Mediastinal Paraganglioma: EUS Findings and a Cautious Reminder of Potential Risks from FNA of Unknown Mediastinal Masses

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Case Report:

A 49-year-old man presented with a one-year history of progressive symptoms of intermittent proximal dysphagia for solids. CT revealed a ~7cm superior mediastinal mass which appeared highly vascular in nature (Figure 1). EGD showed extra-luminal compression of the proximal esophagus and the intraluminal presence of large proximal esophageal varices (Figure 2 and 3). EUS revealed a solid complex heterogenous hypervascular mediastinal mass (Figure 4 and 5) with diffuse vascular component by Doppler and confirmed the presence of varices. FNA was deferred in favor of the ‘safer’ operative approach because of the presence of varices (Figure 6) within the projected needle tract and the highly vascular nature of the lesion. Transcervical open wedge biopsy was performed which was notable for copious bleeding. Intraoperative histologic evaluation favored a neuroendocrine neoplasm; however subsequent immunohistochemistry confirmed paraganglioma (PG) with S-100 positive sustentacular cells outlining nests of cytokeratin negative neuroendocrine cells (Figure 7). The tumor was fortunately non-functional without any complications related to catecholamine release. The patient underwent subsequent successful surgical resection, which was preceded by pre-operative embolization of the feeding vessels. To our knowledge this is the first article to report the EUS findings of a mediastinal PG.
Discussion

Paraganglioma are rare, highly vascular extra-adrenal catecholamine secreting tumors originating from chromaffin cells anywhere along the sympathetic ganglia chain.\(^1\)\(^2\) Most occur sporadically in the 3rd to 5th decade.\(^2\) Although usually intra-abdominal, up to 15% may occur in the intrathoracic region as seen here.\(^2\) These tumors are usually solitary and grow slowly to reach several centimeters in size where they may cause symptoms from mass effect.\(^2\) EUS features of PG have not been well described.\(^1\)\(^3\) Akdamar et al. reported the first case of a retroperitoneal PG in which a 6.6cm paraduodenal cystic like mass was
Unlike the previous case, a predominantly solid lesion was encountered which by Doppler did have a diffuse vascular component. These lesions are usually solid but can undergo cystic degeneration. This lesion was associated with superior vena caval obstruction as manifested by “downhill” varices. Four to five percent are functional and cause symptoms of excess catecholamine secretion particularly after manipulation.

The diagnosis of PG is unlikely to be expected at the time of EUS. FNA of unknown mediastinal lesions is generally effective and safe. However diagnosis of mediastinal masses can be challenging, as cytologic findings are not infrequently undifferentiated and overlapping. In the case of PG, immunohistochemical analysis is needed to make a diagnosis. Further FNA of these lesions may result in catecholamine release with precipitation of life-threatening hypertensive crisis and is therefore contraindicated if suspected without pharmacologic prophylaxis. Pre and intra-operative preparation with alpha-adrenergic blockade (ie. phenoxybenzamine or phentolamine) is most commonly recommended. Hypertensive crisis with associated life threatening hemodynamic instability otherwise is best managed with urgent intervention by an anesthesiologist or cardiologist and intensive care monitoring. When the diagnosis of a catecholamine secreting tumor is suspected by clinical history or imaging, pre-operative evaluation including 24 hour urine collection for catecholamines and metanephrines is recommended along with expert consultation. Management is surgical and frequently challenging due to the vascular nature of lesion. Preoperative embolization of feeding vessels may be performed to minimize the risk of intraoperative bleeding.

Despite its rarity, PG needs to be considered when presented with any unknown vascular mediastinal lesion because it is one of the few exceptions when FNA may pose a serious and unexpected risk. Unfortunately given its non-specific appearance, a catecholamine response to FNA may be the first clue.

References:


9. Bruynzeel H, Feelders RA, Groenland TH, van den Meiracker AH, van Eijck CH, Lange JF, de Herder...


