Case Report On Paraganglioma Duodenale

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Abstract

Paragangliomas are rare tumors that arise from extraadrenal chromaffincells. During embryogenesis, neural crest cells migrate to diverse locations in the body and differentiate into chief and sustentacular cells, which are designated as paraganglia. Presenting a case of paraganglioma who came to our hospital. Preoperatively was diagnosed as GIST but intraop was absolutely different.

Introduction

Extraluminal perigastric masses are frequently considered to be gastrointestinal stromal tumors (GISTs) or other soft tissue sarcomas, when histological confirmation is not possible. Paragangliomas may have a similar radiographic appearance. In the absence of histological diagnosis and symptoms of catecholamine excess, paragangliomas may be mistaken for GISTs. Perigastric abdominal mass presumed to be a GIST after nondiagnostic biopsy, is presented [1,2]. On abdominal CT, there are no unique imaging characteristics specific for paragangliomas [3]. Consequently, these tumors may be mistaken for other primary epithelial or mesenchymal abdominal tumors [3]. For instance, GIST, the most common sarcoma of the gastrointestinal tract, may have a similar radiographic appearance [3,4]. They are often large (84% N 5 cm) tumors arising from the stomach with heterogeneous enhancement (84%) and central necrosis (37%) [5]. While size and location help to differentiate GISTs from paragangliomas, correlation with clinical symptoms is needed. A presumed GIST considered to be resectable does not necessarily warrant preoperative biopsy, and thus further diagnostic workup is not pursued [8].

Case Report(s)

A 48 yr normotensive male came to us with the complaints of pain in the upper abdomen from the last 2 months which was dull aching epigastric non-radiating pricking type of pain no aggravating and reliving factor not associated any other GI complaints. Previous history of gastro jejunostomy 12 yrs back. His general examination and systemic examination was normal except old healed upper midline scar. His blood investigation was within normal limits. Sonographic study suggestive of well defined heterogenous mass lesion 5X4cm in supraumbilical region - ? GIST. His CECT was done and reported to be same.

**Fig 1:** CT picture showing tumor

Patient was planned for laparotomy for excision of tumor. Intraoperative tumor was found in retroduodenal retro pancreatic region in between aorta and duodenum around 5×4 cms. It was seen that with handling of tumor there was sudden rise of blood pressure to 240/120 and an episode of ventricular tachycardia. Following this the vein draining it was ligated first and then the tumor was excised carefully without damaging major vessel.

**Fig 2:** intra op picture showing tumor

**Fig 3:** intra op picture showing tumor

**Fig 5:** intra op picture showing tumor after clamping the vein draining tumor

**Fig 6:** resected specimen cut section

The specimen was sent for histopathological and diagnosed as paraganglioma.

**Fig 7:** microscopic picture

**Fig 8:** microscopic picture

Post operatively patient developed hypotension and was started on vasopressor which was stopped after his BP started rising finally he recovered of his crisis after 3 days and was discharged on 10th post op day.

**Fig 9:** post op picture after removal of sutures

Discussion

Paragangliomas often present with signs of catecholamine excess. The most common catecholamine secreted is norepinephrine, and a classic triad of catecholamine excess (headache, sweating, palpitations) is described [6,7]. However, this triad may be absent, and patients can be asymptomatic or symptoms can be vague (psychiatric disorders, anxiety, facial pallor, weight loss, polyuria/polydipsia, hyperglycemia, secondary erythrocytosis, stroke, and cardiomyopathy) [6]. Paraganglioma was not suspected and preoperative catecholamine levels were not measured. The location of the tumor adjacent to the stomach, the large tumor size, and the absence of classic signs of catecholamine secretion led to the presumed diagnosis of GIST or other sarcoma. A
presumed GIST considered to be resectable does not necessarily warrant preoperative biopsy, and thus further diagnostic workup was not pursued [8].

Sites of paraganglioma

Fig10: showing sites for extra adrenergic pheochromocytoma

Paragangliomas synthesize and store catecholamines, which include norepinephrine (noradrenaline), epinephrine (adrenaline), and dopamine. Elevated plasma and urinary levels of catecholamines and the methylated metabolites, metanephrines, are the cornerstone for the diagnosis. The hormonal activity of tumors fluctuates, resulting in considerable variation in serial catecholamine measurements. Thus, there is some value in obtaining tests during or soon after a symptomatic crisis. On the other hand, most tumors continuously leak O-methylated metabolites, which are detected by metanephrine measurements[9].

Adrenergic blockers (phenoxybenzamine) should be initiated at relatively low doses (e.g., 5–10 mg orally three times per day) and increased as tolerated every few days. Because patients are volume constriicted, liberal salt intake and hydration are necessary to avoid orthostasis. Adequate alpha blockade generally requires 10–14 days, with a typical final dose of 20–30 mg phenoxybenzamine three times per day. Oral prazosin or intravenous phentolamine can be used to manage paroxysms while awaiting adequate alpha blockade. Before surgery, the blood pressure should be consistently below 160/90 mmHg, with moderate orthostasis. Beta blockers (e.g., 10 mg propranolol three to four times per day) can be added after starting alpha blockers, and increased as needed, if tachycardia persists. Because beta blockers can induce a paradoxical increase in blood pressure in the absence of alpha blockade, they should be administered only after effective alpha blockade. Other antihypertensives, such as calcium-channel blockers or angiotensin-converting enzyme inhibitors, have also been used when blood pressure is difficult to control with phenoxybenzamine alone[9]. Finally main stray of treatment is surgical removal or catecholamines in stress-induced cardiomyopathy or Takotsubo cardiomyopathy. Int J Cardiol 2007;114:e15-7.


References


Illustrations

Illustration 1

pictures and figures

Fig 1:- CT picture showing tumor

Fig 2:- intra op picture showing tumor

Fig 3:- intra op picture showing tumor

Fig 5:- intra op picture showing tumor after clamping the vein draining tumor
Fig6:- resected specimen cut section

Fig7:- microscopic picture

Fig8:- microscopic picture
Fig 9: Post op picture after removal of sutures.

Fig 10: Showing sites for extra adrenergic pheochromocytoma.
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