation (dilution)	Immunoreactivity
Neurofilament MAb	DAKO	NF (1:400)	-
Neural CAM	DAKO	CD56 (undiluted)	+++
β-tubulin	SIGMA	βTUB (1:5000)	F++
Synaptophysin MAb	DAKO	SYN (1:25)	++
Chromogranin A MAb	DAKO	CHRG (1:100)	++
Calretinin Poly Ab	Invitrogen	CAL (1:200)	F+
Neuron-specific enolase	DAKO	NSE (undiluted)	+, ++
S100 protein	DACO	S100 (1:4000)	-
Epithelial membrane antigen MAb	DACO	EMA (1:100)	-
Keratins	DACO	CAM5.2, AE1/AE3 (undiluted)	-
GFAP Poly Ab	DAKO	GFAP (1:3000)	++ sustentacular c.
Anti-human melanosome	DAKO	HMB 45 (undiluted)	+ sustentacular c.
Desmin	DAKO	DSM (undiluted)	-
p53	DAKO	p53 (1:200)	-
Ki-67 MAb	DAKO	Ki-67 (1:1000)	++

GFAP: glial fibrillary acidic protein; CAM: cell adhesion molecule; MAb: monoclonal antibody

Table 2: Literature review for intracranial supratentorial primary paraganglioma

Author	Age (gender)	Presentation	Location	Treatment	Outcome
Kruse, 1960 ^[10]	68, male	Behavioral changes	Middle fossa	Resection	Improved
Smith <i>et al.</i> , 1966 ^{*[11]}	17, male	Headache	Pineal region	Resection	Moderate disability
Chytil, 1967 ^[12]	46, male	Visual loss, hypopituitarism	Sellar/suprasellar	Resection + RT	No progression
Bilbao <i>et al.</i> , 1978 ^[13]	37, male	Delayed growth	Sellar	Resection	-
Ho <i>et al.</i> , 1982 ^[14]	65, male	Diplopia	Cavernous sinus	Resection	Moderate disability
Prabhakar <i>et al.</i> , 1984 ^[15]	7, female	Ophthalmoplegia	Parasellar	Resection + RT	-
Steel <i>et al.</i> , 1993 ^[16]	44, female	Headache	Sellar	Resection + RT	No progression
	41, female	Headache, ptosis	Sellar	Resection + RT	No progression
Flint <i>et al.</i> , 1993 ^[17]	17, female	Visual defect	Sellar	Resection	-
Scheithauer <i>et al.</i> , 1996 ^[18]	14, male	Visual defect	Sellar/parasellar	Resection + RT	Left hemiparesis
Nishitani <i>et al.</i> , 1996 ^{*[19]}	41, female	Amenorrhea	Parasellar	Resection	Good recovery
Noble <i>et al.</i> , 1997 ^[8]	71, male	Visual defect	Sellar	Resection	-
Mokry <i>et al.</i> , 1998 ^[9]	76, female	Visual defect	Sellar	Resection	Unchanged
Caro <i>et al.</i> , 1998 ^[20]	84, male	Memory loss	Sellar/suprasellar	Resection	-
Sambaziotis <i>et al.</i> , 1999 ^[21]	54, male	Visual defect	Sellar	Resection	No progression
Yamauchi <i>et al.</i> , 1999 ^[22]	56, female	Headache	Frontal fossa	Resection	No progression
Reithmeier <i>et al.</i> , 2000 ^[23]	42, male	Seizure	Insula	Resection	Hemiparesis
Laquis <i>et al.</i> , 2001 ^[24]	15, female	Oculomotor palsy	Middle fossa	Resection + RT	Improved
Salame <i>et al.</i> , 2001 ^[25]	48, female	Oligomenorrhea	Sellar/parasellar	Resection	No progression
Hertel <i>et al.</i> , 2003 ^[26]	51, female	Facial paresis	Middle fossa	Resection + RT	Oculomotor palsy
Yokoo <i>et al.</i> , 2003 ^[27]	52, female	Behavioral changes	Suprasellar	Resection	-
Arkha <i>et al.</i> , 2003 ^[28]	58, female	Endocrine dysfunction	Sellar/parasellar	Resection	-
Riopel <i>et al.</i> , 2004 ^[29]	66, male	Diplopia	Parasellar	Biopsy	-
Naggara <i>et al.</i> , 2005 ^[30]	47, male	Visual defect	Suprasellar	Resection	-
Zorlu <i>et al.</i> , 2005 ^[31]	37, male	Visual defect	Sellar/suprasellar	Resection + RT	-
Boari <i>et al.</i> , 2006 ^[32]	52, male	Brain ischemia	Sellar	Resection	Pituitary dysfunction
Peltier <i>et al.</i> , 2007 ^[33]	51, female	Oculomotor palsy	Parasellar	Resection	-
Sinha <i>et al.</i> , 2008 ^[34]	18, male	Visual defect	Sellar	Resection + RT	Skull, scalp and femur metastasis
Yoo <i>et al.</i> , 2008 ^[35]	21, female	Headache	Temporal lobe	Resection + RT	-
Ozüm <i>et al.</i> , 2008 ^[36]	70, male	Headache	Sellar/parasellar	Resection + RT	-
Lu <i>et al.</i> , 2009 ^[5]	81, male	Visual change	Sellar/suprasellar	Resection	Died 4 months after (esophageal cancer)
Haresh <i>et al.</i> , 2009 ^[37]	17, male	Visual change	Sellar/suprasellar	Resection + RT	Skull and femur metastasis
Thakar <i>et al.</i> , 2011 ^[38]	40, male	Visual defect	Frontal lobe	Resection	Recurrence (6 months)
Prajsnar <i>et al.</i> , 2011 ^[39]	53, female	Trigeminal neuralgia	Meckel's cave	Resection	Recurrence (2 years)
Albert <i>et al.</i> , 2011 ^[40]	63, male	Proptosis	Sellar/parasellar	Resection + RT	Improved
Nascimento <i>et al.</i> , 2012 ^[41]	33, female	Endocrine dysfunction	Sellar	Resection	Diabetes insipidus
Chaudhry <i>et al.</i> , 2013 ^[42]	44, male	Visual defect	Sellar/suprasellar	Resection	No recurrence

*Found in Yamauchi *et al.* review of literature, 1999. RT: radiation therapy

component makes it unique and the first of this type. The radiological features were not conclusive for primary malignant paraganglioma as only extracranial

primary malignant forms are known. In addition, the tumor was not functional, and it did not associate with any hemodynamic changes intra-operatively to alert for

possible active paraganglioma. Although the resection was complete, but the functional deconditioning that can be attributed to postoperative bleeding impeded early adjuvant therapy.

Radiation therapy for local control has been proposed for incomplete resection of benign paraganglioma. There seems to be a better response to radiosurgery than external beam radiation therapy for such lesions. The treatment is being delivered at a range of 12-15 Gy to the tumor margin.^[44] We would have planned similar treatment for our patient if she was in a better functional state. Chemotherapy appears to play a role in terms of stabilizing the disease and in some it may lead to reduction in tumor size. This could not be applied to our case given the low KPS and stable tumor bed on serial imaging follow-up.

Even with treatment, the prognosis of malignant paraganglioma remains dismal with 50% reported mortality in 5 years.^[45] However, such data were reported for extracranial paraganglioma/pheochromocytoma and we still do not know if this can be applied for intracerebral primary lesions, which are similar to our case. We suggest that such tumors should be treated with upfront postoperative radiation therapy, preferably radiosurgery. Similarly, it is worth considering delivering chemotherapy based on postoperative resection and risk for future recurrence.

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Cite this article as: Al Jishi AA, Lach B, Elgheriani A, Kachur E, Cenic A. Primary supratentorial intracerebral malignant paraganglioma. *Neuroimmunol Neuroinflammation* 2015;2(2):121-6.

Source of Support: Nil. **Conflict of Interest:** No.

Received: 13-09-2014; **Accepted:** 23-12-2014