International Journal Dental and Medical Sciences Research
Volume 3, Issue 2, Mar-Apr. 2021 pp 810-812 www.ijdmsrjournal.com ISSN: 2582-6018

PARAGANGLIOMA: An Uncommon, Yet Interesting Case

Mariyam.M[1], Sherif.A[2], Roshni PS[3], Prasanan.D[4]
Resident doctor, Department of Medical Oncology AOI
Consultant Medical Oncologist, AOI ,Calicut
Consultant Pathologist, Baby memorial hospital , Calicut
Consultant and Head of Medical Oncology,AOI Calicut

Submitted: 30-03-2021 Revised: 05-04-2021 Accepted: 08-04-2021

ABSTRACT: Paragangliomas are uncommon type of neuroendocrine tumours which have the ability to secrete catecholamines. Most of these neoplasms are asymptomatic; but approximately 1 -3% of the cases witness sympathetic hyperactivity and hypertension due to clinically significant catecholamine secretion. It is indeed a task to differentiate paraganglioma from pheochromocytoma, GIST, thyrotoxicosis, migraine headaches, cardiac arrhythmias, anaphylaxis, panic attacks and stroke.

I. INTRODUCTION

Paragangliomas are relatively rare and evasive kind of neuroendocrine tumours that possess the potential to secrete catecholamines; which can occur anywhere between the base of skull and base of pelvis **[1]**. Men are seen to be at a greater predisposition **[2]** and the clinical presentation ranges from being completely asymptomatic, to the classical triad of headache, excessive sweating and palpitations associated with hypertension **[3]**. Though mostly sporadic, about 30 - 40% **[4]** are familial and sometimes maybe associated with genetic diseases such as MEN type 2 A and 2B, Von Hippel Lindau syndrome, Neurofibromatosis type I and Carney stratkis dyad **[5]**.

This is a report of a case of a 24 year old male patient who was seen to have extra adrenal retroperitoneal paraganglioma that was encountered on table during emergency laparotomy. This article aims to make clinicians more aware and to keep in mind paraganglioma as a possible differential for cases presenting with asymptomatic abdominal mass, features of sympathetic hyperactivity, unexplained facial flushing, hyperventilation, etc. especially because these tumours are elusive in the absence of imaging and histopathological examinations. Additionally they pose a threat with disastrous consequences during surgery due to catecholamine hypersecretion if un/miss diagnosed or due to inadequate preoperative preparation.

II. CASE PRESENTATION

A 24 year old male patient, who has no prior known co morbidities had been treated conservatively at a local hospital for complaints of diffuse abdominal pain; however since he had no relief of symptoms, he was taken to a tertiary hospital where he was found to be in hypovolemic shock as evidenced by cold clammy skin, cold extremities and fall in blood pressure. He had worsening of abdominal pain and initially intestinal perforation was suspected in view of rapid fall in BP and massive haemoperitoneum. ABG showed metabolic acidosis. On arrival to our hospital, patient was on inotrope support. Since he was seen to be rapidly deteriorating he was taken for emergency laparotomy. During the procedure a highly vascular mass of size 15x15x15cm was encountered, densely adherent to the left adrenal, compressing and abutting the upper pole of left kidney and left renal hilum and compressing the renal vessels. Originally thought to be an angiomyolipoma the tumour had bleeding from multiple sites. Histopathology examination reported extra adrenal paraganglioma. Upon gross examination, the excised mass was a solid, soft, nodular mass measuring 17x14x8.5cm and weighing 1160grams; Cut section was solid, tan coloured with focal cystic and haemorrhagic areas. Upon Microscopic examination capsulated cellular neoplasm composed of polygonal tumour cells arranged into large nests, trabeculae and in diffuse pattern with interspersed blood vessels (fig A). Cells show abundant granular amphophilic cytoplasm and irregular hyperchromatic to vesicular nuclei with few showing pseudo inclusions. Occasional multinucleated tumour cells were noted. Areas of hemosiderin laden macrophages and thrombosed blood vessels were noted (fig B).

International Journal Dental and Medical Sciences Research
Volume 3, Issue 2, Mar-Apr. 2021 pp 810-812 www.ijdmsrjournal.com ISSN: 2582-6018

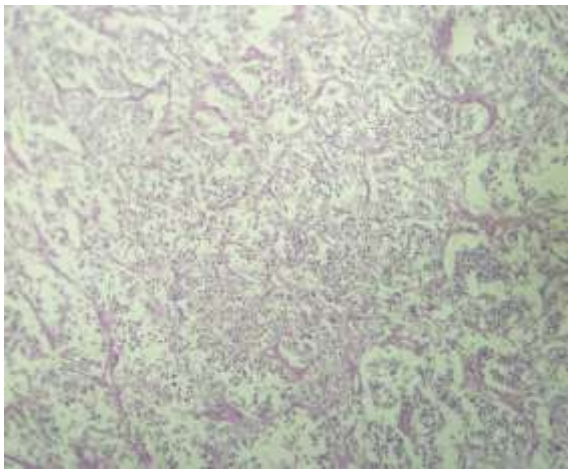

Fig A

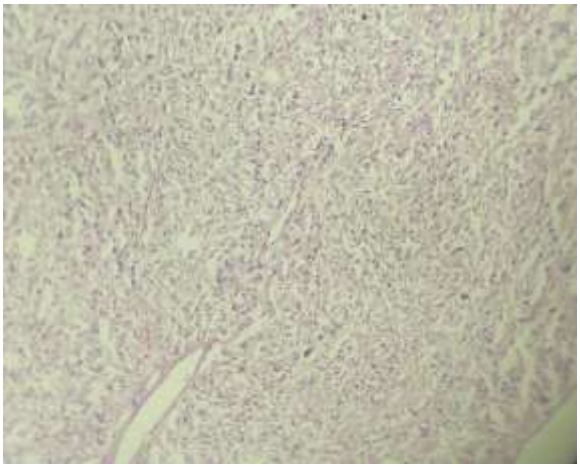

Fig B

IHC: Tumour cells show strong and diffuse positivity for synaptophysin and chromogranin A (fig C and D).

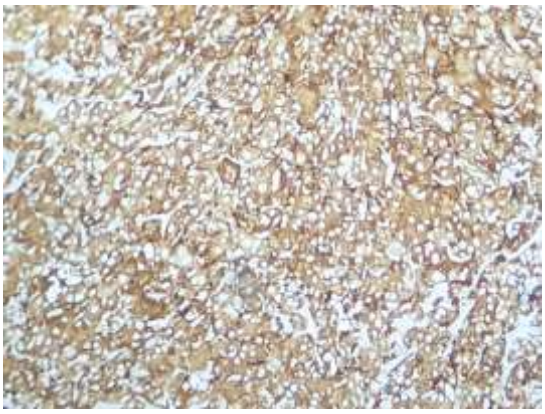

Fig C

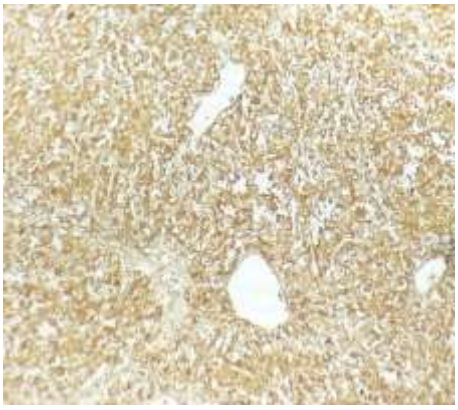

Fig D

Also, 24 hour urine metanephrine - showed normal values. PET CT did not reveal any evidence of distant metastasis or any other tumors. Medical oncology opinion was taken and feasibility of intraperitoneal chemotherapy was discussed.

III. DISCUSSION

Paragangliomas are of two broad types – functional and non-functional **[6]**. Non-functional tumours are most often than not incidental findings upon imaging studies, as in CT scan, where it is seen as a homogenous mass. However for functional Paragangliomas, early diagnosis is crucial as surgical removal of the tumour is often curative **[7]**.

Paragangliomas are often undiagnosed and are often incidental findings, as vast majority of them are asymptomatic. Since these tumours are life threatening, they should be treated with expertise and caution, especially before taking into surgery.

When symptomatic they mimic a variety of other ailments and can easily be mistaken fro them. The symptoms could also be vague like facial flushing, palpitations, excessive sweating etc.

Paragagangliomas are highly vascular tumors due to which they take on a deep reddish hue and they are also found to have firm and rubbery consistency. Microscopically the the tumor cells are seen to be arranged in distinctive balls or nests of cells which are called Zellballen **[8]** nests. The cells in the zellballen structures are positive for chromogranin, synaptophysin, neuron specific enolase, serotonin, neurofilament and neural cell adhesion molecules; which can be identified by immunohistochemistry. Also the paraganglioma cells are argyrophilic, Periodic Acid Schiff negative, Mucicarmine negative and argentaffin negative.

Laparoscopic resection for small non-invasive tumour and laparotomy for large tumour is the preferred route **[9]** of treatment for localised paraganglioma. Pre-operative preparations require

alpha blockers for 7 to 14 days with or without beta blockers, and hydration.

In patients with metastatic disease, palliative chemotherapy with Cyclophosphamide, Dacarbazine and Vincristine (CVD regimen) **[10]** is an option that can be attempted.

Given that the neoplasm is not located in an overly sensitive area, the prognosis is usually reassuring. However, malignant tumours have a much more ominous prognosis. Yearly follow up with serum or urine metanephrine is advised as the chance for recurrence is not completely negligible.

Another interesting fact that has been observed in this case is that the patient had prominent marfanoid features. More study may be required to make a connection between the Marfan syndrome and occurrence of paraganglioma in such individuals.

Conclusively, paraganglioma is a life threatening disease; which most certainly should be put on the differential board in cases presenting with asymptomatic retroperitoneal mass and other features of sympathetic over activity. In diligently keeping in mind such diagnosis, albeit, rare and in mild contradiction with Occam's razor, could save lives and prevent problems; especially intra operatively.

REFERENCES

[1]. Chattoraj AK, Rao UM, Sarkar N, Jakka S. Non-functional retroperitoneal paraganglioma: A case report. J Family Med Prim Care. 2019;8(4):1497-1499. doi:10.4103/jfmpc.jfmpc_189_19

[2]. Date A, Khadav B, Nongmaithem M. Paraganglioma: A case report. J Evid Based Med Healthc 2016; 3(5), 173-175. DOI: 10.18410/jebmh/2016/41

[3]. Tabakin AL, Weintraub MA, Radadia KD, Salazar CG, Sadimin E, Singer EA. Metastatic retroperitoneal paraganglioma: Case report and review of the literature. Clin Oncol (Belmont). 2019 Mar;4:1589. Epub 2019 Mar 8.

[4]. Koopman K, Gaal J, de Krijger RR. Pheochromocytomas and Paragangliomas: New Developments with Regard to Classification, Genetics, and Cell of Origin. Cancers (Basel). 2019;11(8):1070. Published 2019 Jul 29. doi:10.3390/cancers11081070

[5]. William C. Fox, Matthew Read, Richard E. Moon, Eugene W. Moretti, Brian J. Colin, "An Undiagnosed Paraganglioma in a 58-Year-Old Female Who Underwent Tumor Resection", Case Reports in Anesthesiology, vol. 2017, Article ID 5796409, 4 pages, 2017.

[6]. Bacalbasa N, Balescu I, Tanase A, Brezean I, Vilcu M, Brasoveanu V. Successful Resection of a Non-functional Paraganglioma with Celiac Trunk Invasion Followed by Common Hepatic Artery Reimplantation - A Case Report and Literature Review. In Vivo. 2018;32(4):911-914. doi:10.21873/invivo.11328

[7]. Balasubramanian G, Nellaiappan V. Functional paraganglioma. BMJ Case Rep. 2014;2014:bcr2013203425. Published 2014 Feb 20. doi:10.1136/bcr-2013-203425

[8]. Jashnani KD, Patil RD, Balsarkar DJ. Loose cell clusters with vascular coats: Zellballen pattern of paraganglioma on cytology. J Cytol. 2013;30(4):278-279. doi:10.4103/0970-9371.126670

[9]. Alejandro Terrones-Lozano, Alan Hernández-Hernández, Edgar Nathal Vera, Gerardo Yoshiaki Guinto-Nishimura, Jorge Luis Balderrama-Bañares, Claudia Ramírez-Rentería, Judith de la Serna-Soto, Alfredo Adolfo Reza-Albarran, Lesly Portocarrero-Ortiz, "Current Approach of Functioning Head and Neck Paragangliomas: Case Report of a Young Patient with Multiple Asynchronous Tumors", Case Reports in Endocrinology, vol. 2020, Article ID 6827109, 6 pages, 2020.

[10]. Prades CA, Atassi B, Nazeer H. Metastatic Malignant Paraganglioma: A Case Report and Review of Literature. World J Oncol. 2017;8(3):92-95. doi:10.14740/wjon1033w