SJIF Impact Factor: 5.464

WORLD JOURNAL OF ADVANCE HEALTHCARE RESEARCH

ISSN: 2457-0400
Volume: 7.
Issue: 5
Page N. 155-158
Year: 2023

Case Report www.wjahr.com

INTRAABDOMINAL PARAGANGLIOMA-A RARE CASE REPORT

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Received date: 08 March 2023 **Revised date:** 28 March 2023 **Accepted date:** 18 April 2023

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ABSTRACT

Paragangliomas are rare, catecholamine (norepinephrine) secreting neuroendocrine tumors. The parasympathetic tumors are usually asymptomatic and inactive, located mostly in the skull base In contrast, sympathetic lesions are highly active and symptomatic and mainly located in the abdomen and pelvic regions. Surgical biopsy of the lesion is the gold standard to confirm the diagnosis, but it does not differentiate between pheochromocytomas and paragangliomas.[1] Clinical correlation is usually enough for the diagnosis aided by imaging and pathology findings of the lesion. We report a case of 36 year old female with an asymptomatic paragaganglioma with unusual imaging biopsy findings.

INTRODUCTION

Catecholamine (norepinephrine) secreting neuroendocrine tumours, such as paragangliomas, are uncommon and typically found around the base of the skull or in the pre-aortic and paravertebral sympathetic plexus. Less differentiated tumours include head-and-neck paragangliomas in the jugular foramen, ear, or carotid body. In contrast to highly differentiated intraabdominal adrenal medulla tumours such neuroblastoma and pheochromocytoma, they produce norepinephrine. Pheochromocytomas and paragangliomas are extremely vascular and might have sympathetic or parasympathetic characteristics.[2]

The majority of the parasympathetic tumours, which are asymptomatic and dormant, are found in the base of the skull, where the IX and X cranial nerves are distributed. In contrast, sympathetic lesions are primarily found in the abdominal and pelvic areas, where they are highly active, symptomatic, and localised. Compared to the paragangliomas at the base of the skull, they are more functional and hypersecretory (norepinephrine). Although they are often benign, a tiny number of them could develop malignancies and spread.[3] Thus, early detection followed by a full surgical resection is frequently curative and has a good prognosis. The only accurate method for determining the biological aggressiveness of paragangliomas is to look for distant metastases.

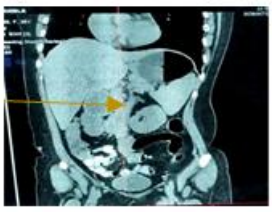
The gold standard for confirming the diagnosis is a surgical biopsy of the lesion, although this procedure does not distinguish between paragangliomas and pheochromocytomas. With the aid of imaging and pathology findings of the lesion, clinical correlation is frequently sufficient for the diagnosis.[4]

CASE REPORT

A 36 year old female came to JJH opd with chief complaints of pain in the lower abdomen since 4 months.No bowel/bladder complaints.No sytemic complaints .Patient is recently diagnosed diabetic and was on medications for past 3 months. Her all routine investigations, chest and abdomen x-ray were within normal limits. On per abdominal finding, no lump or swelling palpable, no tenderness.USG abdomen with pelvis was suggestive of heterogenous, non vascular lesion adjacent to the head of pancreas , near the lower pole of right kidney with multiple thin septations and mural collection seen within.

Patient CECT abdomen was suggestive of well defined exophytic rounded mass lesion, with peripherally placed enhancing soft tissue component and a larger non enhancing necrotic core,likely arising from the right lateral part of the second portion of duodenum.Medially there is loss of fat planes with the ampulla of vater and the uncinate process of pancreas,laterally the fat planes with hepatic flexure and transverse colon is maintained.Posteriorly it is seen compressing the inferior

vena cava.Findings are suggestive of neoplastic etiology and GIST may be a possibility.



Endoscopic Ultrasound was suggestive of cystic pancreatic lesion;hemorrhagic fluid aspirated.

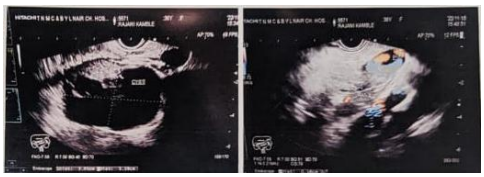


Image guided biopsy from the solid component of the lesion was done Histopathology was suggestive of GIST.

On histopathological diagnosis of GIST, patient was planned for a Whipple's procedure. Patient was induced under general anesthesia and a Chevron incision was taken. Thompson's self retaining retractor was inserted. No free fluid was noted in the abdomen. The greater omentum was lifted off the small bowel, and examined closely. No evidence of any peritoneal metastasis. Hepatocolic ligament divided, and the hepatic flexure of colon pushed down. Kocherisation done, exposing the second and third part of duodenum. Duodenum appears to be normal, no evidence of any exophytic growth or bowel wall thickening. Gastrocolic ligament opened, the pancreas in its entire length examined, no evidence of any mass lesion. C loop of duodenum retracted, Inferior vena cava, Right renal vein visualized. A 6x6 cm greyish lesion with few blood vessels over the surface and free from duodenum, right renal hilum, pancreas was seen. Fine and blunt dissection was done and mass excised in complete totality without capsular breach. Patient's intraoperative blood pressure shot up to a maximum of 180/100 mm of Hg on manipulating the mass. Nitroglycerine drip started and continued post operatively. Blood loss experienced was minimal .No intra or post of complications .Histopathology with IHC report was s/o paraganglioma .Since, the histopathology report was of benign etiology, the surgery was curative, with no requirement for further resection / chemo or radiotherapy. Patient is currently being followed up, 6 month post procedure, the patient is stable, normotensive, relieved of her complaints of pain abdomen.

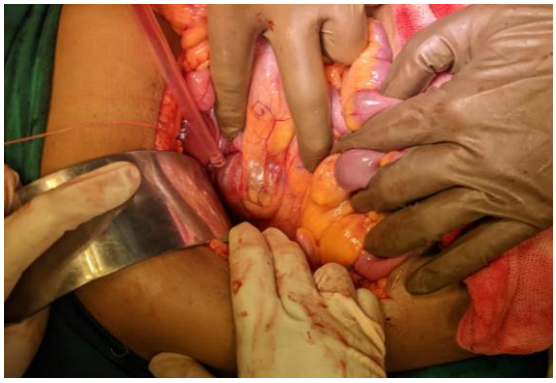


Fig. 3: The suction device in the above figure is pointing towards the mass which is seen to be arising from the retroperitoneum and separate from the right kidney, Inferior vena cava and the bowel wall.

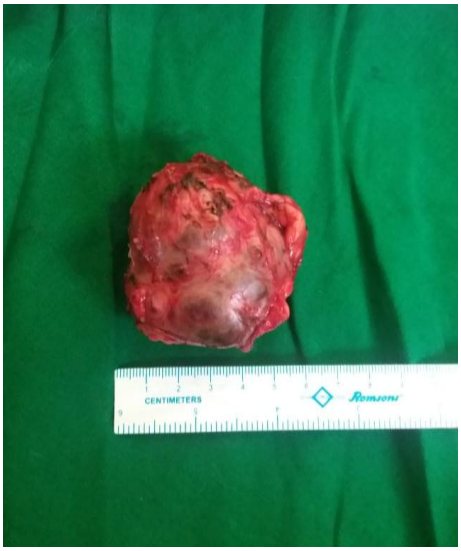


Fig. 4: Excised mass without capsular breach with scale for measure.

DISCUSSION

We report an unusual presentation of paraganglioma symptomatically as well as on imaging modalities. Paragangliomas are highly vascular neuroendocrine tumors that arise from chromaffin cells of the neural crest progenitors located outside of the adrenal gland.Paragangliomas are rare, catecholamine (norepinephrine) secreting neuroendocrine tumors commonly located in pre-aortic and paravertebral sympathetic plexus or skull base. They are usually benign tumors, and a small percentage may become malignant and metastasize. Hence, an early identification leading to complete surgical resection is often curative and carries a favorable prognosis. Detecting distant metastases is the only reliable way to assess the biological aggressiveness of paragangliomas. Surgical biopsy of the lesion is the gold standard to confirm the diagnosis, but it does not differentiate between pheochromocytomas and paragangliomas.

The sympathetic nervous system's chromaffin paraganglia and parasympathetic nervous system are the

sources of paragangliomas (nonchromaffin paraganglia). Chemoreceptors, or parasympathetic paraganglia (chemodecta), are involved in monitoring and controlling blood gas concentrations. The majority of frequent paraganglioma types, such as carotid body paraganglioma, jugulotympanic paraganglioma, and nerve X paraganglioma, originate in these structures. In contrast to only 5% of individuals with paragangliomas of the carotid body, malignant malignancy is frequently seen in paragangliomas arising from the vagus nerve. For distant metastases, glomus tumours of nerve X are most frequently to blame.[5]

Along the sympathetic trunk are chromaffin paraganglia. The largest body of this kind, the spindle-shaped organ of Zuckerandl (para-aortic body), is situated on both sides of the aorta at L3 level, where the inner mesenteric artery emerges from the abdominal aorta. After the age of 40, the Zuckerandl organ, which produces catecholamines, begins to weaken physiologically. The adrenal medulla is frequently used as an illustration of a paraganglial body because of its histological nature. Pheochromocytoma may form as a result of a neoplastic process in the adrenal medulla.[6]

On the basis of the history, physical examination, blood and urine tests, and imaging studies, the diagnosis is formed. The levels of catecholamines (adrenaline, noradrenaline, and dopamine) and their metabolites (metanephrine, VMA) are measured in the blood and 24-hour urine. In neither case were catecholamines or VMA screened using laboratory testing. Because the tumour has strong vascularity, tumour biopsies are not necessary.

Given that these cancers frequently exhibit chemo- and radioresistant characteristics, surgical removal of all malignancies involving tissues is the preferred first treatment.It's possible that distant metastasis to organs like the liver, bone, or lymph nodes is the sole sure sign of cancer. Invasion, vascular and/or capsular, confluent or focal necrosis, diffuse growth or huge nests, high cellularity nuclear pleomorphism, and hyperchromasia are some histological characteristics that are indicative of malignancy[7].Malignancy can be predicted by a number of factors, including tumour weight greater than 80 g, high levels of dopamine within the tumour, and tumour size greater than 5 cm (75% predictive) The majority of malignant tumours have a Ki-67 proliferation score more than 6%.

Moreover, the expression of genes associated to the cell cycle, such as P53 and Ki-67, may be beneficial. Our case had a Ki-67 positive of more than 20%. Due to the difficulty in determining malignancy, all paragangliomas should be thought of as possibly malignant. Malignant paraganglioma patients can only be treated surgically. The transabdominal technique is suggested for big, suspicious tumours that are malignant. Since paragangliomas recur more frequently than pheochromocytomas, long-term follow-up is required. Complete preoperative assessment is advised since preoperative diagnosis of renal or adrenal masses might occasionally provide some diagnostic challenges. Measurements of urine catecholamines and blood pressure must be taken frequently as part of the surveillance.

CONCLUSION

Cells from the parasympathetic and sympathetic nervous systems give rise to paraganglioma. It typically appears as a tumour that grows slowly and causes no pain. Peragangliomas can be unilateral or bilateral tumours, benign or cancerous, inherited, or both. They have good blood circulation. Paraganglioma typically develops near the common carotid artery, however it can also develop inside the middle ear or in the belly.

Prognosis of solitary paraganglioma is usually reassuring if the tumor is not located in a sensitive area and can have safe resection without injury to delicate surrounding structures. Malignant tumors are very rare and usually have a poor prognosis. Yearly follow-up serum or urine fractional metanephrines are advisable.

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