DOI: 10.7860/JCDR/2014/10026.5003

Case Report

Pathology Section

Chondroid Syringoma with Extensive Bone Formation: A Case Report and Review of the Literature

RECEP BEDIR[1], CUNEYT YURDAKUL[2], IBRAHIM SEHITOGLU[3], HASAN GUCER[4], SUPHAN TUNC[5]

ABSTRACT

Chondroid syringoma is a rare, skin appendageal tumour, usually reported at the head and neck region. It is a mostly intradermal and rarely subcutaneous small painless nodule. The histopathological examination is characterized by a combination of epithelial and myoepithelial structures within a chondromyxoid and fibrous stroma. Herein, we present a rare case of chondroid syringoma with extensive bone formation.

Keywords: Benign mixed tumour, Ossification, Skin tumour

CASE REPORT

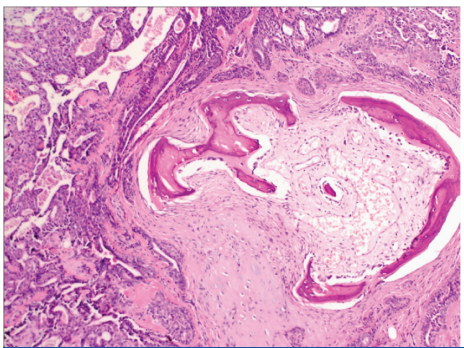
A 20-year-old man presented with a four year history of a slow growing lesion on the forehead. On examination, the lesion was 0.8 cm in size, firm, non-tender, and freely mobile. The lesion was totally excised. Grossly, the tumour was well encapsulated, 0.8x0.5 cm in diameter, firm and glistening with a grey white cut section. Histological examination showed a well-circumscribed tumour composed of lobulated nodules consisting of epithelial–myoepithelial islands with a tubuloglandular pattern, within a chondromyxoid stroma. Tumour in the stroma showed marked bone formation [Table/Fig-1]. Bone formation was surrounding epithelial-myoepithelial islands with a tubuloglandular pattern within a chondromyxoid stroma [Table/Fig-2,3]. The tumour exhibited no pleomorphism, necrosis or increased mitotic activity. In immunohistochemical investigation, pan-cytokeratin (pan-CK) was positive in the inner layer of epithelial cells whereas vimentin and S-100 protein were positive in the outer cell layers [Table/Fig-4,5]. The luminal surface of ductal structures was positive for carcinoembriyonic antigen (CEA) [Table/Fig-6]. Both with morphological and immunohistochemical findings, the case was diagnosed as chondroid syringoma with extensive bone formation.

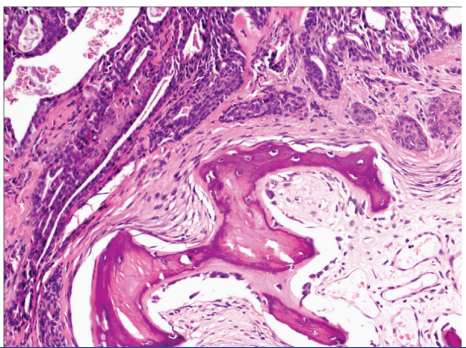
DISCUSSION

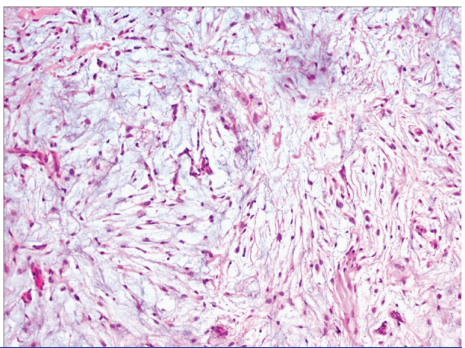
Chondroid syringoma, also known as benign mixed tumour (pleomorphic adenoma) of the skin, is a rare, tumour occurring usually in the head and neck region of adult patients. Tumour is of sweat gland origin [1].The reported incidence is<0.098% among all primer skin tumours. A malignant transformation is very rare. No clinical signs are characteristic of tumour. The definitive diagnosis is made by histopathologic examination. Histopathologic examination displays sweat-gland-like epithelial component in a fibroadipoid, chondroid, hyaline, or mucinous stroma. Bone formation in chondroid syringoma is an extremely rare feature [2,3]. To the best of our knowledge only five cases of chondroid syringoma with bone formation have been reported in published studies upto date. Herein, we report the sixth case of chondroid syringoma with extensive bone formation.

Chondroid syringomas are composed of both epithelial and mesencymal components. In 1961, Hirsch and Helwig first introduced the term chondroid syringoma to describe a mixed tumour of the skin originating from the sweat glands [4]. Characteristically, it is a slow-growing, painless, firm intracutaneous nodule at the head and neck region of adult males. It is often smaller than 3 cm, but may grow larger. Histopathological examinations have shown combination of epithelial and myoepithelial structures within a chondromyxoid and fibrous stroma [5]. Sometimes these tumours may show a differentiation towards various skin adnexal structures (including hair matrix, hair follicle, apocrine, and sebaceous glands), suggesting an origin from a folliculosebaceous-apocrine unit [6]. Bone formation is a rare feature and when present is usually focal and scant. Extensive ossification in chondroid syringoma is very rare, so far, only five such tumours have been reported in the skin [2,3,5,7,8]. The other reported cases of chondroidsyringoma with extensive bone formation are summarized on [Table/Fig-7].

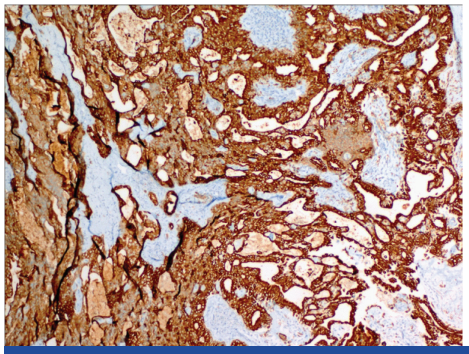
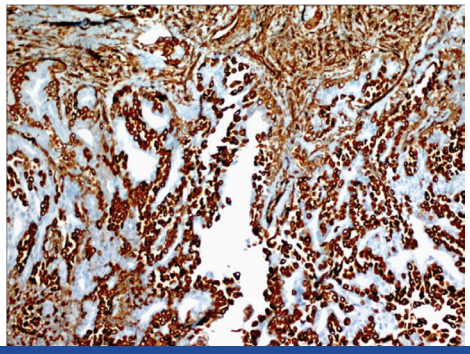
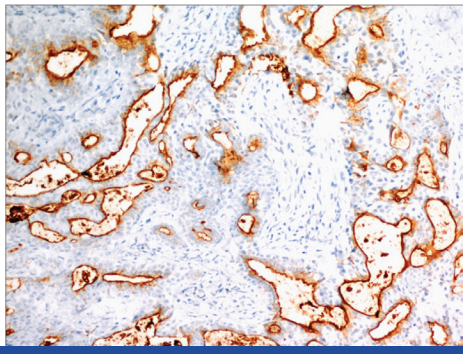
The exact mechanism of bone formation in chondroid syringoma is unknown. Akasaka et al., [8] suggested that ossification in these tumours indicates their development from pluripotential stem cells. On the other hand they have been thought to result from enchondral ossification, by direct laying down of bone by metaplastic myoepithelial cells or by modified myoepithelial cells and as well as by partial enchondral ossification [9]. Shimizu et al., [7] suggested







[Table/Fig-1]: Bone formation within a chondromyxoid stroma(H and E, x100) **[Table/Fig-2]:** Bone formation surrounding epithelial-myoepithelial islands with a tubuloglandular pattern (H and E, X 200) **[Table/Fig-3]:** Epithelial-myoepithelial islands within a chondromyxoid stroma (Immunostaining, X 200)

[Table/Fig-4]: Pan-CK was expressed in inner layer of of epithelial cells (Immunostaining, x100) **[Table/Fig-5]:** Vimentin was expressed in outer layer of epithelial cells (Immunostaining, x200) **[Table/Fig-6]:** CEA was expressed in luminal surface of ductal structures (Immunostaining

References	Year-old	Sex	Localization	Immunohistochemical findings	Treatment
Shimizu S and et al.,[7]	68	F	Upper lip	No	Local excision
Akasaka T and et al.,[8]	58	M	Chin	No	Local excision
Awasthi R et al.,[3]	43	F	Cheek	MIB-1	Local excision
Paul K and et al.,[5]	50	F	Tip of nose	No	Local excision
Eccher A and et al.,[2]	27	M	Frontal region	CK8-18 (+), CKAE1-AE3 (+) CEA(+), S-100 (+)	Local excision
Our case	20	M	Forehead	Pan-CK (+), vimentin (+), CEA(+), S-100 focal (+)	Local excision

[Table/Fig-7]: Clinicopathologic findings of chondroid syringomas with bone formation in the literature

the possibility of direct deposition of osteoids by metaplastic cells. Awasthi et al., [3] reported a case with extensive ossification and marrow formation. Akasaka et al., [8] reported a case of containing ossification, hair matrix, and sebaceous ductal differentiation. Our case did not impart any evidence of enchondral ossification, marrow formation, hair matrix, and sebaceous ductal differentiation in the tumour.

Clinically differential diagnosis includes inflammatory and neoplastic skin lesions such as nevi (particularly on the face osteo-nevusof Nanta), particularly on the face, basal cell carcinomas, pilomatricomas, trichoepitheliomas, hemangiomas, schwannomas, pyogenic granulomas, lipomas, epidermal and dermoid cysts, desmoplastic melanomas, dermatofibromas, and some cutaneous metastases [2,4,8]. Awasthi et al., [3] mentioned that radiological appearances of chondroid syringomas may be confused with malignant tumours because of the presence of irregular speculate structures extending within stroma.

Chondroid syringiomas are seen usually in benign forms and rarely in malignant forms. The malignant form are seen mostly in extremities without relationship with age different from the benign forms [10,11]. Malignant kondroid siringomas behave in aggressive manner. The chances of malignancy increase greater than 3 cm in size of the lesion. Histological features of malignant forms of chondroid syringomas are cytologic atypia, infiltrative margins, satellite tumour nodules, tumour necrosis and involvement in deeper tissues [12,11]. Malignant forms do not arise from a pre-existing condroid syringoma. Excessive amounts of mucoid stroma and poorly differentiated chondroid components give as important signs of the tumour's malignancy and metastatic potential [13]. Bates and Baithun [14] discuss mixed tumours that have signs of malignancy but do not certainly fall into the malignant group. For these they proposed the term atypical mixed tumours. Condroid syringoma in our case is small in size with no findings suggesting malignancy.

Immunohistochemical analysis can be helpful distinguishing the chondroid syringoma from other skin tumours. Tumour shows characteristics of both mesenchymal and epithelial differentiation. The inner layer of tumour has been shown to express cytokeratin (CK), epithelial membrane antigen (EMA) and CEA while the outer layer expresses vimentin, S-100 protein, neuron-spesific enolase (NSE) and in a few cases, glial fibrillary acidic protein (GFAP) [12]. Eccher et al., [2] reported a case that was immunohistochemically stained with CK8-18, CK AE1-AE3, and CEA in the epithelial cells; while S-100 protein was expressed in both stromal and epithelial components. Similarly, in our case immunohistochemical studies revealed expressions with pan-CK, vimentin, CEA and S-100 protein.

Optimal treatment of benign chondroid syringomas is total excision. Functional structures must be preserved during surgery. Fine needle aspiration cytology has been used for diagnosis intention. It may be helpful to determine pathology before excision. However, definitive diagnosis is confirmed by histopathology. It is important to perform tumour excition with normal tissue in surgery margins for preventing recurrence. Adjuvant chemotherapy and radiotherapy have also been tried in malignant chondroid syringomas [5].

CONCLUSION

We present a case of chondroid syringoma of the skin with extensive bone formation. Chondroid syringomas should be included in the differential diagnosis of patients with subcutaneous nodule in the head and neck region. In histopathological examination, presence of tumour with extensive bone formation may cause diagnostic difficulties. For this reason, awareness of this rare entity can be helpful to avoid future diagnostic pitfalls.

REFERENCES

[1] Yavuzer R, Basterzi Y, Sari A, Bir F, Sezer C. Chondroid syringoma: a diagnosis more frequent than expected. *Dermatol Surg*. 2003;29:179-81.
[2] Eccher A, Brunelli M, Gobbo S, Dalfior D, Dvornik G, Barbareschi M, et al. Chondroid syringoma with extensive ossification. *Int J SurgPathol.* 2007:15:385-87.
[3] Awasthi R, Harmse D, Courtney D, Lyons CB. Benign mixed tumour of the skin with extensive ossification and marrow formation: a case report. *J ClinPathol.* 2004;57:1329-30.
[4] Hirsch P, Helwig EB. Chondroid syringoma. Mixed tumour of the skin,salivary gland type. *Arch Dermatol.* 1961:84:835-47.
[5] Paul K, Sreekar H, Dhanraj P, Lamba S, George SM. Chondroid syringoma with extensive ossification. *Ann Maxillofac Surg*. 2011;1:91-92.
[6] Yamamoto O, Yasuda H. An immunohistochemical study of apocrine type of cutaneous mixed tumours with special reference to their follicular and sebaceous differentiation. *J CutanPathol.* 1999;26:232-41.
[7] Shimizu S, Han-Yaku H, Fukushima S, Shimizu H. Immunohistochemical study of mixed tumour of the skin with marked ossification. *Dermatology.* 1996;193:255-57.
[8] Akasaka T, Onodera H, Matsuta M. Cutaneous mixed tumour containing ossification, hair matrix, and sebaceous ductal differentiation. *J Dermatol.* 1997;24:125-31.
[9] Mandal S, Dhingra K, Roy S, Khurana N. Extensive bone with marrow formation in pleomorphic adenoma. Report of a case. *ANZ J Surg.* 2008;78:323-24.
[10] Metzler G, Schaumburg-Lever G, Hornstein O, Rassner G. Malignant chondroid syringoma: immunohistopathology. *Am J Dermatopathol.* 1996;18:83-9.
[11] Malik R, Saxena A, Kamath N. A rarecase of malignant chondroid syringoma of scalp. *Indian Dermatol Online J.* 2013;4:236-38.

[12] Argenyi ZB, Balogh K, Goeken JA. Immunohistochemical characterization of chondroidsyringomas. *Am J ClinPathol* 1988;90:662-69.
[13] 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.
[14] Bates AW, Baithun SI. Atypical mixed tumour 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.

PARTICULARS OF CONTRIBUTORS:
1. Faculty, Deparment of Pathology, Recep Tayyip Erdogan University of Medical Faculty, Rize, Turkey.
2. Faculty, Deparment of Pathology, Recep Tayyip Erdogan University of Medical Faculty, Rize, Turkey.
3. Faculty, Deparment of Pathology, Recep Tayyip Erdogan University of Medical Faculty, Rize, Turkey.
4. Faculty, Deparment of Pathology, Recep Tayyip Erdogan University of Medical Faculty, Rize, Turkey.
5. Faculty, Department of Plastic and Reconstructive Surgery, Rize State Hospital, Plastic and Reconstructive Surgery, Rize, Turkey.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Recep Bedir,
Faculty, Department of Pathology, Recep Tayyip Erdogan University of Medical Faculty, Rize /Turkey.
Phone : +90 5057331695, E-mail : bedirrecep@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **May 18, 2014**
Date of Peer Review: **Jul 14, 2014**
Date of Acceptance: **Jul 24, 2014**
Date of Publishing: **Oct 20, 2014**