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Facial Chondroid Syringoma: First Case Report from a Rural Community in Ghana

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Abstract

Chondroid syringoma (CS), used in place of pleomorphic adenoma of the skin is a rare, benign, skin appendageal tumor. The diagnosis of this rare condition is made on histopathological examination aided by its commonplace clinical presentation. The tumor commonly presents as asymptomatic, indolent lesion and slowly growing mass, typically located in the head and neck region. We present a case report of a CS located on the left cheek of a 33-year-old man believed to be the first case ever from a rural community in Ghana.

Keywords: Chondroid syringoma; Ghana; African; Cutaneous tumour; Mixed tumour

Introduction

Chondroid syringoma (CS) is among the least occurring primary skin tumours with incidence of 0.01%. Also known as mixed tumour of the skin, it is a benign cutaneous sweat gland tumour with both epithelial and mesenchymal components. Although benign, the tumor has the propensity of malignant transformation with metastasis and recurrence. The malignant variants however predominantly involve the extremities as opposed to the benign ones which usually presents as asymptomatic slow growing mass on the head and neck region [1,2]. Common facial locations include cheek, orbit, scalp, chin, upper lip, nose, and forehead [1,3-5]. Other regions of occurrence CS include the torso, and scrotum. Despite its largely asymptomatic nature, depending on its locations, it may presents with some localized symptoms as there has been reported case of intraorbital CS causing exophthalmos with pressure and ocular symptoms [6]. It mostly occurs in the middle-aged population and more prevalent in

females than males [2,4,7].

Histologically, the mass may present with all or some of five basic features: Nests of cuboidal or polygonal cells; intercommunicating tubule-alveolar structures lined with two or more rows of cuboidal cells; ductal structures composed of one or two rows of cuboidal cells; occasional keratinous cysts; and a matrix of varying composition.

The current treatment of choice of CS is complete excision biopsy with close follow up recommended due to the possibility of malignant transformation. Histopathological examination is the gold standard of diagnosis as clinical and radiological investigations may not be distinctive.

We present what in the best of our knowledge is the first case CS reported in an African population on a 33-year-old male who presented with a left cheek swelling.

Case Report

History and Grossing

A 33-year-old male presented with a swelling in the left cheek in the last 2 years. Local examination revealed firm to hard mass with palpable areas of calcification and nodular in nature measuring 5 cm x 5 cm x 5 cm covered by normal skin. The swelling started as a small bump on the left cheek, growing slowly over the years to attain its present size. There was no pain and no neurological deficit. A provisional diagnosis of ossifying fibroma was made. The nodule was excised and sent for histopathological diagnosis.

Gross examination showed a non-encapsulated, hard in consistency, nodular, circumscribed, and non-ulcerated tissue fragment measuring 5 cm x 5 cm x 5 cm. Cut surface showed tanwhite and yellowish area and abundant chondroid components with evidence of calcification. No suspicion of malignancy was made (**Figure 1 and 2**).



Figure 1 Gross features of the mass reveals non encapsulated, nodular, non-ulcerated tissue

A. Physical appearance of patient with mass on the left cheek B. Nodular mass in situ being excised and C. Completely excised nodular mass measuring 5 cm X 5 cm X 5 cm in dimensions. D. Cut surface of the tumour showing a tan-white and yellowish area with abundant chondroid components and evidence of calcification.

Microscopic Features

Section showed a well-circumscribed and unencapsulated multinodular tumor with lobules separated by fibrous septae. Prominent and chondroid stroma with epithelial and myoepithelial cells forming secondary structures. The tumor was composed of glands and ducts as well as tubule-alveoli structures lined by a bi-layered epithelium. Nests of polygonal cells with abundant cytoplasm were observed. The sections also showed keratinous cysts and foci of squamous differentiating epithelial cells and presence of follicular structures of calcification. No atypia, sparse mitosis, no necrosis, and no malignancy seen.

An ultimate diagnosis of chondroid syringoma was made. A follow up was also advised.

No areas of hemorrhage, necrosis or malignancy noted. A. shows lobules separated by fibrous septae with predominant chondroid stroma; B. shows multiple small ducts lined by two layers of cuboidal cells within a chondromyxoid stroma; C shows fibromixoid stroma with glands and ducts lined by single epithelium; D. shows keratinous cysts and foci of squamous differentiation of epithelial cells, no atypia, sparse mitosis.

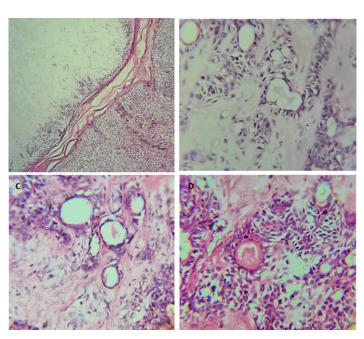


Figure 2: Histological features of the mass

Discussion

Chondroid syringoma is also known as mixed tumor of the skin. The tumor is composed of both epithelial and mesenchymal components [8]. First referred to as mixed tumors, salivary type, Chondroid syringomas (CSs) were described by Hirsch and Helwig in 1994 because of the presence of sweat gland elements in a cartilaginous stroma [9]. Among their series of 188 cases, 150 were associated with head and neck, nineteen involved the extremities, nine the axilla, eight the trunk, and two involved the genitals [9,10]. Consistent with our case, our patient had the tumour in the left cheek.

The reported incidence of CS is as low as 0.01%-0.098%, usually affecting middle-aged or older men than women as in this case, a middle-aged man [11,12]. CS typically presents as firm subcutaneous or intradermal masses often described as erythematous, purple or skin coloured and are usually asymptomatic [10]. Histologically, our patient's tumour presented all the five features of CS namely: the presence of nests of cuboidal or polygonal cells; intercommunicating tubule-alveolar structures lined with two or more rows of cuboidal cells; ductal structures composed of one or two rows of cuboidal cells; occasional keratinous cysts; and a matrix of varying composition [12]. CS may either manifest all the five features or just some of it [9].

Due to the possibility of a malignant transformation and recurrence, our patient is currently under close follow up.

Conclusion

Although CS is a rare tumour, they exist and must still be kept down the list of differentials of a subcutaneous mass especially in the head and neck region. Its propensity of malignant

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transformation makes it imperative for a closer follow up after complete resection.

Consent

We obtained a written informed consent from the patient for the publication of this case report and any accompanying images.

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