Paraganglioma of the Mesenterium: a Case Report

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Summary

Extra-adrenal paragangliomas constitute 10 % or less of phaeochromocytomas/ paragangliomas. Even rarer is the occurrence of paragangliomas outside the usual distribution of paraganglionic tissue. We report a case of extra-adrenal paraganglioma occurring in the small intestine mesentery in a 65-year-old man. To our knowledge, there are only seven case reports of paraganglioma occurring in this non-typical site. Computed tomography showed a solid expansive non-homogenously enhancing mesenteric mass, measuring 10 x 8 cm with peripheral cystic component. Histologically, the tumour had a typical organoid "zellballen" pattern, showed immunohistochemical positivity for synaptophysin, neuron specific enolase, CD-56, chromogranin, and focally vimentin, and was cytokeratin and EMA negative. S-100 protein stained few sustentacular cells. The patient was free from recurrence or metastasis three months after tumour resection. Although rare, paraganglioma should be included in the preoperative differential diagnosis of solid mesenteric tumours, to prevent any potential life-threatening event peroperatively in the case of a catecholamines-producing tumour.

Key words: extra-adrenal paraganglioma – mesentery – phaeochromocytoma

Súhrn
Paraganglióm mezentéria: kazuistika

Extraadrenálne paragangliómy tvoria do 10 % feochromocytómov/paragangliómov. Ešte vzácnejší je výskyt paragangliómov mimo zvyčajnej distribúcie paraganglií. Popisujeme prípad extraadrenálneho paragangliómu mezentéria tenkého čreva u 65 ročného muža. Podľa našich vedomostí, existuje iba sedem popísaných prípadov výskytu paragangliómu v tejto atypickej lokalizácii. Vyšetrenie počítačovou tomografiou zobrazilo solídny expanzívny kontrastom nehomogénne zvýraznený nádor veľkosti 10 x 8 cm. Nádor bol periférne cysticky zmenený. Histologicky mal nádor typickú organoidnú „zellballen" architektoniku, bol imunohistochemicky synaptofyzín, neurón špecifická enoláza, CD-56, chromogranín a fokálne vimentín pozitívny. Cytokeratín a EMA boli negatívne. S-100 proteín zobrazil sporadické sustentakulárne bunky. Pacient bol 3 mesiace po operácii bez známok recidívy alebo metastáz. Napriek tomu, že je výskyt paragangliómu v mezentériu vzácny, mal by byť zahrnutý do predoperačnej diferenciálnej diagnózy, aby sa predišlo potencionálnej život ohrozujúcej príhode počas operácie tumoru produkujúceho katecholamíny.

Kľúčové slová: extraadrenálny paraganglióm – mezentérium – feochromocytóm

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Extra-adrenal paragangliomas constitute 10 % or less of total phaechromocytomas/paragangliomas (6). Knowledge of the distribution of normal paraganglionic tissue is important to a pathologist because of its value in predicting the sites of origin of paragangliomas. These tumours have been reported virtually at all locations where normal paraganglia are found during fetal and adult life and tend to be most frequent in areas where paraganglionic tissue is most abundant. However, paraganglia may occur in locations outside the well-established sympathetic and parasympathetic distributions (11). We describe a case of solitary primary paraganglioma of the mesenterium, incidentally found in a 65-year-old man. To our knowledge, this case represents only the eighth reported case of mesenteric paraganglioma.