DOI: 10.7860/JCDR/2015/13682.6232

Case Report

Ear,Nose and Throat Section

Malignant Cylindroma of Post Aural Region Involving the Temporal Bone

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ABSTRACT

Dermal eccrine cylindroma is a benign adnexal tumour commonly affecting the neck, scalp and skin of elderly individuals. These are poorly circumscribed dermal or subcutaneous lesions consisting of numerous rounded ovoid or cord shaped dermal island that fit together to form a jigsaw pattern. Malignant transformation is not commonly seen. This case highlights malignant transformation of a dermal eccrine cylindroma in the post aural region extending to involve the underlying mastoid bone.

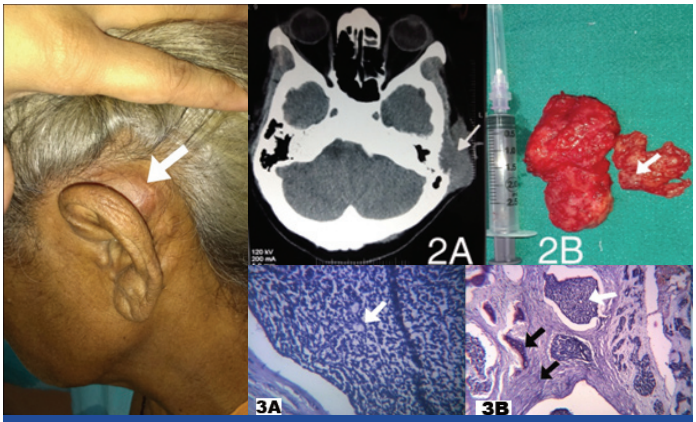
Keywords: Eccrine cylindroma, Malignant transformation, Temporal bone

CASE REPORT

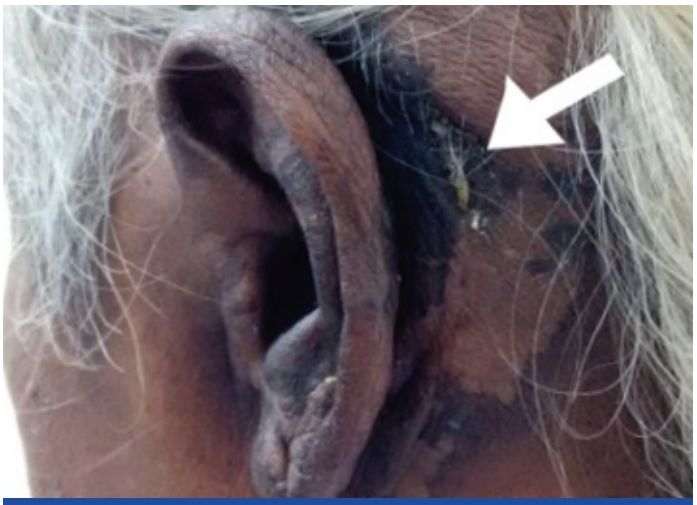
A 60-year-old female presented to the ENT OPD with complaints of left sided post aural swelling since 3 months, which was insidious in onset and gradually progressive in nature. Swelling was associated with pricking type of pain which was intermittent in nature and radiating to pre-auricular region and ear pinna on same side. Patient was a known case of type II diabetis mellitus since 10 years and was on oral hypoglycemic drugs. There was a history of swelling at the same site 2 years back for which she had undergone surgical excision and biopsy was reported as benign dermal cylindroma. There was no history of similar symptoms among the family members.

On examination left side post aural region showed a single smooth swelling over previously healed scar mark which was approximately 4.2 x 3.6 cm [Table/Fig-1]. Skin over the swelling was normal on examination. On palpation mass was tender, hard in consistency, non mobile and there was no local rise of temperature. Skin above the swelling was not fixed and Tragal sign was negative. External auditory canal on inspection was narrowed due to the mass effect on the post canal wall. Tympanic membrane was found to be normal with normal mobility. There were no clinically palpable lymph nodes in the head and neck region. CT scan of the temporal bone showed a single mass on the left post aural region eroding the left mastoid bone [Table/Fig-2a]. Middle ear was normal and with intact ossicles. Inner ear was found to be normal. Needle aspiration cytology showed features of an epithelial neoplasm with basaloid features. Screening CT scan of neck, brain, chest and abdomen did not show any evidence of distant metastasis.

The mass was surgically excised under general anesthesia along with 2.5cm free margins. Left mastoid bone was thinned and eroded by the mass. Mass excised was sent for histopathological examination and was reported as “Malignant transformation of eccrine cylindroma”. Gross tumour specimen showed irregular grayish white areas [Table/Fig-2b] and histopathological slide showed several large irregular islands of small darkly staining cells with basaloid features exhibiting nuclear atypea [Table/Fig-3a]. Typical jigsaw pattern, Peripheral pallisading and hyaline sheath as seen in benign cylindroma was absent and malignant cells were found infiltrating into adjacent fat, skeletal muscles, fibrous tissue and bone [Table/Fig-3b]. Perineural tumour infiltration was present. Postoperative period was uneventful with remnant skin flap in post aural region healing well. Patient was referred for radiotherapy and she is on regular follow up [Table/Fig-4].



[Table/Fig-1]: Clinical picture showing smooth swelling over left post aural region (white arrow) **[Table/Fig-2]:** (a) CT Scan showing a hypodense mass over the left post aural region causing erosion of the underlying mastoid bone (white arrow). (b) Tumour specimen of size 4.2 x 3.6 cm showing irregular grayish white areas (white arrow). **[Table/Fig-3]:** (a) Histopathological section showing deeply staining malignant cells with basaloid feature (white arrow) and nuclear atypea (H & E, 100x) (b) Histopathology showing clusters of deeply staining malignant cells showing basaloid features (white arrow) and infiltrating into the adjacent nerve, bone, stroma, and skeletal muscle (black arrow) (H&E, 400x) with loss of typical jigsaw pattern and peripheral pallisading



[Table/Fig-4]: Healed left post aural region following surgery and radiotherapy

DISCUSSION

Dermal eccrine cylindroma is a benign adnexal tumour that commonly affects the scalp, neck and face of elderly individuals [1]. They present predominantly in older women with 9:1 female to male ratio [1,2]. Molecular studies of familial and sporadic cylindromas have shown frequent alterations at chromosome 16q 12-13 that have recently been found to house the cylindromatosis gene (CYLD) [2]. Defective laminin 5 processing is felt to contribute to the aberrations of the basement membrane [1,3]. The presence of multiple cylindromas

is associated with the Brooke-Spiegler syndrome, in which an autosomal dominantly inherited mutation leads to the development of multiple cylindromas, trichoepitheliomas and spiradenomas primarily located on the head and neck [4]. Cylindromas are usually poorly circumscribed dermal or sub-cutaneous lesions consisting of numerous rounded, ovoid or cord-shaped basaloid dermal islands that fit together like a jig-saw puzzle [5]. Each lobule is surrounded by a distinct basement membrane like hyaline sheath [5]. The tumour islands themselves contain a mixture of small dark and larger pale cells, hyaline droplets and occasional tubular lamina lined by two cell layers. Peripheral pallisading of the small dark cells is often seen [5].

Malignant transformation of single or multiple dermal cylindromas is very rare and there is very little literature evidence available on the subject. The exact aetiology for the malignant transformation is still unclear. Studies have shown that risk factors like prior incomplete surgical excision, exposure to radiation, or frequent trauma at a particular site may be the cause for malignancy [6]. Malignant transformation is clinically seen as recurrence of the lesion, rapid increase in size, and ulceration or bleeding of the lesion [4,6]. Our case presented as a recurrent post aural mass in an elderly female with previous history of surgical excision and rapid increase in size, along with erosion of the underlying mastoid bone. A similar case was reported by Kuklani RM et al., in a 79-year-old female presenting with multiple large swelling over the right ear, scalp and neck region [4].

Malignant cylindromas on histopathological examination show presence of prominent large pale cells. The cells exhibit nuclear atypea, contain pleomorphic nucleoli with increased mitosis. There is absence of a typical jigsaw pattern, along with loss of hyaline sheath and peripheral pallisading [7]. The invasion of tumour cells into the surrounding tissues is frequently noted. These tumours are locally aggressive and can cause regional lymphadenopathy by spreading through lymph vessels. Stomach, thyroid, liver, lungs are the most common sites for distant metastasis, with few cases even reporting of intracranial spread [8]. As both benign and malignant forms of cylindroma express same markers like EMA, CEA, mucin like carcinoma associated antigen (B12), s-100, collagen IV, and CD 34 (QBEND/10), so immunohistochemistry cannot be used for differentiating benign cylindromas from the malignant ones [9,10].

The preferred mode of treatment for solid lesions is wide local excision along with lymph node dissection in isolated lymph node invasion cases without distant metastasis [10,11]. Usually it is followed by chemotherapy (cisplatin + 5 flurouracil) and radiotherapy to achieve complete cure and prevent further recurrence of the disease. Multiple cylindromas may require local excision with skin grafting in multiple settings. Early tumours without distant metastasis or lymph node extension have better prognosis following wide excision and radiotherapy as evident in our case. Batra M et al., have reported a case of rapidly progressive recurrent tumour over the scalp in a 59-year-old female which on histopathological examination showed malignant transformation of dermal cylindroma. Patient was treated with wide local excision and skin grafting and was normal on follow up [2].

CONCLUSION

Eccrine cylindromas are uncommon tumours of the head and neck region with rare malignant transformation. As malignant forms of cylindroma are highly aggressive with local invasion and distant metastasis a thorough knowledge about the tumour is necessary among all head and neck surgeons to detect it at an early stage, as early detection has a better prognosis. A wide local excision of the tumour with skin grafting if needed, along with metastatic lymph node dissection and chemo radiotherapy is the treatment of choice.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Feb 25, 2015**
Date of Peer Review: **May 09, 2015**
Date of Acceptance: **May 26, 2015**
Date of Publishing: **Jul 01, 2015**