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Primary Gallbladder Paraganglioma: A Case Report and Review of Literature



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Abstract

BACKGROUND: Paragangliomas are tumors arising from paraganglia of the autonomic nervous system. They are rare tumors and occurrence inside the gallbladder is exceptionally rare. Biliary paragangliomas are thought to be associated with the parasympathetic fibers and are therefore non-functioning and benign. There are less than 10 cases reported in literature and majority are found incidentally upon cholecystectomy. There is no specific treatment for these tumors and resection is considered sufficient.

CASE PRESENTATION: 63 year old female presented with recurrent biliary colic exacerbated by fatty food. She underwent imaging work up that was consistent with biliary dyskinesia. She underwent uneventful elective laparoscopic cholecystectomy and was doing well post-operatively. Pathology report was significant for chronic cholecystitis, no calculi, and a small focus of paraganglioma.

CONCLUSION: We presented a case in which a primary gallbladder paraganglioma was identified incidentally in a patient who presented with symptomatic biliary dyskinesia. Due to the non-functioning nature of these tumors there are felt to be benign. We know the paraganglia of the gallbladder consists of both parasympathetic and sympathetic fibers. The sympathetic paragangliomas tend to act similar to pheochromocytomas and thus have malignant potential. Due to the rarity of primary gallbladder paraganglioma and limited reported cases, we need more information to determine if further work up and treatment is necessary in such cases.

Background

The paraganglia are neural crest cells that have differentiated into sustentacular and chief cells after their migration during embryogenesis. As a whole, they form what is known as the paraganglionic system, which includes the adrenal medulla and groups of extra-adrenal neuroendocrine cells. The extra-adrenal locations are in close association with the autonomic nervous system. They are primarily located in or near mid-line of the body. Most well-known of these locations include the carotid bodies in the neck and the organs of Zuckerkandl at the bifurcation of the aorta (1,2).

When tumors arise from these neuroendocrine cells, we must differentiate between locations for proper identification. If arising from extra-adrenal locations they are classified as paragangliomas, whereas if originating from within the adrenal medulla, we classify these as pheochromocytomas. Paragangliomas can further be divided based on whether they arise from the sympathetic or parasympathetic ganglia. (3) Sympathetic paragangliomas tend to resemble pheochromocytomas more closely in regards to symptomology and histologic features of "chromaffin reaction". This reaction is based on the brown discoloration of what we know as "chromaffin cells" in the presence of chromate salts. (3) Due to the similarities of sympathetic paragangliomas and pheochromocytomas, their approach in management is the same. This management consists of a biochemical workup along with imaging localization prior to surgical resection. (6)

Paragangliomas are extremely rare tumors with a prevalence estimated around 1:4,500 in the United States. Most commonly found in the fourth to fifth decades of life without a sex predilection. However, parasympathetic paragangliomas are seen more frequently in women. (3) They can occur sporadically or within a hereditary syndrome. If seen in a hereditary fashion, they do tend to appear at an earlier age and are more frequently multicentric. With children, although very rare, you will tend to find paragangliomas that are of the sympathetic variation, multiple, and in association with hereditary syndromes. (3)

Even rarer are the biliary paragangliomas. To date, there are less than 10 reported cases of gallbladder paragangliomas. The majority of these tumors were found incidentally after cholecystectomy. The patients tended to present with symptoms of biliary colic. None of the patients exhibited symptoms of sympathetic system involvement or had a personal history of other endocrine tumors or syndromes. They were predominately found in females in the fifth to sixth decade of life. (1, 4)

Case Presentation

63 year old Caucasian female with past medical history of hypothyroidism presented to the Emergency Department (ED) with right upper quadrant (RUQ) abdominal pain with radiation to right scapula with associated bloating and nausea. Her CT Abdomen revealed a mildly dilated gallbladder. Her blood work was notable for mild hyponatremia and hypokalemia but otherwise unremarkable with normal liver function tests. Her symptoms resolved while in ED and was discharged with recommendation to change diet as well as follow up with Primary Care Physician (PCP) if symptoms persisted.

Over the next couple months, after changing her diet to eating only homemade soups and low fat food, she did notice return of symptoms while eating high fat foods on two separate occasions. Due to recurrent symptoms, a hepatobiliary iminodiacetic acid (HIDA) scan was performed revealing biliary dyskinesia with gallbladder ejection fraction of 4%. She was then referred to general surgery clinic for evaluation. She had lost 13 pounds over the past few months due to change in diet but was otherwise healthy and without complaints beyond symptomatic biliary dyskinesia.

It was recommended she undergo laparoscopic cholecystectomy had an uneventful successful elective surgery. There were no gross abnormalities intra-operatively except chronic cholecystitis. Her pathology returned revealing chronic cholecystitis (Figure 1), no calculi, one benign lymph node, and a subcentimeter focus of paraganglioma at the fundus/body of gallbladder. Paraganglioma was described as organoid, nested island of chief cells with inconspicuous sustentacular cells and capillary network at periphery in the subserosal adipose tissue without mitotic figures identified (Figures 2,3). Immunohistochemical staining was performed on the tumor cells resulting in Synaptophysin positive (Figure 4), Chromogranin positive, and S100 highlights surrounding sustentacular cells.

She was seen a few weeks post-operatively at her follow up visit and was doing well.

Photos

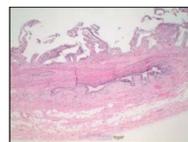


Figure 1. (40x) Representative gallbladder with changes of chronic cholecystitis, including areas of benign-appearing mucosa that are dilated and extend into the muscularis, consistent with Rokitsansky-Aschoff sinuses.

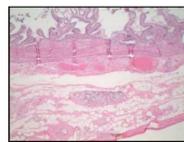


Figure 2. (40x) Gallbladder with subserosal paraganglioma.

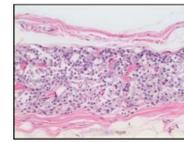


Figure 3. (200x) Paraganglioma showing an organoid, nested island of chief cells with inconspicuous sustentacular cells and capillary network at periphery in the subserosal adipose tissue of the fundus/body of the gallbladder. There are no mitotic figures identified.

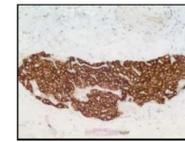


Figure 4. (200x) Positive labeling with Synaptophysin immunohistochemistry

Discussion

There is little known about primary gallbladder paragangliomas. It is believed that they originate from the migration of the paraganglia of the hepatic plexus which in turn innervates the gallbladder via cystic plexus. (6) The cystic plexus contains both parasympathetic and sympathetic fibers. These primary tumors appear to be non-functioning parasympathetic paragangliomas. They do not act similar to pheochromocytomas or sympathetic paragangliomas and therefore are benign tumors. In such, an incidental finding upon cholecystectomy would be adequate treatment. In contrast, sympathetic paragangliomas have a higher risk of a malignant component and is therefore treated similar to pheochromocytomas with resection after staging workup. As these have typically presented with biliary colic and should be in the differential for a gallbladder lesion, one should investigate the potential association with hereditary syndromes if multiple paragangliomas are present.

Conclusion

We reported here a case of primary gallbladder paraganglioma found incidentally after laparoscopic cholecystectomy for recurrent biliary colic and biliary dyskinesia. With only limited published articles regarding similar cases, we currently conclude that these tumors are parasympathetic, benign, and adequately treated with cholecystectomy. We know based on neuroanatomy that the innervation of the gallbladder has both sympathetic and parasympathetic fibers. This raises the question of the possibility of a malignant paraganglioma but has not been reported in literature.

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