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Case Report

A rare case of a benign giant chondroid syringoma of nose

Neemu Hage*, Jaimanti Bakshi, Mayank Rampal

Department of Otorhinolaryngology, Postgraduate Institute of Medical Education and Research, Chandigarh, India

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***Correspondence:**
Dr. Neemu Hage,
E-mail: neemuhage@gmail.com

ABSTRACT

Chondroid syringoma (CS) is a rare benign appendageal tumour of the skin, with histological similarities to pleomorphic adenoma of salivary glands. It typically presents as a slow growing nodular mass generally less than 3 cm, arising from the skin, predominantly in the head and neck region. We report a case of a 61-year-old male with a massive nasal mass attained over a period of 2 years. A fine needle aspiration of the lesion was suggestive of CS. It was excised completely under general anaesthesia. The postoperative histopathology was also consistent with CS. The patient was followed up on a regular basis and was rendered disease free till his last follow up at 4 months. Our purpose of reporting this case is to highlight the rarity of the disease entity, as well as the occasion of it acquiring a large size yet, maintaining its benign nature.

Keywords: Chondroid syringoma, External nose, Benign appendageal tumour

INTRODUCTION

Chondroid syringoma (CS), also known as a mixed tumor of skin, is a benign neoplasm of the sweat gland. It was first described by Billroth as a 'cutaneous mixed tumor', later revised by Hirsch et al, who coined the term, 'chondroid syringoma' in 1961.[1,2] It is an extremely rare tumor of the skin appendage with an incidence reported to be less than 0.01 to 0.098 percent.[3] It is often misdiagnosed as other benign cutaneous lesions such as sebaceous cysts, dermoid cysts and basal cell carcinoma, and histopathology clinches the diagnosis. It is described by a nests of cells and lace-like pattern of strands of epithelial cells and tubuloalveolar structures; differentiation toward a pattern similar to that of sweat glands and ducts; occasionally keratinous cysts and varying amounts of matrix.[1] Generally known to develop up to 3 cm in size, larger lesions have been described in literature arising from the trunk and extremities frequently harbouring malignancy.[4,5]

CASE REPORT

Our patient, a 61 year old gentleman sought ENT consultation for a swelling over the nasal dorsum for just over 2 years. The swelling had an indolent course causing disfigurement of the nose. Our patient denied any pain and nasal complaints. Local examination of the swelling revealed a 6×4 cm firm lobular mass over the nasal dorsum, slightly more towards the left side, with prominent vascularity. The overlying skin was partially adhered to underlying mass and partially mobile (Figure 1).

An FNAC from the lesion revealed epithelial and myoepithelial cells in a background of abundant chondromyxoid stroma consistent with a benign adnexal tumor, likely, chondroid syringoma. A contrast enhanced computed tomography of the nose and paranasal sinuses showed a 4×4×3 cm, moderately homogenous soft tissue mass over nose, with no evidence of underlying necrosis

or calcification, causing partial obliteration of left nasal cavity (Figure 2).

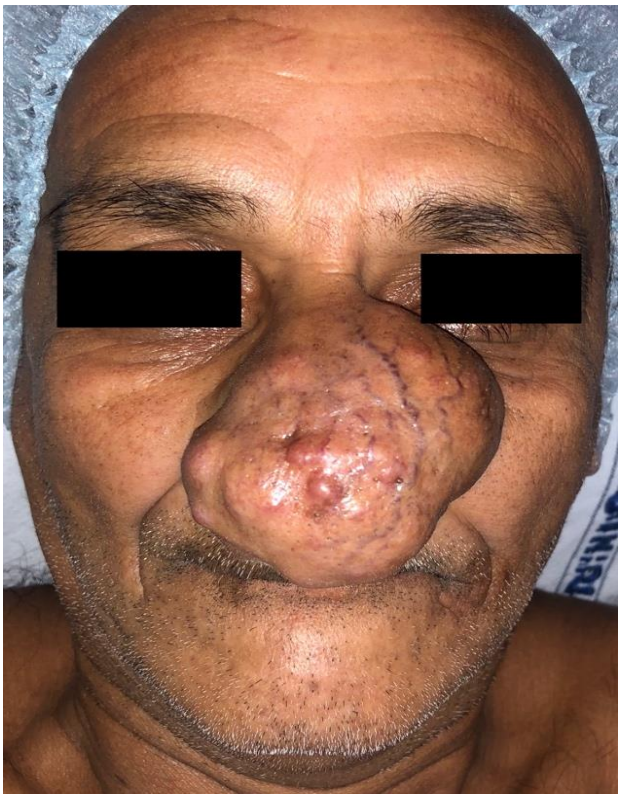

Figure 1: Anterior view of external nasal mass.

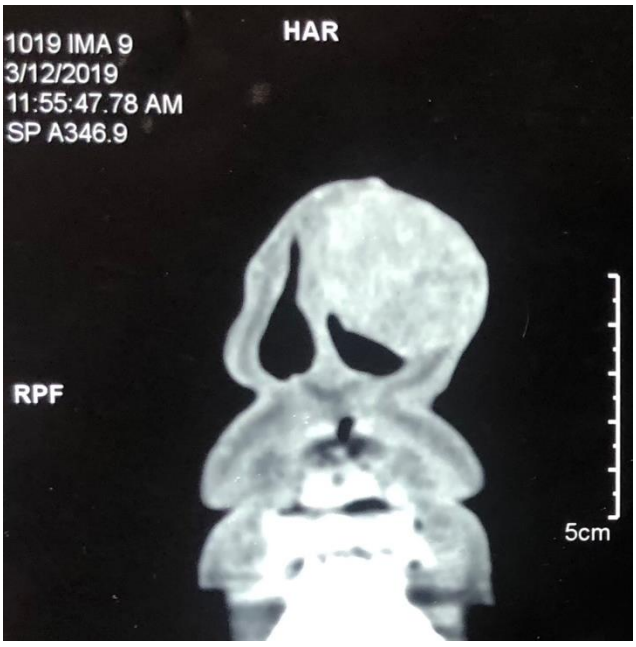

Figure 2: Axial cut of Contrast enhanced computed tomography of nose and paranasal sinuses.

The lesion was excised under general anesthesia. Part of the skin adherent to underlying cartilage was excised. The defect was closed primarily with the redundant skin over two silicone stents tunneled through bilateral nasal cavity to prevent postoperative stenosis (Figure 3).

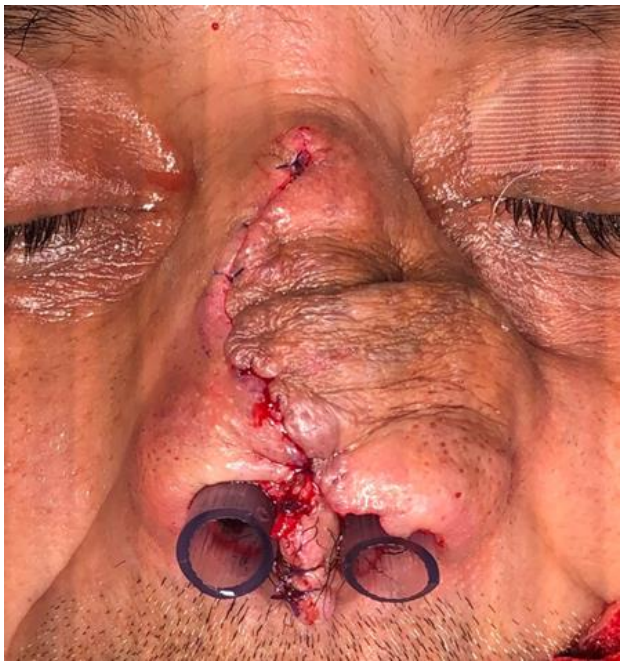

Figure 3: Primary closure of defect over silicone stents in bilateral nasal cavity.

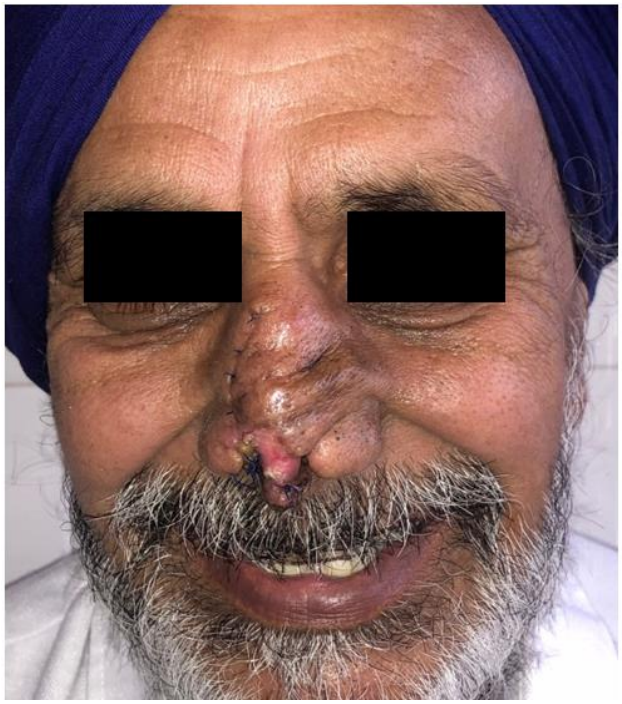

Figure 4: Postoperative appearance at 4 weeks.

A meticulous effort was made to preserve uninvolved skin, which we anticipated would help for secondary reconstruction at a later period. The stents were removed on post-operative day 5. His follow up at 4 weeks revealed a well healed wound over the nasal dorsum with minor nasal tip deformity (Figure 4).

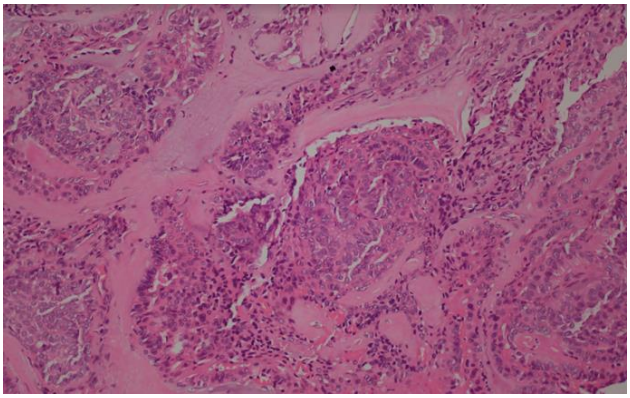

Figure 5: Hematoxylin and eosin stained microphotograph of cut section of tumour.

Histopathology of the excised mass revealed a circumscribed biphasic tumour comprised of, multiple nodules showing cells arranged in cords, tubules, trabeculae, cribriform pattern and pseudo papillae, in a myxoid background. These tubules were filled with inspissate and lined by a dual layer of luminal and myoepithelial cells. All these findings pointed towards a chondroid syringoma. As there were close resection margins in this case, patient has been kept on regular follow up (Figure 5).

There was no evidence of any local recurrence of the lesion at his 4 months follow up visit. Our patient was, to a great degree satisfied with the surgical outcome and did not desire further aesthetic surgery however, we have recommended a plastic surgery consult for the tip deformity.

Patient consent

Informed consent has been obtained from the patient and patient attendants as a standard departmental protocol for all publications.

DISCUSSION

Chondroid syringoma, also known as mixed tumour of the skin, is a rare, biphasic cutaneous neoplasm analogous to pleomorphic adenoma of the salivary glands. In 1961, the term chondroid syringoma was first used by Hirsch et al to describe tumours of mixed origin from the skin.[1,2] They defined these lesions as neoplasms of epithelial origin with the capacity to form sweat gland–like structures capable of producing cartilaginous-like material. Incidence of chondroid syringoma in literature is reported to be less than 0.01 to 0.098 percent. The tumour is almost twice as common in male patients, typically occurring between the ages of 20 and 60 years.[2] Owing to their rarity, many cases are initially misdiagnosed as cysts, dermal nevi, or various other cutaneous adnexal neoplasms and are often correctly identified only after being sent for histopathologic review.

Lesions are typically 0.5 to 3.0 cm in size and generally present on the head and neck as was the case in our patient, but they are also known to occur less frequently on the extremities, abdomen, axilla, and inguinal area.[2] Common sites in head and neck region are nose, cheek and upper lip.[1] Most are benign, but there have been case reports documenting malignant lesions. These malignant tumours are often larger than 3.0 cm and are more likely to manifest in women than men. In addition, they tend to occur on the trunk and extremities rather than the head and neck.[4-6]

Hirsch et al proposed the following as microscopic criteria necessary for the diagnosis of CS: nests of cuboidal or polygonal cells, tubuloalveolar structures with 2 or more rows of cuboidal cells morphologically similar to those in the nests of cuboidal cells, ductal structures composed of 1 or 2 rows of cuboidal cells, occasional keratinous cysts varying in size, and a matrix of varying appearance in hematoxylin and eosin sections.[2] In 1961, Headington grouped CS into eccrine or apocrine types based on their putative epithelial origin.[7] Further study has identified folliculosebaceous differentiation in the apocrine phenotype and although additional morphologic variants have been identified the distinction of eccrine vs apocrine differentiation has persisted in classification of this family of mixed tumours, with the latter proving more common.[8-11] Findings indicative of malignant transformation include cytological atypia, increased mitotic figures, infiltrative margins, satellite tumour nodules, and tumour necrosis.[3]

Although various treatment options for this tumour have been proposed, including electrodessication, dermabrasion, and vaporization with argon or CO2 laser, the treatment of choice is complete surgical excision.[3] As with any excision of facial tumour, aesthetic and functional structures need to be preserved as much as possible. Malignant transformation, though rare and unlikely in head and neck, should be ruled out with close histopathological examination of margins and recognition of malignant cells. If the tumour has been completely excised and is benign, long-term follow-up is not indicated. Follow-up is indicated only if the excision is incomplete or if there is indication of malignant change.[12] Recurrence may be attributed to incomplete excision as the tumour is lobulated and it can be treated by re-excision.[1] As with our case, there are no features suggestive of recurrence till date, however we intend to keep the patient on a long term follow up as there were close margins on histopathology in this case and the recurrence rates reportedly range from 2.4 to 10 percent.[13] For malignant chondroid syringoma, the treatment of choice is wide excision and adjuvant radiation therapy with or without chemotherapy.[14]

CONCLUSION

Chondroid syringoma is an uncommon entity and a surgeon faced with such a tumour is at a conundrum with

the clinical diagnosis. Presentation of a slow growing nodular lesion in the head and neck region of a middle aged patient should alert the surgeon for the possibility of a chondroid syringoma. Accurate history taking and precise histopathologic examination is essential for its diagnosis. Surgical excision is the established treatment of choice. A word of caution needs to be said about the 'complete excision' of the tumour mass, failing which it may recur at a later period. To abate the chances of recurrence a long term follow up is essential. Complete wide local excision with negative margins is necessary if we are dealing with a malignant chondroid syringoma, with thereafter, adjuvant chemoradiotherapy. This, to the best of our knowledge, is the first ever reported case of a giant benign CS of the nose.

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