Incidence of phaechromocytoma and functional paraganglioma in Marne-Ardennes region, a twenty-year study

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Objective : to determine current incidence of pheochromocytoma and to describe its clinical presentation and general characteristics.

Design and method : exhaustive retrospective study of all patients with pheochromocytoma living in Marne and Ardennes french departments observed between January 1988 to December 2008. These data were collected from the files of all the pathologists, endocrinologists and surgeons of the area.

Results : 68 cases (65 pheochromocytomas and 3 functional paragangliomas) were diagnosed giving an incidence of 3.8 cases/ million inhabitants/ year. The diagnosis performance increased with time (incidence 2004-2008 : 5.9 cases/ million inhabitants/ year, incidence 1988-1992 : 3.3 cases/ million inhabitants/ year). 85% of patients suffered from hypertension, 80% had adrenergical symptoms but only 22% with the classic triad (headaches, palpitations, sweating). No case was reported after autopsys. One third of phaechromocytomas were incidentalomas. 15 pheochromocytomas (22%) were associated with specific genetic mutations in 12 different families : 5 MEN2A, 1 MEN2B, 3 neurofibromatosis type 1, 2 SDHB, 2 SDHD, 2 Von Hippel-Lindau. 13 pheochromocytomas (19%) were malignant including 2 SDHB. After a mean 6 years follow-up, we found 5 recurrences and 5 deaths due to pheochromocytoma, including 4 perioperative deaths.

Conclusions : our study shows a 2 times increasing pheochromocytoma incidence compared to previous studies. Moreover, it shows an increasing number of incidentally discovered pheochromocytoma, an improving clinical detection and a disappearance of autopsic diagnosis. Our work confirms that, in general population, 1 pheochromocytoma out of 4 is genetically determined and that 1 out of 5 is malignant.