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Chondroid syringoma – A case report

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Abstract

Chondroid syringoma, also known as “mixed tumour of the skin”, is a rare cutaneous sweat gland tumour with the potential for malignant transformation and distant metastasis. It was first described by Bill Roth. The incidence of chondroid syringoma is reported to be less than 0.01% of all primary skin tumours. The site of predilection for benign chondroid syringoma is the head and neck region. In contrast, malignant chondroid syringoma is more commonly encountered in the extremities and is characterized by rapid growth, local invasion, and distant metastasis. We hereby report an unusual case of benign chondroid syringoma of foot in a 63-year-old male.

Keywords: Chondroid syringoma, primary skin tumours.

Introduction

Skin adnexal neoplasms/tumours (SAT) consist of a wide spectrum of benign and malignant tumours that exhibit morphological differentiation towards one or more types of adnexal structures found in normal skin – pilosebaceous unit, eccrine or apocrine glands. Most adnexal neoplasms are relatively rarely encountered in routine practice. SAT may display more than one line of differentiation (hybrid/composite tumours), rendering precise classification of these tumours difficult. The diagnosis of these mixed SAT relies on histopathological evaluation, and they are usually classified according to predominant morphological component.

The histogenesis of mixed adnexal tumours is still uncertain; however, the possibility of origin from pluripotent stem cells is suggestive. Most SAT are benign, and local complete surgical excision is curative. However, diagnosing some of these tumours has important implications, as they might be markers for syndromes associated with internal malignancies, such as trichilemmomas in Cowden disease and sebaceous tumours in Muir-Torre syndrome.

A malignant counterpart of each SAT has been described. These tumours are rare, locally aggressive, and have the potential for nodal involvement and distant metastasis,

with a poor prognosis. Owing to their different derivation and broad histogenesis, diagnosing these tumours may be challenging. [1]

Tumours of cutaneous sweat glands are uncommon, with a wide histological spectrum, complex classification and many different terms often used to describe the same tumour. Furthermore, many eccrine/ apocrine lesions coexist within hamartomas or within lesions with composite/mixed differentiation.

Chondroid syringoma is one of the uncommon benign cutaneous sweat gland tumours of mixed origin. It presents as a slowly growing, painless, firm nodule, generally occurring in the head and neck region, but most commonly affecting the nose, cheek and upper lip. Less frequently, the extremities, trunk or back may also be affected. Histologically, chondroid syringoma is similar to the benign mixed tumour of the salivary glands, having both epithelial and mesenchymal stromal components. Chondroid syringoma is classified into apocrine and eccrine types, based on the histopathological appearance of the sweat gland lumina within the tumour. Chondroid syringoma is best completely excised because of the possibility of local recurrence and malignant transformation. Malignant chondroid syringoma is very rare, and may arise de novo or from a pre-existing chondroid syringoma which has undergone malignant transformation. Histologically, there is cellular atypia, increased mitotic activity, infiltrative margins, satellite tumour nodules and tumour necrosis. Haematogenous and lymphatic spread to lymph nodes, lungs and bone may occur. The term “atypical chondroid syringoma” is used to describe tumours with histological features of malignancy as well as recurrence, local invasion and satellite tumour nodules, but without proved metastases. [2]

Case report

A 63-year-old male came to surgery out-patient department with chief complaints of swelling over sole of right foot associated with pain since 8 months. There was history of increase in the size of swelling. No history of trauma, fever or discharge from swelling. No history of similar complaints in the past.

On local examination, swelling of size approx. 6 x 5 cm present over right foot sole. It was well defined, non-tender and firm. Fluctuation was absent and slip sign negative (Figure 1). Clinically it was suspected as neoplastic origin, ultrasound guided fine needle aspiration was advised and done which showed sheets, clusters and few scattered epithelial cells. The cells were round to polygonal and, showed overlapping and crowding.

There was mild anisonucleosis and pleomorphism. Occasional moderately enlarged cells with high N:C ratio and hyper chromatic nuclei were seen. Occasional binucleated cells were also seen. Background showed scanty myxoid material and RBCs (Figure 4).

Cytological diagnosis was given as low-grade adnexal tumour.

Later, the tumour was excised under LA and sent for histopathology. On gross examination, we received an encapsulated, globular, brownish, firm tissue mass. Cut surface was yellowish white (Figures 2 & 3).

Microscopy showed a partly encapsulated triphasic tumour mass comoposed of epithelial, myoepithelial and mesenchymal components. The epithelial component showed tumour cells arranged in nests, tubules and ducts showing two layers – outer columnar epithelial layer and inner layer of cells with round to oval hyperchromatic nuclei showing open chromatin, with or without prominent nucleoli and abundant eosinophilic cytoplasm. There is mild anisonucleosis and pleomorphism, and

minimal mitosis. Few cells showed eccentrically placed nuclei. Few ducts showed decapitated secretions.

Scattered myoepithelial cells and plasmacytoid cells were seen at places in the stroma. Mesenchymal component mostly showed chondromyxoid stroma with scattered stellate cells and areas of scattered/clusters of adipocytes. There was NO evidence of severe atypia, markedly increased cellularity, increased abnormal mitoses, compression of surrounding tissue or tumour necrosis (Figures 5 to 9).

Histopathological diagnosis was given as benign mixed adnexal tumour (chondroid syringoma).

Discussion

In 1859, Theodor Bill Roth first described chondroid syringoma as "an entity having the same histopathologic properties as mixed tumours of the salivary glands." [3] However, it was not until 1961 that Hirsch and Helwig coined the term "chondroid syringoma," aptly named because of the histologic appearance of sweat gland features in a cartilage-like stroma. Chondroid syringoma is derived from epithelial and mesenchymal cells and comprises glandular elements of eccrine or apocrine type. Hirsch and Helwig defined the following histopathologic criteria for the characterization of chondroid syringoma: (1) nests of cuboidal or polygonal cells; (2) inter communicating tubulo-alveolar structures lined with 2 or more rows of cuboidal cells; (3) ductal structures composed of 1 or 2 rows of cuboidal cells; (4) occasional keratinous cysts; and (5) a matrix of varying composition. Chondroid syringoma may exhibit either all 5 characteristics or only some, with the most common feature being the nests of cuboidal or polygonal cells. [4]

Chondroid syringoma are mostly benign entities that usually present asymptomatically in middle-aged men with a predilection for the head and neck region. The gross appearance is typically described as a slow-growing, solitary, non-ulcerating mass ranging in size from 0.5 to 3.0 cm. However, cases of benign chondroid syringoma larger than 3.0 cm have been reported. Tumours larger than 3.0 cm are associated with a greater likelihood of malignancy. As of 2013, 30 cases of malignant chondroid syringoma have been described. Malignancy is more common in females, with no age predilection, and are observed more commonly on the extremities. Malignant chondroid syringoma typically arise de novo and rarely from a pre-existing benign chondroid syringoma. Histopathologic features that suggest malignancy include cytologic atypia, tumour necrosis, numerous mitoses, excessive mucoid matrix, and poorly differentiated chondroid components. [4]

It does not have characteristic clinical features; thus, the diagnosis is made based on histopathology. Important considerations in differential diagnosis of chondroid syringoma are neuro fibroma, derma to fibroma, pilomatricoma, histiocytoma, sebaceous cyst, seborrheic keratosis, basal cell carcinoma and squamous cell carcinoma. [5]

Total surgical excision is the treatment of choice for patients with benign chondroid syringoma. The risk of malignancy of this tumour is extremely rare, but it has been reported and therefore patients are recommended to have regular follow-up. [5]

Conclusion

Although chondroid syringoma is an uncommon tumour in the extremities, it should be considered as one of the differential diagnoses, especially when dealing with tumours of this area. Regular follow-up of the patient is recommended to evaluate the risk of malignancy and local recurrence.

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pilosebaceous unit. Journal of clinical pathology. 2007 Feb 1;60(2):129-44.

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Legend Figures

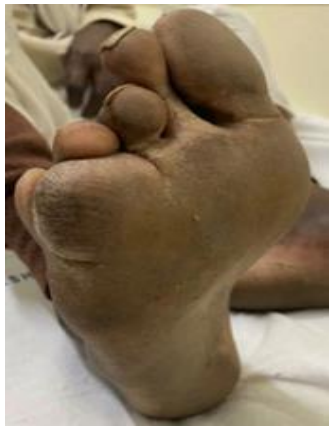

Figure 1: Swelling over sole of right

Figure 2: Grossly, an encapsulated, globular mass was received.

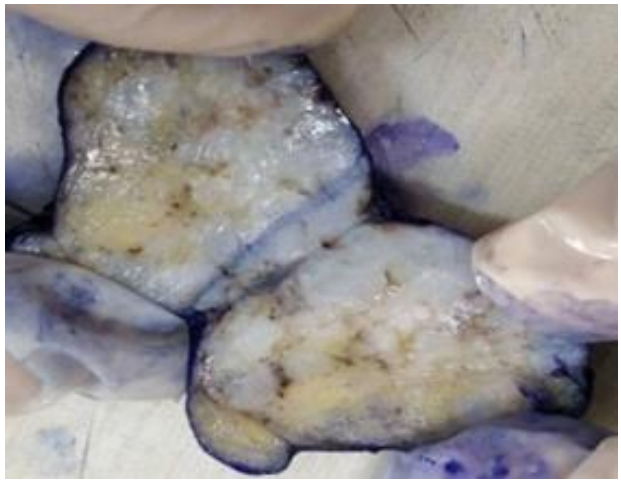

Figure 3: Yellowish white cut surface.

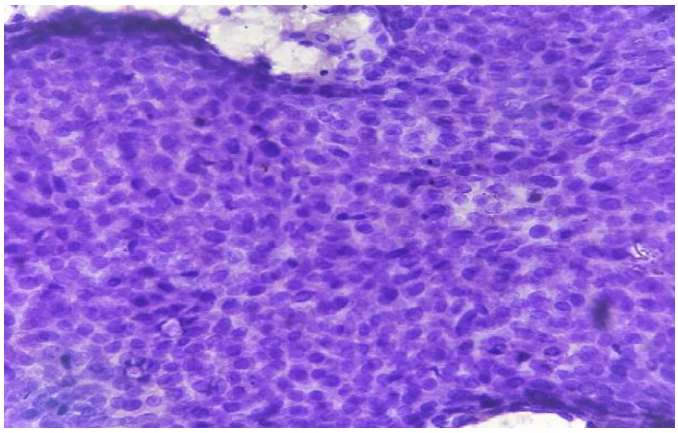

Figure 4: High power view H&E – Photomicrograph of FNAC showing sheet of epithelial cells with

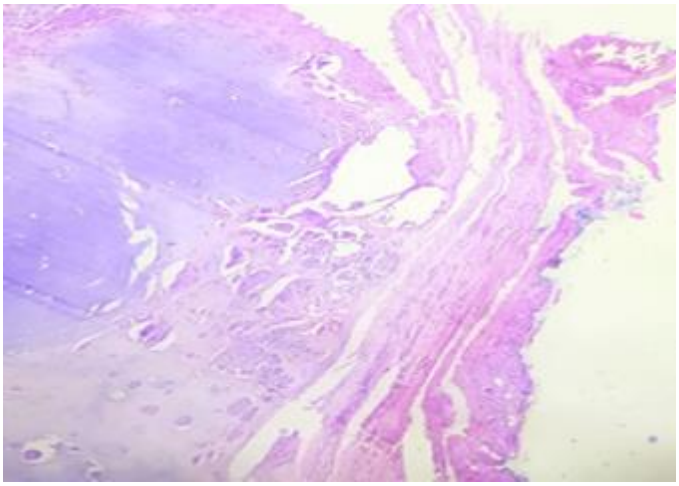

Figure 5: Scanner view H&E: Photomicrograph showing encapsulated triphasic tumour mass.

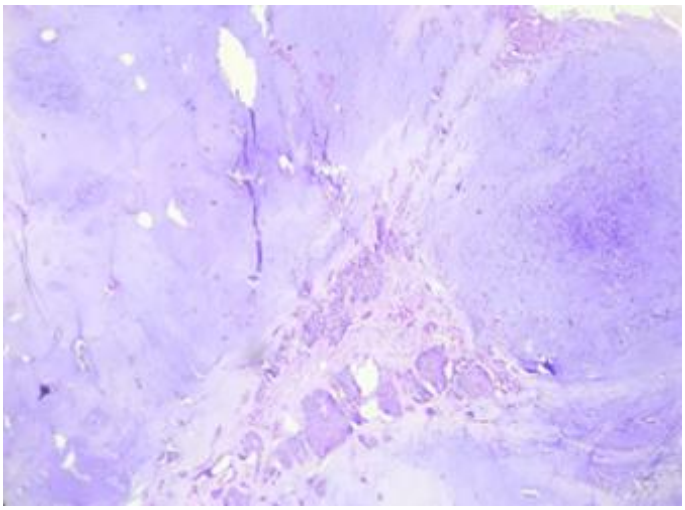

Figure 6: Scanner view H&E: Epithelial nests and chondromyxoid stroma.

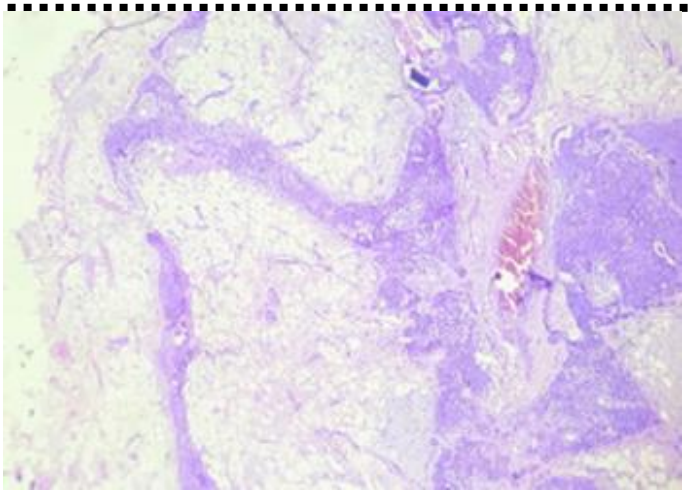

Figure 7: Low power view H&E: Epithelial cells arranged in tubular pattern.

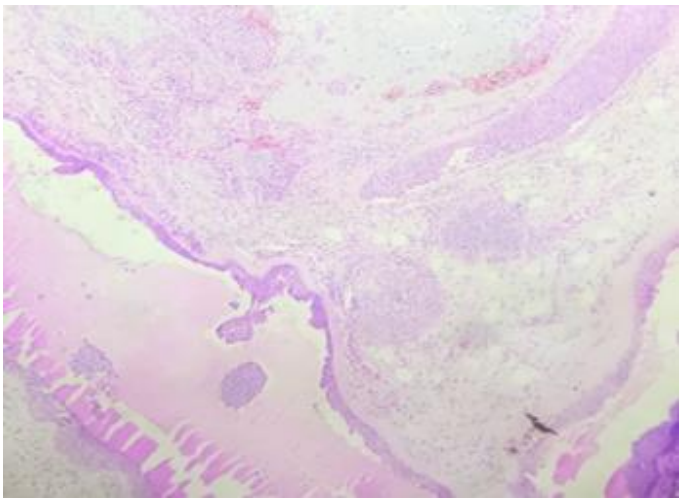

Figure 8: Low power view H&E: Ducts showing decapitated secretions.

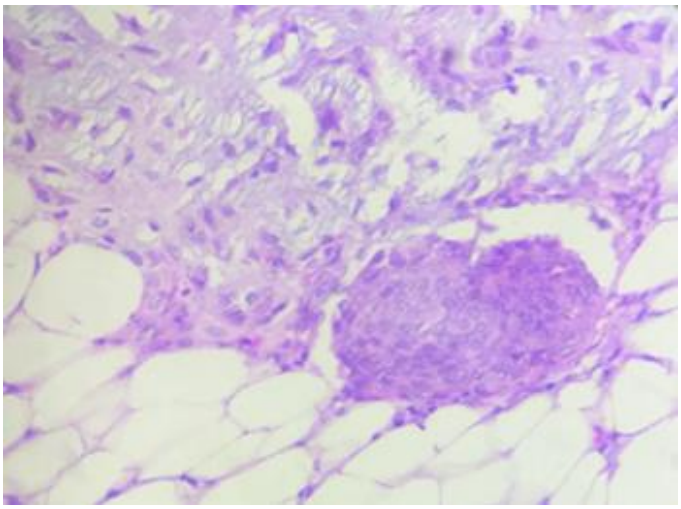

Figure 9: High power view H&E: Epithelial nest and cluster of adipocytes.