Malignant Chondroid Syringoma of Axilla: A rare entity at a rare site

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ABSTRACT

The Chondroid Syringoma (CS) are rare mixed tumours of the skin, benign or malignant, arising from the eccrine sweat glands due to the differentiation of the pleuripotent stem cells of the epithelial and mesenchymal tissues.[1,2] The incidence of CS is very rare accounting to just <0.098% amongst the primary cutaneous tumors. We present a case of recurrent malignant CS arising from the sweat glands of left axilla. This rare case stands as a testimony to the importance of immunohistochemistry in establishing the diagnosis of such rare entities. We have tried to explore the role of clinicians in identifying such rare cases as well as various treatment modalities being used to provide the patient with disease free survival along with good quality of life.

Keywords: Malignant Chondroid Syringoma, Axilla, Adjuvant Radiotherapy, S100, Vimentin, Calponin

INTRODUCTION

The Chondroid Syringoma (CS) are rare mixed tumours of the skin, benign or malignant, arising from the eccrine sweat glands due to the differentiation of the pleuripotent stem cells of the epithelial and mesenchymal tissues.[1,2] The incidence of CS is very rare accounting to just <0.098% amongst the primary cutaneous tumors. We present a case of recurrent malignant CS arising from the sweat glands of left axilla and the role of adjuvant radiotherapy here.

CASE REPORT

A 78 years old male patient presented with a swelling in the anterior axillary fold of left axilla which was painless, nodular and slowly increased in size over a period of 8 months. On examination, the lump was firm, mobile, non tender, slightly inflamed, subcutaneously located and approximately 8 cm x 6 cm in size. On the basis of the clinical picture, the differential diagnosis were epidermoid cyst, sebaceous cyst, adnexal tumour or nodule.

Patient underwent the Excision of the mass on August 9, 2016. On gross examination, the skin covering mass measured 8.0 X 5.5 X 4.5 cm and the overlying skin measuring 5.2 X 3.0 cm was unremarkable. The cut surface showed a solid nodular mass with central area of necrosis. On microscopic examination, the sections showed structure of Chondroid Syringoma. Though there were many areas of benign Chondroid Syringoma, distinct areas of malignant transformation were identified characterised by more cellularity, cellular pleomorphism and necrosis. Lymphovascular permeation or Perineural invasion were not identified. On the
basis of the histopathological examination, the diagnosis was established as Malignant Chondroid Syringioma arising from the skin adnexal origin of the left axilla. No adjuvant treatment was taken.

Patient developed recurrent nodular mass lesion and underwent Wide Excision of it around 9 months from the first surgery, on May 9, 2017. Cut surface showed three well circumscribed nodular lesions within the subcutaneous region measuring 5.0 X 3.4 X 3.0, 2.2 X 2.0 X 2.0 and 1.0 X 1.0 X 0.7 cm, respectively. Overlying skin was free and was 0.2 cm away from the tumour. Also, a single underlying muscle tissue was removed and was free from the tumour. On microscopic examination, invasive sheets of large malignant epithelial cells having large hyperchromatic irregularly contoured nuclei with prominent nucleoli and scanty eosinophilic cytoplasm were seen. Base of resection and skin margins were free. Lymphovascular permeation was seen but Perineural invasion was absent. Further, Immunohistochemistry tests showed CK 7, P63, S 100, Vimentin and Calponin positive. The diagnosis was established as Recurrent Malignant Syringoma of skin adnexal origin of left axilla. He was treated with postoperative radiotherapy with the conventional dose of 2Gy/# to the total dose of 50Gy/25# with the parallel opposed AP/PA (anteroposterior/posteroanterior) fields.
CASE DISCUSSION

The CS was first described by Billroth in 1899. In 1961, the term ‘Chondroid Syringoma’ was termed by Hirsch and Helwig.[3] The term ‘syringoma’ is derived from the Greek word syrinx which means pipe or tube accounting for the tubuloalveolar structures seen in the histological construction of the dermis.[4] The malignant CS is also called sclerosing sweat duct carcinoma, microcystic adnexal carcinoma and syringoid carcinoma. They may be sporadic or associated with rare genetic syndromes, including Birt-Hogg-Dubé syndrome, Brooke-Spiegler syndrome, Cowden syndrome, and Muir-Torre syndrome. [5] According to Basedan and Butler’s classification scheme, 4 variants of CS have been recognized: (1) a localized form, (2) a form associated with Down syndrome, (3) a generalized form that encompasses multiple and eruptive syringomas, and (4) a familial form. Male predilection is seen in malignant CS and most cases present between 4th to 6th decade. The most common site of occurrence are head and neck especially, eyelid. They can also be found in the axilla, trunk, limbs and genitalia. [6]

They usually present as solitary, solid, painless, nonulcerative, subcutaneous or intracutaneous, usually painless, indolent to rapid growing nodule. Though, the recurrent lesions can present as multiple closely placed nodules. In the head & neck, the size can vary from the range of <2 mm to >1 cm. [4-7] But in other sites like axilla and limbs, much larger sizes of tumors are seen. In the most cases of CS, the anaplastic changes are present from the very beginning. So, though the initial histopathological picture may be indicative of the benign tumor only, the sudden malignant transformation of a long standing benign tumor with metastatic spread cannot be overlooked.

The particularly intimidating challenges about the CS are it’s asymptomatic presentation that leads to the delay in presentation of the patient to the clinician. Also, the sites of occurrence and neglect of the self care on the part of patients add to the difficulty of the early presentation. Further, due to the lack of any unique clinical sign or feature as well as the confusing overlapping picture
with many harmless benign skin conditions, they may be misdiagnosed by the clinicians. This leads to the loss of the crucial time period where the lesion is still in the early stages and could have been completely treated. They are locally aggressive and the long neglected cases may lead to skin ulceration and fungation. They have very high propensity for the peri-neural invasion and can lead to paraesthesia or anaesthesia. This can be particularly distressing in cases of the tumors of limbs and may even lead to dysfunctionality based on the site of occurrence. The tumors occurring in Head & Neck have high tendency for metastatic spread due to the rich lymphatic supply.

The mainstay of diagnosis is the histopathological examination which again has overlapping features with other tumors arising from the skin, many of the cases being misdiagnosed as benign. So, here arises a dire necessity for the establishment of robust diagnostic criteria. Histologically, syringomas are characterized by multiple small epithelial collections with central ducts surrounded by a bilayer of cuboidal cells in the superficial dermis. Frequently, a tapering, comma-shaped edge is present at one edge of the epithelial collections with an encompassing fibrotic stroma. The immunohistochemistry tests can emerge as saving grace for the final diagnosis. The most important markers being CK 7, P63, Vimentin, EMA and CD 34.[8-13]

Wide local excision is the first treatment of choice. Moh’s Surgery Grade 2c is the surgery of choice. [11] But accounting to it’s dermal location, the rates of recurrences are extremely high accounting to it’s dermal location. So, the role of adjuvant treatment in the form of radiotherapy becomes essential to prevent recurrences. The role radiation therapy should play in the treatment of malignant CS is uncertain. Case reports suggest that radiation therapy alone is capable of achieving clinical clearance of tumors however, tumors may recur or fail to respond to therapy. Of note, the recurrence of malignant CS in a histologically more aggressive form after radiation therapy has been reported in one patient. The use of radiation therapy as an adjuvant to surgical intervention was evaluated in a retrospective study of 14 patients with malignant CS (11 of whom had positive surgical margins) who were followed for a median of five years. In the study, 13 of 14 patients achieved local control, prompting the authors to suggest that conventional surgical excision plus radiation therapy could yield success rates similar to those observed with Mohs surgery. [12-13]

CONCLUSION
The knowledge about clinical presentation, histopathological picture accentuated by relevant immunohistochemistry tests, adjuvant treatment in the form of radiotherapy as well as patient education plays a crucial role in the effective management of this rare entity with a disease-specific-survival of 10 years. [14]

REFERENCES


DECLARATION:

There are no conflicts of interest.

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