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CHONDROID SYRINGIOMA OVER NOSE: A RARE CASE REPORT

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ABSTRACT

Chondroid Syringioma is a rare skin tumor of mixed type which consist of both mesenchymal and epithelial cell component. Frequently located in head and neck region with incidence of less than 0.1%. Even though malignant transformation occurs rarely, diagnosis and proper management is important. Its presentation may vary from round, firm, nodular or cystic lesion with marginal heterogenicity in sonography. All the lesions were completely removed by excision. Clinical, histopathological, Radiological data of our case report helps the clinician in better diagnosis and management of patients presented with variety of facial lesion.

KEYWORDS

Chondroid Syringioma, Rare skin tumor, Pleomorphic Adenoma of Skin

INTRODUCTION

Skin tumors called chondroid syringomas (CSs) are uncommon and benign. Because of its histological similarity to the benign mixed tumor that originates from the salivary gland, chondroid syringomas were initially identified by Billroth in 1859 as a cutaneous mixed tumor[1]. Of all primary skin tumors, Chondroid syringomas occur in less than 0.01% of cases. In instead of mixed skin tumor, Hirsch and Helwig utilized the term "chondroid syringoma" in 1961. Apocrine type and eccrine type were the two subcategories into which Headington separated chondroid syringomas in 1961, confirming their histological appearance[1].

A solitary, solid, painless, non-ulcerating, subcutaneous or intracutaneous nodule is generally present with chondroid syringomas, which most frequently affect the head and neck and range in size from 2 mm to around 1 cm. With a male to female ratio of 2:11, they frequently afflict individuals in their middle to late years.

Rarely do chondroid syringomas have a malignant subtype. Anaplastic alterations are evident from the beginning in the majority of malignant chondroid syringomas. A mixed cutaneous tumor with mesenchymal and epithelial components, malignant chondroid syringoma primarily affects the trunk and extremities. Rarely, a long-standing chondroid syringoma develops malignant alterations with extensive metastasis[1].

Case Description

A 35 years old female patient presented to E.N.T Department of P.D.U Government Medical College and Civil Hospital, Rajkot with history of right-side swelling over nose. Swelling had been noticed for 1 year which was gradually increasing in size and asymptomatic in nature. Patient did not have any history of trauma / any other medical condition. Rest head and neck and oral examination did not reveal any abnormalities.

On clinical examination single well define swelling of size $10\times10mm^2$ over right side of nose (Fig 1), mobile but little adherent at its base, non-tender with overlying skin normal without any pus/discharge or any punctum over skin or any palpable regional lymphadenopathy. On anterior rhinoscopy of nose there was mild deviation of nasal septum to right without any turbinate's hypertrophy. Nasal mucosa was normal and there was no paranasal sinus tenderness. Patient doesn't have any nasal complaints like nasal blockage, difficulty in breathing, nasal bleeding or discharge.

Biochemistry and other routine blood investigations were in normal range. An X-ray of bilateral nasal bone didn't reveal any bony deformity. An Ultrasonography revealed 10×5mm well define hypo-echoic lesion over local part without any internal vascularity suggestive of benign etiology likely (Sebaceous cyst).

A Fine needle aspiration study shows few groups and clusters of benign epithelial cells intermingled with fibro-myxoid material. Cell shows bland nuclei with well define cytoplasm. Few plasmacytoid cells are seen. Overall findings suggestive of Chondroid Syringioma (Mixed salivary gland tumor of skin) likely.

Surgical excision of cyst done under local anesthesia while keeping safety margin of 1.5cm (Fig 2). Final histopathological examination of specimen shows Chondroid Syringioma (Pleomorphic Adenoma of Skin) (Fig 3).

Post operatively patient does not have any complain of pain or bleeding (Fig 4). Patient doesn't have any recurrence in last 2 years.

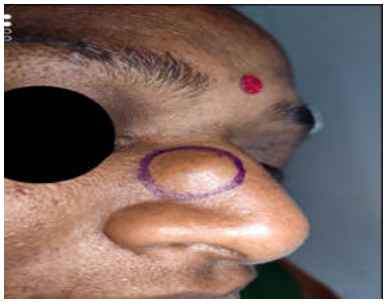

Figure 1: Preoperative image of swelling over right side of nose

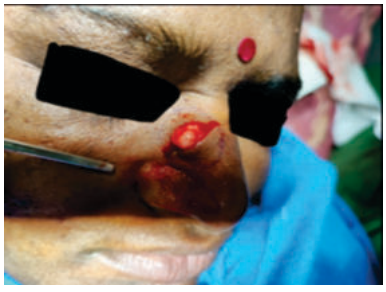

Figure 2: Intraoperative image of swelling over right side of nose

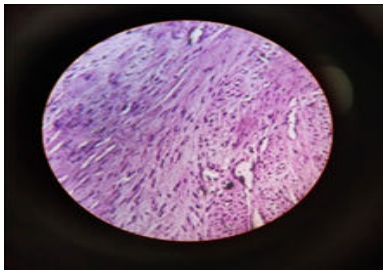

Figure 3: Histological Features of Chondroid Syringioma

Figure 4: Postoperative image of patient with healed scar over right side of nose

DISCUSSION

Chondroid syringomas are uncommon, benign skin tumors that occasionally develop in the head and neck and come from sweat glands. In 1859[2], Billroth classified these lesions as benign mixed skin malignancies.

When examined physically, chondroid syringomas are smooth, solid, non-tender, and subcutaneous or intradermal. Their coloring may resemble the surrounding skins, or it may have a purple or reddish tint. With a male-to-female ratio of 2:1.6, these tumors prefer to affect middle-aged males. From 0.01% to 0.098% of all initial skin cancers are these masses[2].

While chondroid syringoma can occasionally be benign, rare incidences of malignant transformation have also been documented. These cancerous forms are more prevalent in younger girls. They frequently exceed 3 cm in size, spread quickly locally, and favor the trunk or extremities[3].

Hirsch and Helwig outlined five histological criteria for the diagnosis of chondroid syringomas: Cuboidal or polygonal cell nests, linked tubuloalveolar structures bordered by two or more rows of cuboidal epithelial cells, ductal structures lined by one or two rows of cuboidal cells, sporadic keratinous cysts, and a matrix of mixed chondroid and myxoid material are just a few examples of the cell types that may be found in the lungs. Some chondroid syringomas exhibit all five of these traits, whereas others only show a few. Headington's classification of chondroid syringomas into apocrine and eccrine varieties in support of histological variations within luminal morphology. Cytologic atypia, infiltrative margins, satellite tumor nodules, tumor necrosis, and involvement of deep structures are histological characteristics that are thought to indicate malignant transformation[3].

Immunohistochemistry demonstrates that the tubuloalveolar components of chondroid syringomas exhibit cytokeratin (CK), epithelial membrane antigen (EMA), carcino-embryonic antigen (CEA), vimentin, S-100 protein, neuron-specific enolase (NSE), and in a small number of instances, glial fibrillary acidic protein (GFAP)[3].

A malignant chondroid syringoma lesion was found by MRI to have a signal intensity that was slightly higher than muscle on the proton-density sequence, heterogeneously increased on the T2-weighted images, and high with focal regions of intermediate signal on the short T1 inversion recovery (STIR) images. On the T1-weighted pictures, this benign lesion seemed somewhat more intense than muscle, and on the T2-weighted images, the signal had risen in a heterogeneous manner4. It also displayed heterogeneous but diffuse enhancement[4].

The preferred therapy for a mass is surgical excision. Wide local excision of the mass was performed with a 1.5 cm margin of error to prevent recurrence. Recurrence rates range from 2.4 to 10% of cases, according to reports. Recurrence typically results from insufficient excision.

Radiotherapy for malignant chondroid syringomas, Schvili and Rothem described this strategy as an adjuvant therapy following nodal metastatic excision, however they did not specify the methods, dosage, or volume employed. Adjuvant radiation was recommended by Hong et al. following the excision of a local recurrence utilizing 5000 cGy in 25 fractions on the tumor site with 2 cm margins, with no signs of recurrence or metastatic disease after 27 months of follow-up. Watarai et al. described a case of malignant chondroid syringomas recurrence treated with surgery, radiation treatment, and chemotherapy in combination, with no clinical progression after 18 months of follow-up. There was no mention of the radiation dose[5].

Clinical Significance

Chondroid Syringioma is an uncommon, benign cutaneous tumor that develops from the eccrine sweat gland and has a high risk of recurrence. The most straightforward procedure for reducing the likelihood of recurrence is frequently wide local excision. It is necessary to closely monitor the visualization of recurrence.

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Conflict Of Interest

The author declares no conflict of interest in the article reported.

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