

# Nonfunctional Retroperitoneal Paraganglioma: A Histopathological Surprise

N Goel, N Gupta, Hirdaya H Nag, RK Saran, PK Mishra, AK Agarwal

## ABSTRACT

Paragangliomas are the tumors arising from chromaffin tissue which is distributed along the autonomic nervous system. Most of these tumors secrete and store catecholamines. They can be suspected by clinical symptoms and confirmed by biochemical tests. Retroperitoneal paraganglioma mostly arise from adrenal medulla and are also known as adrenal pheochromocytoma. About 10% of abdominal paragangliomas occur at extraadrenal locations and where they are known as extraadrenal paraganglioma or pheochromocytoma (EAP). Like their adrenal counterparts, most EAPs are functional tumors as they secrete and store catecholamines. Very few EAPs do not secrete clinically detectable amount of hormones and present themselves with the pressure symptoms related to their size. These nonfunctional EAPs mimic more commonly occurring tumors of the retroperitoneum and true diagnosis is made only after microscopic examination of the tumor tissue. We had a patient who was suspected to have a pancreatic tumor but found to have a nonfunctional primary EAP completely separate from the pancreas.

**Keywords:** Paraganglioma, Nonfunctional, Retroperitoneal tumor, Extra-adrenal.

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## INTRODUCTION

Extraadrenal paragangliomas (EAPs) are the tumors of the chromaffin cells arising from the sympathetic and parasympathetic nervous system. Most common sites are head, neck, mediastinum and retroperitoneum. EAPs are embryologically related to pheochromocytoma and likewise most of these are functional tumors (functional EAPs). Nonfunctional EAPs are very rare and they can closely mimic other retroperitoneal tumors. Most of the non-functional EAPs are diagnosed after histopathological examination of tumor tissue. We report a case of nonfunctional retroperitoneal paraganglioma mimicking as pancreatic neoplasm which was diagnosed after microscopic examination.

## CASE REPORT

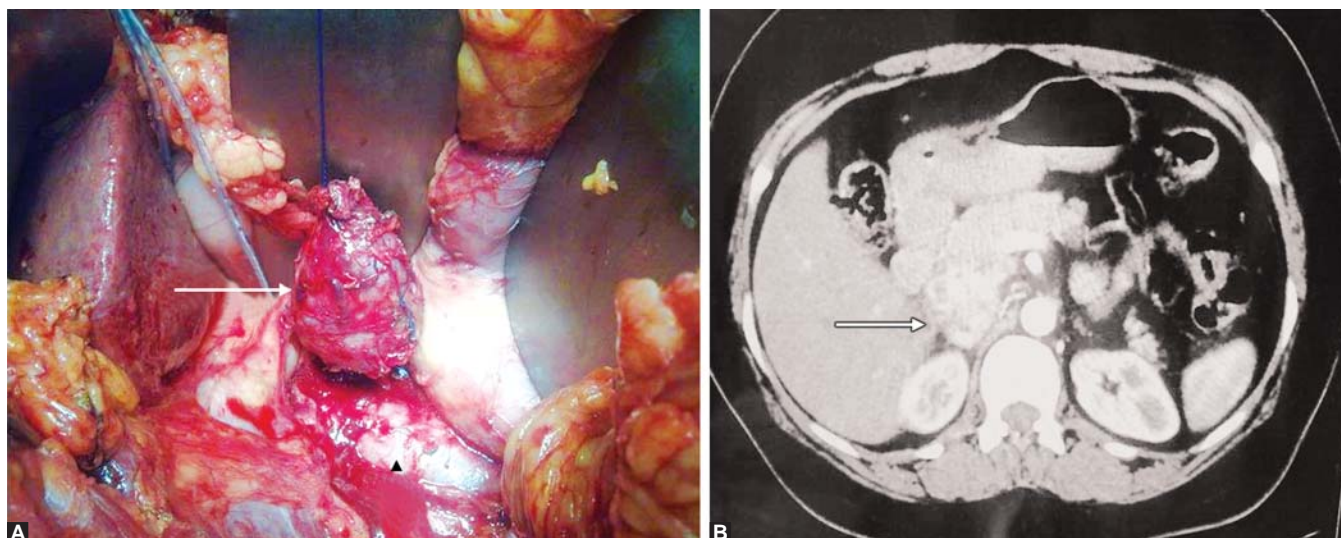
A 50-year-old lady presented with chief complaints of dull aching pain in the central abdomen for last 6 months with

no aggravating or relieving factors. No history of other associated symptoms like fever, jaundice, vomiting, hematemesis and melena. She also gave no history of palpitations, hypertension, headaches or blackouts. Routine hematological and biochemical investigations were within normal limits. Ultrasound examination revealed a 5 × 5 cm tumor in the head of the pancreas with splaying of the superior mesenteric vein. Contrast enhanced computed tomography (CT) of the abdomen revealed lesion in the head and uncinate process of the pancreas which was showing a very good contrast enhancement in the arterial phase and washout in the venous phase. Patient had high serum chromogranin level (6361 IU/ml). Serum gastrin and 48 hours fasting blood sugar levels were within normal range and her gastroduodenoscopy did not reveal any abnormality. Patient was planned for pancreaticoduodenectomy with provisional diagnosis of pancreatic neuroendocrine tumor (nonfunctional). On laparotomy there was a 5 × 5 cm size tumor in the aortocaval groove with well-preserved fat planes with the pancreas. The tumor was densely adherent to the pericaval fascia and could be excised with preservation of the pancreas and adrenal gland (Figs 1A and B), no hemodynamic fluctuations occurred during surgery.

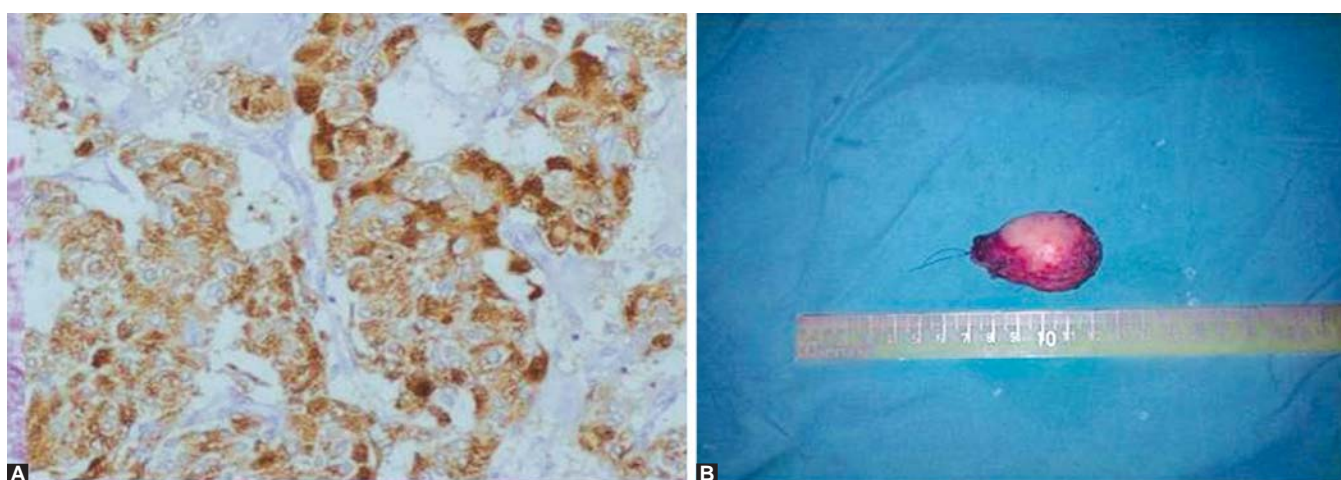
On histological examination, tumor showed cells with round to oval prominent nuclei and abundant granular amphophilic cytoplasm, arranged in small nests (Zell ballen bodies) and on immunostaining tumor cells stained positive for chromogranin (Figs 2A and B), so final diagnosis of nonfunctional primary paraganglioma was made.

## DISCUSSION

Retroperitoneal EAPs are rare neurogenic tumors, arising from sympathetic paraganglia which are distributed along the major blood vessels of the retroperitoneum.<sup>1</sup> These tumors can be confused with other relatively common retroperitoneal neoplasms and it may not be possible to diagnose them without microscopic examination of tumor specially when they are of nonfunctional in nature (endocrine symptoms absent). In a review from John Hopkins institute,<sup>2</sup> out of total nine patients, six were misdiagnosed and treated as pancreatic neoplasms and true



**Figs 1A and B:** (A) Intraoperative photograph showing tumor (arrow) and IVC (arrowhead); (B) CT image showing enhancing tumor on arterial phase in retropancreatic location



**Figs 2A and B:** (A) The immunohistochemical staining for chromogranin; (B) The gross tumor after excision

diagnosis could be made only after histopathological examination. Like other neuroendocrine tumors, EAPs also show a good contrast enhancement and early venous washout on contrast-enhanced CT of the abdomen.<sup>3,4</sup> In most of the cases with functional EAPs it may be possible to suspect them preoperatively due to presence of endocrine symptoms like episodic headache, sweating and hypertension but in cases where these symptoms are absent (nonfunctional) these tumors are usually confused with other tumor of neuroendocrine and nonneuroendocrine nature arising from a retroperitoneal organs like pancreas and true diagnosis is usually made at histology, as was the scenario with our case.<sup>3-5</sup>

Serum levels of chromogranin A, neuron-specific enolase (NSE) or vimentin are elevated with most of the neuroendocrine tumors (NETs) and are helpful to differentiate them from non-NETs. On the basis of endocrine symptoms, NETs can be differentiated by using

hormonal assay in blood or urine (catecholamines, metanephrines, gastrin, somatostatin, etc.) but in cases (like ours) where these symptoms are absent it is not worthy to exhaust battery of all such investigation as it is not cost-effective and it will add very little to the surgical management planning of the patient, it is for this reason we decided not to proceed with these investigations. In cases with nonfunctional solitary NET of retroperitoneum without any detectable metastasis on CT of abdomen, one should go ahead with surgical excision although some of these case may turn out to be functional on operating table so, necessary planning should be done by surgical and anesthetic team and every effort should be made to detect other primary or secondary neoplasm in case tumor is found to be extrapancreatic during operation although we could not find any other tumor. MIBG or FDG PET scan can be done to know the overall spread of the disease in cases with functional paraganglioma but they

are not as useful in cases with nonfunctional tumors.<sup>6,7</sup> About 60 to 70% of these tumors (EAPs) are benign and it is not always possible to separate benign from malignant lesion as metastasis can occur very remotely after the excision of otherwise benign tumor; so, these patients should be followed of for a long period.<sup>7,8</sup>

Microscopically, cellular arrangement in close groups (nests) with larger nuclei and granular cytoplasm is characteristic of paraganglioma and it can further be confirmed by immunohistochemical staining.<sup>2-5</sup>

Treatment of these tumors is surgical excision, which is the best possible modality to achieve complete cure but it may not be possible in cases with extensive local disease, tumor involving vital organs or in cases with distant metastasis at presentation.<sup>1,2,3,8</sup> In such cases other modalities like octreotide therapy, radiotherapy or chemotherapy should be used and they may provide a good control of the disease.<sup>8,9</sup> Prognosis of these tumors is usually good.

## CONCLUSION

In all cases where imaging reveals a highly vascular retroperitoneal tumor in and around pancreas, nonfunctional paraganglioma (EAP) is a real possibility but confirmation is usually possible after microscopic examination.

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