ORIGINAL RESEARCH PAPER

Pathology

CYTODIAGNOSIS OF MALIGNANT CHONDROID SYRINGOMA WITH HISTOPATHOLOGICAL CONFIRMATION – A RARE CASE REPORT

KEY WORDS: Chondroid Syringoma, Malignant Mixed Tumor, Metastatic, Skin Adnexal Tumor

Dr. Ramya Krishnan MD Pathology

ABSTRACT

Chondroid syringoma also known as mixed tumor of the skin as it is composed of epithelial and mesenchymal elements. They exhibit both eccrine and apocrine differentiation. Malignant chondroid syringoma(MCS) is a very rare neoplasm which is very slow growing. It is predominantly seen in females and affects the trunk and extremities, whereas its benign counterpart has a predilection for male gender. As per literature, 51 cases of MCS has been reported so far on histopathology; however only 2 cases had cytological features of MCS described. Here we report a case diagnosed as MCS on cytology followed by confirmation on histopathology

INTRODUCTION

Introduced in 1961 by Hirsch and Helwig, Chondroid syringoma[1] is also known as mixed tumor of the skin analogous to the mixed tumor of salivary gland (Pleomorphic adenoma) since the tumor is composed of epithelial and mesenchymal elements. The tumor may exhibit both eccrine and apocrine differentiation. Malignant transformation can rarely occur in this type of skin adnexal tumor. The clinical diagnosis is challenging. Histopathology is imperative to reach the diagnosis. We report a case of malignant chondroid syringoma with recurrence , diagnosed initially by FNAC and later confirmed by histopathology.

Case Details

A 41 year old female presented with an insidious onset of swelling in the right post auricular region for 7 months duration with history of pain over the swelling for 1 month. No lifting of ear lobule noted. On local examination, the swelling was firm to hard measuring 2x2cm with restricted mobility. On ultrasound it was suspected to be dermoid cyst.

FNAC findings revealed clusters and sheets of epithelial cells with moderate eosinophilic cytoplasm and pleomorphic hyperchromatic nuclei in a background of chondromyxoid matrix suggestive of Malignant chondroid syringoma.

Wide local excision of the lesion with primary closure was done. We received two skin covered gray white soft tissue fragments, the largest measuring 3x0.3x0.1cm and other measuring 1 cm in length. All were embedded.

Microscopy examination showed stratified squamous epithelium with underlying dermis showing circumscribed neoplasm composed of tubules and cords lined by round to polyhedral cells with moderate eosinophilic to vacuolated cytoplasm and pleomorphic hyperchromatic nuclei with occasional cells having vesicular nuclei with prominent nucleoli. Increased mitotic figures and singly scattered tumor giant cells were noted. The epithelial islands were embedded in a background of chondromyxoid material with areas of necrosis and hemorrhage. These features were consistent with FNAC diagnosis of Malignant Chondroid Syringoma.

During the follow up period, the patient presented with recurrence of swelling at the same site within 5 months of surgery. FNAC of this recurrent swelling showed similar features.

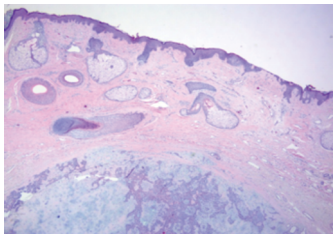

Figure 1: Section Showing Skin with Underlying Well Circumscribed, Nodular Proliferation of Islands of Epithelial Cells in Chondromyxoid Stroma in the Dermis.

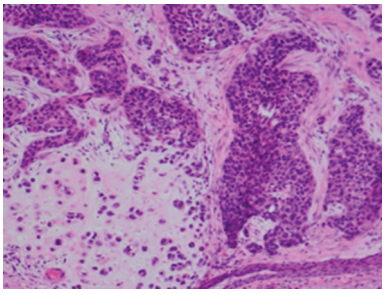

Figure 2: Section Showing Pleomorphic Malignant Epithelial Component and Chondroid Differentiation.

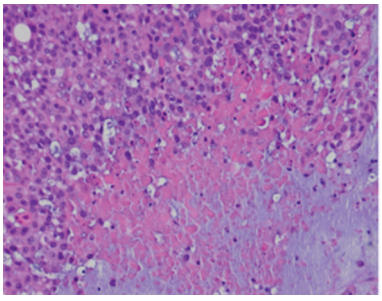

Figure 3: Mitotic Figures and Area of Necrosis.

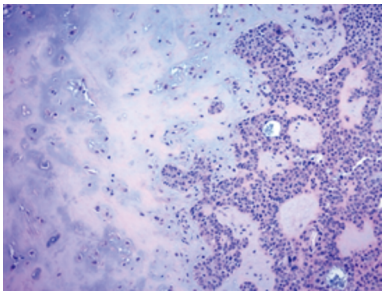

Figure 4: Epithelial Islands in Chondroid Stroma.

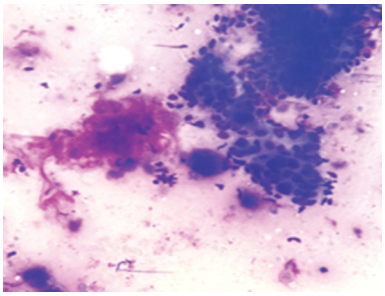

Figure 5: FNAC Smear Shows Epithelial Cell Clusters in Chondromyxoid Stroma.

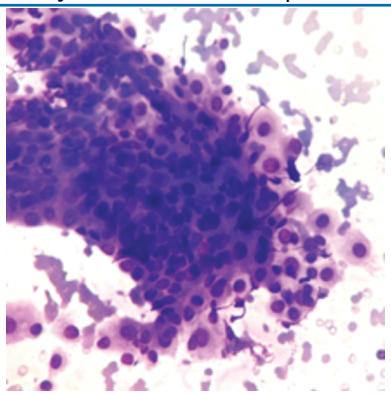

Figure 6: FNAC Smear Showing Epithelial Cells with Scant Cytoplasm, Markedly Pleomorphic Nuclei and Prominent Nucleoli.

DISCUSSION

Malignant CS are rare adnexal tumors with only 51 cases reported in the literature so far.[2] They are considered to be the malignant counterpart of Benign Chondroid Syringoma; biphasic tumors of eccrine cutaneous glands origin comprising of epithelial and mesenchymal components analogous to Pleomorphic adenoma of salivary glands. These usually arise de novo or can rarely develop in a BSC[3,4]. Rapid increase in size and pain draws attention resulting in local destruction, metastasis and also death[9,10].

The age predisposition for these tumors are not clear; mean age at the time of diagnosis was 48.3 years (range 13-86 years)[5,6]. MCS has female predisposition (female: male ratio of 3:2) and affects the extremities commonly[7]. Other sites affected are trunk such as the sacral region and the head and neck region[6]. BCSs are small well-defined lesions, while MCSs may have an infiltrative appearance with median size of 4cm (range 1.4-10cm)[8]. Clinical diagnosis is often challenging due to the lack of defining clinical characteristics and rarity of this condition[3].

In 1988, first attempt to diagnose and document CS on FNAC was made by Masood et al[15]. The aspirate can be thick, mucoid and sometimes gelatinous with moderate cellularity. Scanty stromal elements can be seen in thin aspirate[16]. Alcian blue and mucicarmine stains the mucoid material[15,17]. The epithelial cells can be arranged singly, scattered, in groups or as sheets, attached either loosely or cohesively[18,19,15,20-22]. The individual cells are small to medium sized, monomorphic, round to oval with moderate to dense cytoplasm. The cytoplasm can be eosinophilic to amphophilic giving plasmacytoid appearance[20]. The nuclei appear small, monomorphic, round or oval, central to eccentric in location with fine and evenly distributed chromatin[23,15,20,21,24]. Anisonucleosis, conspicuous nucleoli or nuclei with clear halos can suggest neoplastic changes , but in the absence of other features malignancy can be ruled out[19]. There can be chondroid[15,22], myxoid[17,20,24] or chondromyxoid[23,25,21,26-28] in the background. Myoepithelial cells appear in clusters or aggregates dispersed along with epithelial cells in the stroma and can impart plasmacytoid appearance with dark nuclei[21,29,30].

Studies describing FNAC findings of MCS are also scarce. In 1997, Mishra et al[11] made the first diagnosis of MCS on FNAC. Characteristic findings included were hemorrhagic aspirate, hypercellularity, pleomorphic epithelium, intranuclear and intracytoplasmic vacuolation and pericellular halo. Histopathology confirmed the diagnosis. In 2016, recurrent lesion of MCS was diagnosed by FNAC by Shobhanna et al[10]. The cytology findings were hypercellularity and tissue fragments of malignant appearing round to polygonal cells. Repeat FNAC showed vacuolation, indistinct cell borders, nuclear pleomorphism and multiple prominent nucleoli.

The final diagnosis is based on histopathological examination. The epithelial component has both epithelial and myoepithelial cells with either apocrine or eccrine differentiation. The malignant epithelial component shows hypercellularity, isolated cells, small clusters, solid sheets, glands, infiltrative pattern, necrosis (includes comedonecrosis), nuclear pleomorphism, atypical mitosis, tumor giant cells and perineural or lymphovascular invasion[5,8,7]. The mesenchymal components can show mucinous, cartilaginous or osteoid differentiation[12]. Aggressive behaviour is associated with poor chondroid differentiation and large amount of mucoid matrix. When the subcutaneous/ deep structures are involved along with local recurrences, there is possibility of malignancy in such cases[3,12]. There is no formal grading system due to extreme rarity of MCS.

Utility of immunohistochemical stains may help in confirming the biphasic/ mixed differentiation of these lesions and to rule out metastasis[5,6]. The epithelial component stains positive for cytokeratins including CK5/6, EMA, CEA(glandular differentiation) and p63 (squamous/ myoepithelial differentiation)[6]. S100 and vimentin positivity makes the chondroid differentiation evident[13]

The differential diagnosis can be adenoid cystic carcinoma, mucinous carcinoma and adenoid basal cell carcinoma as they share histopathological features like basaloid cords, tubular or cribriform spaces and specialized stroma.

The clinical course of MCS is considered unpredictable, although it appears to be an aggressive malignancy as per literature. About 50% show local recurrence, 62% show nodal and distant metastasis and about 27% patients die of advanced metastatic disease[3,8]. Distal metastasis commonly involves lung followed by bone and brain[8]. It has also been found that there could be local bone invasion[14].

Mainstay of treatment remains wide local excision and should include the margins.

CONCLUSION

Chondroid syringoma should be included in the differential diagnosis of patients with subcutaneous nodule in the head and neck region. Malignant transformation is a rare event in these tumors. Malignant CS is a very rare neoplasm that is very slow growing with high incidence of recurrence, metastasis and death. FNAC is a very useful tool making preliminary diagnosis of CS along with deep excisional biopsy. However, the final diagnosis is based on histopathological examination. Wide local excision is the mainstay of treatment.

REFERENCES

1. Hirsch P, Helwig EB. Chondroid syringoma. Mixed tumor of skin, salivary gland type. Arch Dermatol 1961;84:835-47.
2. Zufall AG, Mark EJ, Gru AA. Malignant chondroid syringoma: A systematic review. Skin Health Dis 2023;3:e144.
3. Tural D, Selçukbiricik F, Günver F, Karı maz A, Serdengecti S. Facial localization of malignant chondroid syringoma: A rare case report. Case Rep Oncol Med 2013;2013;907980.
4. Mathiasen RA, Rasgon BM, Rumore G. Malignant chondroid syringoma of the face: A first reported case. Otolaryngol Head Neck Surg 2005;133(2):305–7.
5. Lal K, Morrell TJ, Cunningham M, O'Donnell P, Levin NA, Cornejo KM. A case of a malignant cutaneous mixed tumor (chondroid syringoma) of the scapula treated with staged margin-controlled excision. Am J Dermatopathol 2018;40(9):679–81.
6. Nel CE, van der Byl D, Grayson W. Malignant chondroid syringoma: A report of two cases with a sarcomatous mesenchymal component. Dermatopathology (Basel) 2019;6(2):77–84.
7. Krishnamurthy A, Aggarwal N, Deen S, Majhi U, Ramshankar V. Malignant chondroid syringoma of the pinna. Indian J Nucl Med 2015;30(4):334–7.
8. Sánchez Herreros C, Belmar Flores P, De Eusebio Murillo E, Díez Recio E, Cuevas J. A case of cutaneous malignant mixed tumor treated with mohs micrographic surgery. Dermatol Surg 2011;37(2):267 – 70.
9. Mishra K, Agarwal S. Fine needle aspiration cytology of malignant chondroid syringoma: A case report. Acta Cytol 1998;42:1155-8.
10. Shobhanaa PS, Siddaraju N, Jinkala SR, Badhe BA, Ganesh RN. Emphasizing the pivotal role of fine-needle aspiration cytology in a case of recurrent malignant chondroid syringoma. J Cytol 2016;33:103-5.
11. Mishra K, Agarwal S. Fine needle aspiration cytology of malignant chondroid syringoma: a case report. Acta Cytol. 1998;42 (5):1155–1158. doi:10.1159/000332105.

12. Nguyen CM, Cassarino DS. Local recurrence of cutaneous mixed tumor (chondroid syringoma) as malignant mixed tumor of the thumb 20 years after initial diagnosis. J Cutan Pathol 2017;44(3):292–5.
13. Bhosale SJ, Kshirsagar AY, Sulhyan SR, Sulhyan SR, Jagtap SV. Matrix-producing metaplastic breast carcinoma – a rare malignancy. Am J Case Rep 2013;14:213–5.
14. Menéndez RH, Erice SG, Bas CA, Casas G, Dillon HS. Spinal cord compression secondary to metastasis of malignant chondroid syringoma: Case report. J Neurosurg Spine 2015;22(3):310–3.
15. Masood S, Hardy NM. Fine needle aspiration cytology of chondroid syringoma. Report of a case. Acta Cytol. 1988;32(4):482–484.
16. Khan K.Chondroid syringoma: a case with unusual cytological findings. Indian J Dermatol. 2013;58(2):157. doi:10.4103/0019-5154.108072.
17. Gottschalk-Sabag S, Glick T. Chondroid syringoma diagnosed by fine-needle aspiration: a case report. Diagn Cytopathol. 1994;10 (2):152–155.
18. Narasimha A, Kalyani R, Kumar HML, Suresh TN, Supreeth A. Giant chondroid syringoma with divergent differentiation: cyto-histo immuno correlation. Int J Appl Basic Med Res. 2013;3(2):129–131. doi:10.4103/2229-516X.117097.
19. Siddaraju N, Murugan P, Wilfred CD, Choudhury N, Basu D. Preoperative cytologic diagnosis of chondroid syringoma. Acta Cytol. 2009;53(5):607–610. doi:10.1159/000325396
20. Dubb M, Michelow P. Cytologic features of chondroid syringoma in fine needle aspiration biopsies: a report of 3 cases. Acta Cytol. 2010;54(2): 183–186. doi:10.1159/000325005.
21. Kumar S, Ghotekar LH, Thappa DM, Smile R. Diagnosis of chondroid syringoma by fine needle aspiration cytology. Acta Cytol. 2003;47(3): 522–524.
22. Srinivasan R, Rajwanshi A, Padmanabhan V, Dey P. Fine needle aspiration cytology of chondroid syringoma and syringocystadenoma papilliferum. A report of two cases. Acta Cytol. 1993;37(4):535–538.
23. Kumar B. Chondroid syringoma diagnosed by fine needle aspiration cytology. Diagn Cytopathol. 2010;38(1):38–40. doi:10.1002/dc.21159.
24. Rogers R, Zhou F, Grunes D, et al. Chondroid syringoma of the axilla: an unusual tumor diagnosed by fine needle aspiration. Diagn Cytopathol. 2016;44(4):342–346. doi:10.1002/dc.23424.
25. Skoro M, Ostovic KT, Cikara I, Muller D, Novak NP, Virag M. Fine needle aspiration cytology of chondroid syringoma. Coll Antropol. 2010;34(2): 687–690.
26. Lamba S, Nanda A, Kumar U. Chondroid syringoma: fine-needle aspiration cytology of a rare entity at an unusual site. J Clin Diagn Res. 2017;11(7): ED06–ED7. doi:10.7860/JCDR/2017/28405.10135.
27. Nasit JG, Dhruva G. Chondroid syringoma: a diagnosis by fine needle aspiration cytology. J Cutan Aesthet Surg. 2012;5 (3):222–225. doi:10.4103/0974-2077.101404
28. Tokyol C, Aktepe F, Yavas BD, Yildiz H, Aycicek A. Chondroid syringoma: a case report. Acta Cytol. 2010;54(5 Suppl):973–976.
29. Mahantappa H, Ravindra S, Ranganna R, Krishnamurthy T. Cytomorphology of giant chondroid syringoma of the abdominal wall: a case report. Arch Med Health Sci. 2016;4(1):116–118. doi:10.4103/2321-4848.183369.
30. Pal S, Sengupta S, Jana S, Bose K. Fine-needle aspiration cytology of chondroid syringoma of fore arm: report of a rare case. J Cytol. 2014;31(3):171–173. doi:10.4103/0970-9371.145659