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Taufik Aljijakli
ENT Consultant, Dammam Medical Complex

Ramez Atassi
Consultant Histopathologist, Dammam Regional Laboratory

Khalid Dilaijan
ENT Resident, King Fahd Military Medical Complex – Dhahran

Talal Sioufi
ENT Specialist, Dammam Medical Complex, KSA

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Chondroid Syringoma: A Case Report and Literature Review

Taufik Aljijakli,[1] Ramez Atassi,[2] Khalid Dilaijan,[3] Talal Sioufi[4]

[1]ENT Consultant, Dammam Medical Complex, [2]Consultant Histopathologist, Dammam Regional Laboratory, [3]ENT Resident, King Fahd Military Medical Complex – Dhahran, [4]ENT Specialist, Dammam Medical Complex, KSA

Correspondence to: Khalid Dilaijan, Email: kdilaijan@gmail.com

Mixed tumor of the skin, more recently called chondroid syringoma, is a rare benign adnexal tumor of the skin that is characterized by a slowly growing intradermal or subcutaneous nodule, which is usually located in the head and neck.

Recurrence after excision is rare and can be attributed to either incomplete removal, which may occur because of the lobulation of the tumor or more rarely due to malignant transformation. Here we are reporting a female patient who presented with a nasal mass that was previously excised twice and our diagnosis was a benign chondroid syringoma.

Keywords: ***Chondroid syringoma, Mixed tumor of the skin · Pleomorphic adenoma of the skin.***

INTRODUCTION

Chondroid syringoma (CS) is a rare benign adnexal tumor of the skin that was originally described in 1859 by Billroth as a mixed tumor because of the prominent mixture of epithelial and stromal components.[1-3] It is also called pleomorphic adenoma of the skin because it, morphologically, resembles that of salivary gland tissue.[4]

Complete excision of the mass usually ensures no recurrence. Here we are presenting a female patient who presented with a nasal tip mass that was previously excised twice and our diagnosis was a benign chondroid syringoma.

CASE REPORT

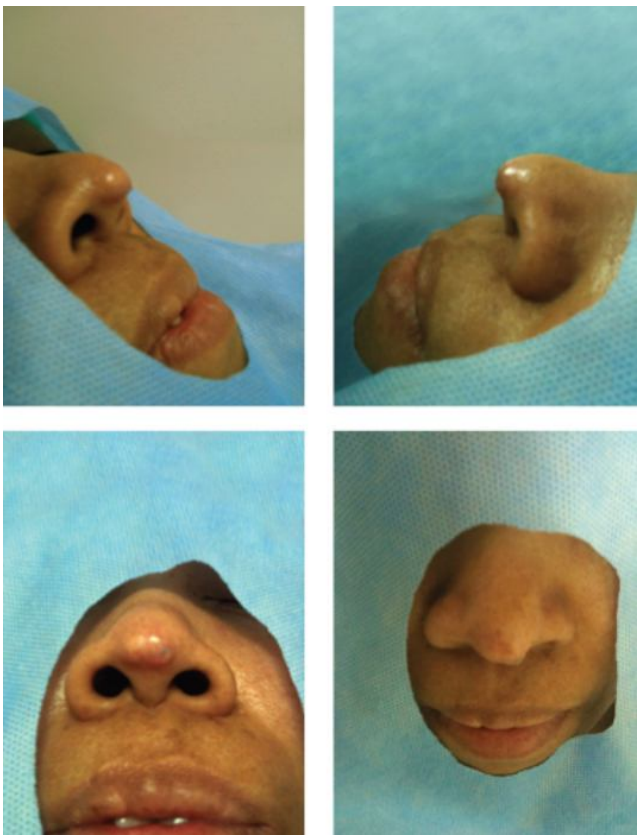
This is a 34 years old Indonesian female patient who is not known to have any chronic medical illness. She presented to ENT clinic at Dammam Medical Complex – Saudi Arabia, complaining of a painless disfiguring nasal tip swelling that was gradually increasing in size over last 5 years.

Patient was worried about the nature of the lesion considering her past surgical history. No history of neck masses or weight loss. No history of previous radiation exposure.

Previously, she underwent excision of the same lesion twice through a transcolumellar incision:

5 and 7 years back in Indonesia. In her 2nd surgery, the mass was excised with part of the medial crus of right lower lateral cartilage & grafted by cartilage from left pinna. We contacted the operating physician to get an idea about surgical details and histopathological diagnoses but it seems that they couldn't get the patient's file.

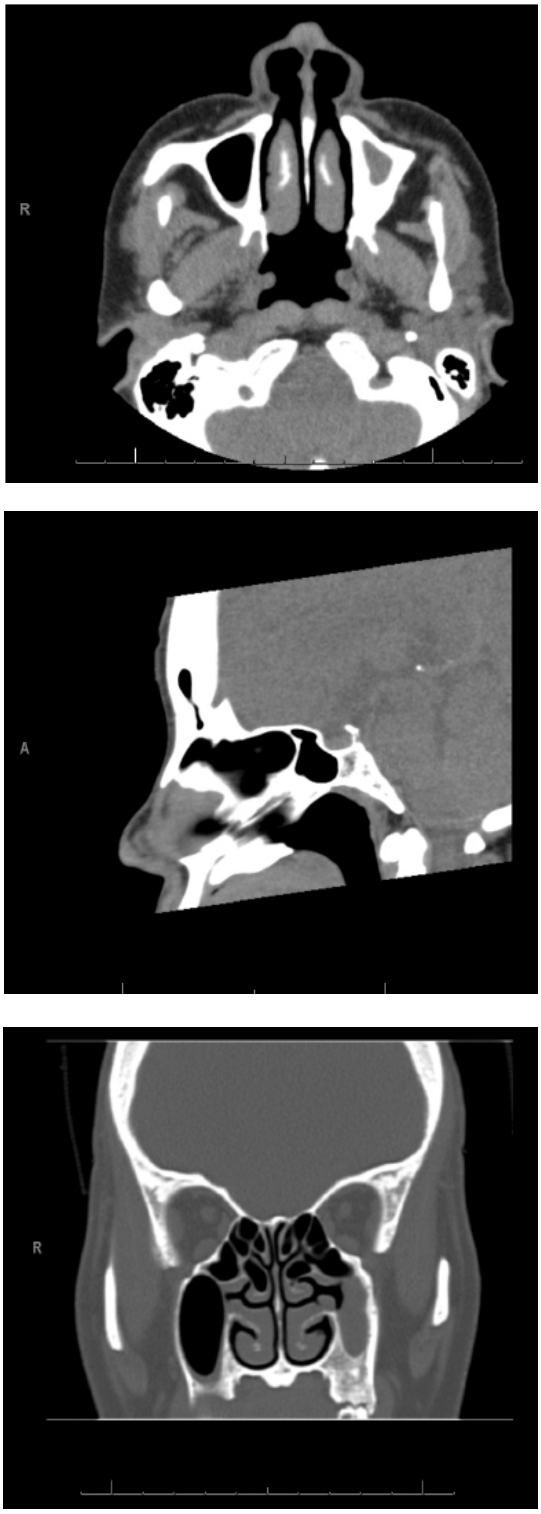
Examination revealed a single 1.0cm x 0.5cm bluish firm non-tender mass at the nasal tip with no discharge or skin ulceration. No asymmetry was appreciated. A transcolumellar scar was present. Endoscopic nasal examination was normal. No palpable neck masses (Figs. 1-4).



Figs 1-4. The nodule.

Sinuses computed tomography scan was done to evaluate the sinuses and nasal cavity (Figs. 5-7). There was a spontaneously, slightly dense soft tissue nodule at the nasal tip area with no calcification, fatty stranding, or peripheral inflammatory changes. Other findings included:

Bilateral (left more than right) maxillary mucosal hypertrophy, bilateral concha bullosa of both superior and middle turbinates as well as hypertrophy of inferior turbinate bilaterally. No other nasal masses. No lytic bony changes.



Figs. 5-7: Axial, coronal, and reconstructed sagittal CT sinuses views, soft tissue window.

For cosmetic reason, we initially tried to excise the mass through a transcolumellar incision (at same scar site). We found that the mass was closely adherent to the skin at the nasal tip area.

So, we decided to do complete excision of the mass with overlying skin (Fig. 8).

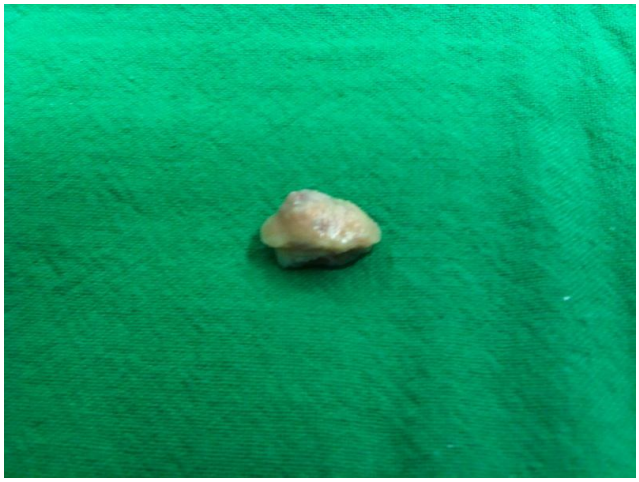
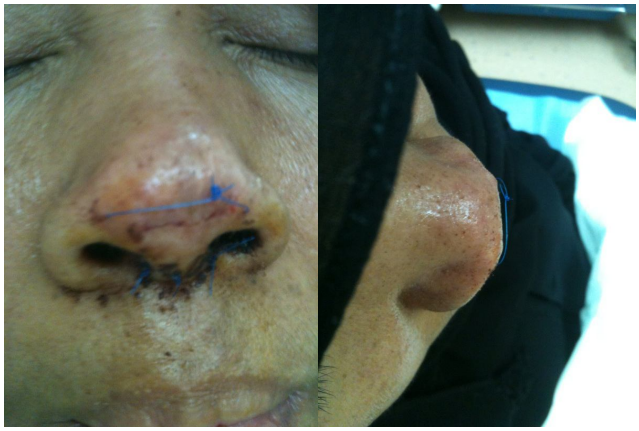


Fig 8. The excised specimen.

Subcuticular closure was done at nasal tip and simple interrupted sutures used for approximating the transcolumellar incision (Figs. 9-10).



Figs. 9-10. Post-operative pictures.

Histopathological examination using hematoxylin and eosin stain (H&E stain) revealed intradermal nodules composed of clusters of myoepithelial cells or tubular formations lined by double epithelial layers surrounded by myxoid and hyalinized stroma which is diagnostic of Chondroid Syringoma (Figs. 11-16). Disease-free margins were confirmed.

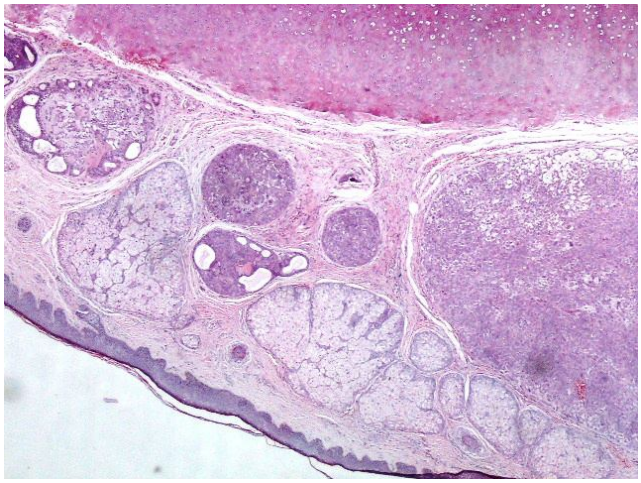


Fig 11. Skin and small part of nasal cartilage withmultiple small nodules in the dermis (H&E stain, 10x4).

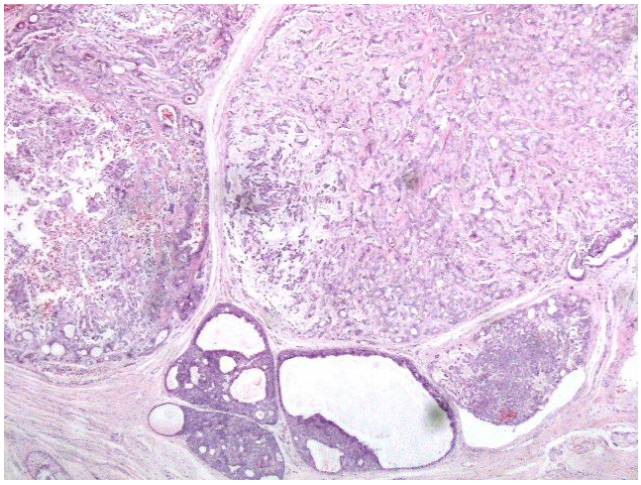


Fig 12. Solid and glandular pattern at the dermis (H&E stain, 10x10).

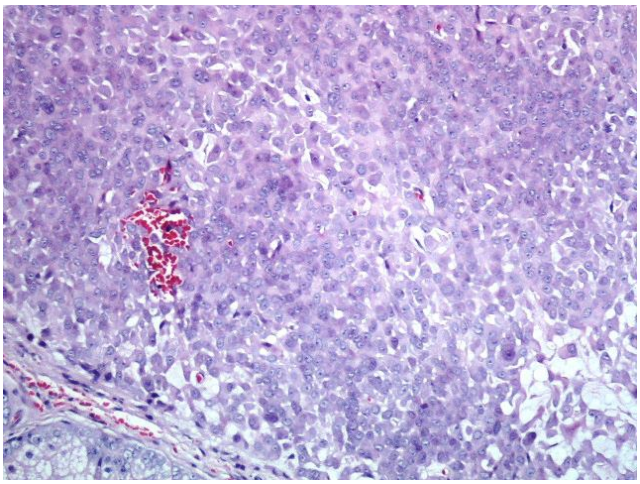


Fig 13. Solid mass of myoepithelial cells (H&E stain, 10x20).

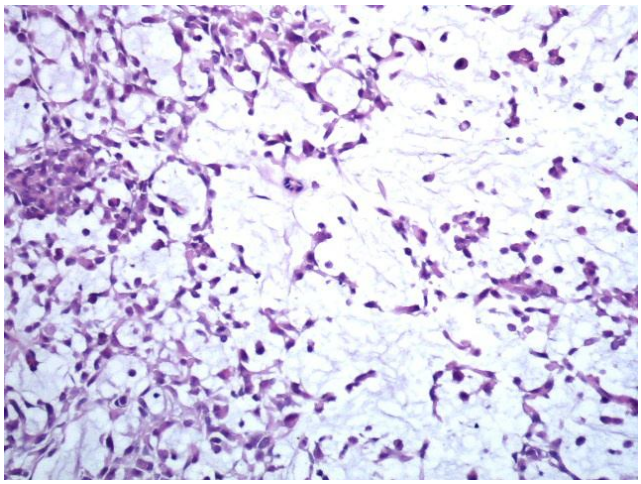


Fig 15. Myxoid stroma (H&E stain, 10x10).

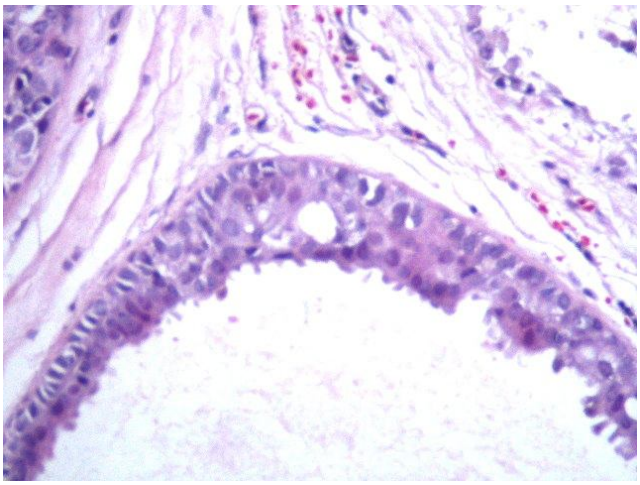


Fig 14. Gland lined by double layers epithelial and myoepithelial cells (H&E stain, 10x40).

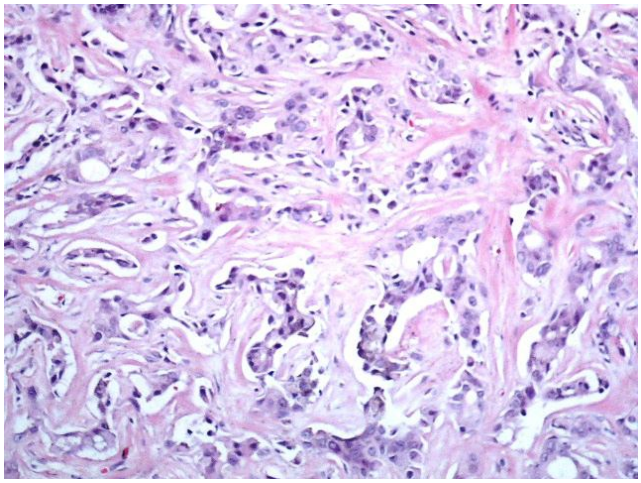
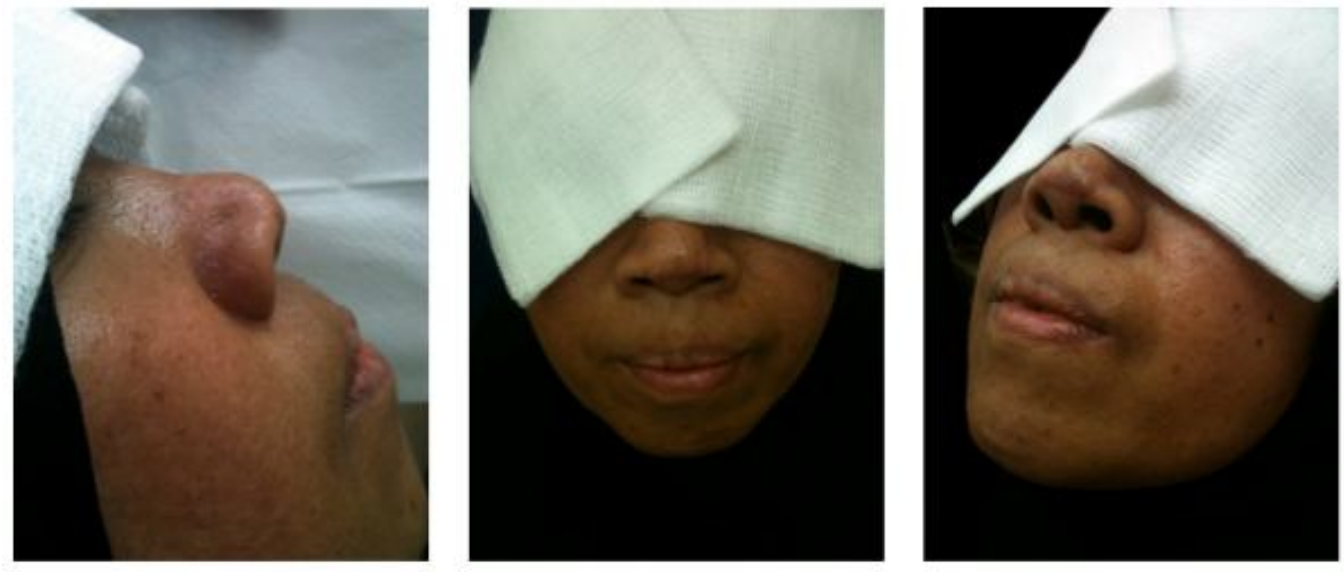


Fig 16. Hyalinized stroma (H&E stain, 10x10).

Patient was followed for 2 months post-operatively and she was doing well (Figs. 17-19).



Figs 17-19. Follow up at 3 weeks post operatively.

LITERATURE REVIEW (DISCUSSION)

Chondroid syringoma (CS) is a rare benign adnexal tumor of the skin that was originally described in 1859 by Billroth as a mixed tumor because of the prominent mixture of epithelial and stromal components.[1-3] It is also called pleomorphic adenoma of the skin because it, morphologically, resembles that of salivary gland tissue.[4] The term "chondroid syringoma" was proposed by Hirsch and Helwig in 1961,[1] given the prominent cartilage-like stromal material in most instances (chondroid) and presence of sweat gland-like elements in the tumor (syringoma).

Patients are usually middle aged and older.[5] With regards to benign CS, gender predilection differs among different studies and textbooks.[1,2,6]

Patients with CS present with a solitary painless well circumscribed slowly growing intradermal or subcutaneous nodule that is usually located in the head and neck.[2,6,7] Glabella and nose were also reported sites.[8-10] Circumscription of CS nodule is given by the effect of greater density of the tumor compared with the tissues around it rather than a definite capsule, which is seldom present.[11] Based on clinical examination alone, it is difficult to diagnose CS and the differential diagnosis may include epidermal cyst, sebaceous cyst, pilar cyst, implantation dermoid, lipoma, compound naevus, clear cell hidradenoma, cystic basal cell carcinoma, neurofibroma, and dermatofibroma.[2,9]

MRI has rarely been used to evaluate suspicious lesions for anatomic extent and to identify tissue of origin, depth of invasion and relation to adjacent structures, such as muscles and bones with no characteristic MRI findings.[12,13] —FNAC can be useful to determine the pathology before excision;[14-16] "however, examination of the excised tissue is most reliable in establishing a definitive diagnosis".[14]

Definitive diagnosis of CS is done by histopathological examination. It is an intradermal or subcutaneous multilobulated tumor with fibrous septa between lobules containing chondroid, myxoid, or hyalinized stroma in variable degrees. Along with epithelial content, it usually contains tubular structures and ductal differentiation, which have a peripheral layer of flattened myoepithelial cells.[6]

The treatment of choice is local excision with a margin of normal tissue, keeping in mind the lobulated nature of this tumor and the possibility of recurrence.[8,9] Patients with a benign chondroid syringoma that was completely excised need no long-term follow up. Long-term follow up is indicated only when the tumor is incompletely excised or if there is any indication of malignancy.[7]

Malignant chondroid syringoma (MCS) is an extremely rare entity.[17] Most MSC were assumed to be malignant from the beginning.[18] Clinically, features that raise suspicion of malignancy include a large size, rapidly growing mass, a mass in the extremities or trunk, or being in female patients.[19] Histological characters of malignancy are irregular, infiltrative borders, vascular invasion, cytologic atypia, nuclear pleomorphism, increased mitotic activity, focal necrosis, and a lesser degree of tubular differentiation than in BCS.[19] It is important to evaluate for presence of large amount of myxoid matrix and poor chondroid differentiation because they may carry risk of malignancy irrespective of other benign pathologic features.[17]

For malignant lesions, early wide excision with a broad margin is the treatment of choice with no enough data to support the use of radiotherapy or chemotherapy in malignant chondroid syringoma patients.[17]

CONCLUSION

Here we are presenting a female patient who presented with a nasal tip mass that was previously excised twice and our diagnosis was a benign chondroid syringoma.

Proper complete excision of the mass with overlying skin is the management of choice for BCS especially when aesthetic and functional characters are preserved. Excision of only the mass underlying the skin may result in residual disease tissues that can lead to recurrence.

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