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Case Report

Dentistry Section

Malignant Transformation of Solitary Cylindroma Involving the Frontonasal Region- A Rarity

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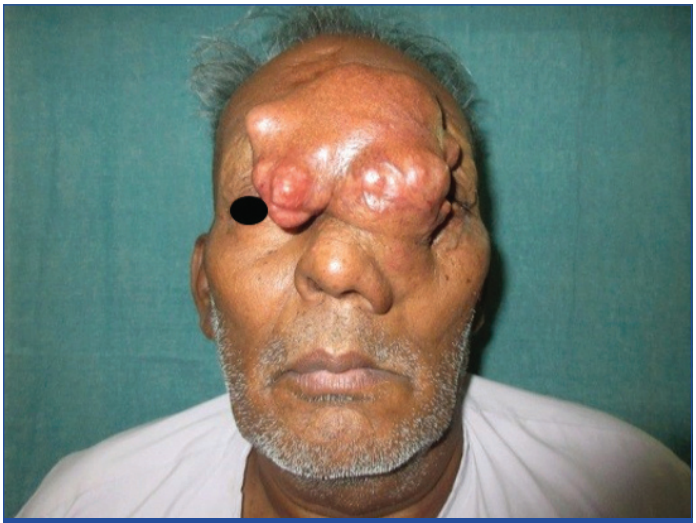
ABSTRACT

Cylindromas are adnexal tumours of either eccrine or apocrine origin. Two common variants- Solitary and Multiple are seen, of which the malignant transformation of a solitary lesion is rare. Solitary tumours occur sporadically in the head and neck and are without a familial history whereas the multiple tumours are inherited in an autosomal dominant pattern occurring in scalp and rarely on trunk. Malignant transformation is observed commonly in multiple tumours, although transformation of solitary lesions has also been reported. We report a rare case of malignant transformation of a solitary cylindroma involving the fronto-nasal region in a 72-year-old diabetic and hypertensive male reported to the Department of Oral and Maxillofacial Surgery, with a chief complaint of rapidly growing, painful growth over fronto-nasal region for 3-years and difficulty in breathing since 3-months. on examination; a single, large, well circumscribed, tender, firm and fixed multinodular growth with focal areas of ulcerations and reddish brown crustations of approx. 9.0×3.5 cm measuring in its greatest dimension was seen in naso-frontal region. A CECT Head showed a well defined, heterogeneously enhancing lobulated soft tissue mass, occupying the left nasal cavity, ethmoid sinus and extending into medial extraconal space of left orbit. The incisional biopsy report suggested low grade skin adnexal tumor of malignant cylindroma. Based on the finding in the biopsy, the CECT surgical excision of the lesion was planned. The patient was kept on follow-up of 3 years and no local recurrence or distant metastases was noted.

Keywords: Adnexal tumours, Canaliculi, Malignant cylindroma, Microcyst, Multinodular

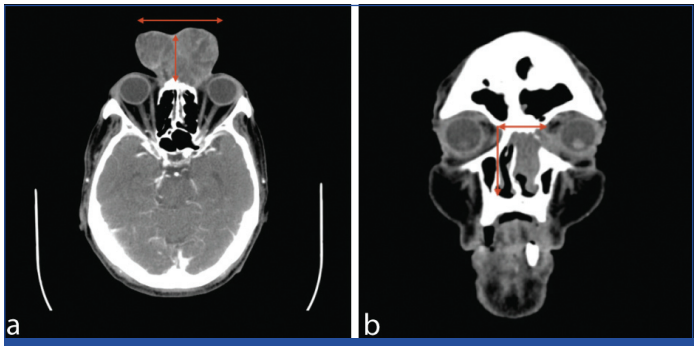
CASE REPORT

A 72-year-old diabetic and hypertensive male reported to the Department of Oral and Maxillofacial Surgery, with a chief complaint of rapidly growing, painful growth over fronto-nasal region for 3-years and difficulty in breathing since 3-months. He gave a history of noticing a small nodular growth which gradually increased to present size. There was history of difficulty in breathing and on examination; a single, large, well circumscribed, tender, firm and fixed multinodular growth with focal areas of ulcerations and reddish brown crustations of approx. 9.0×3.5 cm measuring in its greatest dimension was seen in naso-frontal region crossing the midline [Table/Fig-1]. Excision of a similar mass five years back was also reported. A differential diagnosis of Basal cell carcinoma, trichilemmoma, trichoepithelioma, basaloid follicular hamartoma, spiradenoma, cylindroma and neurofibromatosis was formulated and a biopsy for the lesion was planned following CECT Head.



[Table/Fig-1]: Preoperative frontal view, showing soft tissue mass.

A CECT Head was done, which showed a well defined, heterogeneously enhancing lobulated soft tissue mass, occupying the left nasal cavity, ethmoid sinus and extending into medial extraconal space of left orbit. The lesion had caused destruction of adjacent nasal bone, medial wall of left orbit and was abutting the medial rectus muscle of the left orbit. The brain parenchyma appeared normal [Table/Fig-2a,b].



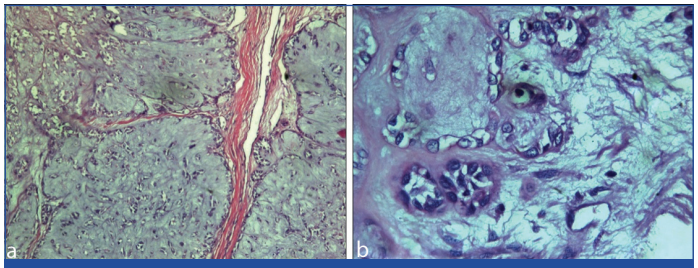
[Table/Fig-2]: a) CECT head- axial view- showing the extension of the lesion; b) CECT head- sagittal view- destruction of the underlying bone.

The incisional biopsy showed myxoid flakes and spindle nuclear cells in small fragments, entrapped within these flakes were basaloid, polygonal epithelial cells which showed hyperchromasia, mild pleomorphism and granular chromatin. At places the hyaline myxoid nature appeared in small cylindrical group surrounded by epithelial cells. The report suggested of low-grade skin adenexal tumour of malignant cylindorma" [Table/Fig-3a].

Based on the finding in the biopsy and the CECT surgical excision of the lesion was planned. Under general anaesthesia, wide local excision of lesion with a safety margin of 2cm was carried out and the defect was reconstructed using a scalp rotational flap based on the superficial temporal artery. The defect over the scalp was covered with split thickness skin graft. The excised specimen was submitted for histopathological examination.

The histological sections showed various sized cylindroid groups

abutting each other made of small to intermediate size cells; Basaloid, cuboidal to polygonal in shape, attempting to form glands, canaliculi and microcyst. Nuclei of the cells were aggressive in appearance. The tumour mass in all sections was in close proximation to the dermis of the skin [Table/Fig-3b].



[Table/Fig-3]: a) Pictomicrograph (H&E, 10X) view showing myxoid flakes and spindle nuclear cells in small fragments, entrapped within these flakes were basaloid, polygonal epithelial cells which showed hyperchromasia, mild pleomorphism and granular chromatin; b) Pictomicrograph (H&E, 40X) view showing irregular dermal cords and nests of highly atypical and pleomorphic cells, which sometimes show ductal differentiation and cyst formation, pleomorphic nuclei, increased abnormal mitoses, loss of hyaline sheaths and loss of peripheral palisading at the tumour island periphery.

The patient was kept on follow-up of 3 years and no local recurrence or distant metastases was noted [Table/Fig-4].

[Table/Fig-4]: Post-operative 3-year follow-up of patient.

DISCUSSION

Cylindroma was first described by Ancell H in 1842 and its malignant nature was described by Weidman in 1929 [1-3]. Cutaneous cylindromas are rare adnexal skin tumours of the head and neck originating from the eccrine or apocrine glands of the skin appendages [2,4]. Two clinical variants are commonly encountered: 1) Solitary lesion without a familial history; and 2) Multiple cutaneous lesions, which is inherited in an autosomal dominant pattern [4]. Multiple cylindroma along with trichoepitheliomas are pathognomic features of Brook speigler syndrome. Solitary tumours occur sporadically in the head and neck and are without a familial history whereas the multiple tumours are inherited in an autosomal dominant pattern occurring in scalp and rarely on trunk. Malignant transformation is observed commonly in multiple tumours, although transformation of solitary lesions has also been reported. Malignant Cylindromas (MC) are locally aggressive, infiltrative, have a tendency for recurrence and exhibits potential for metastasis and therefore demands long term follow-up [3]. Diagnosis can be difficult due to its rarity and varied differential diagnosis such as Basal cell carcinoma, trichilemmoma, trichoepithelioma, basaloid follicular hamartoma, spiradenoma and neurofibromatosis. These can be ruled out by the histopathological examination of the resected specimens.

Cylindromas present as pink to purple-blue nodular lesions of varying sizes commonly present on the forehead and scalp and less frequently on the head and face regions and extremities [2,5]. The lesions are frequently presented in the 5-7th decade of life and have a higher female predilection (F:M::2:1). When the lesions occur over vital anatomic structures like the eye or the nose, the function may be compromised. Histopathologically, the tumour exhibits irregular dermal cords and nests of highly atypical and pleomorphic cells, which sometimes show ductal differentiation and cyst formation, pleomorphic nuclei, increased abnormal mitoses, loss of hyaline sheaths and loss of peripheral palisading at the tumour island periphery. These are characterised by an infiltrative growth pattern, without connection to the epidermis. Residual benign cylindromatous foci, characterised by a dual cell population, organised in to basaloid islands, surrounded by a thick, hyaline basal membrane with a typical 'jigsaw' pattern are scattered though the tumour [6,7]. Immunohistochemically, Malignant cylindromas express markers indicating their dual origins from eccrine and apocrine glands. CMA 5.2, EMA and CEA are expressed in tubular structures, while positivity to S100 and GCDFP-15 is variable [6,8]. performed Immunohistochemistry in the present case, Vimentin and S-100 protein was found to be positive in proliferating cell component of the tumour and EMA was negative except in some areas of skin epidermis. The solitary variant occurs more frequently than the multiple variant and has a lower malignant transformation potential. Malignant transformation is characterised by presence of ulceration, the potential for rapid growth, colour change, presence of pain and fixation of the lesion. However, once the transformation has taken place, the tumour in its aggressive forms, invades the underlying tissues, causing local destruction and potentiates distant metastases. Local, infiltrative growth shows distant metastases with a rate of approximately 67% compared to smaller lesions [9,10]. Metastases is most commonly found in cervical lymph nodes, liver and spine, however lung, rib, stomach and thyroid may also be affected. The prevalence of malignant cylindroma is not well documented as only a few are reported in the literature. Malignant transformation is usually seen in multiple cylindromas but may rarely be seen in solitary form [11]. Among 36 cases described in the literature only 9 solitary lesions transforming to the malignant variant were reported [2]. The present case demonstrated the transformation of a solitary lesion into a malignant variant.

Author	Age/Sex	Size (cm)	Lesion characteristics	Treatment	Follow-up period (Months)
Lyon JB and Rouillard LM (1961) [13]	59/M	10×8	Left posterior scalp: Reddish, penetrating the dura mater. Necrotic and hemorrhagic.	Excision+Adjuvant RTx	12
Urbanski SJ et al., (1985) [14]	-	6.5	Scalp: initial lesion invaded to calvaria	Excision	30
Galadari E et al., (1987) [15]	66/M	2.5×2.5	Scalp: Nodular lesion with hemorrhagic crust growing over 6 yrs		
Lin PY et al., (1987) [16]		3	Scalp		24
Iyer PV and Leong AS (1989) [17]	60/M	a) 5×3×3 b) 3×3×3	Left side of the scalp: Fleshy and pedunculated	Excision and RND	

Hammond DC et al., (1990) [18]	69/F	3	Right post-auricular: Ulcerating, bulbous, firm.	Excision and RND	36
Lo JS et al., (1991) [19]	71/F	8.8×7×4	Left parieto-occipital scalp: Rapidly enlarging plaque on scalp, first noticed 5y ago	Mohs Surgery	84
Lotem M et al., (1992) [20]	81/F	1-1.5	Scalp: Multiple nodules, that were ulcerated, hemorrhagic, necrotic	Excision + grafting	12
Donner LR et al., (1995) [21]	81/F	3×1.2×1.1	Right mastoid: Growing over 2 yrs, increasingly painful; hard thickening of right mastoid skin	Excision, including mastoid and incus and stapes; neck dissection	36
Pizinger K and Michal M (2000) [22]	68/F	10	Neck: Red, firm, tender, poorly circumscribed. CT scan revealed other tumours in spinal column, kidney	No intervention	6
Mashkevich G et al., (2006) [23]	58/F	13×9	Left external auditory canal: Patient presented with aural fullness, otalgia, tinnitus	Excision with SSG	24
Akgul GG et al., (2013) [24]	52/M	20×10	Scalp: Hyperemic, firm. Needle aspiration of submandibular LN revealed metastasis	Excision of entire scalp, bilateral modified radical neck dissection. Adjuvant CTx and RTx	60
Abedi SM et al., (2014) [25]	63/M	1.8	Right neck: Painless nodule that increased in size over weeks. No ulceration, no necrosis, no hemorrhage	Excision	
Borik L et al., (2015) [3]	83		Scalp: Rapidly growing; poorly circumscribed, with infiltrative growth pattern at the base	Excision	12
Tripathy SM et al., (2015) [9]	60/F	4.2×3.6	Left post-aural: Painful, insidious onset and progressive over 3 m	Excision	
Pal S et al., (2016) [26]	52/F	3.5×3	Left parietal region	Excision	12
Roncevic R and Roncevic D (2016) [27]	65/M	0.5-6	Multiple over scalp	Excision	108
Portincasa A et al., (2018) [28]	18/F	8×5-20×10	Multiple over scalp, Frontal area, occipito-temporal and retro-auricular	Excision	72
Kim SM et al., (2019) [11]	60/M		Right parietal region	Excision and Primary closure	6

[Table/Fig-5]: Previously published cases of cylindroma in literature [3,9,11,13-28].
RTx: Radiotherapy; CTx: Chemotherapy; RND: Radical Neck Dissection; SSG: Split thickness skin graft

Treatment for solitary lesions entails the surgical excision with wide safety margins of 2 cm laterally and 1 cm basal margin keeping in account for the recurrence and malignant transformation. Other reported treatment modalities are simple excision, scalping, Mohs' microscopic surgery and laser ablation [2,5,12]. The defect can be reconstructed by local flaps or free grafts. Multiple lesions require a carefully planned, multi-step surgical approach. In the present case/patient case, we performed a wide local excision of the tumour with 2 cm safe margins and reconstruction by scalp rotational flaps. Recurrence following surgical excision is about 42% [4,11].

An electronic literature review was conducted on PubMed to identify case reports describing cylindroma using the search terms such as "malignant cylindroma," "cylindrocarcinoma", "head and neck cylindroma" and "transformation cylindroma". A summary of these cases is shown in [Table/Fig-5] [3,9,11,13-28].

The reported case is on follow-up for 3-years and no recurrence or metastasis was recorded.

CONCLUSION(S)

Malignant cylindromas are rare tumours seen in head and neck region. They are locally invasive and may show distant metastasis. Early diagnosis and treatment gives best results. Regular follow-up to monitor recurrence and distant metastasis is required.

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