A rare case of malignant chondroid syringoma of scalp

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ABSTRACT

Chondroid syringoma represents the cutaneous counterpart of mixed tumor (pleomorphic adenoma) of salivary glands, therefore it is also termed “mixed tumour of the skin.” It is generally accepted that there are both eccrine and apocrine variants of mixed tumors of skin. Malignant chondroid syringoma is a malignant eccrine neoplasm that is very rarely encountered in radio-pathological and clinical practice. Unlike the benign variety which occurs most often on the head and neck, malignant chondroid syringomas are most often found on the trunk and extremities. The usual presentation is that of an asymptomatic slow-growing mass in the region of head or neck. We present here a case report of histopathologically proven malignant chondroid syringoma of scalp in occipital region, with dural invasion. Malignant chondroid syringomas clinically appear as very large nodules and they metastasize at a very high rate. The treatment of these malignancies is surgical excision.

Key words: Chondroid syringoma, pleomorphic adenoma, scalp tumors, skin tumors

INTRODUCTION

Pleomorphic adenoma, or chondroid syringoma (CS), is a rare, benign, skin appendageal tumor.\textsuperscript{[1]} The case is rare and unique because the incidence of this tumor is very low; the reported incidence is <0.098% amongst all primary skin tumors. Moreover, the malignant counterpart is even more rare (30 cases worldwide till now). Finally, most malignant CSs have been reported in the extremities and trunk except for two, which were reported in face.\textsuperscript{[2,3]} To our knowledge, our case is only the third case of malignant CS reported in the region of head.

CASE REPORT

A 61-year-old female presented with a large, fungating, and ulcerated growth over the occipital region of scalp [Figure 1]. The history was that of a painless, slow growing mass over a 9-month time period. Clinically the patient was suspected to have a squamous cell malignancy of scalp (which was the most confusing differential diagnosis). Initial biopsy of the lesion favored a benign mixed eccrine tumor of the scalp, that is, a benign CS. The patient was then referred for contrast enhanced computerized tomography (CECT) scan of head. The scout image was suggestive of a soft tissue lesion in the occipital region with erosion of the adjacent occipital bone [Figure 2]. CECT revealed a moderately and heterogeneously enhancing soft tissue density mass lesion of size approx. 8.0 1.4 cm, arising from the occipital scalp, causing lytic destruction of the adjacent occipital bone. Also, the underlying dura appeared focally thin,
with white matter edema in the occipital region, suggesting dural invasion [Figure 3]. Wide local excision with primary closure was done. Histopathology revealed small groups as well as scattered pleomorphic epithelial cells having hyperchromatic nuclei and scanty cytoplasm with one to two mitotic figures per high power field, surrounded by abundant basophilic chondromyxoid stroma, diagnostic of malignant CS [Figure 4]. The patient developed high-grade fever and altered sensorium, and died on the 2nd postoperative day, possibly due to meningitis. This infection could have spread to the meninges from operative site, as there was certain evidence of meningeal invasion on CECT.

**DISCUSSION**

First described by Hirsch and Helwig in 1961, CSs are rare, benign tumors of the skin that arise from the eccrine sweat glands.\(^4\) Lesions are typically located on the head and neck, and are non-ulcerating, slow growing, subcutaneous, or dermal nodules. These affect middle-aged men more than women.\(^5,6\) Although most are benign, malignant forms have been reported.\(^2,7,8\) Unlike its benign counterpart, the malignant form occurs predominantly in females, has no age related predilection, and is observed more commonly on the extremities.\(^2,7,9\) Only two cases have been reported to occur in the head.\(^2,3\) Differentiation toward various skin adnexal structures (including hair matrix, hair follicle, apocrine, and sebaceous glands) is rare. CS may be confused clinically with various skin lesions, including benign tumors of epidermal or mesenchymatous appendages, such as dermoid or sebaceous cyst, and neurofibroma. Various treatment modalities have been proposed for CS, namely electrodissection, dermabrasion, and vaporization with argon or CO2 laser.

Malignant CS is one of the rarest subtypes and appears to behave in an aggressive manner. The reported cases occurred in the extremities and torso of young female patients. Tumors greater than 3 cm in size have a greater likelihood of malignancy. Histological features suggestive of malignancy include cytologic atypia, infiltrative margins, satellite tumor nodules, tumor necrosis, and involvement of deep structures.\(^2,10\) For malignant lesions, the initial treatment modality is aggressive surgery. Adjuvant radiotherapy, with or without chemotherapy, may be recommended.

To conclude, in the evaluation of neoplastic growths of scalp, CS should be considered in the differential diagnosis. Invasion
into adjacent structures (such as calvarium, dura, etc.) should
suggest strong possibility of malignancy. However, excisional
biopsy causing minimum possible destruction of aesthetic
and functional structures is the final diagnostic, as well as
therapeutic approach.

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Cite this article as: Malik R, Saxena A, Kamath N. A rare case of malignant
chondroid syringoma of scalp. Indian Dermatol Online J 2013;4:236-8.

Source(s) of Support: Nil, Conflict of Interest: No