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Epidemiology of Meningioma

Adelheid Wöhrer

Abstract: Meningiomas are among the most common intracranial tumours, their incidence rising with increasing age. The majority of meningiomas show a benign clinical behaviour, which might in part explain the former lack of systematic registration with only imprecise esti-

mates of incidence and survival. With the introduction of specialized brain tumour registries, detailed epidemiological information has become available. Herein, besides demographic disease characteristics, the latest findings on

aetiological risk factors are reviewed. **Eur Assoc NeuroOncol Mag 2013; 3 (3): 95–6.**

Key words: meningioma, epidemiology, risk factors, aetiology

■ Background and Incidence

Meningiomas account for approximately $\frac{1}{3}$ of primary intracranial tumours in adults [1], with an age-adjusted incidence rate of approximately 7 per 100,000 person-years [1, 2]. While in Scandinavia the incidence seemingly increased between 1968 and 1997 from 2.6 to 4.5/100,000 in women and from 1.4 to 1.9/100,000 in men [3], no such increase was observed in Italy or the United Kingdom, where the disease rate has remained stable for decades [4, 5]. Small meningiomas are often asymptomatic and, thus, constitute frequent incidental autopsy findings (in 1.4 % of a Swedish autopsy-based cohort) [6].

■ Histological Subtypes and Prognostic Considerations

Meningiomas exhibit a wide range of morphologic appearances. According to the WHO classification, up to 15 histopathological variants are distinguished [7]. Of the various histological subtypes meningothelial, fibrous, and transitional meningiomas (all WHO grade I) constitute the most common variants [7]. Whereas the majority of meningioma subtypes (> 80 %) show a benign clinical behaviour (WHO grade I), those variants which fall into the WHO grade-II and -III categories (atypical and anaplastic meningiomas) are associated with a higher likelihood of tumour recurrence and a more aggressive disease course. Five-year overall survival rates of 55 % have been reported for malignant meningioma [8]. In contrast, a recent study in the United States found 3-year overall survival rates of > 85 % for non-malignant intracranial meningiomas [9].

■ Age and Gender Distribution

The majority of meningiomas occur in middle-aged and elderly individuals, the incidence rising with increasing age [1]. In contrast, meningiomas are extremely rare in children [10]. However, childhood examples tend to include more aggres-

sive meningioma subtypes with 5-year overall survival rates of 83.9 % [10]. Whereas benign meningiomas show a strong predilection for women (ie, twice as common in females as compared with males) [2], malignant meningiomas tend to occur more frequently in males [2, 11].

■ Tumour Site

Meningiomas usually occur in an intracranial, intraspinal, or orbital location, the cerebral convexity being the most common site [11]. Still, rare meningiomas have been observed in almost all organs [7]. Moreover, atypical and anaplastic variants have the potential to produce systemic metastases most often to the lung [12–16]. In addition, tumour-to-tumour metastasis (of a systemic cancer to meningioma) has been reported [17, 18]. In fact, meningiomas are the most common “recipients” among intracranial tumours. However, the exact mechanisms of this rare phenomenon are not yet fully understood [17, 18].

■ Aetiology

Meningiomas are well-known to be induced by low-, moderate-, and high-dose radiation, most commonly administered for childhood malignancies, with median times to tumour occurrence of 35, 26, and 19–24 years, respectively [19]. Younger patients often exhibit a longer latency period [20]. In fact, meningiomas are the most common form of radiation-induced neoplasms reported [21]. Compared with their sporadic counterparts, radiation-induced meningiomas are often aggressive or malignant, generally occur in younger age groups, are likely to be multiple, and associated with higher recurrence rates [22]. In contrast to ionizing radiation, the association of meningioma risk with non-ionizing low-frequency electromagnetic fields from mobile and cordless phone use is less consistent. A recent meta-analysis of the Hardell group and IARC Interphone studies did not yield an increased hazard ratio for meningioma [23].

The predominance of women among meningioma patients suggests an aetiological role for sex hormones. However, so far, little evidence has been found for reproductive and menstrual factors such as age at menarche, age at menopause, or parity [24]. Still, at first operation, 88 % of meningiomas exhibit progesterone, 40 % oestrogen, and 39 % androgen receptors [25]. However, the higher incidence of meningiomas in women cannot be explained by differences of sex hormone expression [25].

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■ Genetic Susceptibility

Meningiomas usually occur in a sporadic setting. However, if they occur at multiple sites, they are mostly (> 90 %) associated with familial tumour syndromes [7]. They are common in neurofibromatosis type 2 (NF2) families with 50–75 % of affected individuals developing a meningioma during their lifetime [26]. Outside the setting of NF2, a number of families have been reported to be at increased risk. Meanwhile, several additional genes have been implicated in those multiple meningiomas, such as mutation of the SMARCB1 gene [27, 28] or loss-of-function mutations in SUFU [29]. Most recently, an association of loss-of-function mutations in the SMARCE1 gene with multiple spinal meningiomas of clear-celled phenotype has been described [30].

■ Summary and Conclusion

Meningiomas account for approximately $\frac{1}{3}$ of all intracranial tumours, their incidence rising with increasing age. The majority of tumours are of benign behaviour, whereas atypical and anaplastic variants are associated with a higher likelihood of disease recurrence. Benign meningiomas are twice as common in females as compared with males, although the aetiological role of sex hormones is not yet fully understood. Ionizing radiation constitutes an established risk factor. The vast majority of meningiomas occur sporadically, whereas multiple meningiomas are most often associated with familial tumour syndromes such as NF2.

■ Conflict of Interest

None.

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