Case Report

Giant chondroid syringoma with divergent differentiation: Cyto-histo-immuno correlation

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Abstract

Chondroid syringoma is a rare benign skin adnexal tumor of eccrine/apocrine origin affecting commonly the head and neck region. It is also called as mixed tumor of skin because of the presence of both the epithelial and mesenchymal components. There are very few case reports of chondroid syringoma diagnosed on fine needle aspiration cytology (FNAC). We hereby report a case of giant chondroid syringoma occurring over the lower back in a 50-year-old male, diagnosed initially on cytology and confirmed by histopathology. This case is reported for its unusual size and site of occurrence. We describe the clinical features, cytology, histopathology, immunohistochemistry, and differential diagnosis of giant chondroid syringoma along with review of literature.

Key words: Adnexal tumor, chondroid syringoma, fine needle aspiration cytology, immunohistochemistry

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Introduction

Chondroid syringoma is a rare benign skin adnexal tumor of eccrine/apocrine origin. It is also called as mixed tumor of skin because of the presence of both the epithelial and mesenchymal components.[1] The reported incidence is <0.098%.[2] The commonest sites are head and neck regions and the extremities. The usual size is less than 3 cm.[3] There are very few case reports of chondroid syringoma diagnosed on fine needle aspiration cytology (FNAC). This case is reported because of unusual site of occurrence, large size, and showing varied histological features.

Case Report

A 50-year-old male presented with painless, progressively enlarging swelling over the lower back since 3 years. Examination revealed a firm non-tender swelling measuring 12 × 8 × 5 cm. Surface showed two nodules, largest measuring 6 × 5 × 4 cm and smallest 1.5 × 1 cm [Figure 1a]. Aspiration yielded thick, mucoid, and gelatinous material.

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Cytology

Smears showed cells arranged in loose cohesive clusters and in discretes, amidst which was chondromyxoid material [Figure 1b]. Cells were round to oval, centrally located nuclei having fine chromatin with moderate to abundant cytoplasm, a few showed one to two prominent nucleoli [Figure 1c] consistent with chondroid syringoma.

Histopathology findings

Gross

Received a skin covered soft tissue mass which measured 14 × 9.5 × 4 cm. Skin measured 12.5 × 8.5 cm. External surface showed two nodules, largest measured 6 × 5 ×4 cm and smallest 1.5 × 1 cm. Cut surface revealed a lobulated mass showing grey white myxoid areas [Figure 1d].

Microscopy

Revealed a well-demarcated neoplasm composed of round to oval cells arranged in sheets, tubuloalveolar, and branching cleft-like pattern. Tumor showed bands of eosinophilic material along with fibromyxoid stroma and chondroid areas. Areas showing lipomatous and clear cell changes [Figure 2a], cartilage [Figure 2b], osteoid bone

formation [Figure 2c], and foci of calcification [Figure 2d] were also observed.

Immunohistochemistry

The tumor cells arranged in tubuloalveolar pattern showed expression of IHC markers. The inner layer of epithelial cells were positive for pancytokeratin (AE1 + AE2) [Figure 3a] and the outer layers expressed vimentin (V9) [Figure 3b]. Mouse monoclonal antibodies manufactured by Biogenix were used for our study.

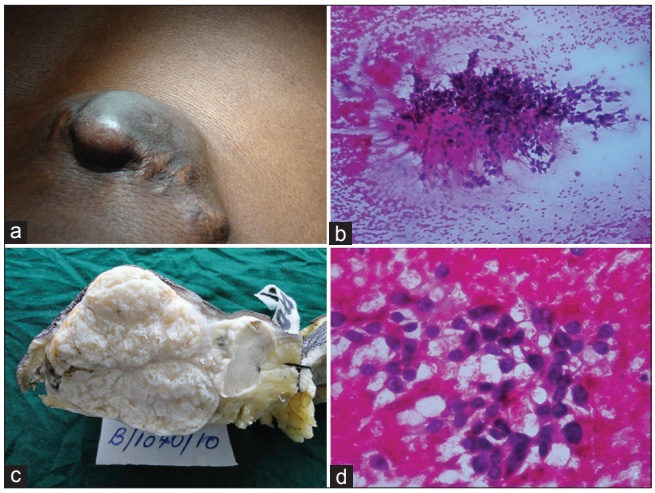

Figure 1: (a) Swelling over the lower back with surface showing two nodules. (b) Smear showing clusters of round cells with moderate to abundant cytoplasm embedded in chondromyxoid ground substance (H and E, ×100). (c) A cut-surface revealed a lobulated mass showing grey white myxoid areas. (d) A few tumor cells showed 1-2 prominent nucleoli (H and E, ×400)

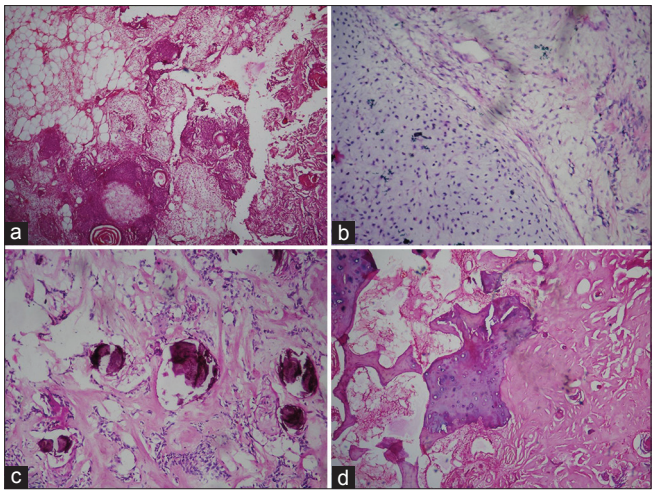

Figure 2: Microphotograph showing (a) chondroid syringoma with lipomatous differentiation and clear cell changes (H and E, ×100). (b) Cartilage (H and E, ×400). (c) Focal areas of calcification (H and E, ×400). (d) Osteoid bone formation (H and E, ×100)

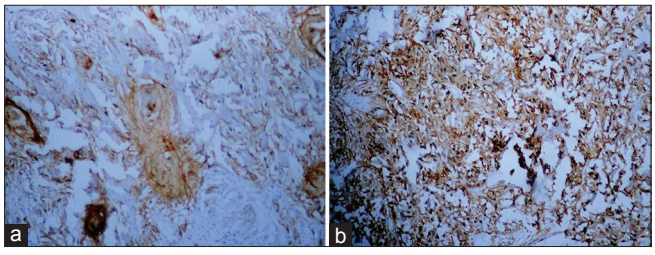

Figure 3: Microphotograph of tumor cells showing (a) cytokeratin positivity (CK, ×100). (b) Vimentin positivity (VIM, ×100)

Discussion

Hirsch and Helwig introduced the term *chondroid syringoma* in 1961 because of the cartilaginous matrix (chondroid) and sweat gland elements (syringoma).[4] It is usually seen in middle aged or older male patients. Clinically, they present as slow growing, painless, firm, subcutaneous, or intracutaneous nodule. The lesion usually measures 0.5-3 cm.[2,3,5] The commonest sites are head and neck region; particularly, cheek, nose, or lip. There are reported cases occurring over the scalp, eyelid, orbit, auditory canal, hand, foot, axillary region, abdomen, penis, vulva, and scrotum.[6,7] Giant chondroid syringomas occurring at unusual sites such as axilla,[8] arm,[2] and shoulder[9] have been reported.

Fine needle aspirate yields thick, mucoid, and gelatinous material. Cytological examination shows epithelial and myoepithelial cells embedded in metachromatic chondromyxoid ground substance. The nuclei are monomorphic with finely dispersed nuclear chromatin, some may show eccentrically placed nuclei.[3,4,6]

Hirsch and Helwig[4] proposed the following five histological criteria for diagnosis: 1) Nests of cuboidal or polygonal cells; 2) intercommunicating tubuloalveolar structures lined with two or more rows of cuboidal cells; 3) ductal structures composed of one or two rows of cuboidal cells; 4) occasional keratinous cysts; and 5) a matrix of varying composition.[4] Headington[10] recognized two types, apocrine and eccrine. The apocrine type demonstrates irregular branching tubules (tubulocystic pattern) lined by at least two cell-thick epithelium. The eccrine type is characterized by rather uniform, small, round tubules that are evenly spaced within a myxoid-chondroid matrix. Histologically, follicular and sebaceous differentiations along with presence of Merkel cells were first described by Mohamed *et al*.[11] However, lipomatous differentiation and clear cell changes are also described. Osteoid formation cartilage and calcification[12] can occur. All these changes were seen in our case also.

Malignancy in chondroid syringoma (CS) is rare. However, there are reported cases of malignancy occurring in young female patients in the extremities and torso.[9] Tumors greater than 3 cm in size have a greater likelihood of malignancy. Histological features that suggest malignant transformation in a chondroid syringoma include cytologic atypia, infiltrative margins, satellite tumor nodules, tumor necrosis, and involvement of deep structures.[13]

Hence, careful follow-up is essential in cases where the tumor size is big as in our case.

The tubuloalveolar components of chondroid syringomas are composed of two layers of cells with different immunophenotypes. The inner layer expresses cytokeratin (CK), epithelial membrane antigen, and carcinoembryonic antigen, that is characteristic of cells with an epithelial lineage.[12] The outer layers express vimentin, S-100 protein, neuron-specific enolase, and in a few cases, glial fibrillary acidic protein[12] that is characteristic of both mesenchymal and epithelial differentiation.

Mills opined that mixed tumors are monoclonal neoplasms with replicating cells that have the ability to differentiate toward epithelium or mesenchyme, and may account for the histologic variability of mixed tumors of the skin.[14]

Differential diagnosis for chondroid syringoma clinically includes dermoid or sebaceous cysts, neurofibroma, dermatofibroma, basal cell carcinoma, pilomatricoma, histiocytoma, and seborrheic keratosis.[2,3,5]

Chondroid syringoma may be associated with hydrocystoma like changes.[15] Apocrine or eccrine hydrocystomas are positive for CK, human milk fat globulin 1.[16] But they are negative for vimentin. Hence in our case, we employed both pancytokeratin and vimentin to confirm chondroid syringomas with tubuloalveolar pattern showing biphenotypic expression, thus ruling out hydrocystomas.

The optimal treatment for chondroid syringoma is surgical excision. Other modalities of treatment such as electrodesiccation, dermabrasion, and vaporization with argon or CO_2 laser have been used.[5] FNAC has been used for diagnostic purposes and may prove useful to determine pathology before excision; however, examination of excised tissue is most reliable in establishing a definitive diagnosis and to rule out malignancy. Because of the lobulated nature of the tumor, it is important to include a margin of normal tissue with the excision to ensure complete removal of the tumor, otherwise there are chances of local recurrence.

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