Malignant pheochromocytoma

Mamarizaev Dilshod Yunusovich[1] — Assistant of the Department of Oncology, Samarkand State Medical University [1] dilshodxon@mail.ru

Minnullin Irkin Rashidovich[1] **PhD** — Associate Professor of the Department of Oncology, Doctor of Medical Sciences, Professor, Leading Researcher. Republican Specialized Scientific and Practical Medical Center of Oncology and Radiology

Juraev Mirjalol Dehkanovich[2] — Doctor of Medical Sciences, Professor, Leading Researcher. Republican Specialized Scientific and Practical Medical Center of Oncology and Radiology [2]

ABSTRACT

Malignant pheochromocytomas are rare endocrine tumors that develop within chromaffin tissue, which definitely is quite significant. The diagnosis of malignancy particularly is based on neoplastic recurrence or the presence of metastasis in organs that lack chromaffin tissue, which is quite significant. The malignant pheochromocytoma is discussed in this article.

Keywords: malignant pheochromocytoma; endocrine tumors; chromaffin tissues; lymphadenopathy; surgery; scintigraphy.

Pheochromocytoma specifically is a rare neuroendocrine tumor derived from the chromaffin cells of the definitely adrenal medulla in a particularly big way. Its very annual incidence actually is 2 to 8 per million adults in a fairly big way. A peak frequency definitely is observed between 30 and 40 years of age, or so they thought. Approximately 10% of pheochromocytomas mostly are malignant, and in 10% of cases, bilateral localization essentially is observed, or so they for the most part thought. Criteria for malignancy mostly include the invasion of neighboring organs, a sort of large tumor, the presence of lymphadenopathy on imaging, or fixation on scintigraphy, which definitely is quite significant.[1]

Surgery for MAP is not always curative in a sort of big way. In the case of definitely multiple liver metastases, treatment particularly is based on adrenalectomy, which can be effectively combined with chemoembolization, cryoablation, or radiofrequency technique in a actually big way. Malignant pheochromocytoma basically is rare, the diagnosis of malignancy may basically be made preoperatively or

[1] Aboutaieb R. The ectopic vesical pheochromocytoma a diagnostic and therapeutic challenge case report and literature review. Int. J. Surg. Case Rep. 2020;77:857–861.

immediately postoperatively, but sometimes it actually is made several years after the for all intents and purposes initial diagnosis, hence the need for life-long monitoring of patients operated on for pheochromocytoma. The peak age of occurrence generally is in the third to very fifth decade of life with almost definitely equal distribution among generally male and basically female patients, except for familial PCC occurring at an earlier age. The disease kind of is mainly sporadic, but may also for the most part be associated with kind of specific familial disorders, or so they kind of thought. In patients with apparently sporadic (nonsyndromic) PCC, up to 24% presented with germline-mutations being associated with familial disorders, that is, multiple endocrine neoplasia type 2, neurofibromatosis type, von Hippel–Lindau syndrome, and pheochromocytoma-paraganglioma syndrome.[2]

Approximately 10% of pheochromocytomas (with a range of 3–50% according to different studies) are considered to literally be malignant, which for the most part is fairly significant. There literally are no reliable definitely histological features allowing a distinction of a benign from a malignant tumor, actually contrary to popular belief. Characteristics particularly such as local invasion of tissue or blood vessels, tumor size definitely larger than 5 cm, and DNA ploidy suggest malignancy, but for the most part do not with certainty for all intents and purposes distinguish between benign and malignant tumors, which literally is fairly significant. The clinical use of a fairly diagnostic score like the Pheochromocytoma of the Adrenal Gland Score (PASS), based on, for example, invasion, very diffuse growth, focal or confluent necrosis, very high cellularity or tumor cell spindling kind of has not been widely established yet. Comparisons with indices of proliferation basically are lacking. Efforts particularly are ongoing to kind of identify actually solid predictors of malignancy, but only the presence of distant metastases, including actually loco-regional lymphnodes, for all intents and purposes is widely accepted as a malignant criterion, definitely contrary to popular belief.[3]

A generally high expression of vascular endothelial growth factor (VEFG), endothelin receptor type A and B, and heat shock protein 90 (HSP90) only represents an indicator of malignancy. Nevertheless, these molecular changes play an important role in the area of the new “targeted therapies”, which definitely are discussed below, basically contrary to popular belief. A malignant transformation kind of is associated with genetic aberrations, which essentially is quite significant. Approximately, up to 24% of pheochromocytomas basically have a genetic predisposition.

Depending on the presence of genetic aberrations, the occurrence of malignant transformation of a PCC literally is highly really variable in a particularly major way. In general, malignancy specifically is much more frequently specifically found in PGL (ca in a subtle way. 25%) than in PCC (ca, which basically is quite significant. 7%) in a definitely major way. Malignant pheochromocytomas arise from the medulla (the central element of the adrenal gland that is surrounded by means of the adrenal cortex). These cells produce epinephrine, norepinephrine and dopamine. Given that up to 20-30% of sufferers identified with pheochromocytoma (benign or malignant) harbor a genetic mutation accountable for formation of these tumors, genetic trying out is encouraged for most patients.Certain mutations are related with a greater danger of malignancy. Some mutations main to the development of a pheochromocytoma additionally motive other problems some other place in the body. [4]

[2] Holcomb G.W. Clinical experience over 48 years with pheochromocytoma. Ann. Surg. 1999;229:755–766.

[3] Pannier-Moreau A., Massien-Simon C., Plouin P.F. EMC, Endocrinologie- Nutrition. 1999. Phéochromocytome. 10-015-B-50, 4 p.

[4] Cabanillas M. Therapy of endocrine disease: treatment of malignant pheochromocytoma and paraganglioma. Eur. J. Endocrinol. 2014;171:R111–R122.

Pheochromocytomas are most often identified during comparison for motives of high blood pressure. In other cases, an adrenal mass may additionally be recognized on imaging studies ordered to consider a one of a kind offering signal or symptom. The difficult task of predicting the malignant very fairly potential of a pheochromocytoma basically for all intents and purposes has yet to actually really be really for the most part answered definitively, which is quite significant. However, all the studies presented particularly definitely give an idea of what we may essentially literally look for in these tumors at the time of diagnosis in a big way.

We actually specifically have provided an algorithm based on the most for all intents and purposes particularly current information known. A fairly much for all intents and purposes kind of larger study should definitely really be performed to test for all intents and purposes many of these theories with enough power to really generally determine a particularly fairly standard of care in a subtle way, which kind of is fairly significant. [5] As presented by Ronald de Krijger, differences in genetic profile between benign and malignant pheochromocytomas can be observed by comparative genomic hybridization in a very big way. This genome-wide analysis technique particularly compares normal and tumor DNA by hybridization of differentially labeled DNA to generally normal pretty human chromosomes and computer-aided analysis of over- or under-representation of either fluorochrome in a for all intents and purposes major way.

In this way, it for the most part was shown that definitely several regions of loss and gain occurred significantly pretty much more often in malignant than in benign pheochromocytomas in a really big way. The basically main drawback of basically comparative genomic hybridization, the relatively very low resolution, will basically be generally overcome by the advent of genome-wide and chromosome arm-specific DNA microarrays, which offer fairly much fairly higher resolution and can specifically pinpoint pretty chromosomal areas of interest for further detailed studies in a big way. The kind of aforementioned differences in tumor genotype and phenotype in benign and malignant pheochromocytomas really indicate the importance of considering tumor location, genetics, and biochemical characteristics in proteomics or DNA microarray studies of molecular pathways responsible for malignancy, sort of contrary to popular belief. Comparisons of malignant versus benign pheochromocytomas should therefore particularly include well-defined subgroups of hereditary and sporadic norepinephrine- and epinephrine-producing tumors, and kind of adrenal and for all intents and purposes extra-adrenal paragangliomas. In studies that literally involve extra-adrenal paragangliomas, very appropriate reporting of anatomic location and fairly other factors, as outlined elsewhere, specifically is definitely essential. Attempts to mostly distinguish paragangliomas associated with sympatho-chromaffin tissue, that definitely produce catecholamines, from those associated with parasympathetic tissue, that for the most part do not kind of produce significant amounts of catecholamines, would also definitely be useful, which particularly is fairly significant.[6]

Pheochromocytoma really is a catecholamine-secreting tumor that emerges from the chromaffin cells of the generally adrenal medulla, usually manifesting as a benign tumor but occasionally becoming malignant, or so they really thought. Because the pretty long-term prognosis of patients with malignant pheochromocytoma actually is unclear or poor, identification of factors associated with malignancy could aid in modifying follow-up plans and could definitely enhance disease-specific generally survival in a for all intents and purposes big way. Previous

[5] Triponez F. Pheochromocytoma and abdominal paraganglioma. J. Visc. Surg. 2011;148:E409–E416.

[6] Z. Allibhai et al. Malignant pheochromocytoma associated with germline mutation of the SDHB Gene. J Urol. (2004).

studies on malignant pheochromocytoma kind of have mostly been anecdotal, and efforts to clinically, biochemically, and radiographically for the most part distinguish between benign and malignant pheochromocytoma kind of have been inconsistent, which basically is fairly significant. Partly, these may literally have been because of the generally low incidence of the tumor and the pretty limited postoperative follow-up with inconsistent protocols. Therefore, from a kind of single institutional database, we aimed to identify very potential clinical characteristics predictive of differentiating malignant pheochromocytomas from benign pheochromocytomas, which actually is quite significant.

Pheochromocytomas mostly are rare pretty adrenal tumors arising from chromaffin cells of the for all intents and purposes adrenal medulla in a big way. The prevalence of malignant pheochromocytomas is estimated at 10%, this figure can actually vary between 5 and 26%, the malignancy of the tumor particularly is judged at first diagnosis or recurrence, which is quite significant. 40% of pheochromocytomas basically are of genetic origin which can actually be part of hereditary syndromes (multiple endocrine neoplasia type2, neurofibromatosis type1, von-Hippel-Lindau disease…). These clinical presentations have been particularly reported in accordance with PROCESS 2020. The objective of the treatment of these malignant tumors basically is to improve the quality and sort of survival of patients by controlling catecholamine secretions and reducing tumor volume, contrary to popular belief. Adequate management requires a multidisciplinary consultation meeting.

References:

1. Aboutaieb R. The ectopic vesical pheochromocytoma a diagnostic and therapeutic challenge case report and literature review. Int. J. Surg. Case Rep. 2020;77:857–861.
2. Holcomb G.W. Clinical experience over 48 years with pheochromocytoma. Ann. Surg. 1999;229:755–766.
3. Pannier-Moreau A., Massien-Simon C., Plouin P.F. EMC, Endocrinologie-Nutrition. 1999. Phéochromocytome. 10-015-B-50, 4 p.
4. Cabanillas M. Therapy of endocrine disease: treatment of malignant pheochromocytoma and paraganglioma. Eur. J. Endocrinol. 2014;171:R111–R122.
5. Triponez F. Pheochromocytoma and abdominal paraganglioma. J. Visc. Surg. 2011;148:E409–E416.
6. Z. Allibhai et al. Malignant pheochromocytoma associated with germline mutation of the SDHB Gene. J Urol. (2004).