Langenbecks Arch Surg (2012) 397:145–146
DOI 10.1007/s00423-011-0893-5

EDITORIAL

Positional statement of the European Society of Endocrine Surgeons (ESES) on malignant adrenal tumors

Jean-François Henry · Jean-Louis Peix · Jean-Louis Kraimps

Published online: 29 December 2011

Malignant adrenal tumors are very rare, and in addition, the criteria determining whether an adrenal neoplasm is benign or malignant are not precise. This explains that it is difficult to give clear recommendations, and in many cases, it is easier to recommend what should not be done than to recommend what should be done.

The European Society of Endocrine Surgeons organized in Lyon (May 13–14, 2011) a workshop on malignant or potentially malignant adrenal tumors. In the year prior to the workshop, three working groups reviewed the available literature on pheochromocytomas (PCCs)/paragangliomas (PGLs), adrenocortical carcinomas, and metastases, respectively. In their respective field, these three working groups tried to grade the evidence for recommendations in accordance with accepted international standards. Results of their extensive update were presented during this workshop, and recommendations were discussed in three plenary sessions with input from all the participants. Articles were amended on the basis of the discussion at the workshop.

J.-F. Henry
Department of Endocrine Surgery, University Hospital La Timone, Marseilles, France

J.-L. Peix
Department of Endocrine Surgery, Lyon Sud Hospital, 69495 pierre Benite, France

J.-L. Kraimps (✉)
Department of Endocrine Surgery,
Jean Bernard Hospital,
86021 Poitiers, France
e-mail: j.l.kraimps@chu-poitiers.fr

Adrenocortical carcinomas

The indeterminate adrenal mass [1]

There is very limited evidence on which to base recommendations for the management of the small number of adult patient with an indeterminate/lipid poor (on CT/MRI) adrenal mass in whom there are no critical, biochemical, or radiological indications for surgery. Further investigation with MRI (chemical shift/contrast enhanced) is advised. Negative uptake on ^{18}F-FDG PET scan or a positive scan with appropriately low tumor SUV_{max} or tumor/liver SUV_{max} ratio supports nonintervention. At the present time, the added value and role of image-guided FNA are unclear.

If adrenalectomy is not to be recommended or performed on the basis of the clinician's opinion and patient choice, appropriate follow-up according to local protocols is essential. A subsequent increase in tumor size is an indication for surgery.

Adrenocortical carcinoma: which surgical approach? [2]

There are no randomized studies comparing open adrenalectomy with laparoscopic adrenalectomy for ACC. There is no evidence of postoperative benefit for the patients undergoing laparoscopic adrenalectomy compared to open adrenalectomy (level B).

Results from comparison of oncological outcomes in ACC between open and laparoscopic approaches are equivocal: increased risk of local recurrence and peritoneal carinomatosis by the laparoscopic route (level D), identical results between the two approaches in terms of survival, and recurrence and peritoneal carcinomatosis (level C). An open approach is recommended in case of local invasion, with a view to achieving an R0 resection (level D).

Laparoscopic resection of ACC/potentially malignant tumors, which includes removal of surrounding periadrenal fat and results in an R0 resection without tumor capsule rupture, may be performed for pre-and intraoperative stage 1–2 ACC and tumors with diameter <10 cm (level C).

Malignant pheochromocytomas and paragangliomas: a diagnostic challenge [3]

According to the WHO classification, malignancy of PCCs and PGLs is defined by the presence of metastases at non-chromaffin sites distant from that of the primary tumor and not by local invasion. Preferably, the discrimination between malignant and benign PCCs/PGLs should be made preoperatively. From the clinical point of view, a lack of the "typical signs" (hypertension, palpitations, headache and diaphoresis) may raise the suspicion that one is dealing with a malignant case.

Biochemically, malignant tumors may lack various enzymes. Patients with high preoperative 24-h urinary dopamine levels may therefore have an increased likelihood of having malignant PCC. A markedly increased preoperative chromogranin A plasma level in patients with malignant PCCs in comparison to patients with benign pheochromocytomas has also been reported. Genetically, PCCs/PGLs associated with SDHB germline mutations warrant closer follow-up due to high (up to 40%) frequency of malignancy and a very aggressive behavior of those tumors.

Concerning imaging techniques, no technique is the gold standard, and specific sequences of exams might be needed for each tumor type. A combination of two or more imaging techniques is often required for diagnosis and staging. Usually, radiological techniques (such as ultrasound, CT, or MRI) are useful in the localization of the primary tumor, particularly if nonfunctioning, while nuclear medicine aids in the evaluation of the extent of disease, staging, and therapy decision making.

Concerning surgery, no consensus currently exists regarding the optimal quality (adjacent organ, node dissection) of the surgical procedure to resect a malignant PCC or PGL. Complete removal has to be achieved, and open accesses are still the standard.

Postoperatively, multiparameter scoring systems have been used to predict malignancy but their value is questionable. For example, there is currently no agreement on the utility and reproducibility of the pheochromocytoma of adrenal scaled score (PASS). At this time point, a high PASS score should not be considered as diagnostic of malignancy as some tumors with high score never metastasize. Despite extensive research, no single parameter has been identified yet that can distinguish benign from malignant tumors.

Due to the rarity of malignant PCCs/PGLs and the obvious difficulties in distinguishing benign and malignant PCCs/PGLs, any patient with a PCC/PGL should be treated in a specialized center where a multidisciplinary setting with specialized teams consisting of radiologists, endocrinologist, oncologists, pathologists, and surgeons is available. This would also facilitate future studies to address the above-mentioned diagnostic and/or therapeutic obstacles.

Adrenal metastases [4]

Imaging techniques

In the evaluation of potential adrenal metastasis, three imaging studies are of particular interest: CT, MRI, and PET. PET/CT is the test of choice to use in patients with suspected adrenal metastasis. If the primary cancer was FDG-avid, a PET/CT with benign characteristics is sufficient to exclude adrenal metastasis. If the primary cancer was not FDG-avid, an adrenal biopsy can be proposed if the CT characteristics are not clear for an adenoma. MRI is not better than CT and should only be used when CT cannot (pregnant women for instance).

Open or laparoscopic approach?

Achieving wide surgical margins with en bloc excision of periadrenal fat is a primary requirement during radical adrenalectomy for metastatic cancer. Laparoscopic adrenalectomy for metastases can be performed in appropriately selected cases with equal oncologic outcomes to open approaches while providing advantages in patient morbidity. Caution must be taken to avoid tumor entry or spillage because of the potential for local recurrence, port site recurrence, and carcinomatosis that can occur with these aggressive tumors. In these three fields (adrenocortical carcinomas, pheochromocytomas, and adrenal metastases), finally, most recommendations given in this review can only be considered as level IV, grade C.

Conflicts of interest None.

References

1. Harrison B (2011) The indeterminate adrenal mass. Langenbecks Arch Surg 397(2):147–154
2. Carnaille B (2011) Adrenocortical carcinoma: which surgical approach? Langenbecks Arch Surg 397(2):195–199
3. Gimm O, DeMicco C, Perren A, Giammarile F, Walz MK, Brunaud L (2011) Malignant pheochromocytomas and paragangliomas: a diagnostic challenge. Langenbecks Arch Surg 397(2):155–177
4. Sancho JJ, Triponez F, Montet X, Sitges-Serra X (2011) Surgical management of adrenal metastases. Langenbecks Arch Surg 397(2): 179–194