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INTERESTING LESIONS OF SKIN IN PAEDIATRIC AGE GROUP

[1]Dr. Shyamala Srujana*, [1]Dr. Syed Sajjad Saifullah Quadri, [2]Dr. Teki Satyasri and [3]Dr. O. Shravan Kumar

[1]Assistant Professors of Pathology, Department of Pathology, Gandhi Medical College, Secunderabad, Telangana.
[2]Assistant Professor, Department of DVL, Gandhi Medical College, Secunderabad, Telangana.
[3]Professor &HOD, Department of Pathology ,Gandhi Medical College, Secunderabad, Telangana.

Corresponding Author: Dr. Shyamala Srujana
Assistant Professors of Pathology, Department of Pathology, Gandhi Medical College, Secunderabad, Telangana.

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ABSTRACT
Benign skin tumors of childhood tend to regress spontaneously within few years. But some rare tumors in children, such as Pilomatricoma, Syringocystadenam papilleferum and xanthofibroma, which need to be excised completely should be kept in mind, for early diagnosis and better prognosis. Only 1-2% of skin tumors excised in children tend to be malignant. The ability to promptly diagnose and treat common benign tumors and to distinguish them from malignant lesions is very important in childhood. We hereby present three interesting lesions encountered in children.

KEYWORDS: Skin tumors, Pilomatricoma, Syringocystadenam papilleferum and xanthofibroma.

INTRODUCTION
Benign skin tumors of childhood tend to regress spontaneously within few years. but some rare tumors in children which need to be excised completely should be kept in mind, for early diagnosis and better prognosis. Only 1-2% of skin tumors excised in children tend to be malignant. The ability to promptly diagnose and treat common benign tumors and to distinguish them from malignant lesions is very important in childhood. We hereby present three interesting lesions encountered in children.

MATERIAL AND METHODS
All the three cases were investigated for routine surgical profile. Excission biopsy was done in all the cases. Specimen was sent in formalin fixative for routine histopathology processing. H&E stain was done for all the cases. Special stains like PAS and Van Geison was done in xanthofibroma case to confirm the diagnosis.

DISCUSSION
CASE 1: A 3 year old male child presented with complaints of swelling on the scalp since 1 year. No history of itching or bleeding. The swelling started as a nodule of size 1cms, gradually increasing and attained present size with ulceration. O/e: single swelling of 3x1cms noted on the occipital region of the scalp, central ulceration +. Borders are regular. Base is not fixed to the deeper structures. No regional lymphadenopathy or other skin lesions noted.

Excision of the lesion was done and sent for histopathological examination.

Gross
Received single skin covered soft tissue mass measuring 3x1x0.5cms with central ulceration measuring 0.5cms in diameter. c/s: grey white with cystic areas noted.

Microscopic examination: sections showed epidermis and dermis. Epidermis shows focal ulceration with papillomatosis with cystic invaginations into the dermis. These invaginations showed many papillae which were lined by cuboidal to columnar epithelial cells with oval nuclei and pale eoisnophilic cytoplasm. (Figs 1,2).There is dense lymphoplamacytic infiltrate in the stroma.

Features were suggestive of Syringocystadenoma papilleferum.

DISCUSSION
Syringocystadenoma papilliferum is an rare uncommon adnexal sweat gland neoplasm mostly involving the head and neck region, most commonly on the scalp or the face.Syringocystadenoma papilliferum is a rare neoplasm, commonly appearing at birth, during infancy or around the time of puberty[1]

Syringocystadenoma papilliferum occurs with equal frequency in both sexes[2]

The lesion usually measures between 1 to 3 cm and <4cms in diameter[3]

Three clinical types have been described

a. Plaque type

Presenting as an alopecic patch on the scalp and may enlarge during puberty to become nodular, verrucous or crusted Plaques commonly tend to be associated with a naevus sebaceous of Jadassohn in one-third of the cases.

b. Linear type

Consists of multiple reddish pink firm papules or umbilicated nodules 1-10 mm in size commonly occurring over face and neck.

c. Solitary nodular type

They are domed pedunculated nodules 5-10 mm in size with a predilection for the trunk shoulder and axillae.

It may occur de-novo or within a nevus sebaceous. In the majority of the cases, there is a coexistent nevus sebaceous. The precursor lesion for it is organoid naevi, in which it commonly arises it.[2]

Regarding origin of tumor, Yamamoto *et al*.[4] postulated it to be of pluripotent cells based on immunohistochemical and ultrastructural grounds,.

Böni *et al*.[5] showed evidence for mutations in PTCH or P16 tumour suppressor genes

Syringocystadenoma papilliferum can exhibit both apocrine and of eccrine differentiation, as shown by positive immunoreactivity with gross cystic disease fluid proteins 15,24 and 13, suggestive of apocrine differentiation[6] and for cytokeratins, suggestive of differentiation of eccrine glands[7]

It is probable that these tumor arises from undifferentiated cells with capacity to exhibit both apocrine and eccrine secretion.

Basal Cell Carcinoma (BCC) development has been reported in upto 10% of the cases.[8]

Squamous cell carcinoma (SCC) may also develop, but much less frequently. Till now, only two cases of verrucous carcinoma in conjunction with Syringocystadenoma papilliferum, have been published.[9]

Our case is a 3 year old male child who presented with lesion in the scalp.

Histopathologically, the epidermis shows varying degrees of papillomatosis. One or several cystic invaginations extend downward from the epidermis. Commonly malformed sebaceous glands and hair structures are seen in syringocystadenoma papilliferum.

The only treatment for syringocystadenoma papilliferum is excision biopsy.

CONCLUSION

Syringocystadenoma papilliferum is a rare neoplasm, present most commonly on head and neck area, occuirng in paediatric age group. The excision of the tumor is sufficient in most of the cases.

Case 2: A one year old child girl presented with a firm swelling in the left ear lobe of 4 months duration. The lesion gradually increased in size. No significant history was noted. Physical examination revealed a firm mobile nodular mass measuring 1.2x1cms. overlying skin showed reddish appearance. clinical diagnosis of vascular lesion was considered. Swelling was excised and sent for histopathological examination.

Gross

Recieved specimen as greyish brown soft tissue bits altogether measuring 1x1cms.

Microscopic examination showed tumor composed of an epithelial component exhibiting the mixed type of basaloid and ghost cells (Fig 4) along with fibroblastic connective tissue stroma. The basaloid cells are characterized by round to oval, hyperchromatic nuclei and scanty cytoplasm. Ghost cells (Fig 5) appear eosinophilic with a central unstained shadow in the site of the lost nucleus. Scattered multinucleated giant cells, focii of calcification noted.

Based on these histopathological findings, diagnosis of pilomatricoma was made.

DISCUSSION: Pilomatrixoma is a rare, benign skin tumor arising from the hair follicle, commonly involving the head and neck region[10] It is seen in scalp, eyelids, arms and preauricular area.[11]

It typically presents as a slow-growing, freely mobile mass of the dermis. It is usually solitary, but multiple lesions have been reported.[12]

Pilomatrixoma, or calcifying epithelioma of Malherbe, was first described in 1880 by Malherbe and Chenantais.[13] Dubreuilh and Cazenave described the unique histopathologic characteristics of this neoplasm, including islands of epithelial cells and shadow cells.[14] Forbis and Helwig proposed the term pilomatrixoma[15] It shows female predominance.[16]

Other locations include the upper extremity, trunk and lower extremity in decreasing order of frequency.[17] They can develop at any age,but demonstrate bimodal peak presentation during the first and sixth decades , however, majority occur within the first two decades of life.[18]

Four distinct morphological stages of pilomatricoma are defined as: (a) early: small and cystic lesions, (b) fully developed: large and cystic, (c) early regressive: foci of basaloid cells, shadow cells and lymphocytic infiltrate

with multinucleated giant cells, (d) late regressive: numerous shadow cells, absence of basaloid and inflammatory cells, calcification and ossification may be present. Our case fit in the early regressive stage.

Histopathologically, pilom atricoma consists of lobules and nests of epithelial cells composed of two major cell types: basophilic cells and eosinophilic shadow cells. Early lesions show a predominance of basophilic cells grouped in islands at the tumor periphery. With tumor maturation, the basophilic cells acquire more cytoplasm and gradually lose their nuclei to become eosinophilic shadow cells. These latter cells constitute the central portion of the tumor and frequently calcify.

Treatment of pilomatricoma is complete surgical excision as spontaneous regression is uncommon. Malignant transformation to a pilomatrix carcinoma should be suspected in cases with repeated local recurrences.[19]

CONCLUSION: The main purpose of this article is tomake the clinicians aware of pilomatricoma.

Case 3: A 8 year old male child presented with a nodule on his right upper back since 1½ years, gradually increasing in size. No associated symptoms. On examination a swelling of size 3x2cms noted, just below the right scapula, soft to firm in consistency, freely mobile and non-tender. Overlying skin appears normal. FNAC done revealed fibroblastic tissue and few scattered foamy cells s/o? adnexal tumor?? histiocytic tumor.

Specimen was excised and sent for histopathological examination

Gross
Received skin covered soft tissue mass measuring 3x2cms.c/s shows grey white and yellow areas.

Microscopic examination showed tumor consisting of hypercellular proliferation of spindle cells (Figs6,7) and foamy histiocytes, located in the dermis (Fig8). The cells are arranged in fascicles and in sheets. The lesions extended through the full thickness of the dermis into the subcutis. Occasional spindle shaped cells showing atypia and occasional mitotic figures, suggesting the possibility of Atypical xanthofibroma.

Special Stains
PAS negative in the foamy xanthomatous cells(Fig 9).

Van Geison
Showed positivity for the collagen (Fig10).

DISCUSSION
Atypical xanthofibroma of the skin is the recently recognized pseudosarcomas. According to all the available data, it is benign, probably representing a reactive process in the dermis.[20] It characteristically occurs in the older age group in sun exposed areas of head and neck. The lesion is most frequently misdiagnosed as sarcoma.[21]

Our case is a child of 8 years old, which is a rare occurrence. The recognition of the atypical fibroxanthoma,which does not behave clinically in an aggressive manner, is of utmost importance.[22]

A major criteria stressed by Krupsan and Mc Gavran, for distinguishing Atypical xanthofibroma from sarcoma, as well as other malignancies, is the presence of monomorphic tumor cell populations in the malignant lesions,in contrast to the polyphasic tumor cell populations in the atypical xanthofibromas.[23]

The aetiology of atypical xanthofibroma of the skin is not known, although it most likely reperesents a reactive process of the dermis.

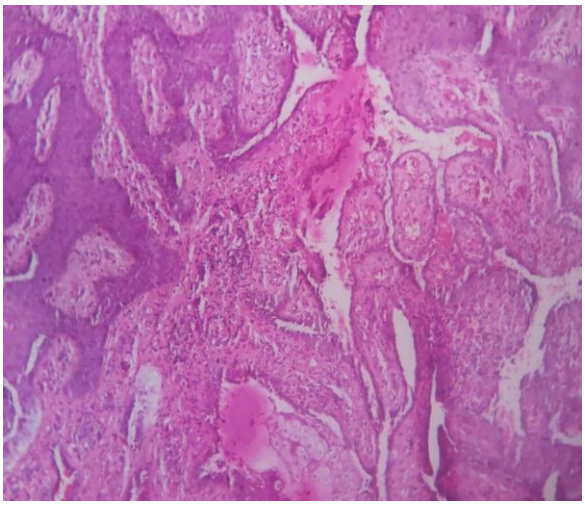

Fig 1: 4x: Syringocystadenoma papilleferum showing cystic invaginations into the dermis.

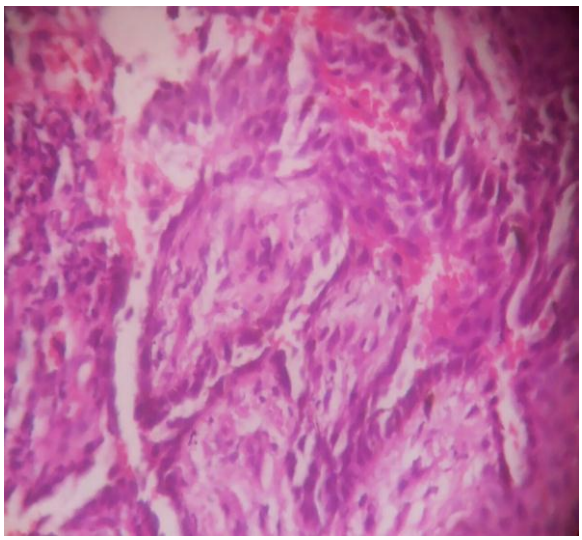

Fig 2: 40x: These invaginations showing many papillae lined by two types of cells.

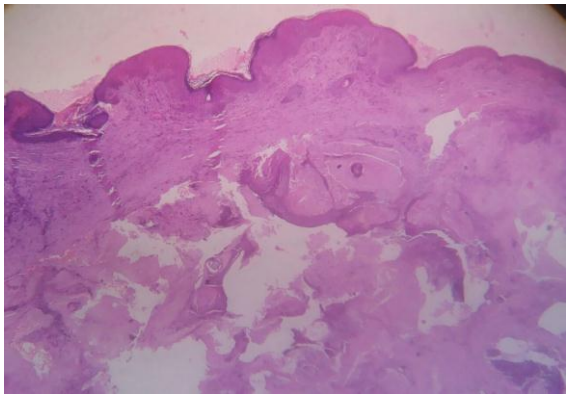

Fig 3: H&E: 4x: Scanner view of pilomatricoma

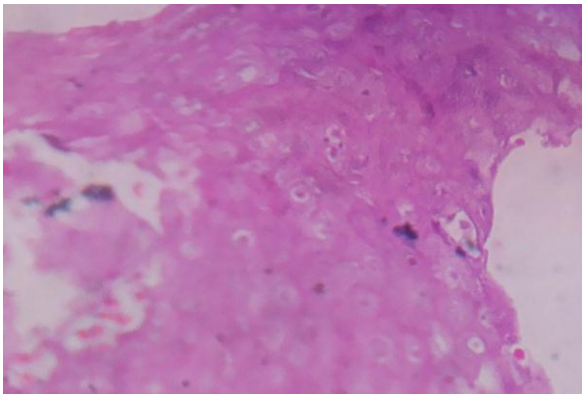

Fig 5: H&E: 40x: Tumor tissue showing ghost cells

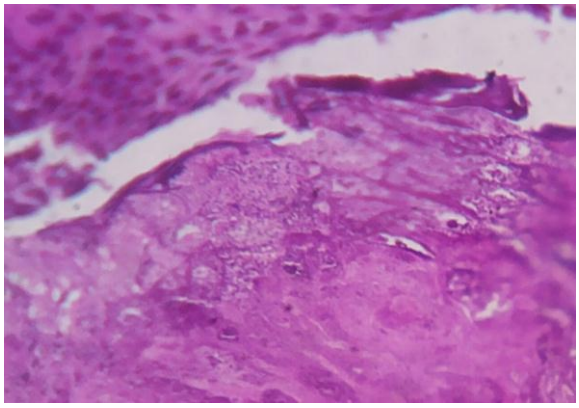

Fig 4: H&E: 4x: Pilomatricoma showing both ghost cells and basasloid cells.

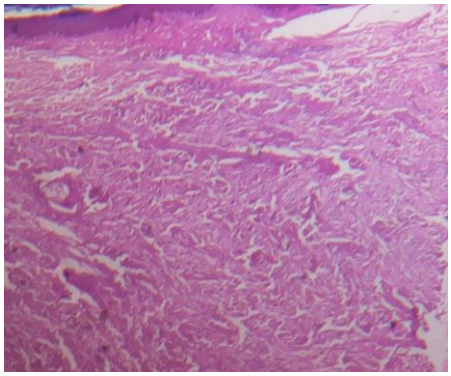

Fig 6: 4x- H&E: tumor in the dermis

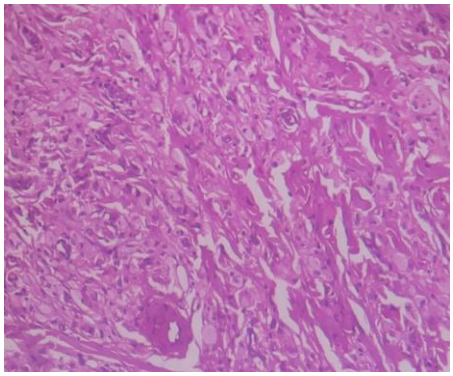

Fig 7: 10x -H&E: Tumor arranged in fascicles and sheets

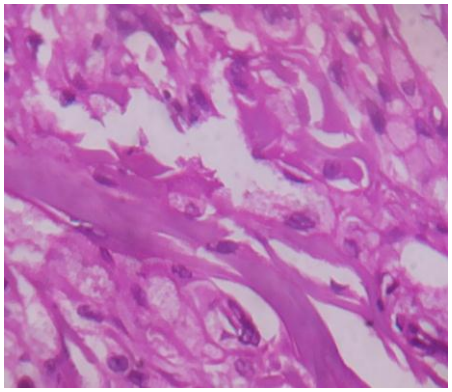

Fig 8: H&E: Foamy histiocytes located in the dermis

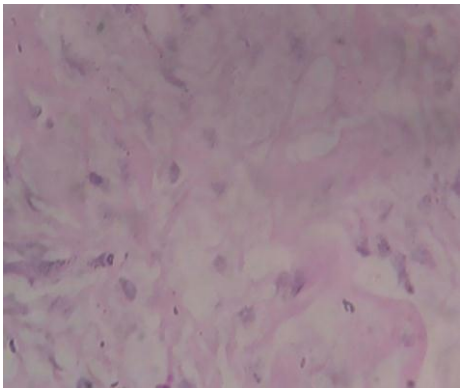

Fig 9:PAS stain:Negative in histiocytes

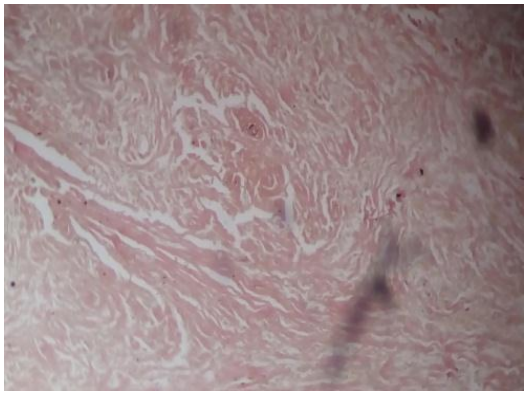

Fig 10: Van Geison stain:positive for the collagen (red colour)

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