

Malignant chondroid syringoma of the pinna

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ABSTRACT

Chondroid syringoma (CS) represents the cutaneous counterpart of mixed tumor (pleomorphic adenoma) of salivary glands. The malignant counterpart of CS, termed as “malignant CS” is a malignant eccrine neoplasm which lacks distinctive clinical features, often delaying initial diagnosis. Unlike its benign counterpart which often localizes in the head and neck region, malignant CS most often encountered in the trunk and the extremities. We report a rare case of an aggressive malignant CS of the left pinna with cervical lymph node metastasis. Our patient, to the best of our knowledge, possibly is the first case of malignant CS of the pinna and the fourth to arise in the head and neck region. The diagnostic challenges with an added emphasis on the role of positron emission tomography-computed tomography in aiding the management of this rare tumor are discussed.

Keywords: Ear tumors, malignant chondroid syringoma, pleomorphic adenoma, skin tumors

INTRODUCTION

Chondroid syringomas (CS), first described by Hirsch and Helwig in 1961, is a rare, benign, skin appendageal tumor,^[1] the reported incidence of which is <0.098% of all the primary cutaneous neoplasms. Being a counterpart of mixed tumor (pleomorphic adenoma) of salivary glands, it is also termed as “mixed tumor of the skin” The malignant variant of CS is believed to be even rarer and has commonly been reported in the extremities and trunk and extremely rarely in the head and neck region.^[2-4] Our patient, to the best of our knowledge, is the first case of malignant CS of the pinna and the fourth to arise in the head and neck region.

CASE REPORT

A 41-year-old gentleman without any known comorbid illnesses was initially evaluated at an outside center for multiple swellings of the left neck of 4 months duration. A fine needle aspiration cytology (FNAC) from one of the swelling was suggestive of a poorly differentiated carcinoma and he was referred to our center for further evaluation.

Clinical examination at our center revealed the multiple enlarged left cervical lymph nodes involving levels II, III, and IV of varying sizes, the largest measuring 2.5 cm × 2.5 cm in the left level IIA. The thyroid gland was found to be minimally enlarged with multiple ill-defined nodules along both the lobes. Clinical examination further revealed a painless 3 cm × 3 cm swelling in the upper part of the left helix (of the left pinna) which was firm, fixed to the skin, but was freely mobile over the underlying ear cartilage [Figure 1]. When further probed, the patient gave a history of a long-standing swelling in the left pinna with a slight increase in size over a period of 4 months. Examination of the rest of the upper aero-digestive tract was normal.

An ultrasound neck showed multiple nodules in both lobes of thyroid alongside multiple significant nodes in the left neck along levels II, III, and IV. An ultrasound-guided FNAC from the thyroid showed benign follicular cells with occasional inflammatory cells, but no atypical cells. An FNAC done from left level II lymph node, however, was suggestive of carcinoma of glandular origin (immunohistochemistry for thyroid transcription factor-1 was negative). A provisional diagnosis of an adenocarcinoma from an unknown primary with an incidental sebaceous cyst in the left pinna was made. Further evaluation with a positron emission tomography-computed tomography (PET-CT) showed avidity in the multiple cervical lymph nodes involving the left level IIA (standardized uptake value [SUV] 3.8), left level III and left level IV (SUV 2.9). Further, positron emission tomography avidity was also noted in the lobulated soft tissue lesion with necrosis arising from left

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pinna (SUV 2.9) and multiple hypo dense lesions in both lobes of thyroid (SUV 4.2) [Figures 2 and 3].

A repeat FNAC from the thyroid showed no atypical cells. However, the FNAC from the left ear helix swelling interestingly showed few clusters of atypical epithelial cells suggestive of epithelial malignancy. A preoperative diagnosis of a possibly skin adnexal tumor with cervical lymph node metastasis was made. The patient underwent a wide excision of ear helix tumor and left radical neck dissection. The defect in the pinna was bridged by a local advancement flap [Figure 4]. His final histopathology was suggestive of a malignant CS of the left pinna resected with clear margins and metastasis in 48/53 left cervical lymph nodes [Figure 5]. The patient after a multi-disciplinary board discussion was considered for adjuvant radiotherapy to the tumor bed and neck.



Figure 1: Clinical photograph at presentation

DISCUSSION

Chondroid syringomas are rare skin tumors which are known to arise from the eccrine sweat glands.^[1] They belong to the family of myoepithelial tumors, which consists of morphologically similar tumors that includes, pleomorphic adenoma of the salivary gland, CS of skin and mixed tumor of somatic soft tissue and bone.

They usually present as a painless, solid, subcutaneous or dermal nodule with a normal margin. The mean age of the patients at the time of diagnosis was 48.3 years (range 13–84 years). Most CS are benign with a slow progression over years.^[5] They can be confused clinically with various other skin lesions including dermoid or sebaceous cysts and neurofibromas. Malignant CS are extraordinarily rare tumors which may originate de novo or can rarely develop in a CS.^[4,5] Unlike its benign counterpart, the malignant CS occurs predominantly in females (3:2), has no definite age predilection and has been reported to occur more commonly in the extremities.^[4] Malignant lesions are usually >3 cm; however benign tumors measuring 10 cm have also been reported.^[5]

A few reports have described the magnetic resonance imaging findings of benign and malignant CS, which are in fact nonspecific. There are hardly any reports of the use of PET-CT as a staging modality in the management of malignant CS.

The histological features suggestive of malignancy in a CS include cytologic atypia, infiltrative margins, satellite nodules, necrosis, and involvement of the deeper structures.^[4] Malignant tumors that may bear similar characteristics with malignant CSs include well-differentiated and myxoid liposarcomas, extraskeletal

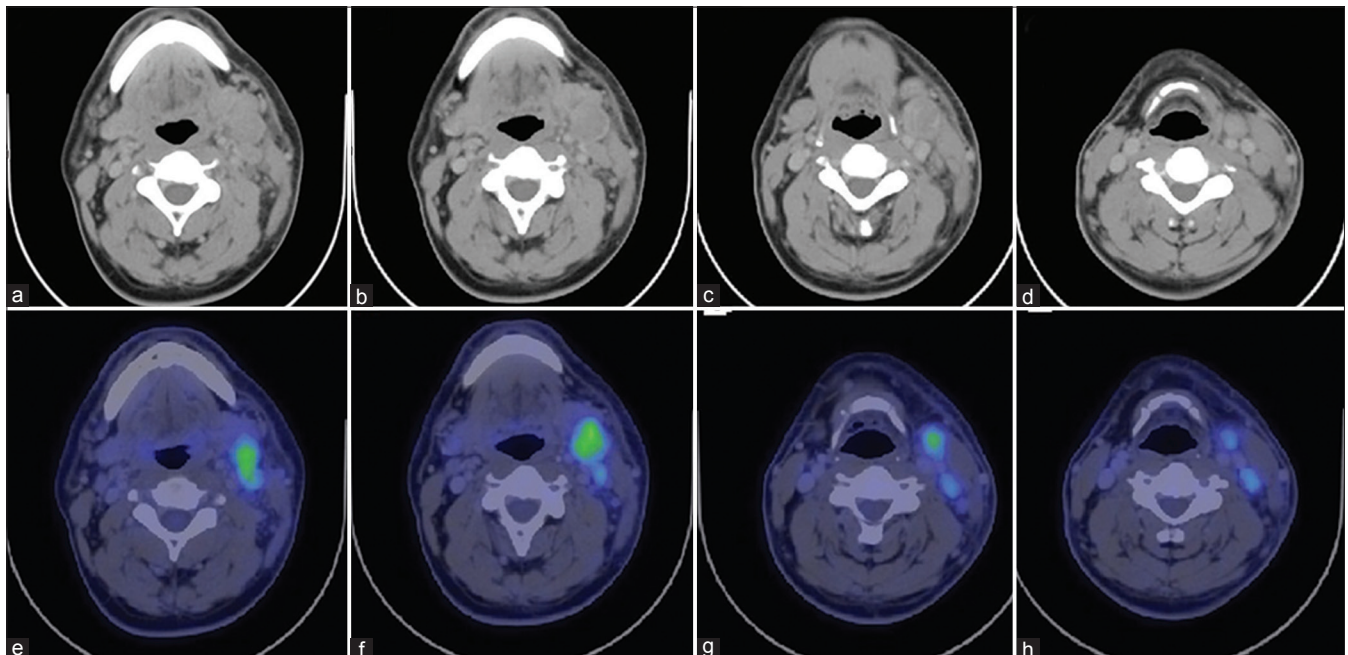


Figure 2: (a-d) Axial computed tomography (CT) images, (e-h) the corresponding axial positron emission tomography-CT image showing uptake in the multiple cervical lymph nodes involving the left levels II to IV

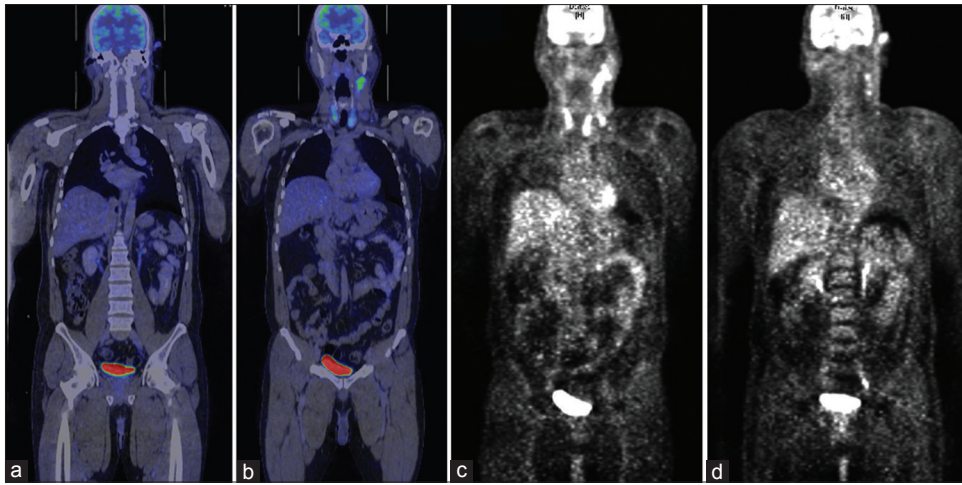


Figure 3: (a and b) Coronal positron emission tomography-computed tomography images, (c and d) coronal positron emission tomography (PET) images: Showing uptake in the multiple cervical lymph nodes involving the left level IIA (standardized uptake value [SUV] 3.8), left level III and left level IV (SUV 2.9). Further, PET avidity was also noted in the lobulated soft tissue lesion with necrosis arising from left pinna (SUV 2.9) and multiple hypo dense lesions in both lobes of thyroid (SUV 4.2)



Figure 4: Postoperative clinical photograph of the patient following the wide excision of left ear helix tumor and left radical neck dissection. The defect on the pinna was bridged by a local advancement flap

myxoid chondrosarcoma, myxofibrosarcoma, and malignant peripheral nerve sheath tumors.

The management of CS is predominantly surgical and consists of complete excision of the tumor. For malignant CS, the initial treatment modality is aggressive surgery aimed at achieving negative margins.^[6-9] Adjuvant radiotherapy, with or without chemotherapy have been used but with limited success.^[7,8]

Malignant CS tend to behave in an unpredictable manner; deaths have been reported to occur as early as 9 weeks following surgery; on the other hand, the longest reported survivor has been 12 years.^[4] Of the reported cases, nearly half of the patients have had local recurrences and 39% had nodal metastases.^[6] Distant metastases were reported in about 36% of the cases, the most common site was the lung, followed by bone and brain.^[6] There are no uniform guidelines with regards to the management of malignant CS, especially in the metastatic setting. A PET-CT

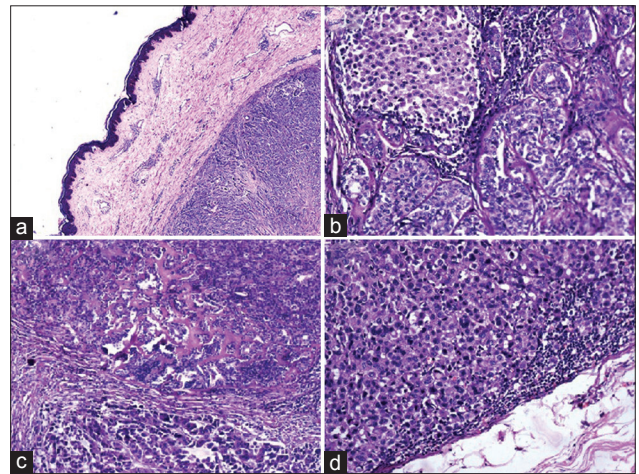


Figure 5: (a and b) Section shows a malignant neoplasm composed of lobules of cells arranged in a glandular pattern forming cystic lumina admixed with lobules composed of solid sheets of plasmacytoid cells with eosinophilic to clear cytoplasm, numerous tumor giant cells, comedonecrosis and increased mitosis (H and E $\times 20$), (c) shows chondroid syringoma with frank carcinomatous areas, (d) chondroid syringoma showing metastasis in the cervical lymph nodes

may be a useful tool in staging and in the follow-up of patients with malignant CS considering its high propensity for nodal and distant metastasis. Targeted therapies with PI3K/AKT/mTOR pathway inhibitors, currently in early clinical trials, are being explored as potential treatment options.^[10]

CONCLUSION

Malignant CSs, being biologically aggressive tumors, presents with many diagnostic and management challenges. A PET-CT scan may be a useful modality in staging as was seen in our patient. However its role needs to be better defined. Achieving a negative surgical margin using wide excision technique remains the mainstay of management of malignant CSs; these patients need to be further monitored closely due to its heightened metastatic potential.

REFERENCES

1. Hirsch P, Helwig EB. Chondroid syringoma. Mixed tumor of skin, salivary gland type. *Arch Dermatol* 1961;84:835-47.
2. Malik R, Saxena A, Kamath N. A rare case of malignant chondroid syringoma of scalp. *Indian Dermatol Online J* 2013;4:236-8.
3. Araújo JL, de Aguiar GB, do Prado Aguiar U, Mayrink D, Saade N, Veiga JC. Malignant chondroid syringoma with central nervous system involvement. *J Craniofac Surg* 2012;23:514-5.
4. Mathiasen RA, Rasgon BM, Rumore G. Malignant chondroid syringoma of the face: A first reported case. *Otolaryngol Head Neck Surg* 2005;133:305-7.
5. Sungur N, Uysal A, Gümüş M, Koçer U. An unusual chondroid syringoma. *Dermatol Surg* 2003;29:977-9.
6. Takahashi H, Ishiko A, Kobayashi M, Tanikawa A, Takasu H, Tanaka M. Malignant chondroid syringoma with bone invasion: A case report and review of the literature. *Am J Dermatopathol* 2004;26:403-6.
7. Seenu V, Goel AK, Shukla NK, Prakash MB, Mohan R. Malignant chondroid syringoma of scalp with cervical lymph node metastasis. *J Assoc Physicians India* 1996;44:565-6.
8. 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;22:310-3.
9. Shashikala P, Chandrashekar HR, Sharma S, Suresh KK. Malignant chondroid syringoma. *Indian J Dermatol Venereol Leprol* 2004;70:175-6.
10. Le LP, Dias-Santagata D, Pawlak AC, Cospes AK, Nguyen AT, Selim MA, *et al.* Apocrine-ecrine carcinomas: Molecular and immunohistochemical analyses. *PLoS One* 2012;7:e47290.

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