ORIGINAL RESEARCH PAPER | Medical Science | Volume : 6 | Issue : 8 | August 2016 | ISSN - 2249-555X | IF : 3.919 | IC Value : 74.50

Skin Adnexal Tumors – Still a Pandora's Box

KEYWORDS: adnexal neoplasm, eccrine, apocrine

* P.Umarani	C.Swathi	K.Rama murthy
Assistant Professor, Department of Pathology,Alluri Sitarama Raju Academy of Medical Sciences, Eluru – 534005,W.G.Dist, Andhra Pradesh. * Corresponding Author	Assistant Professor, Department of Pathology,Alluri Sitarama Raju Academy of Medical Sciences, Eluru – 534005,W.G.Dist, Andhra Pradesh.	Professor & HOD, Department of Pathology,Alluri Sitarama Raju Academy of Medical Sciences, Eluru – 534005,W.G.Dist, Andhra Pradesh.

ABSTRACT *Evaluation of skin biopsies with special emphasis on adnexal neoplasms in regards to incidence, morphological sub-types and also few prognostically indeterminate mimics in adnexal neoplasms.In a 3year retrospective study, total no. of skin biopsies received were 282, of which 5% were adnexal tumors. All the tissue specimen received were formalin fixed, processed in paraffin wax, stained with Haematoxylin & Eosin along with special stains wherever necessary & reported. Among adnexal tumors, the tumors with eccrine differentiation constituted 50% & apocrine were 21.42%. Tumors with follicular differentiation were 21.42% & single case with sebaceous differentiation accounting to 7.14% . M:F ratio in benign tumors is 1:1, and 2 cases of malignant tumors in males are reported at ASRAMS. Clinically adnexal neoplasms should be regarded as precursor of malignancy. So critical evaluation of the skin biopsies is essential.*

Introduction :

The concept of appendageal tumorigenesis is described by the mechanism governing the proliferation and differentiation of pluripotent stem cells situated within various niches in epidermis and associated adnexae.[1] These stem cells when undergoing neoplastic transformation, may express one or more lines of appendageal differentiation to varying degrees which are considered under traditional headings of hair follicle tumor, sebaceous tumors, apocrine tumors, eccrine tumor, complex adnexal tumors.[2] Historically immunhistochemistry is of little value in definitely distinguishing among the phenotypic patterns of adnexal neoplasms, however identification of specific mutations in genes is useful. From clinical point of view adnexal neoplasms should be always regarded as precursors of malignancy.[1]

Materials and Methods :

In a 3 year retrospective study, 232 skin biopsies were received, from the year 2009 to 2011 in the department of Pathology, ASRAMS, Eluru of which 14(5%) were adnexal tumours. All the tissue specimen received were formalin fixed, processed in paraffin wax, stained with Haematoxylin & Eosin along with special stains where ever necessary and examined. The adnexal tumours are classified according to WHO classification(Table 1).

Table1 : Classification of Adnexal tumours

Lesion type	Follicular differentiation	SebaceousDifferentiation	Apocrine differentiation	Eccrine differentiation
Hyperplasias	Hair follicle nevusDilated pore Hair follicle hamartoma	Nevus sebaceusSebaceous Hyperplasia	Apocrine nevus	Eccrine nevus
Benign neoplasms	Trichofolliculoma Pilar sheath acanthomaFibrofolliculoma TrichoepitheliomaTrichoblastoma Trichoadenoma PilomatricomaTrichilemmomaProliferating trichilemmal tumor-Trichilemmal horn	Sebaceous adenoma Sebacoma	Apocrine hidrocytoma Hidradenoma papilliferum Syringocystadenoma papilliferum Tubular apocrine adenoma Apocrine cylindroma	Eccrine hidrocytoma Syringoma Eccrine cylindroma Eccrine poroma Eccrine syringofibradenoma Eccrine spiradenoma Papillary eccrine adenoma Nodular hidradenoma Chondroid syringoma
Malignant neoplasms	Pilomatrix carcinoma Malignant proliferating trichilemmal tumor Trichilemmal carcinoma Trichoblastic carcinoma	Sebaceous carcinoma	Malignant apocrine cylindroma	Porocarcinoma Malignant eccrine spiradenoma Malignant nodular spiradenoma Eccrine adenocarcinoma Adenoidcystic carcinoma Mucinous eccrine carcinoma Syringoid eccrine carcinoma Malignant eccrine cylindroma

Observations and results :

Total no. of skin biopsies were 232. Of these, adnexal tumors constituted 14 in number. The total benign tumors were 85.69% and malignant lesions were 14.29%. M:F ratio in benign cases constituting 1:1 and malignant variant consists of 2 cases, both in male patients.

Eccrine tumors constitute 50% and sebaceous tumors were 7.14%. The benign tumors were 85.69% and the malignant tumors constituted 14.29%.

Volume : 6 | Issue : 8 | August 2016 | ISSN - 2249-555X | IF : 3.919 | IC Value : 74.50

Table 2: Various adnexal tumors at ASRAMS

Types	Follicular	Sebaceous	Apocrine	Eccrine
Hyperplasia				
Benign	Pilomatricoma(3)	Sebaceous Adenoma(1)	Apocrine fibroadenoma(1) Hidradenoma papilliferum(1) Syringocystadenoma papilliferum(1)	Eccrine poroma(1) Eccrine spiradenoma(3) Clear cell hidradenoma(1)
Malignant				Malignant clear cell hidradenoma(1) Mucinous eccrine carcinoma(1)

Of the 14 cases ,three were Pilomatricomas(Figure1), one of which presented on the left eyelid, and the other two cases involved the scalp region. Of the three, largest biopsy received was a skin covered soft tissue bit measuring 1cm. A single case of sebaceous adenoma presented in a female patient of age 40yrs in the right lower eyelid.

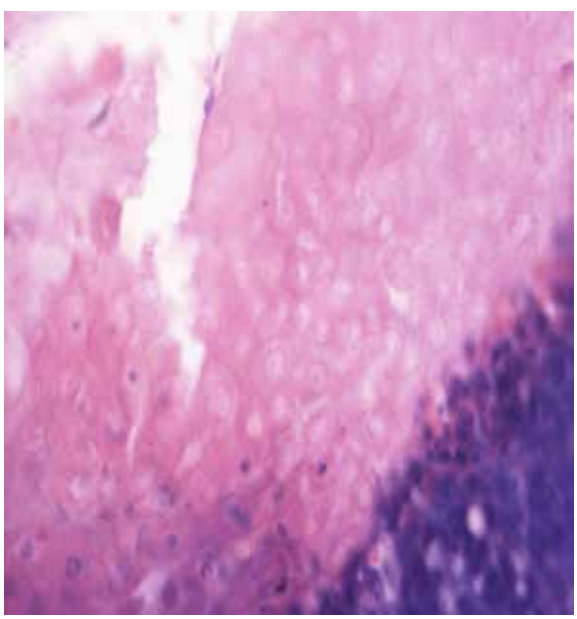

Fig1 - Pilomatricoma 6987-10 (H&E, x40) The photomicrograph shows the transfomation of basophilic cells into shadow cells is associated with loss of nuclei .

Apocrine tumours which constituted around 21.42% of the total, all being benign variants. One case of apocrine fibroadenoma was noted in a male patient of age 70yrs who presented with a haemarrhoidal mass in the anal canal at 3'Oclock position. A case of hidradenoma papilliferum(Figure2) in a female patient of 50yrs age was received as an excision biopsy specimen from vulval region. A case of syringocystadenoma papilliferum(Figure3) in a male patient of age 16yrs , as a swelling over the scalp.

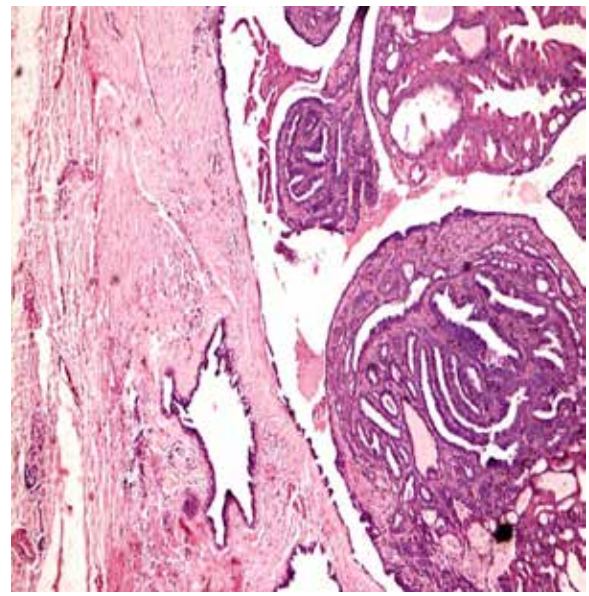

Fig 2 - Hidradenoma papilliferum. 4467-08 (H&E, x10) The photomicrograph shows the tumour composed of well circumscribed dermal nodule with scattered cystic and branching papillary projections. The papillary folding are lined by one layer of cylindrical cells which shows active 'decapitation' secretion like that of apocrine glands.

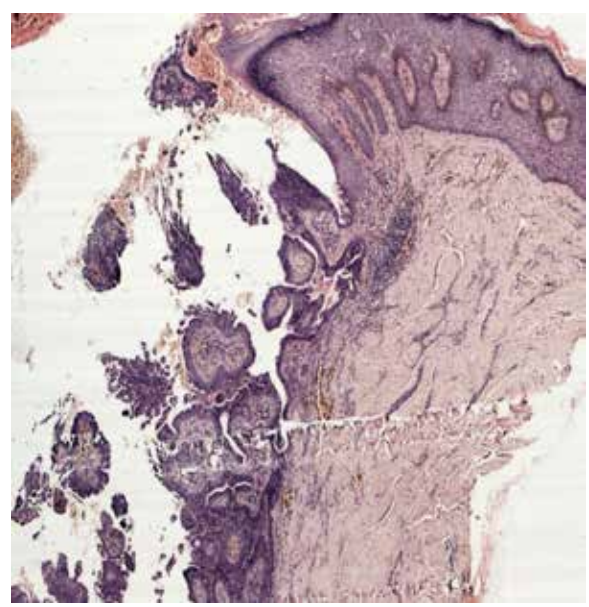

Fig3 - Syringocystadenoma papilliferum 7110-10 (H&E, x10). The photomicrograph shows tumour exhibiting cystic degeneration and invagination into the dermis, with numerous papillary projection with a focally covered stratified squamous epithelium.

Tumors with Eccrine differentiation comprises the largest group in our study(Table1). Eccrine spiradenoma(Figure4) constituited 3 cases at ASRAMS. A case was presented as right lower lid swelling, others were on face & neck. A female patient, age 45yrs with swelling on right lower leg, at ASRAMS was diagnosed as eccrine poroma. Another tumour of Eccrine differentiation, Clear cell hidradenoma was reported in a male patient of 36year age, presented as a solitary lesion on scalp.

Volume : 6 | Issue : 8 | August 2016 | ISSN - 2249-555X | IF : 3.919 | IC Value : 74.50

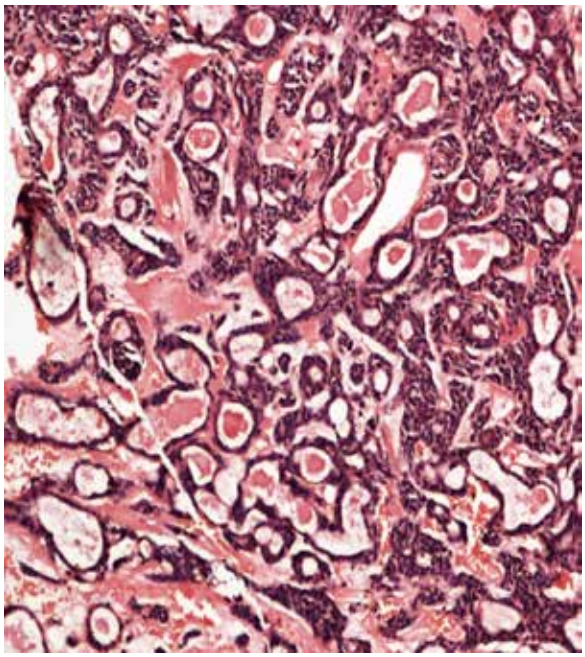

Fig 4 - Eccrine spiradenoma 4385-09(H&E, x10) The photomicrograph shows well circumscribed aggregates of blue tumor cells and collections of hyalinised material.

Malignant tumors constituted two cases of which one was eccrine variant. Malignant clear cell hidradenoma(Figure5) arising from the anterior abdominal wall, above the umbilicus. We received a skin covered mass measuring 9x4x2.5cm. External surface is nodular and cut section showed an ill circumscribed solid mass, grey-white in colour infiltrating into the subcutaneous tissue. The patient had a past history of excision of the mass in the same location, 5years back. It showed infiltration into the deeper subcutaneous tissue.

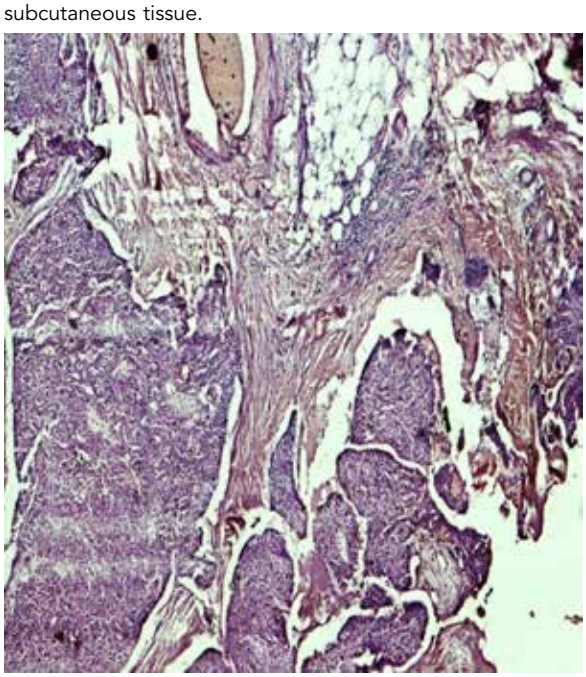

Fig 5a - Malignant Clear cell Hidradenoma 4300-11 (H&E, x10) The photomicrograph shows a cellular tumour tissue with infiltrative borders into the subcutis.

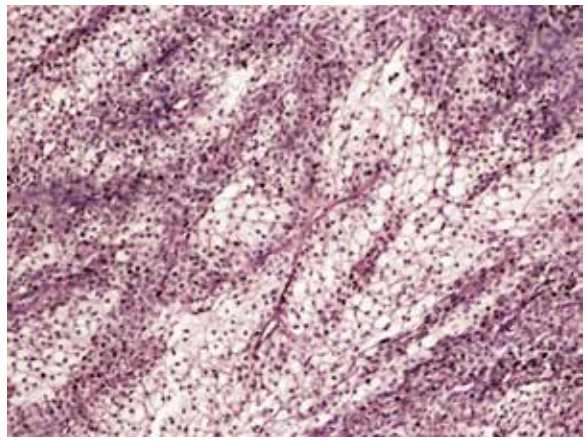

fig 5b - Malignant Clear cell Hidradenoma 4300-11 (H&E, x400) The photomicrograph shows tumor tissue comprising of both clear & polygonal cells along with atypical mitoses.

A case Mucinous eccrine carcinoma(figure 6) is reported, in a male patient of age 60yrs who presented with a swelling of size 4x3x2 cm ,in anterior axillary line. Cut section of the tumour showed solid grey-white mass with mucinous areas.

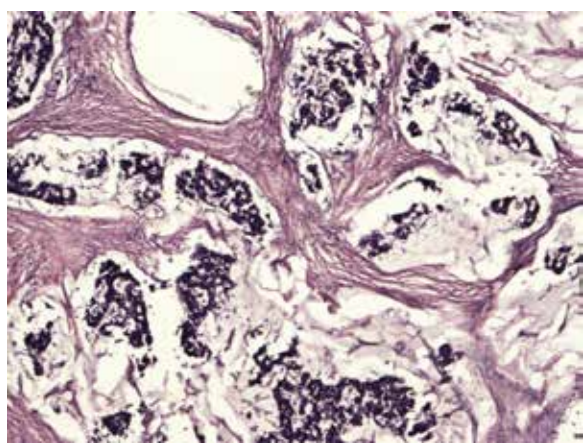

Fig 6 - Mucinous Eccrine carcinoma 330-11 (H&E, x100) The photomicrograph shows tumor, divided into numerous compartments containing abundant amount of mucin surrounding the small islets of tumor cell

Discussion :

Adnexal neoplasms are rare tumours. The 3year study was taken on 282 skin biopsies of which 14(4.96%) cases were adnexal tumours. 85% were benign and 15% were malignant which was also seen in Ankit Sharma etal study[15]. The commonest were of eccrine origin consisting of 7cases (53.3%). Tumours of sebaceous origin are least common, comprising a single case(6.66%). Adnexal tumours classification is voluminous, hence few commonest tumours received by us are discussed.

Pilomatricomas were the commonest tumours of trichilemmal origin at ASRAMS, in which the shadow cells are characteristic having distinct cell border and central unstained area as a shadow of lost nucleus. Pilomatricoma should be differentiated from the wall of trichilemmal cyst, in which the peripheral basophilic cells show a palisading pattern and absence of typical shadow cells.

Sebaceous adenoma is characterised by irregular lobules

Volume : 6 | Issue : 8 | August 2016 | ISSN - 2249-555X | IF : 3.919 | IC Value : 74.50

of both undifferentiated basaloid cells and mature sebaceous cells. It should be differentiated from sebaceous hyperplasias, which consist of predominantly mature sebaceous cells in the lobules, and sebacoma which consists of mainly undifferentiated basaloid cells. Tamer etal and Smith J etal has commented that this lesion is usually documented in patients with Muir-Torre syndrome.[1,3]

Tamer etal and Smith J etal has reported that apocrine fibroadenoma occurs mostly in perianal and axillary region, histologically resembling breast tissue, consisting of predominantly stromal component & apocrine lumina with decapitation secretions.[1,2] These features were similar to apocrine fibroadenoma in our case. Ek cib etal also reported that apocrine fibroadenoma can be associated with perianal fistula even.[4] Syringocystadenoma papilliferum consists of cystic invagination extending from the epidermis into the dermis, with numerous papillary projections. The papillary projections lined by 2 rows of cells with luminal columnar cells showing active decapitation and an outer cuboidal cells. Plasma cells are characteristically seen in papillary core, which are clearly noted in our case. It is commonly seen in scalp or face, and known to occur in other sites. As compared to E1 Fatoiki F etal and Pawal P etal[5,6] study, this lesion was presented in the scalp region. Hidradenoma papilliferum displayed an adenoma with apocrine differentiation, located in the dermis with tubular cystic structures and papillary infoldings. It is common in females, in the perineal region and perianal region. Tammer etal, Rosmaninho AD etal, Morinura S etal documented a rare location of hidradenoma papilliferumm at eyelids and abdomen.[1,7,8] Luminal cells of the tumour contain large PAS positive, diastase resistant granules.

Tumours with eccrine differentiation constituited 7cases(50%), of the total adnexal tumours in our study. Eccrine spiradenoma consists of well demarcated tumour lobules with sheets of epithelial cells, arranged in intertwining cords. Eccrine spiradenoma at ASRAMS was presented on the right lower lid, scalp and neck in the 3 different cases. Tamer etal, Churcil Livingstone etal, Rodrignez – Martin M etal has reported eccrine spiradenoma with rosteriform and linear pattern of arrangement.[1,9,10] Eccrine poroma consists of broad anastamosing bands emanating from the epidermis. The tumour comprises of uniformly small cuboidal cells & are connected by intercellular bridges. It usually presents on the plantar region, leg, hands & fingers as a solitary lesion. Cases can also present in other rare sites as in Kang Mc etal study.[11]. A case of Eccrine poroma was presented as right lower leg swelling in a female patient at ASRAMS. Clear cell hidradenoma comprises of multiple lobules of tumour cells in the dermis, with focal cystic change. The tumour cells are clear & polygonal cells with foci of eosinophilic material & small ductal lumina. Our case showed similar picture along with nuclear grooves in both clear & polygonal cells as compared to Kurshid A etal.[12] Single case of this tumour was evaluated in a male patient as a swelling on scalp region at ASRAMS.

Malignant clear cell hidradenoma is a large tumour with infiltrative borders. There are sheets of atypical tumour cells consisting of both clear cells and polygonal cells, exhibiting mitotic activity.The tumour proper revealed lobules of clear and eosinophilic polygonal cells, with mitotic figures, invading the lower dermis and subcutis. Souvatzidis P etal noticed that the survival time is inversely proportional to the size of the tumour.[13] It usually presents on the head, trunk and distal extremities.

Mucinous eccrine carcinoma histologically shows tumour tissue which is divided into numerous compartments with abundant pools of mucin surrounding the small tumour islets with eccrine differentiation. The tumor islands show mild cytological atypia. Similar features were noted in our case. Tamer etal and Akinicim etal concluded that mucinous eccrine carcinoma is an uncommon tumor that should be differentiated from primary & metastatic mucinous adenocarcinoma.[1,14]

Conclusion :

Clinically adnexal neoplasms should be regarded as precursor of malignancy. So, critical evaluation of the skin biopsies is essential. The commonest tumour encountered were of eccrine differentiation, followed by apocrine and trichilemmal differentiation and the least common in our study are sebaceous tumors.

References:

1. Tamer S.S. Ahmed, Joseph Del Priore, John T.Seykora In:Lever's Histopathology of skin .Tenth edition Philadephia;Wolters Kluwer(India),2009 ;851-909.
2. Churchil,Living stone,Elsevier. Tumors of cutaneous appendages. In: Weedons skin Pathology.3rd edition. British library cataloguing in publication data, 2010;758 - 807.
3. Smith J, CroweK, Mc Gaughran J,etal. Sebaceous adenoma arising with in an ovarian mature cystic teratoma in MuirTorre syndrome. Ann Diagn Pathol 2011, June16.
4. Ek ciB,GokeeO,Ozkan F. Apocrine fibroadenoma of perianal region associated with perianal fistula[Journal article].J Clinical med Res2010,Oct11;2(5).239-42.
5. E1 fatoiki F,Kadir K,Quakadi A,etal. Syringocystadenoma papilliferum:Unusual location[Journal article].dermatol Res Pract 2010;709371
6. PalwaP,KaushalS,GuptaS,etal.Linear Syringocysadenoma papilliferum:an unusual location[Journal article].Pediatr Dermatol 2011;17(6);7
7. Rosmaninho AD,de AlmediaMT,CostaV,etal. Ectopic hidradenoma papilliferum [Journal article].Dermatol Res Pract2010;709371
8. MorinuraS,KadonoT,SugayaM,etal.Ectopic Hidradenoma papilliferum on abdomen.Eur J Dermatol ,2011Mar16.
9. A lfonso-Trujilo-1,Arteaga-Hernandez E,perez suarezJC(Eccrine spiradenoma is a zosteriform distribution:Presentation of a case).Actas Dermosifiliogr 2009 sep;100(7);619-20.
10. Rodriguez-MartinM,SandreZ GonzalezR,Saez- RodriguezM,etal. An unusual case of congenital linear eccrine spiradenoma [Journal article].Pediatr Dermatol2009,Mar-Apr;26(2):180-3.
11. KangMC, Kim SA, Lee KS etal. A case of an unusual eccrine poroma on left forearm area. [Journal article].Ann Dermatol 2011 May;23(2):250-3
12. Kurshid A, YaqoobN,DevanHA,PervezS, 'Nuclear grooves' in Nodular hidradenoma:frequency & significance of an unrecognised histopathological feature[Journal article].JCutan Pathol2007 Nov;34(11):871-5.
13. SouvatzidisP,SbanoP,Mandato F,FimianiM,CastelliA.Malignant Nodular hidradenoma of skin:report of 7cases. J Eur Acad Dermatol Venereol,2008May;22(5);549-54
14. AkinciM,SahanA,CetinB,etal.Primary mucinous eccrine adenocarcinoma of skin in a 69yr oldman[case report,Journal article].Chirurgia(Bucur)2010Jan-Feb;105(1):109-11.
15. Ankit sharma, DeepakG.Paricharak, Jitendra Singh Nigam etal Histopathological study of skin adnexal tumors – Institutional study in South India. Journal of skin cancer,2014;feb