

# Benign chondroid syringoma affecting the upper lip: Report of a rare case and review of literature

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## Abstract

Chondroid syringoma (CS) (mixed tumor of the skin) is a rare neoplasm of the sweat glands, which presents itself as a slow-growing, painless, nonulcerated, subcutaneous or intracutaneous mass often occurring in the head and neck region. The clinician may miss the diagnosis of this lesion due to its rarity. CS should be considered in the differential diagnosis of any subcutaneous nodules, especially in the head and neck region. The diagnosis of CS is mainly based on the histopathologic examination. This article presents a 35-year-old male with a mass on the upper lip that was histopathologically diagnosed as an apocrine variant of benign CS with squamous metaplasia after surgical excision. No sign of recurrence is evident till date during the follow-up. We report this case because of its rarity. A brief literature review and all the reported cases in the lip have been listed.

**Keywords:** Apocrine type, benign, chondroid syringoma, diagnosis, histopathology, lip, malignant potential, mixed tumor, skin, treatment

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## INTRODUCTION

Chondroid syringoma (CS) is a tumor arising from the sweat glands or pilosebaceous unit. CS was first described by Billroth in 1859.<sup>[1]</sup> These lesions were initially referred to as mixed tumors of the skin of both epithelial and mesenchymal origin.<sup>[2]</sup> The term “Chondroid syringoma” was coined by Hirsch and Helwig in 1961 based on the histologic presentation.<sup>[3]</sup> CS is a rare tumor with an incidence of <0.098%. The diagnosis of CS is mainly based on the histopathologic evaluation. A search through the existing literature was carried out for all the reported cases of CS at various sites which revealed very few cases of CS on the upper lip [Table 1].<sup>[4-14]</sup>

## CASE REPORT

A 35-year-old male patient reported to the outpatient department of our institute with a complaint of a mass on the left side of the upper lip. The lesion was present for a duration of 10 years. The lesion was small, to begin with and slowly progressed to the present size. Clinical examination revealed an asymptomatic, firm subcutaneous mass measuring 3.5 cm × 1 cm × 0.7 cm on the left side of the upper lip [Figure 1]. The patient wanted it to be removed due to esthetic reasons. Intraoral examination failed to reveal any other abnormality. A biopsy was advised for diagnostic confirmation. After obtaining an informed patient consent, a complete surgical excision was done under local anesthesia

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using an intraoral approach to avoid scarring [Figure 2]. The wound was closed primarily, and the healing was uneventful. There is no sign of recurrence till date.

On gross examination, the lesion was grayish white in color with a smooth and glistening surface and was firm in consistency. Cut surface revealed whitish solid areas with small foci of hemorrhage. Histopathologic examination of the tissue specimen revealed a well-encapsulated lesion [Figure 3] composed of benign epithelial cells arranged in the form of sheets, nests and tubuloductular structures with a prominent chondroid and myxoid stroma [Figure 4]. Tubuloductal structures of varying size and shape lined by a double layer of epithelial cells and adjacent stromal hyalinization was evident [Figure 5]. Some showed cystic dilatation filled with keratin flakes and squamous metaplasia [Figure 6]. Few mucoid faint basophilic areas with scattered stromal cells surrounded by halo resembling cartilage (pseudo cartilage) were also seen [Figure 4]. Occasional hair shafts were also noted. Based on these findings, a final diagnosis of an apocrine variant of benign CS with squamous metaplasia was made.



**Figure 1:** Solitary, asymptomatic subcutaneous mass on the left side of the upper lip

## DISCUSSION

CS is a benign tumor derived from skin appendages commonly affecting the middle-aged and elderly males. CS is a slow-growing painless lesion and patients get it removed mainly for esthetic reasons. These lesions present themselves as solitary, well-delineated, slow-growing, painless, nonulcerated, subcutaneous or intracutaneous nodules sometimes with surface telangiectasia and sessile base. Nose is the most common site in the head and neck region followed by the skin of the cheek and the upper lip. Reports of CS on the eyes, eyelid, hands, foot, axillary area, abdomen, penis, vulva, scrotum, scalp and back have also been published in the literature.<sup>[4,15]</sup>

Clinically, the diagnosis of CS is quite challenging and most often overlooked or mistaken with other lesions such as dermoid or sebaceous cysts, compound nevus, neurofibroma, histiocytoma, pilomatrixoma, dermatofibroma, basal cell carcinoma and seborrheic keratosis.<sup>[5,16]</sup>

CS manifests in two forms. Benign form is more frequently reported in males than females with a ratio of

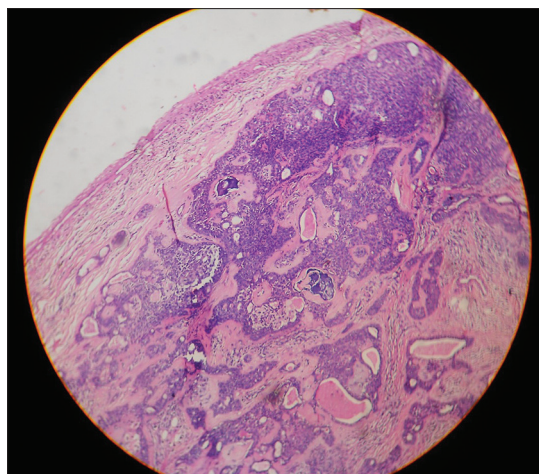


**Figure 2:** Intraoperative photograph showing the surgical procedure through an intraoral approach

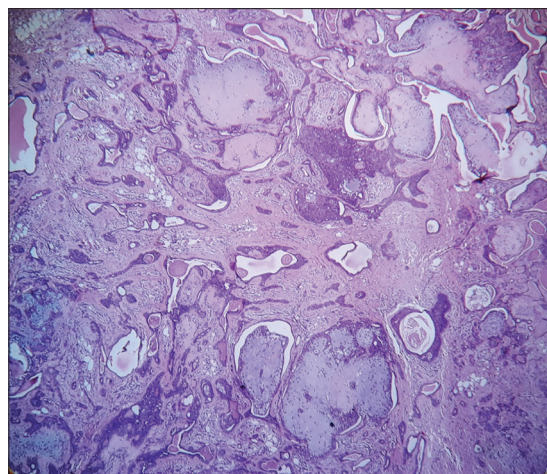
**Table 1: Reported cases of benign chondroid syringoma in the lip**

Reported year	Author	Number of cases	Location	Patient details (age in years)
1959	Stout and Gorman <sup>[4]</sup>	16	Upper lip	-
		2	Lower lip	
1986	Triantafyllou and Ravidis <sup>[5]</sup>	1	Upper lip	-
1986	Adlam and Wood <sup>[6]</sup>	1	Upper lip	-
2002	Bekerecioglu <i>et al.</i> <sup>[7]</sup>	3	Upper lip	27/male, 28/female, 24/female
2003	Shimizu <i>et al.</i> <sup>[8]</sup>	1	Upper lip	68/male
2004	Arikan <i>et al.</i> <sup>[9]</sup>	1	Upper lip	73/male
2010	Dubb and Michelow <sup>[10]</sup>	1	Upper lip	58/female
2015	Girgis <i>et al.</i> <sup>[11]</sup>	1	Upper lip	23/male
2016	Shilpa <i>et al.</i> <sup>[12]</sup>	1	Upper lip	48/male
2016	Kundu <i>et al.</i> <sup>[13]</sup>	1	Upper lip	46/male
2016	Min <i>et al.</i> <sup>[14]</sup>	5	Upper lip	44/male, 39/male, 47/male, 64/male, 65/female
2017	Reddy <i>et al.</i> (present case)	1	Upper lip	35/male

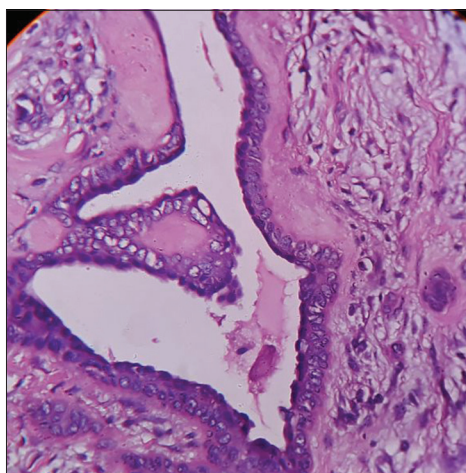




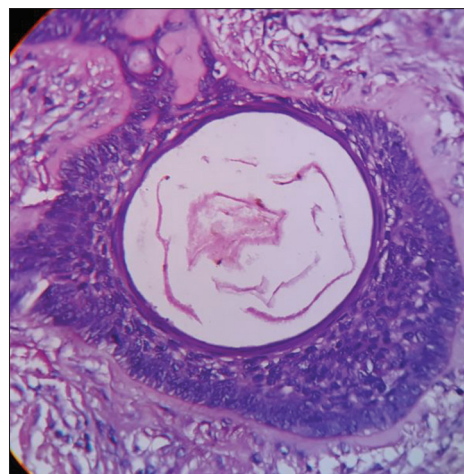
**Figure 3:** Photomicrograph showing well-circumscribed tumor with the peripheral capsule (H & E,  $\times 10$ )



**Figure 4:** Photomicrograph showing the lesion composed of nests and irregular tubuloalveolar structures composed of epithelial cells embedded in a myxoid to chondroid (pseudocartilage) stroma (H & E,  $\times 10$ )



**Figure 5:** Photomicrograph showing irregular tubulocystic structure lined by double rows of epithelial cells with adjacent stromal hyalinization (H & E,  $\times 40$ )



**Figure 6:** Photomicrograph showing cystic dilatation filled with keratin flakes and squamous metaplasia (H & E,  $\times 40$ )

1.3:1 to 1.5:1 while the malignant form occurs more often in females.<sup>[2,4,17]</sup> This condition can affect 20 to 70-year-old individuals but is more common in the middle and elderly age group. CS has been reported in children as well.<sup>[18]</sup>

Imaging modalities such as magnetic resonance imaging, ultrasound<sup>[19]</sup> and computed tomography are not very suggestive of CS and can be used to determine the origin, depth of invasion and relation of the lesion with the adjacent structures.<sup>[20]</sup>

Fine-needle aspiration cytology and cytologic examination under May–Grünwald–Giemsa staining, hematoxylin and eosin staining and Papanicolaou staining are also been reported.<sup>[13]</sup>

Definitive diagnosis can be established only through histopathologic examination.

Five histological criteria have been proposed for the diagnosis of CS as follows:<sup>[3]</sup>

- Nests of cuboidal and polygonal cells
- Intercommunicating tubuloalveolar structures lined with two or more rows of cuboidal cells
- Ductal structures composed of one or two rows of cuboidal cells
- Occasional keratinous cysts
- A matrix of varying composition.

CS may display all these or few of these features. It has histologic similarities with the pleomorphic adenoma of the salivary gland. Some contrasting features between CS and pleomorphic adenoma have been summarized in Table 2.<sup>[12,21]</sup>

CS is composed of both the epithelial and mesenchymal components. Epithelial cells are arranged in sheets, islands,

**Table 2: Contrasting features between benign chondroid syringoma and pleomorphic adenoma**

Features	Benign chondroid syringoma	Pleomorphic adenoma
Origin	Sweat glands	Salivary glands
Incidence	Very rare tumor	Most common benign salivary gland tumor
Histogenesis	Myoepithelial cell, Merkel cell <sup>[21]</sup>	Intercalated duct reserve cells, myoepithelial cells
Site	Can occur in sites other than head and neck area (like extremities)	Rare in sites other than head and neck region
Age	Middle aged and elderly individuals	Middle-aged individuals
Sex	Benign form is more common in males	More common in females
Size	Usually <0.5-3 cm	Usually 2-6 cm
Histopathology	Epithelial cells show differentiation towards adnexal structures (i.e., hair follicle, sebaceous gland, apocrine sweat gland and eccrine sweat gland)	Adnexal structures not commonly seen
Histopathology	Serous and mucous acinar cells not evident	Serous and mucous acinar cells are evident
Histopathology	Epithelial cells arranged in tubulocystic structures lined by single or double rows. The cells of the peripheral layer showed proliferation into the stroma which has a mucoid, faintly basophilic appearance suggestive of chondroid syringoma <sup>[12]</sup>	Epithelial cells arranged in sheets, strands, ducts

and nests of squamous cells or glandular cells, cords, tubular structures embedded in a fibroadipoid, chondroid, hyaline and mucinous stroma.<sup>[6,9,16]</sup> Extensive ossification and marrow formation have been reported in CS by Awasthi *et al.*<sup>[22]</sup>

Two histologic variants of CS have been described by Headington,<sup>[23]</sup> namely:

- Eccrine type with smaller round lumens, lined by a single row of cuboidal epithelial cells evenly spaced within a myxoid-chondroid stroma and
- Apocrine type with irregular or haphazard distribution of tubular and cystic branching tubules and lumina lined by double rows of epithelial cells - inner cuboidal cells and outer flattened cells.

The inner layer expresses positivity for the epithelial markers such as cytokeratin, epithelial membrane antigen and carcinoembryonic antigen.<sup>[24]</sup> While the outer layer expresses positivity for mesenchymal markers such as vimentin, S-100 protein, neuron-specific enolase, and glial fibrillary acidic protein. The chondroid area was earlier believed to be derived from myoepithelial cells. However, recently, it was concluded to be true cartilaginous tissue with typical chondrocytes confirmed ultrastructurally in the subsequent studies.<sup>[25]</sup> Mills suggested that CS are monoclonal tumors with replicating cells having the ability to differentiate into epithelial and mesenchymal tissues accounting for the histologic variability of CS.<sup>[26]</sup> The presence of Merkel cells (CK20 positivity) in CS showing follicular differentiation supports the hypothesis that Merkel cells may be an integral constituent of follicles. The presence of Merkel cells as a proliferative form in apocrine CS may support the derivation of Merkel cells from folliculo-sebaceous-apocrine germ or their precursor cells, although the origin of Merkel cells (cells with neuroendocrine differentiation) in the skin is not limited to follicular germ.<sup>[21]</sup> Some CS as in the foot may be preceded by trauma. Immunohistochemistry may vary depending on whether CS is an eccrine type or apocrine type. However, some tumors may display both features.<sup>[27]</sup>

Malignant CS may arise *de novo* or in pre-existing benign CS. Although rare, few cases of malignant CS have been reported in the past which show characteristics of aggressiveness, metastasis and recurrence. Features such as increased mitosis, cellular and nuclear pleomorphism, invasive margins, satellite tumor nodules, necrosis, fixation to underlying tissues, excessive mucoid stroma, poorly differentiated chondroid components, rapid growth, and size of more than 3 cm might indicate malignant potential.<sup>[28-30]</sup> Malignant CS is more likely to occur on trunk and extremities and are more common in females.<sup>[12,19]</sup> There may be local lymph node metastasis (48%) as well as distant metastasis to lung, bone, and brain.<sup>[31-35]</sup> Treatment of choice for CS is complete surgical excision with a rim of 4 mm normal tissue. Adjuvant radiotherapy with or without chemotherapy may be advised but is not very beneficial.<sup>[12,36]</sup> Other treatment options include dermabrasion, electrodissection and vaporization with argon or carbon dioxide laser.<sup>[2]</sup> Due to the paucity of CS cases, the accuracy and efficacy of various treatment modalities are yet to be ascertained.

## CONCLUSION

CS should be considered in the differential diagnosis of any subcutaneous nodules, especially in the head and neck region. The diagnosis is often not thought and missed considering the rarity of this adnexal neoplasm. Clinicians must be aware of this condition and should also rule out malignant CS. A careful histopathologic examination is the key to accurate diagnosis of CS.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.



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## Conflicts of interest

There are no conflicts of interest.

## REFERENCES

- Billroth T. Beobachtungen über geschwulste der speicheldrüsen. (Observations on the tumors of salivary glands). *Virchows Arch Pathol Anat* 1859;17:357.
- Yavuzer R, Başterzi Y, Sari A, Bir F, Sezer C. Chondroid syringoma: A diagnosis more frequent than expected. *Dermatol Surg* 2003;29:179-81.
- Hirsch P, Helwig EB. Chondroid syringoma. Mixed tumor of skin, salivary gland type. *Arch Dermatol* 1961;84:835-47.
- Stout AP, Gorman JG. Mixed tumors of the skin of the salivary gland type. *Cancer* 1959;12:537-43.
- Triantafyllou AG, Rapidis AD. Chondroid syringoma of the upper lip: Report of a case. *J Oral Maxillofac Surg* 1986;44:744-8.
- Adlam DM, Wood GA. The chondroid syringoma (mixed tumor of skin). Report of a case in the upper lip. *Oral Surg Oral Med Oral Pathol* 1986;61:69-72.
- Bekerecioglu M, Tercan M, Karakok M, Atik B. Benign chondroid syringoma: A confusing clinical diagnosis. *Eur J Plast Surg* 2002;25:316-8.
- Shimizu M, Kawano K, Fujiwara S, Noguchi T, Goto Y. Mixed tumor of the skin (chondroid syringoma) occurring in the upper lip: Report of a case. *Dermatol* 2003;15:37-41.
- Arikan OK, Erdoğan S, Muluk NB, Koç C. Chondroid syringoma of the upper lip: A case report. *Kulak Burun Bogaz Ihtis Derg* 2004;13:25-7.
- Dubb M, Michelow P. Cytologic features of chondroid syringoma in fine needle aspiration biopsies: A report of 3 cases. *Acta Cytol* 2010;54:183-6.
- Girgis S, Gillan G, Piper K. Rare benign mixed tumour of the upper lip: A case report. *Ann Med Surg (Lond)* 2015;4:380-3.
- Shilpa K, Leelavathy B, Divya G, Lakshmi DV. Chondroid syringoma: Histopathology a cornerstone tool in diagnosis. *Indian J Dermatopathol Diagn Dermatol* 2016;3:20-1.
- Kundu R, Punia RS, Handa U, Dalal U. Chondroid syringoma: Cytomorphology of four cases and review of literature. *Arch Cytol Histopathol Res* 2016;1:63-7.
- Min KH, Byun JH, Lim JS, Lee HK, Lee WM, Joo JE, *et al.* Chondroid syringoma on face. *Arch Craniofac Surg* 2016;17:173-5.
- Bhastin TS, Mannan R, Bhatia PK, Sharma M, Bhalla A. Fine needle aspiration cytology diagnosis of the eccrine variant of chondroid syringoma: Case report of a rare entity with review of literature. *J Clin Diagn Res* 2010;4:2641-4.
- Chen AH, Moreano EH, Houston B, Funk GF. Chondroid syringoma of the head and neck: Clinical management and literature review. *Ear Nose Throat J* 1996;75:104-8.
- Abila S, Bouhllaba J, Zarkika S, Lamchahaba FZ, Saidib A, Ismailia N, *et al.* Syringome chondroïde de la face (chondroid syringoma of the face). *Ann Dermatol Venereol* 2012;139:681-3.
- Turhan-Haktanir N, Sahin O, Bukulmez A, Demir Y. Chondroid syringoma in a child. *Pediatr Dermatol* 2007;24:505-7.
- Tural D, Selçukbiricik F, Günver F, Karışmaz A, Serdengeçti S. Facial localization of malignant chondroid syringoma: A rare case report. *Case Rep Oncol Med* 2013;2013:907980.
- Nicolaou S, Dubec JJ, Munk PL, O'Connell JX, Lee MJ. Malignant chondroid syringoma of the skin: Magnetic resonance imaging features. *Australas Radiol* 2001;45:240-3.
- Salama ME, Azam M, Ma CK, Ormsby A, Zarbo RJ, Amin MB, *et al.* Chondroid syringoma. Cytokeratin 20 immunolocalization of Merkel cells and reappraisal of apocrine folliculo-sebaceous differentiation. *Arch Pathol Lab Med* 2004;128:986-90.
- Awasthi R, Harmse D, Courtney D, Lyons CB. Benign mixed tumour of the skin with extensive ossification and marrow formation: A case report. *J Clin Pathol* 2004;57:1329-30.
- Headington JT. Mixed tumours of skin: Eccrine and apocrine types. *Arch Dermatol* 1961;84:989-96.
- Sheikh SS, Pennanen M, Montgomery E. Benign chondroid syringoma: Report of a case clinically mimicking a malignant neoplasm. *J Surg Oncol* 2000;73:228-30.
- Argenyi ZB, Balogh K, Goeken JA. Immunohistochemical characterization of chondroid syringomas. *Am J Clin Pathol* 1988;90:662-9.
- Mills SE. Mixed tumor of the skin: A model of divergent differentiation. *J Cutan Pathol* 1984;11:382-6.
- Mandeville JT, Roh JH, Woog JJ, Gonnering RS, Levin PS, Mazzoli RA, *et al.* Cutaneous benign mixed tumor (chondroid syringoma) of the eyelid: Clinical presentation and management. *Ophthalmic Plast Reconstr Surg* 2004;20:110-6.
- Bates AW, Baithun SI. Atypical mixed tumor of the skin: Histologic, immunohistochemical, and ultrastructural features in three cases and a review of the criteria for malignancy. *Am J Dermatopathol* 1998;20:35-40.
- Gündüz K, Demirel S, Heper AO, Günalp I. A rare case of atypical chondroid syringoma of the lower eyelid and review of the literature. *Surv Ophthalmol* 2006;51:280-5.
- Ishimura E, Iwamoto H, Kobashi Y, Yamabe H, Ichijima K. Malignant chondroid syringoma. Report of a case with widespread metastasis and review of pertinent literature. *Cancer* 1983;52:1966-73.
- Barnett MD, Wallack MK, Zuretti A, Mesia L, Emery RS, Berson AM, *et al.* Recurrent malignant chondroid syringoma of the foot: A case report and review of the literature. *Am J Clin Oncol* 2000;23:227-32.
- Watson JA, Walker MM, Smith NP, Hunt DM. Malignant chondroid syringoma - A rare cause of secondary bone tumour. *Clin Exp Dermatol* 1991;16:306-7.
- Kiely JL, Dunne B, McCabe M, McNicholas WT. Malignant chondroid syringoma presenting as multiple pulmonary nodules. *Thorax* 1997;52:395-6.
- Watarai A, Amoh Y, Aki R, Takasu H, Katsuoka K. Malignant chondroid syringoma: Report of a case with lymph node metastasis 12 years after local excision. *Dermatol Online J* 2011;17:5.
- Araújo JL, de Aguiar GB, do Prado Aguiar U, Mayrink D, Saade N, Veiga JC, *et al.* Malignant chondroid syringoma with central nervous system involvement. *J Craniofac Surg* 2012;23:514-5.
- Belfquih H, El Mostarchid B, Oukabli M, Akhaddar A, Boucetta M. Benign chondroid syringoma of the orbit: A rare cause of exophthalmos. *Head Face Med* 2012;8:8.