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Report 2013: Tumors of the pineal region

Incidence of pineal tumours. A review of the literature

Incidence des tumeurs pinéales. Revue de la littérature

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ABSTRACT

The knowledge of the incidence of pineal tumours is important not only for diagnostic care but also for its therapeutic programme. We reviewed the incidence of pineal tumours reported in literature in an attempt to establish if a difference existed between pineal gland tumours and the pineal region tumours as different authors use both expressions to indicate the same thing. The rate of frequency of these tumours is useful to guide the therapeutic choice for patients as the decisional tree is completely different for either germ cell tumours, pineal gland tumours or pineal gliomas and eventually papillary tumours of the pineal region. According to the French Register of pineal tumours, true pineal tumours represent: 27% pineal parenchymal tumours (PPT), 27% germ cell tumours, 17% gliomas, 8% papillary tumours. True pineal gland tumours are represented by: pineocytomas - (13%), pineal parenchymal tumours with intermediary differentiation PTT-ID - (66%) and pinealoblastomas - (21%). There was no statistical difference found between the French register and the Lyon series concerning histological diagnosis. It seemed to us important to discover its true incidence by comparing the data published in the literature and to stress the utility of the French Register for these uncommon tumours not only for recording new histological cases but also to document clinical symptomatology, therapeutic programmes, length of follow-up and clinical results for each patient treated. A better understanding of their natural history and improved evaluation of different treatments and their complications should contribute to improve clinical results.

Mots clés :
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Gliomes pinéales

RÉSUMÉ

Il est important d'établir l'incidence des tumeurs propres de la glande pinéale et de la région pinéale. Le fait de connaître l'incidence de ces tumeurs peut aider d'un point de vue pratique à une meilleure prise en charge de celles-ci. La stratégie thérapeutique est différente selon qu'il s'agit d'une tumeur du parenchyme, d'une tumeur germinale ou d'une tumeur papillaire. L'étude des cas recensés par le Registre français des tumeurs pinéales a mis en évidence que 27 % des cas sont des tumeurs du parenchyme pinéal, 27 % des cas des tumeurs germinales, 17 % des gliomes de la glande pinéale et 8 % des cas des tumeurs papillaires. Les tumeurs de la glande pinéale sont réparties en pinéalocytomes (13 %), tumeurs pinéales à différenciation intermédiaire (66 %) et pinéaloblastomes (21 %). Il n'y a pas de différences statistiques importantes en ce qui concerne la distribution anatomo-pathologique entre les chiffres du Registre français et ceux de Lyon. La nécessité et l'importance d'un Registre pour ces tumeurs rares regroupant différents types histologiques et nécessitant différentes stratégies thérapeutiques, semblent donc évidentes pour le recensement des nouveaux cas histologiques, pour l'établissement des données cliniques, des protocoles thérapeutiques et pour la vérification de courbes de suivi et de résultats cliniques. Dans ce contexte, le Registre français prend toute son importance, en particulier, parce qu'il permet la confrontation des études de ces tumeurs aux différentes expériences de la littérature.

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1. Introduction

The knowledge of the true incidence of pineal tumours and pineal region tumours is of great importance because different histological entities are represented and consequently for a particular tumour an adapted therapeutic programme has to be established.

In other words, the histological diagnosis can condition the choice of surgical or complementary treatment except for the treatment of hydrocephaly.

Generally, in the literature, when pineal or pineal region tumours are reported there is some mention of the incidence of different histological specimens that can develop in the pineal region.

In order to define the true incidence of pineal tumours from that of the pineal region we were prompted to review various papers in the literature that reported the incidence of pineal and pineal region tumours.

In the pineal tumours group, we have included true pineal tumours, germ cell tumours, pineal gliomas and papillary tumours of the pineal region that are considered the most frequent pineal tumours.

2. Anatomy of the pineal gland and pineal region

The pineal gland develops in a deep-seated region, the pineal region, which is located in the posterior part of the incisural space [1]. The pineal gland belongs to the posterior part of the third ventricle and it is located between the posterior commissure at the top and the tectal plate below. The gland that is a small reddish brown structure has a size which varies between 10 and 14 mm. The pineal gland develops as a diverticulum in the diencephalic roof of the third ventricle and is attached to the posterior board with a stalk. The pineal region is surrounded by different structures that at the same time defines its boundaries: the roof, anterior wall, lateral wall and the floor of the posterior *incisura*. The roof of the pineal region is formed by the lower surface of the *splenium* of the corpus callosum, the hippocampal commissure and the crus of the fornix. The pineal body, the tectal plate and the posterior third ventricle make up the anterior wall of this region. In the lateral wall, we find the pulvinar, the crus of the fornix and the medial surface of the cerebral hemisphere. The floor of the pineal region is formed by the superior surface of the cerebellar vermis. The particular anatomical relationship of the pineal gland and the pineal region render the surgical approach difficult.

3. Data from the literature of the incidence for pineal tumours

The incidence of pineal tumours is 0.4 to 1% of all intracranial adult tumours [2,3]. Jouvet reported an incidence of 1% [4] (Table 1).

For Zhu, they represent less than 1% of all intracranial neoplasms [5]. For Chibbaro, they account for 0.6–0.9% of all brain tumours [6].

For Dahll, they represent 0.4% of all central nervous system tumours in adults [7]. Al-Hussaini reported 0.8% for all ages from the SEER data base [8].

In children, the incidence is higher, i.e. between 2.7 to 11% [2,3,8–12]. Raimondi and Tomita [13] reported an incidence of 9.4% corresponding to one of the most elevated incidences in the literature.

For Villano and Shibui, the incidence varied between 3 to 5% among children [14,15]. For Bruce, pineal tumours represented 3 to 8% of all brain tumours in the paediatric age [16] whereas Wilson reported an incidence of 11% of all paediatric tumours considering lesions of the posterior part of the third ventricle [17].

The incidence reported by Wong from the Brain Tumour Registry of Japan was of 3.2% in children and adults [18].

For Dahll, the incidence was 2.8% in children up to 19 years of age [7].

There is no theoretical explanation for the more significant frequency of pineal tumours in children; it seems to be more an epidemiological factor. Russel and Rubinstein reported that pineal tumours represented 0.1 to 1% of intracranial growths and affirmed that they were much more frequent in children [19]. As regards pineal germ cell tumours that occur more frequently in adolescence one could evoke possible hormonal factors but with no scientific proof [20]. The peak incidence observed during adolescence and early adulthood suggests that endocrinological changes which occur during puberty may be involved in the awakening of dormant cells. Melatonin, the hormone of the pineal gland involved in the sleep pattern, interfering with follicle-stimulating luteinizing hormone release, may play a role in the activation of germ cell tumours within the pineal region [20].

4. Data from the literature regarding the incidence of pineal region tumours

Pineal region tumours accounted for 0.6–0.9% in North America and Europe whereas they represented 3%–3.2% in Japan and in Southeast Asia and 3% in China [21] (Table 2).

According to the centralized brain tumour registries the incidence of pineal tumours varies from 0.4 to 1% among adult patients [22].

These deep-seated tumours consisted in only 0.5% to 1% intracranial neoplasms in adults in different European and American series, with an unexplained higher incidence of 3.2 to 4% in the Japanese literature [23,24]. For Herrada–Pineda the incidence of pineal region tumours was between 3 and 8% [25]. The frequency in Japan is 5 times higher than in the Western countries [26] (CBTRJ, 2008). For Tseng pineal tumours represented 0.4–0.5 of intracranial lesions [27]. Shin reported a frequency of less than 1% for adults and between 3 and 8% in children [28].

Pineal region tumours accounted for 0.4 to 1% of all central nervous system tumours in adults [29] and the same the incidence of 0.4% in adults was reported by the CBTRUS [30]. Senapati reported an incidence of less than 1% in adults and from 3 to 8% in children [31]. Cuccia also reported an incidence of 0.4 to 1% in adults and 3 to 8% in children [32].

For Smith, pineal region tumours represented 3% to 8% of intracranial neoplasms in children [22] and Mandera reported an incidence of between 3 and 11% in children [29].

The same incidence of 2.8% in children up to 19 years of age was reported in the statistical report on brain tumours in the United States (CBTRUS 2000–2004, reported in 2008).

These tumours are rare, consisting in less than 11% of all paediatric tumours, and approximately 60% of pineal region tumours were of germ cell origin [33].

Oi reported that the epidemiological characteristics of Japanese patients were completely different from those of Caucasian or even other Asian populations such as the Chinese [11,34] and confirmed that the patient population in Japan and Korea were almost identical [11,35]. Also, Konovalov reported a higher unexplained incidence of 3.2% in the Japanese literature [36].

5. Discussion

The data reported in the literature are sometimes confusing concerning the incidence of pineal or pineal region tumours as well as in relation to other factors such as age, sex and racial origin.

Concerning racial differences, recent studies have not been able to demonstrate this higher incidence as reported by Cheng and Oi

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