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ORIGINAL ARTICLE

Clinicopathological study of 47 cases of sebaceoma

Étude anatomoclinique de 47 cas de sébacéome

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suppl.
Informations

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Sebaceoma;
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Sebaceous gland;
Sebaceous tumour;
Sebaceous adenoma

Summary

Background. — Sebaceoma is a rare and poorly understood form of sebaceous tumour, and it is of great significance since it may reveal Muir-Torre syndrome (MTS). Herein, we present a series of cases with details of the histopathological appearance.

Patients and methods. — We examined records of cases labelled as sebaceous tumour recorded at the Strasbourg Dermatopathology Laboratory between 1991 and 2015. We include cases of benign sebaceous tumour predominantly involving immature basophilic cells. The clinical and histological data were collected as well as screening for a history of MTS.

Results. — We studied 47 cases of sebaceomas (26 men), in patients of mean age 67.6 years, located primarily in the head or neck (32 cases). Of the 17 patients followed up, 6 had MTS. Different types of architecture were seen: dermal nodule (9 cases) or cystic nodule (9 cases), multiple dermal nodules (22 cases), exophytic tumour (4 cases) and an appearance intermediate with sebaceous adenoma (3 cases). The cells involved were basophilic, with the presence of round ducts exhibiting an eosinophilic cuticle and, in rare cases, mature sebocytes. Mitoses were observed: mean 6.6/10 fields (0 to 19). In all cases, there was expression of CK17, EMA and androgen receptors, but not of BerEP4.

Discussion. — Sebaceoma is a small benign tumour but identification is imperative due to association with MTS. A knowledge of the associated cytological and architectural elements – particularly cysts and labyrinthine patterns – and immunolabelling enable differential

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diagnosis with respect to other tumours. The extra-facial and cystic forms in particular require screening for MTS. If there is any doubt, immunolabelling of androgen receptors provides a precious tool.

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MOTS CLÉS

Sébacéome ;
Syndrome de
Muir-Torre ;
Glande sébacée ;
Tumeur sébacée ;
Adénome sébacé

Résumé

Objectif. — Le sébacéome est une tumeur sébacée rare, mal connue, de signification capitale puisque cette lésion peut révéler un syndrome de Muir-Torre (SMT). Nous présentons une série de cas en détaillant les aspects anatomocliniques.

Méthodes. — Nous avons relu les cas étiquetés « tumeur sébacée » recensés au laboratoire de dermatopathologie de Strasbourg entre 1991 et 2015. Ont été inclus les cas de tumeur sébacée bénigne à prédominance de cellules basophiles immatures. Les données clinico-histologiques ont été collectées en recherchant en outre des antécédents de SMT.

Résultats. — Nous avons inclus 47 sébacéomes (26 hommes), d'âge moyen 67,6 ans, principalement localisés à la tête ou au cou (32 cas). Parmi les 17 patients avec suivi, 6 étaient porteurs d'un SMT. On distinguait différents types d'architecture : nodule dermique (9 cas) ou kystique (9 cas), nodules dermiques multiples (22 cas), tumeur exophytique (4 cas) et aspect intermédiaire avec l'adénome sébacé (3 cas). Les cellules étaient basophiles, avec présence de canaux ronds à cuticule éosinophile et de rares sébocytes matures. On voyait des mitoses : en moyenne 6,6/10 champs (0 à 19). Il existait toujours une expression de CK17, EMA et des récepteurs androgènes, mais pas de BerEP4.

Discussion. — Le sébacéome est une petite tumeur bénigne de diagnostic impératif car associée au SMT. La connaissance des éléments cytologiques et architecturaux — notamment les kystes et les formes labyrinthiques — et les immunomarquages permettent le diagnostic différentiel avec d'autres tumeurs. Les formes extrafaciales et kystiques doivent attirer l'attention pour la recherche du SMT. En cas de doute, l'immunomarquage des récepteurs androgènes est d'une aide précieuse.

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The name “sebaceoma” was first coined in 1984 by Troy and Ackerman to refer to an unusual tumour of the sebaceous glands that was already known, but was the subject of confused terminology, and for which no consensus existed concerning diagnostic criteria [1]. Indeed until that point, the term “sebaceous epithelioma” was used to refer to a benign lesion, insufficiently differentiated to be considered a sebaceous adenoma, but too highly differentiated to be assimilated with a basal cell carcinoma or a sebaceous carcinoma [2,3]. Sebaceoma also differs from sebaceous hyperplasia, which involves an increase in the volume of the sebaceous lobules, which are linked by an excretory duct with a single infundibulum [4].

A sebaceous adenoma comprises several lobules clearly delineated by connective septa [5]. It contains fewer immature cells than mature sebocytes. Sebaceous carcinoma is more or less well differentiated but contains atypical cells, frequent mitoses and broad nuclei with prominent nucleoli [6].

The former term “sebaceous epithelioma” thus lay somewhere between “basal cell carcinoma with sebaceous differentiation” and “sebaceous adenoma”, with fewer mature sebaceous cells than immature cells [7]. The presence of cystic formations has been reported in such

“sebaceous epitheliomas”. Other authors have proposed the term “sebomatricomas” for the lesions typically associated with Muir-Torre syndrome (MTS), the architecture of which is difficult to qualify, or where several types of differentiation coexist within a single lesion [8]; however, this name has never really caught on.

From the time it was first used, the word sebaceoma was understood to refer to a lesion associated with MTS, in common with sebaceous adenoma [9,10]. It is extremely important to be aware of this association with regards to diagnosis of MTS, the most characteristic cutaneous elements of which are sebaceous tumours, frequently multiple [11], with the exception of sebaceous hyperplasias, which are banal and insufficient to evoke this syndrome. MTS is a phenotypic variant of Lynch syndrome [12,13]. It concerns a predisposition of autosomal dominant inheritance associated with a mutation on one of the genes coding for mismatch repair (MMR) proteins, responsible for microsatellite instability [14–16]. Germinal mutations are situated mainly on genes MLH1, MSH2, MSH6 and PMS2 with, according to the study by Mangold et al., predominance on MSH2 [17,18]. MTS is defined by an association of a malignant internal tumour and sebaceous cutaneous tumours (sebaceoma, sebaceous adenoma or sebaceous carcinoma)

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