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Pathology

Primary Cerebellar Paraganglioma – An Incidental Finding on Autopsy, A Case Report and Review of Literature

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Abstract | **Case Report**

Paraganglioma are extra – adrenal neuroendocrine tumors derived from neural crest. They arise from paraganglia present along the paravertebral sympathetic chain as well as from parasympathetic ganglia such as aortic body, carotid body, and vagal nerve. Primary Intracranial origin represents a rare and unusual location for such tumors. We report a rare and incidental intracerebellar paraganglioma in a 37-year-old lady as very few cases are reported in the literature and none is reported on autopsy.

Keywords: Paraganglioma, Intracerebellar, Neural crest, Zellballen pattern, Neuroendocrine tumor, Autopsy.

INTRODUCTION

Paraganglioma are rare neuroendocrine tumors of neural crest origin. They are benign mostly, but aggressive malignant tumor with distant metastasis can also occur [1]. Paraganglioma of head and neck accounts for only 0.6% of all tumors. They mostly arise from carotid body [app. 75%] and jugular foramen [app. 16%]. Other origins include larynx, thyroid, vagus nerve and nasal cavity. Approximately 90% of paraganglioma in the brain arises from jugular foramen followed by tympanic nerve in the temporal bone [2]. Autonomic dysfunctional symptoms may occur such as excessive sweating, hypertension, and tachycardia secondary to vasoactive substance release. They are known to be primary tumor with WHO Grade 1 [3].

CASE REPORT

Here, we report an incidental autopsy finding of Cerebellar Paraganglioma in a 37-year-old female. The viscera of the patient were received in the department of pathology, Guru Gobind Singh Medical College Faridkot, Punjab. We have received part of brain [cerebellum] measuring 9x6x3.5cm weighing 200 gms. Cut section shows a grey tan area measuring 1.5 cm in diameter. Histological examination revealed the characteristic cellular arrangement of paraganglioma generally designated as Zellballen pattern on light microscopy. Typical morphology includes monomorphic cells with abundant, eosinophilic, granular cytoplasm with round to ovoid, vesicular, salt and pepper nuclei arranged in a zellballen pattern separated by fibrovascular septae with sustentacular cells. Mild degree of pleomorphism, hyperchromatic nuclei and low mitotic activity are seen. There were no significant gross and microscopic findings in other organs received.

DISCUSSION

Paragangliomas are neural crest–derived tumors that arise from the paraganglia of the autonomic nervous system. Although they are generally considered benign, malignant variants do occur occasionally. These tumors may be functional, with active secretion of catecholamines; manipulation of such lesions can precipitate significant hemodynamic instability. Due to their rarity and shared histopathological features, the incidence of paragangliomas is often reported together with pheochromocytomas. In the United States, the combined annual incidence is estimated at 500–1,600 cases, with approximately 50% of patients presenting with hypertension [4].

Most paragangliomas are sporadic; however, several genetic and syndromic associations have been identified. Germline mutations are detected in approximately 27–32% of cases, with alterations in the succinate dehydrogenase [SDH] complex genes being among the most frequently implicated [5,6]. Furthermore, recent studies have demonstrated a strong association between SDHB gene and malignant behavior of paraganglioma [7-10].

There have been only 45 reported cases of primary intracranial supratentorial paragangliomas since 1960, and they all harbor benign growth features [3].

Literature review for intracranial supratentorial primary paraganglioma

Author	Age [gender]	Presentation	Location	Treatment	Outcome
Kruse, 1960	68, male	Behavioral changes	Middle fossa	Resection	Improved
Smith *et al.,* 1966*	17, male	Headache	Pineal region	Resection	Moderate disability
Chytil, 1967	46, male	Visual loss, hypopituitarism	Sellar/suprasellar	Resection + RT	No progression
Bilbao *et al.,* 1978	37, male	Delayed growth	Sellar	Resection	-
Ho *et al.,* 1982	65, male	Diplopia	Cavernous sinus	Resection	Moderate disability
Prabhakar *et al.,* 1984	7, female	Ophthalmoplegia	Parasellar	Resection + RT	-
Steel *et al.,* 1993	44, female	Headache	Sellar	Resection + RT	No progression
	41, female	Headache, ptosis	Sellar	Resection + RT	No progression
Flint *et al.,* 1993	17, female	Visual defect	Sellar	Resection	-
Scheithauer *et al.,* 1996	14, male	Visual defect	Sellar/parasellar	Resection + RT	Left hemiparesis
Nishitani *et al.,* 1996	41, female	Amenorrhea	Parasellar	Resection	Good recovery
Noble *et al.,* 1997	71, male	Visual defect	Sellar	Resection	-
Mokry *et al.,* 1998	76, female	Visual defect	Sellar	Resection	Unchanged
Caro *et al.,* 1998	84, male	Memory loss	Sellar/suprasellar	Resection	-
Sambaziotis *et al.,* 1999	54, male	Visual defect	Sellar	Resection	No progression
Yamauchi *et al.,* 1999	56, female	Headache	Frontal fossa	Resection	No progression
Reithmeier *et al.,* 2000	42, male	Seizure	Insula	Resection	Hemiparesis
Laquis *et al.,* 2001	15, female	Occulomotor palsy	Middle fossa	Resection + RT	Improved
Salame *et al.,* 2001	48, female	Oligomenorrhea	Sellar/parasellar	Resection	No progression
Hertel *et al.,* 2003	51, female	Facial paresis	Middle fossa	Resection + RT	Occulomotor palsy
Yokoo *et al.,* 2003	52, female	Behavioral changes	Suprasellar	Resection	-
Arkha *et al.,* 2003	58, female	Endocrine dysfunction	Sellar/parasellar	Resection	-
Riopel *et al.,* 2004	66, male	Diplopia	Parasellar	Biopsy	-
Naggara *et al.,* 2005	47, male	Visual defect	Suprasellar	Resection	-
Zorlu *et al.,* 2005	37, male	Visual defect	Sellar/suprasellar	Resection + RT	-
Boari *et al.,* 2006	52, male	Brain ischemia	Sellar	Resection	Pit.dysfunction
Peltier *et al.,* 2007	51, female	Occulomotor palsy	Parasellar	Resection	-
Sinha *et al.,* 2008	18, male	Visual defect	Sellar	Resection + RT	Skull, scalp and femur metastasis
Yoo *et al.,* 2008	21, female	Headache	Temporal lobe	Resection + RT	-
Ozüm *et al.,* 2008	70, male	Headache	Sellar/parasellar	Resection + RT	-
Lu *et al.,* 2009	81, male	Visual change	Sellar/suprasellar	Resection	Died 4 months after [esophageal cancer]
Haresh *et al.,* 2009	17, male	Visual change	Sellar/suprasellar	Resection + RT	Skull and femur metastasis
Thakar *et al.,* 2011	40, male	Visual defect	Frontal lobe	Resection	Recurrence [6 months]
Prajsnar *et al.,* 2011	53, female	Trigeminal neuralgia	Meckel's cave	Resection	Recurrence [2 years]
Albert *et al.,* 2011	63, male	Proptosis	Sellar/parasellar	Resection + RT	Improved
Nascimento *et al.,* 2012	33, female	Endocrine dysfunction	Sellar	Resection	Diabetes insipidus
Chaudhry *et al.,* 2013	44, male	Visual defect	Sellar/suprasellar	Resection	No recurrence
Zhao-Jian Li *et al.,* 2014	16, male	Headache, dizziness	Intracerebellar	Resection	No recurrence
Ahmed A *et al.,* 2015	48, female	Headache, slurred speech	supratentorial	Resection +RT	Died after 2 months
Sean B.Lyne *et al.,* 2019	73, female	Visual defect	Sellar	Resection	No recurrence
Elizabeth A. *et al.,* 2020	81, male	Visual defect	Sellar	Resection	No recurrence

Author	Age [gender]	Presentation	Location	Treatment	Outcome
Stefan Stojanoski *et al.,* 2021	31, male	Hypertension, headache	Sellar/ parasellar	Resection	Recurrence [4 years]
Ailsa Maria Main *et al.,* 2022	39, female	Respiratory distress and hypotension	Skull base	Resection	No recurrence
Nadeem Akhtar *et al.,* 2024	42, female	Hypertension, tachycardia, headache	Cerebellopontine angle	Resection	No recurrence
Shota Yoshimura *et al.,* 2025	75, male	Asympatomatic	Sellar	Resection	No recurrence

The mean age at presentation was 47 years, with a female-to-male ratio of 1:1.3. In approximately three-quarters of cases, the sellar/parasellar region was the most common site of involvement, and patients commonly presented with headache, ophthalmoplegia, and endocrinopathy. All reported lesions demonstrated benign behavior, although a few cases required adjuvant radiotherapy following incomplete surgical resection [11].

In this report, we presented a rare and incidental finding of primary intracerebellar Paraganglioma in an autopsy of 37-year-old female in the department of pathology, Guru Gobind Singh medical college Faridkot, Punjab with characteristic Zellballen pattern on light microscopy. No case has been reported yet on autopsy.

CONCLUSION

1. Intracerebellar Paraganglioma is a rare tumor and histopathlogy is the gold standard.
2. Autopsy specimens should be thoroughly grossed for any incidental finding.

REFERENCES

1. Cai P, Mahta A, Kim RY, Kesari S. Paraganglioma with intracranial metastasis: a case report and review of the literature. *Neurol Sci.* 2012;33[3]:685–9.
2. Yamauchi T, Kubota M, Saeki N, Aihara N, Iwadate Y, Yamaura A. Paraganglioma in the frontal skull base: case report. *Neurol Med Chir [Tokyo].* 1999;39[4]:308–12.
3. 3] Al Jishi AA, Lach B, Cenic A, *et al.,* Primary supratentorial intracerebral malignant paraganglioma. *Neurosciences [Riyadh].* 2015;20[2]:121–6.
4. Chen H, Sippel RS, O'Dorisio MS, Vinik AI, Lloyd RV, Pacak K; North American Neuroendocrine Tumor Society [NANETS]. The North American Neuroendocrine Tumor Society consensus guideline for the diagnosis and management of neuroendocrine tumors: pheochromocytoma, paraganglioma, and medullary thyroid cancer. *Pancreas.* 2010;39[6]:775–83.
5. Amar L, Bertherat J, Baudin E, Ajzenberg C, Bressac-de Paillerets B, Chabre O, *et al.,* Genetic testing in pheochromocytoma or functional paraganglioma. *J Clin Oncol.* 2005;23[34]:8812–18.
6. Lu JQ, Khalil M, Hu W, Sutherland GR, Clark AW. Tumor-to-tumor metastasis: esophageal carcinoma metastatic to an intracranial paraganglioma. *J Neurosurg.* 2009;110[4]:744–8.
7. Ellis RJ, Patel D, Prodanov T, Nilubol N, Pacak K, Kebebew E. The presence of SDHB mutations should modify surgical indications for carotid body paragangliomas. *Ann Surg.* 2014;260[1]:158–62.
8. Lee KY, Oh YW, Noh HJ, Lee YJ, Yong HS, Kang EY, *et al.,* Extraadrenal paragangliomas of the body: imaging features. *AJR Am J Roentgenol.* 2006;187[2]:492–504.
9. Noble ER, Smoker WR, Ghatak NR. Atypical skull base paragangliomas. *AJNR Am J Neuroradiol.* 1997;18[5]:986–90.
10. Mokry M, Kleinert R, Clarici G, Obermayer-Pietsch B. Primary paraganglioma simulating pituitary macroadenoma: a case report and review of the literature. *Neuroradiology.* 1998;40[4]:233–7.
11. Voulgaris SG, Partheni M, Tzortzidis F, Ravazoula P, Pessach IS, Papadakis N, Polyzoidis KS. Suprasellar and intrasellar paragangliomas. *Clin Neuropathol.* 2006; 25:221–6.