



Original Research Article

Study of clinicohistopathological correlation of skin adnexal tumours

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ARTICLE INFO

Article history:

Received 20-04-1992

Accepted 10-06-2022

Available online 30-06-2022

Keywords:

Appendageal tumours

Trichoepithelioma

Syringoma

Pilomatricoma

Trichilemmal cyst

Hidrocystoma

Syringocystadenoma papilliferum

Trichofolliculoma

ABSTRACT

Background: Appendageal tumours (Appendageal tumours) are neoplasms which differentiate towards or arise from pilosebaceous apparatus, apocrine gland or eccrine sweat glands. Overall incidence of Appendageal tumours is low, yet they can be challenging to diagnose.

Aim: The study aims at determining the frequency, profile, and distribution of skin adnexal tumours and association of their distribution with patient's age and sex.

Materials and Methods: Cross-sectional descriptive study, conducted over 1-year period. All clinically suspected cases of Appendageal tumours were evaluated and subjected to histopathological examination. Confirmed cases of Appendageal tumours were finally analyzed.

Results: Out of 43586 new patients attending OPD, 64(0.14%) cases of Appendageal tumours were suspected. And 60 underwent histopathological examination. Histopathology was confirmatory in only 42 (70%) cases. Out of 42, the hair follicle tumors comprised of trichoepithelioma 18/42(42.85%), trichofolliculoma 1/42 (2.38%), pilomatricoma 4/42 (9.52%), and trichilemmal cyst 4/42 (9.52%). The sweat gland tumors are comprised of chondroid syringoma 12/42(28.57%), hidrocystoma 2/42(4.76%) and syringocystadenoma papilliferum 1/42(2.38%). Females (61.90%) outnumbered males (38.09%) in our study population.

Conclusion: The incidence of benign skin Appendageal tumours is more as compared to the malignant ones as found in our study. Histopathology remains the gold standard for diagnosis of these tumours.

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1. Introduction

Appendageal tumours (Appendageal tumours) are neoplasms which differentiate towards or arise from pilosebaceous apparatus, apocrine gland or eccrine sweat gland.¹ These tumors basically originate from undifferentiated pluripotent stem cells and finally differentiate to specific tumors influenced by genetics, local vascularity, and the microenvironment of the epidermis and dermis. Head and neck region is unique because of its rich distribution of pilosebaceous apparatus, apocrine as well eccrine sweat glands. It has also been previously

documented that Appendageal tumours predominate over head and neck area.¹ Clinical diagnosis of different entity is often difficult, as most of the Appendageal tumours present as asymptomatic papules or nodules. Anatomic location, number and distribution of these lesions provide important clue to clinical diagnosis but histopathology is invaluable in conformation of the diagnosis.²

Majority of these tumours are benign^{3,4} and the malignant ones are usually irregularly shaped, solitary, rapidly growing plaques or nodules that have a tendency to ulcerate. Risk of malignant degeneration varies with individual lesions.² Local recurrence is well recorded but metastases is rare with the exception of the malignant eccrine and apocrine gland derived tumors and sebaceous

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carcinoma.³ However, diagnosing some of these tumors has important implications, as they might be markers for syndromes associated with internal malignancies, such as trichilemmomas in Cowden disease and sebaceous tumors in Muir-Torre syndrome.⁵

The study aims at determining the frequency profile, and distribution of skin adnexal tumours and association of their distribution with patient's age and sex.

2. Materials and Methods

It was a cross-sectional, descriptive study, conducted over a period of 1 year from November 2018 to November 2019. Total numbers of patients attending dermatology OPD were recorded and clinically suspicious cases of Appendageal tumours were evaluated. Histopathological examination was advised in all suspected cases and only those subjects consenting for histopathology were included for the study. Confirmed cases of Appendageal tumours were considered as the finally analyzable subjects.

2.1. Inclusion criteria

Patients irrespective of age and sex presenting with skin adnexal tumours willing to give consent for enrollment into the study, and In case of paediatric age group, those patients whose parents willing to sign the consent form.

2.2. Exclusion criteria being

Patients clinically diagnosed as having appendageal tumours but not proven histologically as having adnexal tumour. And Patients not willing to participate in the study.

3. Results

Among 43586 new patients attending OPD, 64(0.14%) cases of Appendageal tumours were suspected. And 60 underwent histopathological examination. Histopathology was confirmatory in only 42 (70%) cases, with an incidence of 0.096%. In the remaining eighteen patients, histopathology revealed either milia or epidermal cyst or basal cell carcinoma. In our study, 70% of clinically suspicious cases were proved to be of appendageal in origin. The hair follicle tumors constituted the largest group involving 26/42(61.90%), followed by the sweat gland tumors 16/42 (38.09%). The hair follicle tumors are comprised of trichoepithelioma 18/42(42.85%), trichofolliculoma 1/42 (2.38%), pilomatricoma 4/42 (9.52%), and trichilemmal cyst 4/42 (9.52%). The sweat glands tumors are comprised of chondroid syringoma 12/42(28.57%), hidrocystoma 2/42(4.76%) and syringocystadenoma papilliferum 1/42(2.38%) [Table 1].

Mean presenting age of our study population was 32.64 years (ranging from 11 to 55 years). Most common site affected was head and neck 31/42(73.80%) followed by

Table 1:

Tumours	Number (n=42)	Percentage
Trichoepithelioma	18	42.85%
Syringoma	12	28.57%
Pilomatricoma	4	9.52%
Trichilemmal cyst	4	9.52%
Hidrocystoma	2	4.76%
Syringocystadenoma papilliferum	1	2.38%
Trichofolliculoma	1	2.38%
Total	42	100%

trunk 9/42(21%), and upper limb 2/42 (5.20%)[Table 2].

Table 2:

Site of the tumour	Number	Percentage
Head and neck	31	73.80%
Trunk	9	21%
Upper limb	2	5.20%

Female's outnumbered (65.21%) males (34.78%) in our study population with the male to female ratio being 1:1.62[Table 3].

Table 3:

Tumours	Number of tumours	Male	Female	Male: Female
Trichoepithelioma	18	10	8	5:4
Syringoma	12	2	10	1:5
Pilomatricoma	4	1	3	1:3
Trichilemmal cyst	4	3	1	1:3
Hidrocystoma	2	0	2	0:2
Syringocystadenoma papilliferum	1	0	1	0:1
Trichofolliculoma	1	0	1	0:1
Total	42	16	26	1:1.62

4. Discussion

The incidence of Appendageal tumours is very low and only 0.096 % of patients attending the OPD were found to be suffering from Appendageal tumours our study. Skin appendageal tumors differentiate along one or more adnexal line and their overall incidence is low in our Indian population.

Appendageal tumours are classified into 4 types according to their differentiation toward apocrine gland, eccrine gland, sebaceous gland and hair follicle.⁶

Importance of diagnosing Appendageal tumours lies in the fact that the presence of Appendageal tumours in some instances may lead to the recognition of a genetic syndrome, like Muir-Torre syndrome associated with sebaceous

tumors, Cowden's syndrome with trichilemmomas, etc.³ It is currently believed that Appendageal tumours are derived from cells that have the ability to differentiate toward any of the appendages. In many lesions, the differentiation is uniform and the tumor can be recognized and categorized based on its resemblance to a normal appendage or part of it. In other cases, the pluripotent cell may differentiate toward more than one type of appendage giving rise to a tumor that contains element of two or more appendage in varying degrees of maturation.³ In our study, though such combined nature was not detected. The clinical appearance of appendageal tumor is mostly non-specific¹ and their diagnosis lie firmly in the realm of histopathologists. In the present study, 30% (18 /60) of clinically suspected cases were proved to be non-AT after histopathological examination. Milia were mistaken as syringoma, basal cell carcinoma as desmoplastic trichoepithelioma. Hence, these entities should be kept in mind before making a diagnosis of Appendageal tumours.

In our study, trichoepithelioma [Figure 1a,b] was the most common hair follicle Appendageal tumours constituting 18/42(42.85%) and were distributed around nose suggesting that trichoepithelioma remains the primary differential diagnosis of Appendageal tumours centered around nose. Trichoepithelioma was found to be multiple and familial only in four cases, while solitary variety was found to be more common than multiple. Other hair follicle Appendageal tumours in our study were trichofolliculoma [Figure 2a,b] constituting 1/42 (2.38%), pilomatricoma [Figure 3a,b] constituting 4/42 (9.52%), and trichilemmal cyst [Figure 4a,b] constituting 4/42 (9.52%).

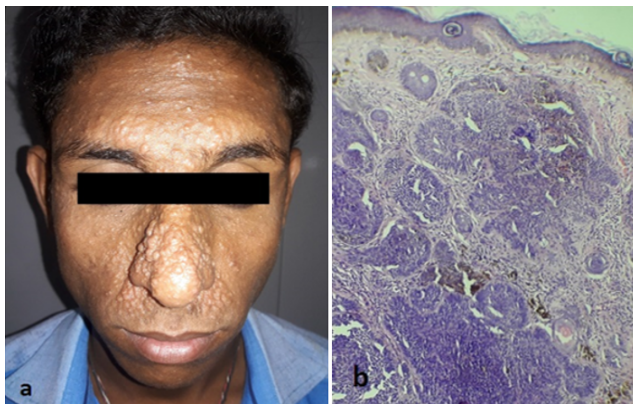


Fig. 1: **a:** Multiple trichoepitheliomas over the nasolabial fold, Nose and forehead in 34 years old male; **b:** Histopathology of trichoepithelioma showing variably sized tumor masses with cribriform appearance horn cyst (H & E stain, 40X).

Among sweat gland tumours chondroid syringoma [Figure 5a,b] was the most common Appendageal tumours constituting 12/42(28.57%), hidrocystoma[Figure 6a,b] constituting 2/42(4.76%), followed by syringocystadenoma papilliferum [Figure 7a,b] constituting 1/42(2.38%).

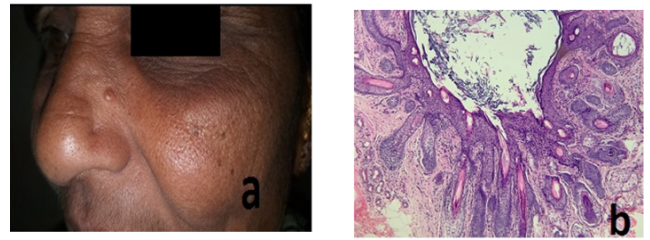


Fig. 2: **a:** Trichofolliculoma in a 52 year of female; **b:** Histopathology of Trichofolliculoma showing central dilated follicle with keratin and vellus hair shafts.

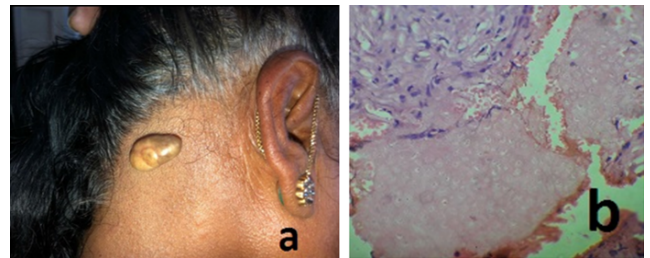


Fig. 3: **a:** Trichoepithelioma in a 32 year old female; **b:** Histopathology of trichoepithelioma enucleated "shadow" or "ghost" cells

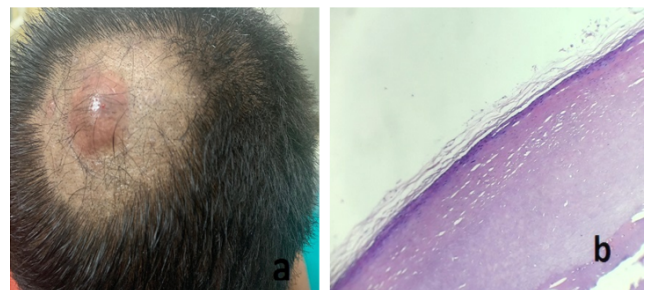


Fig. 4: **a:** Trichilemmal cyst on the scalp of 32 year old male; **b:** Histopathology of Trichilemmal cyst shows cyst wall with trichilemmal differentiation.

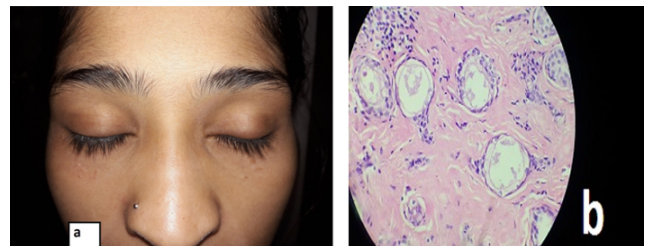


Fig. 5: **a:** Syringoma in a 18-year-old female; **b:** Histopathology of syringoma showing cords of epithelium resembling tadpole (H & E stain, 100X)

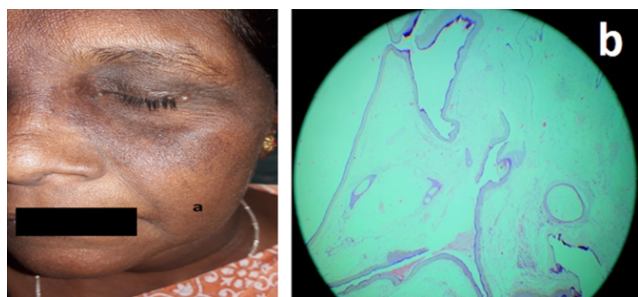


Fig. 6: **a:** Hidrocystoma on the upper eyelid in a 56 year old female; **b:** Histopathology of Hidrocystoma showing multilocular cystic lined by bilaminar epithelium.

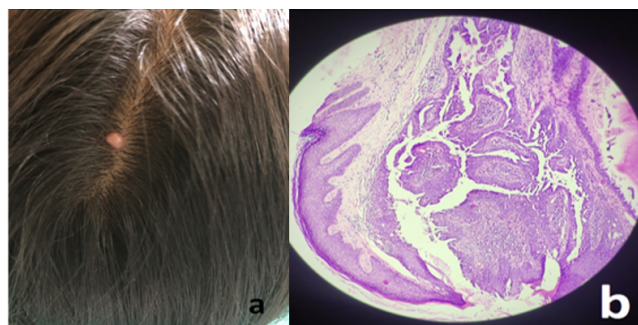


Fig. 7: **a:** Syringocystadenoma papilliferum on the scalp in a 26 year old female; **b:** Histopathology of syringocystadenoma papilliferum showing cystic invagination of surface epithelium with papillary folds (H & E stain, 40X).

No sebaceous gland Appendageal tumours were identified in our study. Incidence of benign tumours is more as compared to malignant cases. In our study histopathological features of malignancy was absent in all cases and supports the usual benign course of the Appendageal tumours. In present study, most common tumours were Trichoepithelioma and Syringoma followed by Pilomatricoma and Trichilemmal cyst.^{7–12}

To summarize the incidence of benign skin AT's is more as compared to the malignant ones. Skin adnexal tumours can occur anywhere in the body; however head and neck region constitutes the most common site. Histopathology remains the gold standard for diagnosis of these tumours. The higher incidence among females of any age group than males.

5. Conflict of Interest

None.


6. Source of Funding


None.

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Cite this article: Budamakuntla L, Ansari MS. Study of clinicohistopathological correlation of skin adnexal tumours. *IP Indian J Clin Exp Dermatol* 2022;8(2):114-117.