



Case Report

SEBACEOUS CARCINOMA OF SCALP- RARE SKIN CANCER - A CASE REPORT

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Introduction:

Sebaceous gland carcinoma is an aggressive, uncommon, cutaneous tumor. This tumor is thought to arise from sebaceous glands in the skin and thus may arise anywhere on the body where these glands exist. Approximately 75% of these tumors arise in the periocular region, an area rich in sebaceous glands. This tumor exhibits an aggressive clinical course with a significant tendency for both local recurrence and distant metastasis.

Diagnosis and treatment tend to be delayed because sebaceous carcinoma is frequently mistaken for more common benign entities, further complicating treatment of this aggressive malignancy. In addition to its varied clinical appearance, a varied histologic appearance may occur, and delayed diagnosis or misdiagnosis following a biopsy is not uncommon.

When arising in the periocular region, the clinical presentation is often variable, and sebaceous gland carcinoma is often not initially suspected. Instead, patients may receive multiple courses of incision and drainage for chalazion before a definitive biopsy is performed.

Most sebaceous gland carcinomas have no obvious etiology. Only a few are associated with Muir-Torre syndrome which is an autosomal dominant genodermatosis consisting of sebaceous neoplasms

(sebaceous adenomas, sebaceous carcinomas, and sebaceous epitheliomas) with or without keratoacanthomas and one or more low-grade visceral malignancies. In approximately 40% of cases with Muir-Torre syndrome patients develop some type of sebaceous tumor before or concurrent with visceral malignancy. An evaluation for this syndrome is advisable once sebaceous gland carcinoma is diagnosed.

Case summary:

Mrs Nafsia Begum a 69-year-old women presented with the history of a nodular swelling on occipital region of scalp for one year. It was gradually increasing in size during this period but otherwise asymptomatic. There was no significant past medical history and drug history. There was also no family history of malignancy.

On examination there was a nodular swelling over the scalp in occipital region which was 1cmx1cm in diameter, hard in consistency, fixed with the skin but not with underlying structures and the surrounding area was normal. There was no regional enlarged lymph-node and general examination including abdominal examination was normal.

The differential diagnoses include: Neurofibroma, lipoma and sebaceous cyst.

The nodule was excised on 23th January 2011 and sent for histopathological examination which revealed the epidermis is unremarkable, the dermis presents a malignant tumor composed of atypical large cells having clear cytoplasm and pleomorphic hyperchromatic nuclei arranged in nests, suggestive of sebaceous differentiation. Large areas of necrosis also seen. The neoplasm contained lobules of

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undifferentiated cells with focal vacuolated sebocytic differentiation and sebaceous ductal differentiation (Figure 1). The histological diagnosis was sebaceous carcinoma (SC). There was no comment on resection

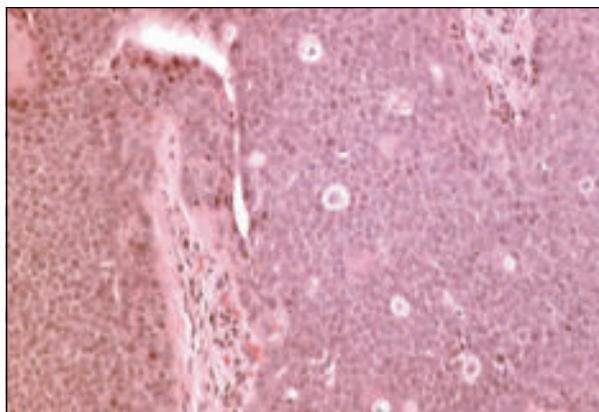


Fig.-1: Histopathological appearance of Sebaceous Carcinoma.

margins. The slide was reviewed which revealed same report.

On 05th February 2011 wide local excision (2cm from the margin) done under the guidance of frozen section biopsy and wound was covered by four wings flap. There was no residual tumor on histopathological report.

As sebaceous carcinoma may be associated with Muir-Torre syndrome, screening for underlying neoplasia was performed. Results of complete blood picture, ESR, renal and liver function tests, carcino-embryonic antigen, alpha-fetoprotein were normal. Faecal occult blood test was negative. Ultrasonography of abdomen and pelvis showed normal. Chest X-ray and CT scan of brain revealed normal. The patient was referred for colonoscopy to exclude underlying colonic neoplasm which was normal.

The patient was reviewed 6 months interval regularly for one year. On history and physical exam there was no local recurrence or no regional lymph-adenopathy. For assessing distal metastasis we did chest X-ray, ultrasonography of abdomen and pelvis, CEA, complete blood picture, faecal occult blood test and colonoscopy which revealed normal.

Discussion

Sebaceous carcinoma is a malignancy of the sebaceous glands first well-described by Allaire in 1891¹. It consists of a single yellow or orange nodule.

The lesion grows slowly and often appears after 40 years of age. The face particularly the eyelids and scalp are the most commonly affected sites. It has also been reported to arise from actinic keratoses² and Bowen's disease of the vulva³. Metastases are common in sebaceous carcinomas affecting the upper eyelid, often spreading to the regional lymph nodes and viscera. Metastases are uncommon in extra-ocular lesions. Sebaceous carcinoma may be a marker for the Muir-Torre syndrome which is an autosomal dominant genodermatosis consisting of sebaceous neoplasms (sebaceous adenomas, sebaceous carcinomas, and sebaceous epitheliomas) with or without keratoacanthomas and one or more low-grade visceral malignancies⁴.

Extra orbital sebaceous carcinoma is very rare. So far only 150 cases of extra orbital sebaceous carcinoma were reported in the literature⁵. The extra orbital sebaceous carcinoma commonly occurs on head and scalp due to the abundant sebaceous glands⁶ the other sites involved by Sebaceous carcinoma are external genitalia and extremities⁷. However the incidence of metastasis is less than 10% in extra orbital sebaceous carcinoma. Sebaceous carcinoma is a disease of 6th and 7th decade of life occurs in men and women equally. The incidence varies from 0.2% to 4.6% of all skin cancers, the incidence is about 0.6% in people below 20 years of age⁸.

In a study of 59 cases of sebaceous gland neoplasms, 42% were found to have associated malignancies.⁹ Sebaceous carcinoma may therefore be a marker of this syndrome and may precede the onset of malignancy for more than 20 years. It is more common in males (male: female ratio 3:2) and affects patients between 31 and 89 years of age but may occur in early age. There is a family history of malignancy in 87% of cases¹⁰.

In another study of 147 patients with Muir-Torre syndrome and internal malignancy, colorectal carcinoma was the most common visceral malignancy (53% of cases), genito-urinary neoplasms were the next most commonly associated malignancy (25%)¹¹. Other associated visceral malignancies include breast, head and neck, haematological, lung and gastrointestinal tract malignancies are occasionally reported.

Extra orbital sebaceous carcinoma has a different behaviour compared to the orbital one which is

considered less aggressive than orbital Sebaceous carcinoma¹². Extra ocular sebaceous carcinoma is known to recur locally and spread to lymph nodes and visceral organs¹³A review of 91 cases of extraocular sebaceous carcinoma by Bailet et al. reported a 21% rate of metastasis constituting 15% of regional lymph node metastasis and 10% of distant metastases that include pulmonary, visceral or central nerve system. A 29% recurrence rate following treatment was observed¹⁴.

Initial evaluation in the history and physical examination should be aimed at searching for a possible affected organ. Investigations including chest X-ray and CT abdomen, etc. should be based on the results of the physical examination. As colonic carcinoma is commonly associated with this syndrome, carcino-embryonic antigen and stool for occult blood are often performed. Colonoscopy or barium enema is recommended at the initial evaluation and at three to five-yearly intervals. Life-long surveillance for malignancy is required in patients with sebaceous gland tumours. Family members are also screened for sebaceous tumours and for visceral malignancies. Once detected sebaceous lesions (sebaceous adenoma or epithelioma) should be excised with wide margins.

We conclude that sebaceous carcinoma is an aggressive skin malignancy and the management of this is challenging. Due to its rare occurrence, excision with wide margins is the current standard of care. Patients with sebaceous carcinoma have to be followed up closely for assessment of recurrence and distant metastasis.

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