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Case Series

A rare case series of chondroid syringoma in three young patients

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ABSTRACT

Chondroid syringoma is also known as mixed tumour of the skin. It is a rare, biphasic cutaneous neoplasm similar to pleomorphic adenoma of salivary glands. Because of its rarity, many cases are misdiagnosed in the initial stages as cysts or other cutaneous adnexal neoplasms and are often identified only after being sent for histopathological examination. Although it usually presents in middle and older aged patients, we are presenting three younger patients with Chondroid syringoma, located over the back, nose and cheek, respectively.

Keywords: Chondroid syringoma , Mixed tumor, Cutaneous neoplasm

INTRODUCTION

Chondroid syringoma is a rare tumor of the skin.[1] The incidence of this tumor among primary skin tumors is reported to be less than 0.01%.[2] It was first described by Hirsch and Helwig.1 It is also known as mixed tumor of the skin as it has both epithelial and mesenchymal stromal components. It occurs as a slow-growing, small, cutaneous nodule or mass.[3] Eighty percent of the tumors are seen in older patients with male predominance.[4] It usually involves head and neck region and presents as asymptomatic slow growing, firm subcutaneous or intra dermal nodule. Histopathology reveals differentiation towards the adnexal ductal epithelium with chondromyxoid differentiation in the stroma.[5]

CASE SERIES

Case 1

A 42 year old female presented in surgery OPD with swelling on right side of the cheek since 1.5 year. Clinical diagnosis was made as hemangioma. Excisional biopsy was done and sent to histopathology lab.

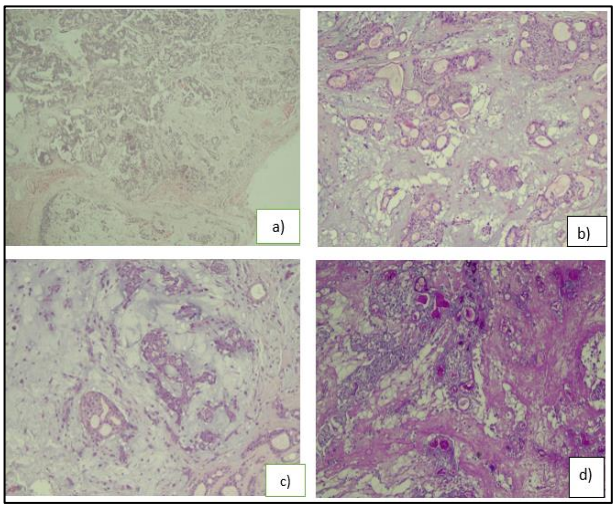

Figure 1: (a, b) Sections showed well encapsulated and circumscribed mass comprising of small and large group of epithelial cells arranged in ductules, branching tubules and scattered singly. (×40). c) Abundant myxoid stroma. d) Intraluminal amorphous eosinophilic material which is PAS positive and diastase resistant.

Grossly received a greyish brown globular structure measuring 2×1×0.8 cm. On microscopic examination sections showed well encapsulated and circumscribed mass comprising of small and large group of epithelial cells arranged in ductules, branching tubules and scattered singly. Tubules are lined by two layers of cells. Luminal cells are cuboidal and peripheral cells are flattened and are embedded in an abundant mucoid stroma. The lumen contains amorphous eosinophilic material which is PAS positive and diastase resistant. Occasional mitosis seen. Histopathological features are consistence with chondroid syringoma.

Case 2

A 24 years old male presented in surgery OPD with cystic swelling over dorsum of the nose. Clinical diagnosis was made as sebaceous cyst. Excisional biopsy was done and sent to histopathology lab. Grossly received single soft tissue piece, firm and greyish white in color measuring 0.9×0.7 cm. Microscopic examination showed keratinized squamous epithelium with follicular plugging. Mid dermis shows well circumscribed nodule comprising of branched tubular lumina, focally cystically dilated with eosinophilic amorphous secretions which are PAS positive and diastase resistant. Tubules are lined by two layers of epithelium, outer layer is flattened and inner cuboidal. The stroma is fibrotic and myxoid. Histopathological features are consistence with chondroid syringoma.

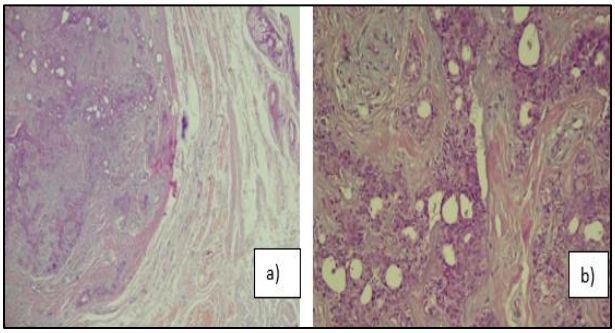

Figure 2: a) Well circumscribed nodule comprising of branched tubular lumina with adjacent skin adenexa. b) Group of epithelial cells arranged in ductules, branching tubules with myxoid stroma.

Case 3

A 31 years male presented in surgery OPD with cystic swelling on the back. Clinical diagnosis was made as infected sebaceous cyst. Excisional biopsy was done and sent to histopathology lab. Received multiple cyst wall linings greyish white in color collectively measuring 3×2×1 cm. Microscopic examination showed a well circumscribed nodule revealing predominantly epithelial and stromal component. The epithelial component comprised of small, round to oval cells with bland nuclei arranged in tubules, glands, cystic spaces, nests and focal papillae formations. Cystic spaces containing eosinophilic secretions that are PAS positive and diastase resistant. Stromal component comprised of myxoid material with focal hyalinized areas. Dilated thin-walled vascular spaces are also present. One of the sections shows stripes of fibrous wall of the cyst. Histopathological features were consistent with chondroid syringoma.

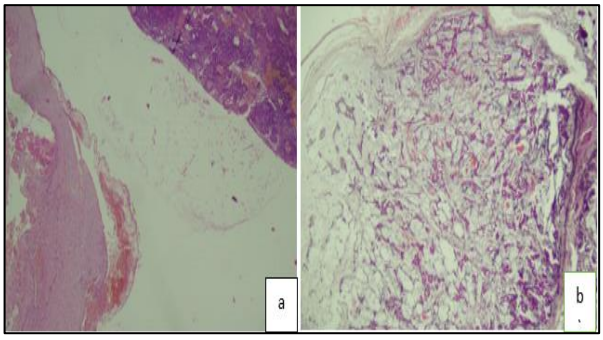

Figure 3: a) The sections shows stripes of fibrous wall of the cyst. b) The epithelial component comprised of small, round to oval cells with bland nuclei arranged in tubules, glands, cystic spaces, nests and focal papillae formations.

DISCUSSION

The Chondroid syringoma is a rare mixed tumor of sweat gland. It has both a benign and malignant form.[3] Virchow and Minssen coined the term mixed tumors for neoplasms with microscopic features that indicated both epithelial and mesenchymal origin.[2] Chondroid syringoma commonly presents as a single, firm, circumscribed, non-ulcerated, asymptomatic, slow growing, lobulated nodule within the deep dermis or subcutaneous tissue in middle or older aged male patients.[1] Its usual size is 0.5 cm to 3 cm. Some cases of large and even multifocal tumors have been noted. Head and neck area, particularly cheek, nose, or skin above the lip, are the common sites for occurrence of chondroid syringoma, though it may occur anywhere over the body.[3] Clinical manifestations are non-specific.

The diagnosis is typically made histopathologically. Hirsch and Helwig have proposed five histopathological criteria for the diagnosis of chondroid syringoma, which include: nests of cuboidal or polygonal cells, tubuloalveolar structures, that is gland like elements lined with 2 or more rows of cuboidal cells (depending on plane of sectioning) morphologically similar to those in the nests of cuboidal cells. In serial sections the tubuloalveolar structures are seen to intercommunicate. Ductal structures which are composed of 1 or 2 rows of cuboidal cells, either occurring alone or connected with tubuloalveolar structures. The periphery of the ductal structures sometimes showed tail-like protuberances. Occasionally keratinous cysts of varying sizes from very small to large. The epithelial wall was lined with flattened squamoid cells. Matrix of varying appearance in hematoxylin and eosin-stained sections (a) homogeneous faintly bluish chondroid substance was the most common; (b) bluish,

foamy appearing material was very likely the precursor of the chondroid substance; (c) Least common was the eosinophilic, homogeneous, and hyaline material.[6] The stroma of chondroid syringoma is myxoid, chondroid, mucoid, adipose, and rarely osteoid.[5] The stroma is stained positive for periodic acid-Schiff, and Alcian blue, and is focal positivity shown for keratin, desmin, vimentin, and S-100 protein with an immunohistochemical study.[2]

In 1961, Headington divided Chondroid syringoma into two groups, based on their histopathological appearance which includes apocrine type and eccrine type.[3] The eccrine type with uniform small round tubules, lined by a single row of epithelial cells and the apocrine variant with tubular and cystic branching lumina, lined by two rows of epithelial cells. The malignant form usually occurs predominantly in females. It has no age-related predilection and is observed more commonly on extremities. Poorly differentiated chondroid components and excessive amounts of mucoid matrix and are important indicators of the tumor malignancy and metastatic potential.[1] Multiple treatment modalities are available, including dermabrasion, vaporization by laser, electrodessication, and conventional excision, with the last preferred for complete removal.[7] Fine needle aspiration cytology (FNAC) may be used to diagnose the pathology before excision.[8] The optimal treatment of benign chondroid syringomas is surgical excision. It is important to include a margin of normal tissue with the excision of complete removal of the tumor because of the lobulated nature of the tumor, otherwise local recurrence may occur.[1] It is difficult to diagnose chondroid syringoma clinically or radiologically. All of our cases were misdiagnosed. Excisional biopsy with histopathological examination and histochemical stains remains the gold standard.[2]

CONCLUSION

Chondroid syringoma usually occurs in middle to older age men but all our cases were patients of younger age group. It is difficult to diagnose chondroid syringoma clinically and radiologically. All our cases were misdiagnosed clinically so histopathological examination ssis very important in reaching the diagnosis.

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