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ORIGINAL ARTICLE



Syringoma: A clinicopathological study of 244 cases

Syringomes : étude anatomoclinique de 244 cas

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KEYWORDS

Syringoma;
Adnexal tumour;
Clear cells;
Vulvar pruritus

Summary

Background. – Syringomas are benign tumours of the sweat glands, the most familiar clinical presentation of which is the presence of multiple lesions on the eyelids. The aim of our study was to determine the clinical and histological characteristics of a large series of patients and to examine anatomoclinical correlations.

Patients and methods. – This was a retrospective study conducted in all of the cases of syringoma analysed at the cutaneous histopathology laboratory in Strasbourg between 1970 and 2008. The clinical elements, patient history and diagnostic data were collated. All slides were re-read in order to determine the microscopic characteristics of the lesions.

Results. – Two hundred and forty-four lesions were included. The sex ratio was 0.27 and the mean age was 42 years (8 to 85 years). Multiple syringomas were noted in 76% of cases, of which 29.2% were eruptive, and one case occurred in a setting of metastatic melanoma. The sites of predilection were the face (56.7%, of which 36.3% on the eyelids), the chest (18.1%) and the neck (17.5%) for the multiple forms. The lesions were in the form of papules (67%), either brown (34.2%) or flesh-coloured (19.8%). Pruritus was reported in 14 cases, including 4 at vulvar sites (out of a total of 8). A diagnosis of syringoma was made by the clinician in only 30.2% of the multiple forms, with mastocytosis being proposed in 7.1% of cases. The clear-cell forms (18 cases) presented no special clinical features.

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Conclusion. – Syringomas are frequently multiple and are seen mainly in women. They are found predominantly on the face and trunk, and lesions are generally brown and pruritic, a little-known feature that accounts for the degree of diagnostic confusion with mastocytosis. The vulvar forms, which are often pruritic, are poorly known. The eruptive forms may include a hormonal component.

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MOTS CLÉS

Syringome ;
Tumeur annexe ;
Cellules claires ;
Prurit vulvaire

Résumé

Introduction. – Les syringomes sont des tumeurs eccrines bénignes, dont l'aspect clinique le mieux connu est celui de lésions multiples des paupières. Le but de cette étude était de déterminer les caractéristiques cliniques et histologiques d'une grande série de cas et d'en étudier la corrélation anatomo-clinique.

Matériel et méthodes. – Il s'agit d'une étude rétrospective réalisée sur tous les cas de syringome analysés au laboratoire d'histopathologie cutanée de Strasbourg entre 1970 et 2008. Les éléments cliniques, anamnestiques et les diagnostics évoqués ont été recueillis. Toutes les lames ont été relues pour déterminer les caractéristiques microscopiques des lésions.

Résultats. – Deux cent quarante-quatre lésions ont été incluses. Le sex-ratio était de 0,27 et l'âge moyen de 42 ans (8 à 85 ans). Les syringomes étaient multiples dans 76% des cas, dont 29,2% étaient éruptifs, avec un cas survenu au cours d'un mélanome métastatique. Les localisations préférentielles étaient le visage (56,7%, dont 36,3% aux paupières), le thorax (18,1%) et le cou (17,5%) pour les formes multiples. Il s'agissait de papules (67%), de couleur brune (34,2%) ou chair (19,8%). Un prurit était signalé dans 14 cas, dont 4 cas de localisation vulvaire (sur 8 au total). Le diagnostic de syringome était évoqué par le clinicien dans 30,2% des formes multiples seulement, une mastocytose étant proposée dans 7,1% des cas. Les formes à cellules claires (18 cas) n'avaient pas de particularité clinique.

Conclusion. – Les syringomes sont le plus souvent multiples et prédominent chez la femme. Leurs localisations électives sont le visage et le tronc et les lésions sont souvent brunes et prurigineuses, notion mal connue, expliquant la confusion avec une mastocytose. Les formes vulvaires, souvent prurigineuses, sont méconnues. Les formes éruptives pourraient avoir des déterminants hormonaux.

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Eccrine sweat glands occur only in mammals and play a key role in temperature regulation. Their structure comprises a deep secretory unit, the secretory coil, and an excretory duct that leads via a complex tubal configuration to the skin surface [1]. Syringomas are benign eccrine tumours which, although quite common, are not often seen in clinical practice; microscopic diagnosis of these tumours is straightforward. They may be associated with several different syndromes [2].

The term "syringoma" comes from the Greek word *syrinx*, meaning tube. In the 19th century, syringomas were thought to be glandular, but immunohistochemistry and electronic microscopy have subsequently provided evidence of their eccrine origin, and this view is now widely accepted [3,4]. Contrary to what was long held, syringomas do not derive from the intraepidermal section of the sweat duct but from the deep dermal region of the same duct located at the junction with the secretory coil of the gland [3]. The proliferation of cells in the lumen of the duct results in the development of spiral structures in which sweat can no longer move freely or exit through the skin surface.

Clinically, they comprise small papules that are commonly found on the lower eyelids or the trunk. They may occur in episodes, and are referred to as the eruptive form [5–7], or they may occur in isolation [8]. They are reputedly common in trisomy patients [9,10]. The key reasons for consulting are aesthetic considerations or pruritus [11]. In most cases, diagnosis requires histological confirmation and biopsy samples reveal small dilated duct formations edged by a double row of cuboid or flattened cells. A clear cell variant exists and appears to be more common in diabetic patients [2].

There have been numerous publications in the literature of isolated cases, certain of which are particular in terms of their clinical presentation. At best, studies exist involving only small series of cases, with possible overestimation of certain forms. The purpose of our study was to present the clinical and histological characteristics of syringomas based on a very large series of cases in order to facilitate their identification, particularly in supposedly rare forms or forms having an atypical clinical presentation.

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