Case Report

Malignant chondroid syringoma: An unusual scalp tumor

Geeta Sundar[1], Harish K Rao[2]

From [1]Junior Resident, [2]Professor and Head of Unit, Department of General Surgery, Kasturba Medical College, Mangalore, Karnataka, India

ABSTRACT

Chondroid syringomas are similar to mixed parotid tumors and present in both benign and malignant forms. Malignant chondroid syringoma (MCS) is a rare skin tumor that has a predilection for extremities, particularly in young women. It is even rarer to present as a scalp tumor with very few reported cases in the literature. We present a middle-aged woman, with a history of increasing fatigability of her right arm for the past 3 months who, on examination, was found to have scalp swelling and matted right posterior triangle lymph nodes. The working diagnosis on her was a large sebaceous cyst with secondary in the neck from an occult primary/non-Hodgkin's lymphoma. Preliminary fine-needle aspiration was inconclusive. Imaging followed by wide excision of the tumor and the nearby occipital node was done. The final histopathological diagnosis was MCS with secondary in the lymph node. These tumors are aggressive and metastasize early. Radical surgery is the only hope of cure as adjuvant treatment is yet to be standardized.

Key words: Aggressive, Eccrine tumor, Malignant chondroid syringoma, Scalp, Rhabdoid cells

Malignant chondroid syringoma (MCS), the malignant form of the benign chondroid syringoma (CS), is a very rare, aggressive mixed eccrine tumor, with an increased risk of metastasis to the lungs, bones, brain, and with a higher predilection in younger females. It is noted in the extremities and trunk, with literature showing only very few cases of head-and-neck involvement [1-4]. It is found that these tumors are clinically unpredictable, grow rapidly, and have a big size with histological features showing epithelial and mesenchymal components, necrosis, satellite lesions, atypia, and invasion [1-3]. Moreover, while trauma may be an inciting factor, MCS can arise *de novo* or be attributed to a malignant change in a long-standing CS [2,4]. The treatment includes radical surgical intervention with adequate margins. Adjuvant therapy of chemotherapy/radiotherapy has not been found to be of any benefits [1-5].

MCS presenting as scalp swelling is rarer. We report this unusual case because of the nature of its presentation followed by a rapid progression to death. The patient reported swelling in the scalp for many years, she was not aware of the other swellings in the neck and the only reason for which she sought medical attention was because of the fatigue in the right arm.

The case report was approved by the Institutional Ethical Committee of the Kasturba Medical College Mangalore, India. No information regarding the patient's identities has been used in the case study report or in the images. No real names or identifiers have been used.

Access this article online	
Received- 21 June 2022 Initial Review- 09 July 2022 Accepted- 05 August 2022	Quick Response code
DOI: 10.32677/ijcr.v8i8.3530	

CASE REPORT

A 44-year-old homemaker presented with the complaints of increased fatigability of the right arm for 3 months. She was able to carry out her normal work and had no muscular weakness or sensory disturbances or deficits.

On examination, she was healthy, vitals were stable, and was found to have swelling over the right parieto-occipital area of about 5×7 cm. The skin over the swelling was depigmented and had a peau d' orange appearance. It was sparsely covered by hair and there was no visible punctum. The swelling showed no pulsation and the surrounding skin appeared normal and was covered by normal looking hair. On further inquiry, the patient reports having the swelling for a long time and was not troubled by it. It was slowly enlarging and has not undergone any unusual change in the recent past. Palpation revealed a uniformly soft, non-tender swelling that was moving with the skin and attached to it all around. The swelling was not fluctuant and did not indent. There was no palpable pulsation. A lymph node measuring 1.5×1.5 was palpable just below the swelling. The rest of the scalp was normal on palpation (Fig. 1). The neck examination revealed several matted mobile lymph nodes in the right posterior triangle, measuring about 6×7 cm. The supraclavicular fossa was full but a distinct mass was not felt. The nodes were non-tender and not fixed to the skin. The rest of the neck examination was

Correspondence to: Dr. Geeta Sundar, No.33, NGEF Layout, Nagarbhavi, Bengaluru - 560 072, Karnataka, India. E-mail: harry_geeta@yahoo.co.in

normal. Oral cavity examination was also normal. Examination of the arm and hand revealed no abnormality. Rest of the systemic examination was normal. A working differential diagnosis of sebaceous cyst of the scalp and neck secondary from unknown primary/non-Hodgkin's lymphoma/tuberculosis adenitis was considered.

The patient was advised to undergo fine-needle aspiration (FNA) of the mass. FNA suggested the possibility of amelanotic melanoma, rhabdomyosarcoma, or high-grade carcinoma. The patient was thereupon sent for a full workup including serum lactate dehydrogenase (LDH), computed tomography (CT) scans of the head, neck, and chest with ultrasonography of the abdomen. Her blood investigations were normal except for a marginal rise in serum LDH. CT scan of the head-and-neck region was reported as a large sebaceous cyst with an enlarged occipital lymph node (Fig. 2).

The rest of the findings were as reported on the clinical examination. No additional lesions were discovered elsewhere in the body. The patient underwent excision of the scalp swelling and the lymph node in the scalp under local anesthesia. Skin closure was easy and tension free (Fig. 3).

The histopathological (HPE) report of two sections was studied – one from the node and one from the lesion. The section from the node revealed fragments of lymphoid tissue with proliferating cells with areas of hemorrhage and necrosis; and the section from the lesion showed skin with dermis displaying a partly circumscribed biphasic lesion with proliferating cells arranged in tubules and ductular pattern, lined by bilayered epithelium in a chondromyxoid stroma (Fig. 4). An abrupt transition is noted to solid islands and fascicles of markedly pleomorphic epithelioid and spindled cells. These cells showed moderate eosinophilic cytoplasm, vesicular nucleus, and prominent eosinophilic micronuclei. Few bizarre cells and multinucleate cells were noted. Cells displayed rhabdoid morphology in the myxoid matrix. The intervening stroma showed lobules of cartilage and osteoid with areas of hyalinization. Many mitotic figures were seen. Large geographic areas of necrosis were noted. Margins were all clear and lymphovascular invasion was present.

Immune histochemistry markers such as cytokeratin (CK) and epithelial membrane antigen showed strong focal positivity in the tumor cells; desmin, LCA, and HMB45 were negative; S100 showed strong positivity in the myoepithelial cells of the tubules.

Skin sutures were removed on the 10th day and the wound seemed to have healed well. The neck nodes were deemed inoperable and the patient was referred to a medical oncologist for further management. However, the patient did not report to the oncologist out of fear and misconception about chemotherapy.

Instead, she sought treatment from a homeopathy doctor. However, during the next 3 months of homeopathic treatment, the patient steadily worsened with fatigue and then the onset of breathlessness, following which, she sought oncology consultation at another local tertiary care center. According to available records from that tertiary care; she was admitted with a history of increasing generalized weakness and breathlessness. On examination, she was found to be moderate built and nourished, not anemic, cyanosed, jaundiced, and had no peripheral pitting pedal edema. Her pulse and blood pressure were normal and she had a normal respiratory rate at rest. The scalp wound had healed well and there was no evidence of any residual swelling. The lymph nodes in the right posterior triangle were enlarged and matted with restricted mobility. Supraclavicular fossa was full but no distinct mass was felt. No neurological deficit was noted in the right arm. No other swellings were noted in the neck or elsewhere in the body. The respiratory system on examination was normal. Abdominal examination revealed no organomegaly or free fluid. As per the documented CT neck and positron emission tomography (PET) scan findings; CT showed anterior septae and coarse calcifications (3.8×6.3×5.7 cm) in the subcutaneous plane

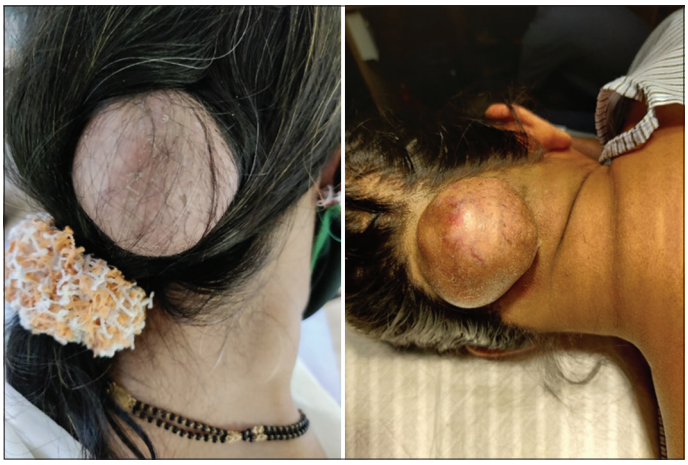

Figure 1: Left side of the image showing the scalp swelling with the right supraclavicular swelling, and the right side showing it preoperatively

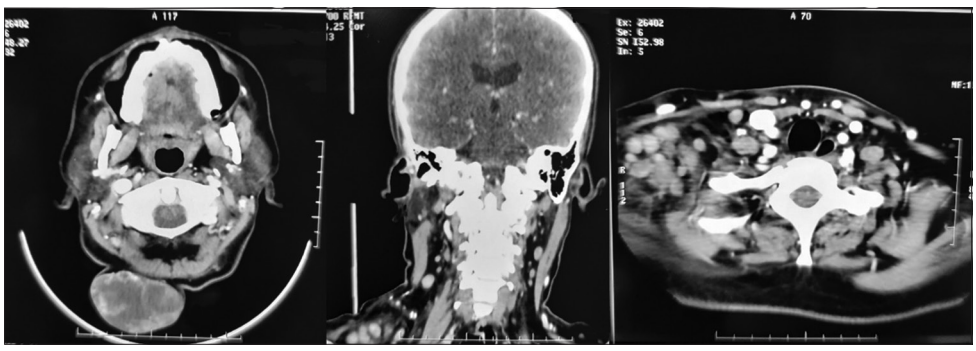

Figure 2: Computed tomography scan of head and neck showing the scalp swelling and enlarged lymph nodes

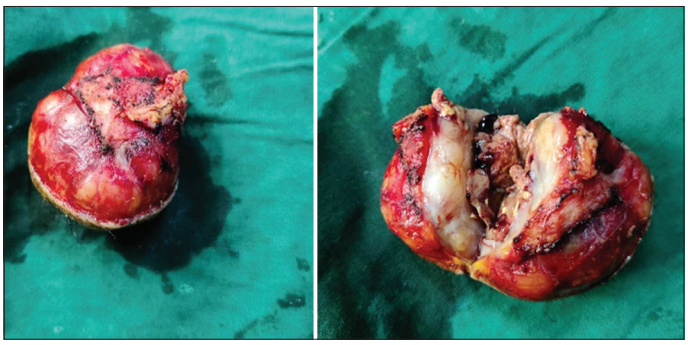

Figure 3: Gross appearance of the lesion

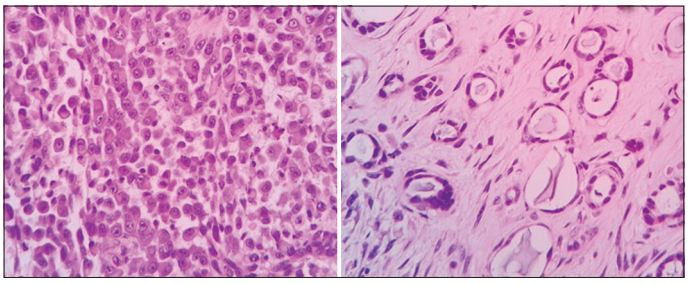

Figure 4: The photomicrograph in ×40 shows pleomorphic cells with epithelioid morphology on the left side and photomicrograph ×400 view showing cells arranged in tubules and ductular pattern lined by a bilayered epithelium in a chondromyxoid stroma on the right side

in the right occiput region extending till the upper body of C3 and multiple right neck nodes; and PET scan showed metabolically active disease with large hypodense lesion extending from the right cervical level 4 to the right axillary region with multiple bilateral hypermetabolic levels 2, 3, and 4 nodes and posterior cervical nodes right>left, with few right axillary subpectoral lymph nodes and intramuscular deposits in the upper back and multiple liver metastases.

After consulting with the hospital oncologist, her primary consultant decided to start her on Tab. Gefitinib 250 mg OD. On taking the medication, the patient had several episodes of vomiting and later that night suffered a cardiac arrest. All attempts at revival failed and she was declared dead. The cause of death as reported in the discharge summary was aspiration pneumonia from persistent vomiting.

DISCUSSION

As compared to its benign counterpart, MCS, with only 50 odd cases reported in the literature [6], tends to occur in younger females, and more commonly in the extremities and trunks. They are known to occur *de novo* or develop in an incompletely resected or pre-existing benign chondroid syringoma [5-7]. These tumors are usually asymptomatic, non-specific, and hence not actively reported by the patients. It is the rapid growth of the tumor or associated pain that induces patients to seek help [8].

There are no established risk factors, for malignant transformation; history of trauma is implicated to probably play a role [6], MCS follows an unpredictable course and commonly metastasizes through lymphatic and hematogenous routes to the lung, bone, and brain [5,9]. MCS is determined by increased growth, size >3 cm, is aggressive, and can present with metastasis [6].

Chondroid syringomas are made up of (a) epithelial components with eccrine and apocrine sweat glands and (b) mesenchymal parts having chondroid, osteoid, adipose, or fibrous characteristics which are situated around epithelial cells with variable glandular differentiation. Fine-needle aspiration cytology can play a role in identifying the diagnosis [6]. Differential diagnoses of MCS can be a sebaceous cyst, dermoid cyst, neurofibroma, pilomatrixoma, amelanotic nevus, and basal cell carcinoma.

Histopathology in MCS shows – atypical cytology, elevated mitotic activity, increased mucoid matrix, nuclear pleomorphism, epithelial cells in ill-forming cords with a hemorrhagic background, poor differentiation, necrosis, capsular invasion, and satellite nodules with immunohistochemistry showing possible combinations of CKs, carcinoembryonic antigen, CK, PS100, vimentin, actin, calponin, and myoepithelial cells [5-8].

Imaging studies are routinely carried out to identify the lymph nodal involvement or the extent of metastasis, with magnetic resource imaging playing a bigger role in identifying the extent and dimensions of the tumor, tissue of origin, and site [7]. According to a systematic review by Zufall *et al.*, the site and size of the primary tumor are more to play a role in prognosis and mortality [7].

Our patient is a middle-aged female who presented with a large swelling over the right side of her scalp with associated lymph nodal swellings in the neck, and no other associated symptoms. A differential diagnosis of sebaceous cyst of scalp and neck secondary from an unknown primary was assumed. Imaging investigations showed a possibility of a sebaceous cyst and enlarged nodes. The patient underwent wide local excision of the scalp swelling with node under local anesthesia with primary closure. The final HPE report showed MCS with clear margins. She was referred for a medical oncologist's opinion, but it was delayed and she developed progression of disease with systemic metastases due to delay in seeking a timely opinion and appropriate treatment.

The treatment is total adequate surgical excision with free margins to achieve disease control and prevent recurrence because it takes care of the microscopic satellite nodules [5-7]. The literature has shown that around 49% of MCS can recur. Radiotherapy and chemotherapy (such as 5-fluorouracil, Adriamycin, cisplatin, and cyclophosphamide) [2] have not yet been proven to have value in treatment, but are given in advanced cases of chondroid syringoma because of its unpredictable nature and technical difficulty in completely excising over sensitive areas. Given the recurrence rate of the tumors, long-term patient follow-up is to be maintained [5,6,9].

CONCLUSION

The report represents one of the rarer findings of an aggressive scalp MCS that on appearance appeared benign. It was possible

that the tumor was benign to start with and recently turned malignant which can be often seen in such tumors.

A malignant tumor of the scalp would have reasonable in view of the large size, peau d' orange appearance of the skin, enlarged lymph node in the proximity of the tumor, and further nodes in the posterior triangle of the neck. And least we are aware of the condition and its prognosis, misjudgment is likely that may lead to poor outcomes.

REFERENCES

1. Malik R, Saxena A, Kamath N. A rare case of malignant chondroid syringoma of scalp. Indian Dermatol Online J 2013;4:236-8.
2. Nel CE, van der Byl D, Grayson W. Malignant chondroid syringoma: A report of two cases with a sarcomatous mesenchymal component. Dermatopathology (Basel) 2019;6:77-84.
3. Ka S, Gnangnon F, Diouf D. Malignant chondroid syringoma in a West African cancer institute: A case report. Int J Surg Case Rep 2016;25:137-8.
4. Tural D, Selçukbiricik F, Günver F, Karışmaz A, Serdengecti S. Facial localization of malignant chondroid syringoma: A rare case report. Case Rep Oncol Med 2013;907980:3
5. Takahashi H, Ishiko A, Kobayashi M, Tanikawa A, Takasu H, Tanaka Md M. Malignant chondroid syringoma with bone invasion: A case report and review of the literature. Am J Dermatopathol 2004;26:403-6.
6. Favareto SL, Pellizzon AA, Lopes Pinto CA, Bertolli E, Castro DG. The role of radiation therapy as an adjuvant treatment in nodal metastasis of malignant chondroid syringoma. Cureus 2020;12:e11360.
7. Zufall AG, Mark EJ, Gru AA. Malignant chondroid syringoma: A systematic review. Skin Health Dis 2022:e144. Doi: 10.1002/ski2.14412of12-ZUFALLETAL.
8. Chauvel-Picard J, Pierrefeu A, Harou O, Breton P, Sigaux N. Unusual cystic lesion of the eyebrow: A case report of malignant chondroid syringoma. J Stomatol Oral Maxillofac Surg 2018;119:232-5.
9. Sekar R, Kalaiarasi R, Ganesan S, Alexander A, Saxena SK. Malignant chondroid syringoma with nose and paranasal sinus extension: A case report Allergy Rhinol (Providence) 2019. Doi: 10.1177/2152656719893367.

Funding: *None;* **Conflicts of Interest:** *None Stated.*

How to cite this article: Sundar G, Rao HK. Malignant chondroid syringoma: An unusual scalp tumor. Indian J Case Reports. 2022;8(8):249-252.