Original Article

Histomorphological Spectrum of Skin Adnexal Tumours : A Retrospective Study in a Tertiary Care Centre

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ABSTRACT

Background: Skin Adnexal Tumours (SATs) are large and divergent group of tumours which are classified based on their appendageal differentiation into eccrine,follicular,sebaceous and apocrine.They pose daunting diagnostic challenges to both clinicians and pathologists alike.This study aims to evaluate the histopathological charecterestics of skin adnexal neoplasms and correlate with their clinical profile.

Methods: This is a retrospective study of skin adnexal tumours (28 cases) diagnosed on histopathological examination over a period of two years .(January 2014 to December 2015).

Results: Skin adnexal tumours are uncommon lesions with an incidence of 0.27%.These tumours were common in the 51 to 60 age group and showed a female preponderance. Head and neck region particularly the scalp was commonly involved. Benign tumours were more common (78.6%) than the malignant ones (21.4%). .Sweat gland tumours constituted the largest group (61%) followed by hair follicle tumours (21%)and sebaceous tumours (14%).Apocrine gland tumours were less common. Nodular hidradenoma was the most common benign tumour and sebaceous carcinoma was the most common malignant tumour encountered in the present study.

Conclusion: SATs are relatively uncommon lesions and have distinct histopathological features. Clinical diagnosis is difficult as most of these lesions are nondescript and histopathological examination is essential for its diagnosis.

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Introduction

Skin adnexal tumours (SATs) poses a diagnostic challenge to both the treating clinician as well as the dermatopathologist since a similar looking nodule or papule show a wide histopathological spectrum. The spectrum ranges from a benign adnexal tumour that is cured with excision to a malignant counterpart that is locally aggressive and has a potential for distal metastasis. These adnexal tumours arise from the pluripotent stem cells that differentiate along one or more primary adnexal structures including eccrine ,follicular, apocrine and sebaceous glands or may differentiate along multiple cell lines .[1]Although the site and distribution of the lesions provide some clue to the diagnosis, histopathology is indispensable in the diagnosis of SATs.[2] Few clinicopathological studies on skin adnexal tumours are available in the literature. This study aims at histopathological analysis and clinicopathological correlation of the skin adnexal tumours in our tertiary care hospital.

Materials and Methods

This is a retrospective study spanning over a period of two years from January 2014 to December 2015 in our tertiary care teaching hospital. The clinical records of the cases diagnosed as Skin adnexal tumours in our department of Pathology, Madurai Medical College were retreived . All the slides from these cases were reviewed by a panel of pathologists. Special stains like PAS stain was done wherever necessary. The age, gender, site, clinical presentation, associated syndromes if any ,were tabulated and correlated with the histopathologic diagnosis.

Result

Out of total 10,303 specimens received in our department during this period ,28 cases were skin adnexal tumours ,incidence being 0.27%. In the present study ,SATs were observed in all age groups ranging from 10 to 96 years. The highest incidence was observed in the 51 to 60 age group. (9/28 cases , 32.1 %) followed by 61 to 70 age group (6/28 21.4 %).out of the 28 cases 16 were females with a male :female ratio of 0.75:1 (Table 1)

Histologically 22 were benign (78.6%) and 6 malignant tumours.(21.4 %). (Figure 1). Sweat gland tumours constituted the largest group (17/28 cases, 61 %) followed by hair follicle tumours (6/28, 21 %) and sebaceous tumours .(4/28,14%).(Figure 2). Clinically most cases presented as nodules and cysts. Head and Neck region , particularly the scalp was commonly involved. Table 2 depicts the various types of skin adnexal tumours encountered in the present study and their clinical presentation and diagnosis.

Nodular hidradenoma was the most common benign tumour in the present study (6/28 cases). Out of the 6 malignant tumours reported in the present study ,3 were sebaceous carcinoma and 3 were malignant sweat gland tumours. All the 6 malignant tumours encountered in the present study were seen above 50 years of age.

Table 1: Age and Sex Wise Distribution

ADNEXAL TUMOURS	MALE	FEMALE	0-10 YRS	11 -20 YRS	21 -30 YRS	31-40 YRS	41 -50 YRS	51-60 YRS	61 -70 YRS	71 -80 YRS	81-90 YRS	91 AND ABOVE
SWEAT GLAND TUMOURS												
BENIGN												
1. Nodular hidradenoma	2	4	1				1	2	1	1		
2. Cylindroma	1	3			1			3				
3. Chondroid syringoma	2				2							
4. Spiradenoma		1						1				
5. Eccrine poroma	1							1				
MALIGNANT												
6. Porocarcinoma		1						1				
7. Ductal eccrine adenocarcinoma		1								1		
8. Adenoid cystic carcinoma	1								1			
HAIR FOLLICLE TUMOURS												

ADNEXAL TUMOURS	MALE	FEMALE	0-10 YRS	11 -20 YRS	21 -30 YRS	31- 40 YRS	41 -50 YRS	51-60 YRS	61 -70 YRS	71 -80 YRS	81-90 YRS	91 AND ABOVE
BENIGN												
9. Trichoepithelioma	2	1							2		1	
10. Pilomatricoma	1	1		2								
11. Trichilemmoma		1					1					
APOCRINE GLAND TUMOURS												
BENIGN												
12. Syringocystadenoma papilliferum	1											1
SEBACEOUS GLAND TUMOURS												
BENIGN												
13. Sebaceoma		1				1						
MALIGNANT												
14. Sebaceous carcinoma	1	2						1	2			
TOTAL	12	16	1	2	3	1	2	9	6	2	1	1

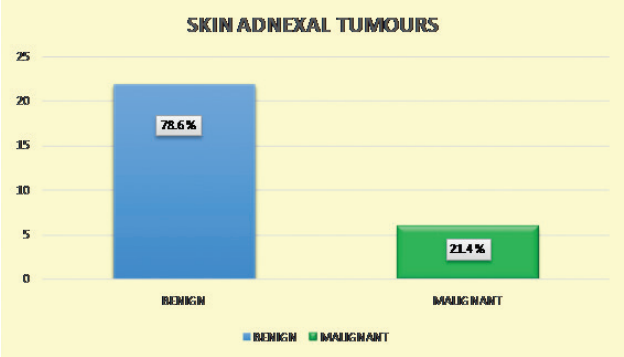

Fig. 1:

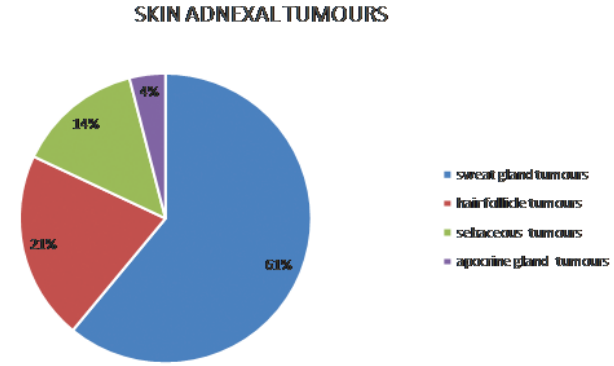

Fig. 2:

Table 2: Clinical Presentation and Diagnosis

ADNEXAL TUMOURS	NO. OF CASES	SITE	CLINICAL PRESENTATION/CLINICAL DIAGNOSIS
SWEAT GLAND TUMOURS			
BENIGN			
1. Nodular hidradenoma	6	Scalp(3) Face (1) Luteal region (1) Abdomen(1)	Dermato fibroma,(3),sebaceoma(1) sebaceous cyst (2)
2. Cylindroma	4	Face (3) Scalp (1)	Nodule (2) Cyst (1) Associated with Brooke Spiegters syndrome.(1)
3. Chondroid syringoma	2	Upper lip (1) Ear (1)	Cyst (2)

ADNEXAL TUMOURS	NO. OF CASES	SITE	CLINICAL PRESENTATION/CLINICAL DIAGNOSIS
4. Spiradenoma	1	Arm (1)	Nodule (1)
5. Eccrine poroma	1	Scalp(1)	Nodule(1)
MALIGNANT			
6. Porocarcinoma	1	Thigh (1)	Squamous cell carcinoma(1)
7. Ductal eccrine adenocarcinoma	1	Chest(1)	Skin nodule(1)
8. Adenoid cystic carcinoma	1	Scalp (1)	Cyst (1)
HAIR FOLLICLE TUMOURS			
BENIGN			
9. Trichoepithelioma	3	Scalp (1) Nape of neck(1) Thigh(1)	Cyst(1) Nodule (2)
10. Pilomatricoma	2	Scalp (2)	Dermoid cyst(1) Sebaceous cyst(1)
11. Trichilemmoma	1	Scalp(1)	Squamous cell carcinoma(1)
APOCRINE GLAND TUMOURS			
BENIGN			
12. Syringocystadenoma papilliferum	1	Umbilicus(1)	Nodule(1)
SEBACEOUS GLAND TUMOURS			
BENIGN			
13. Sebaceoma	1	Scalp (1)	Squamous cell carcinoma(1)
MALIGNANT			
14. Sebaceous carcinoma	3	Eyelid (1) Scalp (2)	Squamous cell carcinoma (3)

Discussion

Skin appendageal tumors are large and divergent group of tumours which are classified into 4 main groups based on their differentiation into adnexal structures present in normal skin: hairfollicle, sebaceous, apocrine and eccrine glands.[3]Some exhibit multilineage differentiation as the tumour originate from multipotential undifferentiated cells present within the epidermis or appendageal structures.[1]

SATs are relatively uncommon tumours.In our study the incidence was 0.27% which is in concordance with the study by Chayanika Pantola et.al.[4] Wide range of age distribution was observed in the present study with the commonest age group being 51 to 60 years. Similar observations were made in the study by Ankit et.al.[5]There was a female preponderance in the present study with a male:female ratio of 0.75:1 which correlated with other studies.[6,7]

Clinical diagnosis of SATs is often rendered difficult as most of them present as nodules ,cysts and papules. In the present study correlation between clinical and histological diagnosis was observed in only few cases similar to studies by Radhika et.al.[7] Skin adnexal neoplasms are clues to hereditary tumour syndrome such as Brooke- Spiegter (Cylindroma), Cowdens syndrome (Trichilemmoma) and Muir Torre syndrome (sebaceous tumours). [8]In our study we received a case of cylindroma associated with Brooke – Spiegter syndrome. SHTs were common in the head and neck region as in the studies by Ankit et.al and Radhika et.al.[5,7]

Most SATs are benign tumours and incidence of malignant tumours is low .In our study too benign tumours were more common (78.6 %) which was in par with studies by Ankit et.al and Vani et.al .[5,6] In the present study tumours with eccrine differentiation were the commonest (61%) followed by hair follicle (21%),sebaceous (14%) and apocrine differentiation . (4%) Similar observations were reported in studies by Vani et.al and Radhika et.al. [6,7]Nodular hidradenoma was the most frequent tumour

encountered in the present study as in studies by Chayanika Pantola et.al,Vani et.al and Radhika et.al . [4,6,7]

Tumours with eccrine differentiation : In the present study, 6 cases of nodular hidradenoma were reported. They were common in females (4/6 cases) and occurred in the head and neck region commonly involving the scalp. Similar observations were reported by Chayanika Pantola et.al and Gayathri et.al. [4,9] In the present study, clinically 3 cases were diagnosed as dermatofibroma,one as sebaceoma and two as sebaceous cyst. Histologically these tumours are composed of nodules and nests of monomorphous polyhedral cells with small ductular lumens. Clear cell change may be seen.

4 cases of cylindroma were encountered in the present study. They were also common in females involving head and neck region. In the present study, one case was associated with Brooke-Speigter syndrome. It is an inherited syndrome associated with cutaneous adnexal lesions such as spiradenoma, cylindroma, trichoepithelioma and basal cell adenoma of salivary gland.[10] Histologically cylindromas are charecterized by many well defined variably sized islands of central large cells and peripheral small basaloid cells surrounded by thick PAS positive basement membrane like material imparting "jijsaw puzzle "pattern.(Figure 3)

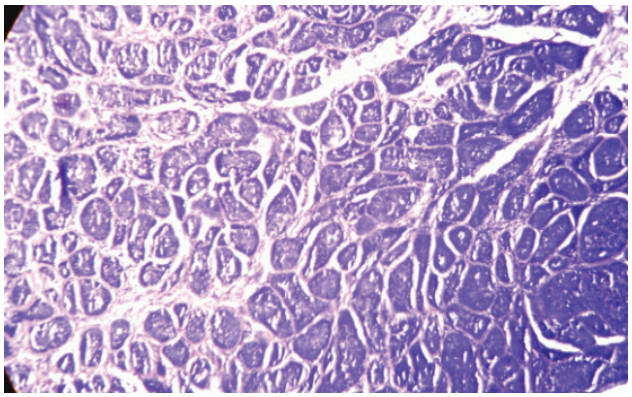

Fig. 3: Cylindroma- Shows irregular nests of basaloid cells arranged in a "jigsaw" manner surrounded by a thick eosinophilic basement membrane.(10 x ,H & E)

Chondroid syringomas also called as mixed tumour of the skin were the next common tumor with eccrine differentiation reported in the present study.They commonly affect nose,cheek and upper lip.In the present study,out of the two cases reported one involved upper lip and other occurred in the neck region. Similar observations were made in the study by Gayathri et.al.[9] Histologically ,these tumours are similar to benign mixed tumour of salivary glands .

One case of spiradenoma and one case of eccrine poroma were seen in the present study.Spiradenoma orginate from the straight intradermal eccrine duct and present as a solitary painful nodular lesion in young adults commonly involving upper extremities and trunk. However in our study it was seen in an elderly female presenting as a nodule involving the arm. They occur as a well circumscibed nodule in the dermis composed of large cells with vesicular nuclei ,pale abundant cytoplasm and small cells with hyperchromatic nuclei and scant cytoplasm. Perivascular space which is an important histological clue first described byVan Den Oord and Chris De Wolf Peters in 1998 [11] was observed in our case.

Poromas orginate from the outer cells of intraepidermal excretory ducts of sweat gland commonly involving palms and soles.They are composed of nodules of basaloid cells. (Figure 4). In our study, poroma presented as a nodule involving the scalp in an adult male.

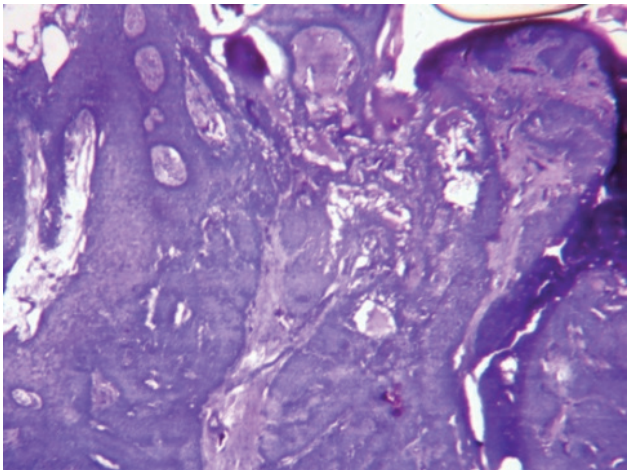

Fig. 4: Eccrine poroma -Tumor cells grow in the form of cords and nest arising from the epidermis. (10 x. H & E)

One case of Porocarcinoma was reported in the present study. It occurred as a growth involving the thigh and clinically it was diagnosed as squamous cell carcinoma. Porocarcinoma usually arise in a pre-existing benign poroid tumour and are found in the extremities as in our case. Histologically these tumours have infiltrative borders and tumour cells exhibit varying degrees of nuclear atypia and brisk mitotic activity.

One case of ductal adeno carcinoma presenting as a nodule in the chest was reported in the present study. It is a malignant sweat gland tumour with the tumour cells exhibiting cytonuclear atypia, mitotic activity, lymphovascular invasion, perineural invasion and infiltrative growth pattern.

One case of adenoid cystic carcinoma encountered in the present study presented as a cyst involving the scalp in an elderly male. It is a very rare malignant tumour affecting older adults .It can occur in any anatomical site but more common in the scalp [12]which correlated with the present study. Histological appearance is similar to its salivary gland counterpart. Before diagnosing primary cutaneous adenoid cytic carcinoma, Salivary gland adenoid cystic carcinoma and adenoid Basal cell carcinoma should be excluded.

Tumours with follicular differentiation: In the present study 6 cases of SATs with hair follicle differentiation were reported. Of these, 3 cases were Trichoepitheliomas. One case presented as a cyst in the scalp and other two presented as nodule involving scalp and thigh. Trichoepitheliomas can be found in any area of hair bearing skin commonly involving head and neck region in adults. These tumours are composed of nests of basaloid cells with peripheral palisading surrounded by dense stroma. Keratin filled horn cysts are usually seen within the nests. Artefactual retraction is uncommon.

2 cases of Pilomatricoma both presenting clinically as a cyst involving scalp were encountered in the present study. Pilomatricoma commonly affects children and adolescents .They have predilection for head and neck and upper extremities. Early lesions are cystic. Histologically they are composed of uniform basaloid cells transformed into pale eosinophilic anucleated shadow cells admixed with keratin. (Figure 5)Familial occurrence and multiple lesions are associated with Turners syndrome,Gardner syndrome and Rubinstein Taybi syndrome. They were not encountered in the present study.

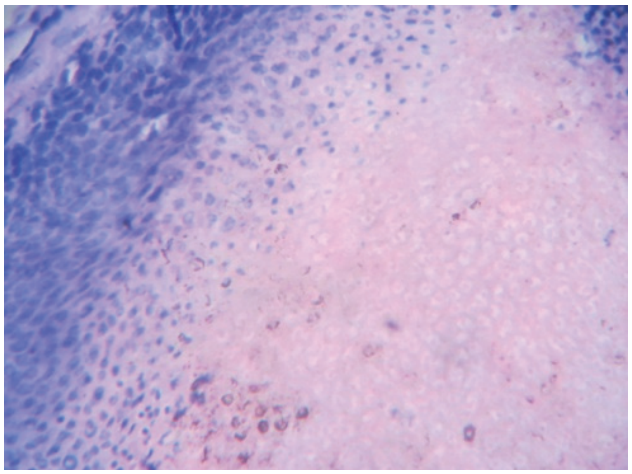

Fig. 5: Pilomatrixoma (Calcifing epithelioma of Malherbe).Showing two basic cell types: basaloid cells and "shadow" or ghost cells.(10 x, H & E)

One case of Trichilemmoma involving scalp was reported in the present study. These are benign tumours arising from the outer root sheath of hair follicles. Clinically they present as a small papule or veruccous lesion involving the face of older adults. The tumour is composed of lobules of uniform cells with clear cytoplasm with basophilic cells arranged in a palisaded manner at the periphery ,surrounded by PAS positive diastase resistant Basement membrane material.(Figure 6)

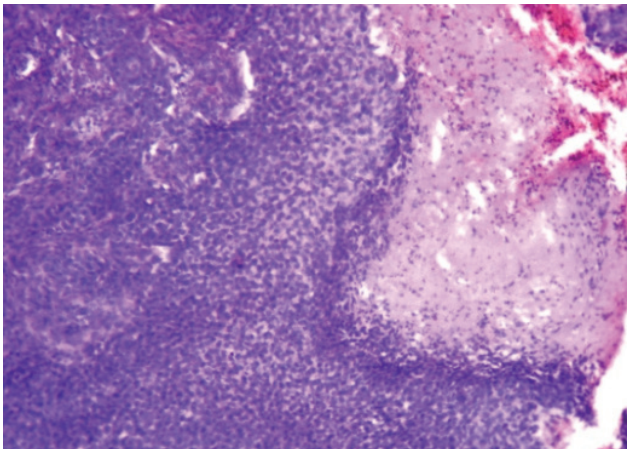

Fig. 6: Trichilemmoma. Shows plate like growth of glycogen rich clear cells.(10 x ,H & E)

Tumours with apocrine differentiation :One case of syringocystadenoma papilliferum presenting as a nodule in the umbilical region of a 96 year old male was reported in this study. These are benign tumours usually affecting young adults common in face and scalp. Histologically it is charecterized by papillary structures with a dermal fibrovascular core lined by two layers of epithelial cells. (Figure 7)Marked plasmacytic infiltrate is present in the fibrovascular core.

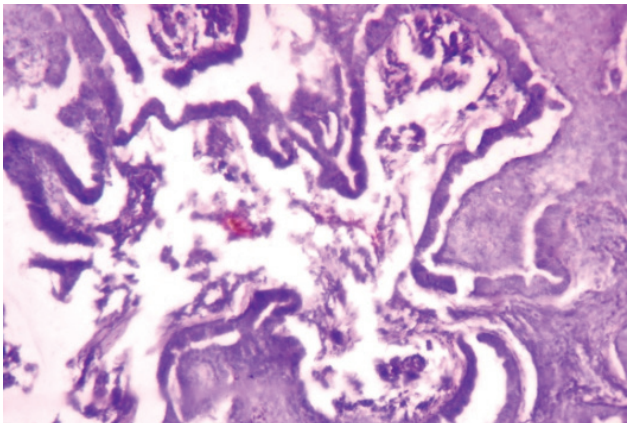

Fig. 7: Syringocystadenoma papilliferum – Shows papillae lined by two layers of epithelial cells. (10 x ,H & E)

Tumours with sebaceous differentiation: One case of sebaceoma was reported in the present study. Sebaceoma formerly called as sebaceous epithelioma is a benign tumour composed of basaloid epithelial cells admixed with single or clusters of mature sebaceous cells.

Sebaceous carcinoma was the most common malignant tumour encountered in the present study. Similar observations were made in the study by Hesari K et.al. [13] Sebaceous carcinoma predominantly arise from Meibomian glands or glands of Zeis in the eyelids. In our study one case had eyelid involvement and other two had involvement of scalp .All the three cases were clinically diagnosed as squamous cell carcinoma. Unlike sebaceous adenoma and sebaceoma, sebaceous carcinoma is charecterized by infiltrative growth pattern and composed of pleomorphic basaloid cells exhibiting increased mitotic activity. Sometimes it is difficult to differentiate it from clear cell squamous cell carcinoma and clear cell basal cell carcinoma. Cytoplasmic glycogen is less abundant in sebaceous carcinoma when compared to clear cell basal cell carcinoma and clear cell squamous cell carcinoma which can be demonstrated by PAS.[14] Immunohistochemically ,sebaceous carcinoma shows strong positivity for EMA (Epithelial Membrane Antigen)whereas squamous cell carcinoma stains weakly and Basal cell carcinoma is negative for this marker.

Conclusion

Skin adnexal tumours are infrequent lesions most commonly occurring in the fifth to sixth decade of life. They commonly involve the head and neck region. Benign tumours are more common than malignant ones. The clinical presentation is often nondescript with most of these lesions presenting as nodule and papule. Histologically it poses diagnostic difficulties to pathologists alike due to their wide spectrum and frequency of differentiation along different cell lines in the same lesion. Since histopathology serves as the gold standard for diagnosing skin adnexal tumours ,familiarity with the various presentation of these lesions is essential for the surgical pathologist.

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Competing Interests

None
Declared

Reference

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