

CHONDROID SYRINGOMA – A RARE MIXED APPENDAGEAL TUMOUR AT MULTIPLE SITES

Haritha Komeravelli¹, Parthasaradhi Anchala¹
¹Consultant Dermatologist

Corresponding Author:

Dr Haritha K

Anchala 'S Skin Institute & Research Centre, Road No B20, Journalist Colony, Jubilee Hills,, Hyderabad 500033, Telangana, India

Email: haritha_komeravelli@yahoo.com

Abstract

Chondroid syringomas are rare, generally benign, mixed tumors of the skin, composed of sweat gland elements. The tumor usually presents as benign asymptomatic, slowly-growing, subcutaneous or intradermal, single nodule occurring commonly over head and neck [3],[4]. We report a case of multiple, benign eccrine variant of Chondroid Syringoma, occurring on the scalp in 72yr old man, for its rarity and unique mode of presentation.

Key words: Chondroid syringoma, multiple, eccrine

Introduction

Chondroid syringomas are rare, generally benign, mixed tumors of the skin which were first described by Billroth (1859), that have both a benign and malignant form with an incidence of 0.01%–0.098% and of unknown etiopathogenesis. Hirsch and Helwig (1961) gave them the appellation chondroid syringoma, because of the presence of sweat gland elements which are set in a cartilaginous stroma.^{1,2} Morphologically it is considered to be the cutaneous counterpart of the pleomorphic adenoma of salivary glands, but differs from it in that it rarely recurs.³ Chondroid syringoma is often overlooked because of its rarity and the unremarkable clinical presentation. Hence, the tumour is typically diagnosed retrospectively from histopathological examination, and further classified as – apocrine and eccrine variants, apocrine being more common^{3,4}.

Case Report

A 72 yr old man presented with multiple, asymptomatic, painless, hard raised skin lesions on scalp of 30 years duration (fig 1-3). Lesions started as a few, small pea sized ones which progressively increased to the present size and were seen over scalp and forehead. There was no h/o any other skin or systemic

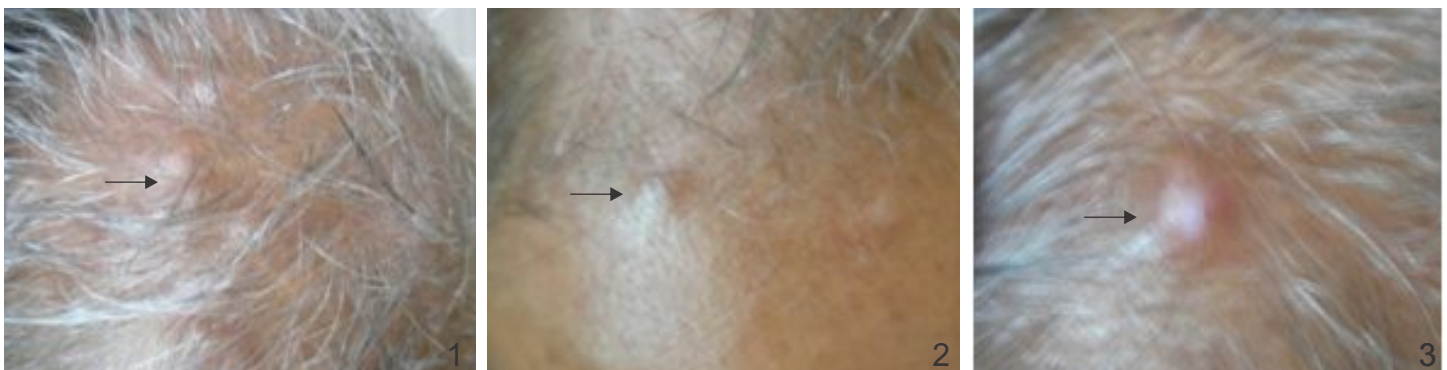
complaints. Cutaneous examination revealed multiple (12), polysized (0.5cm to 2cm), round to oval shaped, skin coloured, hard to soft, non tender papules and nodules over vertex (fig 1), forehead(fig 2), right temporal area of scalp(fig 3).

Skin over the lesions is not pinchable, and is mobile horizontally not vertically - not attached to the underlying structures. Excision biopsy was done and sent for histopathological examination keeping differential diagnoses:

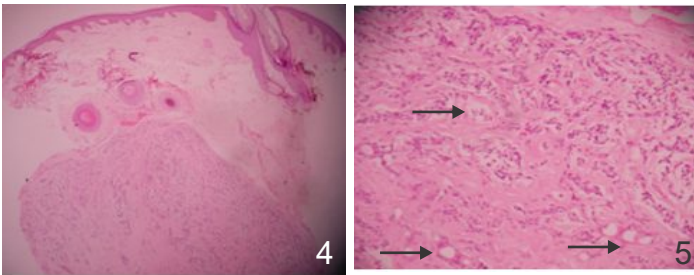
- (1) Sebaceous cyst (calcified) (2) Cylindroma (3) Spiradenoma.
- (4) Hidradenoma (5) Metastatic nodules (6) Calcinosis cutis.

Microscopic examination showed skin with underlying lesion proper, comprised of cells arranged in cords and trabecular pattern and with focal glandular pattern(fig 4-8).

Cords are separated by abundant mucoid stroma (fig 10). The cells are polygonal with bland nuclei and clear cytoplasm (fig 5 - 9). Few glands show bilayering of cells (fig: 6-8) No mitosis and no atypia. Eccrine variant- tadpole like acrosyringium (fig: 5-8) and negative for PAS diastase (rules out the apocrine variant). Positive Special stains for Chondroid Syringoma (fig: 9-12) S/O Benign Chondroid Syringoma having Eccrine variant.



Figs 1-3: Polysized, round to oval shaped, Papules and nodules.



Figs 4,5: Revealing tubule-cystic spaces in dermis (H&E;10x,40x)

Discussion

Hirsch & Helwig¹ proposed 5 criteria for diagnosis of chondroid syringoma: 1] Nests of polygonal/cuboidal cells. 2] Intercommunicating tubuloalveolar structures lined with 2 or more cuboidal cells. 3] Ductal structures composed of 1/2 rows of cuboidal cells. 4] Occasional keratinous cysts. 5] Matrix of mixed chondroid and myxoid composition. In the present case it almost fulfilled the criteria.

Headington^{3,4} divided CS into apocrine and eccrine variants based on luminal morphology.

1) The apocrine type: Is the most common type and demonstrates irregular branching tubules (tubulocystic pattern) lined by at least 2-cell-thick epithelium.

2) The eccrine type: characterized by rather uniform, all, round tubules that are evenly spaced within a myxoid-chondroid matrix. The present case was of eccrine type according to the Headington's criteria.

With constellation of clinical, histopathological and immunohistochemistry findings, following diagnosis was concluded: "Benign Multiple Chondroid Syringomas on scalp with Eccrine differentiation". Because of malignant potential, the usual first-line treatment is total excision of tumor without destroying aesthetic and functional structure and regular follow up.²

We are presenting this rarest case of benign chondroid syringoma, with multiple nodules without any atypia over long duration. Our case is of interest because histopathologically both the luminal variants and the tadpole (comma shaped) appearance of tubules are classically seen in eccrine variant of Chondroid Syringoma, which itself is a rare entity.

Though 400 cases of chondroid syringoma are reported worldwide there are very few cases of eccrine variant; still

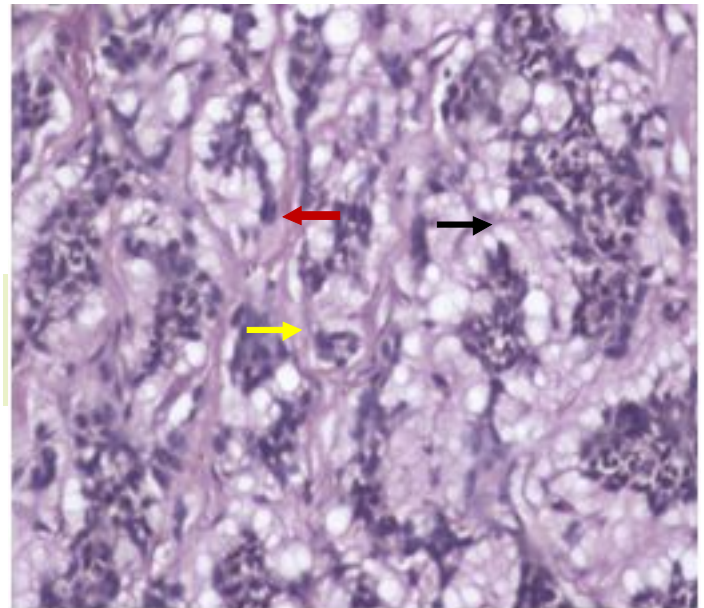


Fig9: Epithelial cells (black) and myoepithelial clear cells (red), against chondromyxoid matrix (yellow) (H&E, 100x)

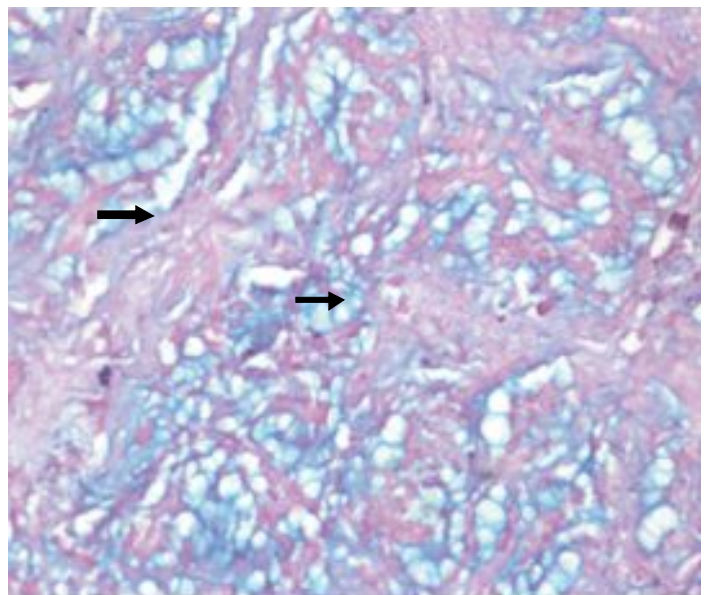
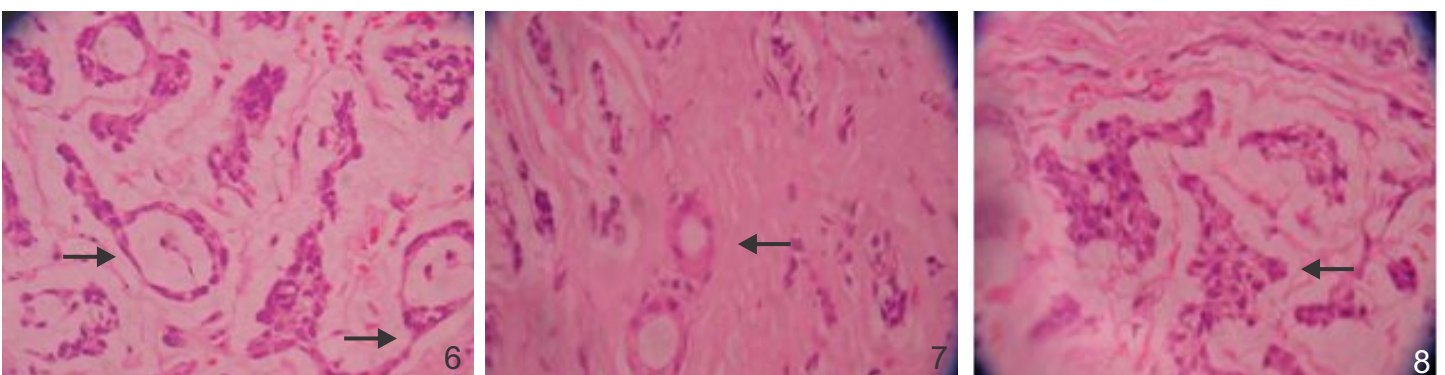


Fig10: Highlighting chondromyxoid matrix (ALCIAN PAS;100X)

further, eccrine variant presenting with multiple lesions are sparse.^{5,6,7}

When an elderly male patient presents with multiple asymptomatic lesions over scalp, neck or forehead, chondroid



Figs 6-8: Tadpole like acrosyringium- [eccrine variant (red)] and trabecular pattern (blue) (H&E;100x)

syringoma has to be considered as one of the diagnosis. Histopathology helps in confirmation and treatment of the disease.

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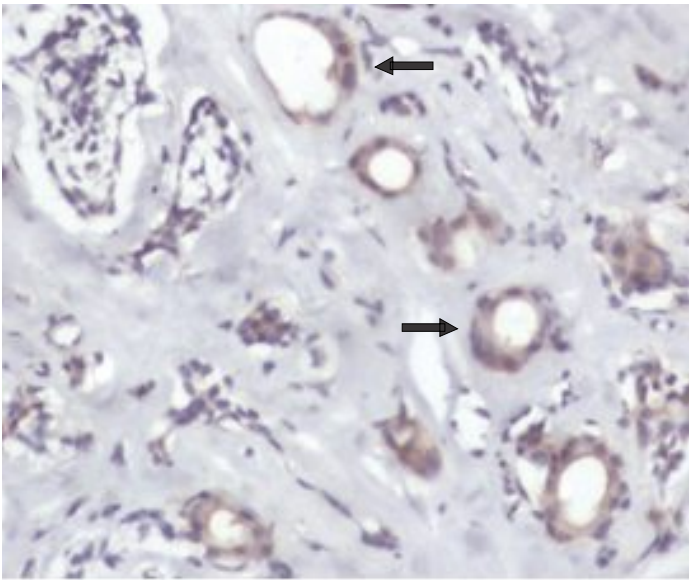


Fig 11: Highlighting the epithelial component (IHC with CYTOKERATIN;100X)

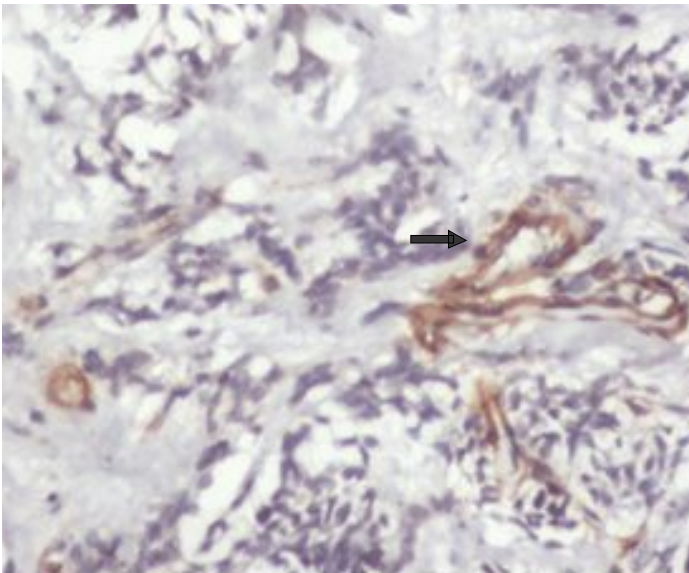


Fig 12: Highlighting the myoepithelial component (IHC with SMA;100X)