

## RADIOLOGICAL AND PATHOLOGICAL EVALUATION OF TRICHILEMMAL CYSTS OF THE SCALP

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### ABSTRACT

#### BACKGROUND

Trichilemmal cysts or pilar tumours are slow growing scalp lesion commonly found in elderly women. These slow growing lesions may cause morbidity and even mortality. Recurrence of the lesions after local excision is common. These trichilemmal cyst usually indolent with benign nature or may transform to proliferating trichilemmal cyst or Proliferating Pilar Tumour (PPT) or may show malignant transformation.

The aim of the study is to study the radiological and pathological evaluation of trichilemmal cysts of scalp.

#### MATERIALS AND METHODS

A hospital-based cross-sectional retrospective study was conducted. The study group comprised of 20 patients presenting to the Departments of Radiodiagnosis, Radiotherapy, General Surgery, Plastic Surgery and Dermatology in a tertiary care hospital from July 2015 to August 2017. All patients were initially evaluated clinically followed by cross-sectional imaging modality like Computed Tomography (CT) or Magnetic Resonance Imaging (MRI).

#### RESULTS

In 20 patients, a total of 51 trichilemmal cysts were evaluated where 46 (90.2%) were benign trichilemmal cysts and 5 (9.8%) showed malignant transformation. The mean age of presentation was 47.9 yrs.  $\pm$  1.5 (SD) with male:female ratio of 1:3. The mean duration of presence of trichilemmal cyst was 5.9 yrs.  $\pm$  3.2 (SD). Out of 5 malignant trichilemmal cysts 3 patients (15%) showed bony calvarial erosion and 2 patients (10%) showed intracranial extensions. Statistical significance with 'p' value of 0.003 was noted between the size of largest dimension of trichilemmal cyst and their histopathology without any statistical significance between duration of swelling and their histopathology.

#### CONCLUSION

Even though, the trichilemmal cysts of scalp are denoted as benign lesion, as they usually shows recurrence and its affinity to become locally aggressive and turn into malignancy is there. Hence, clinical, radiological and pathological correlation is necessary to decreased morbidity or even mortality from these scalp lesions.

#### KEYWORDS

Pilar tumour, Proliferating, Magnetic Resonance Imaging (MRI), Histopathological.

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#### BACKGROUND

Trichilemmal Cyst (TC) or pilar tumour is derived from the outer root sheath of a hair follicle.<sup>1</sup> This trichilemmal cyst occurring in 5-10% of the population with female preponderance.<sup>2</sup> The trichilemmal cyst can be familial and may have autosomal dominant inheritance.<sup>3</sup>

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These trichilemmal cyst or proliferating trichilemmal cyst are well circumscribed dermal or subcutaneous neoplasm with trichilemmal type keratinisation of squamous epithelium.<sup>4</sup> The trichilemmal cyst can undergo transformation into proliferating trichilemmal cysts or Proliferating Pilar Tumour (PTT), which appears as larger lobulated scalp lesion.<sup>2</sup> The proliferating trichilemmal cyst are often benign and more prone for recurrence after incomplete excision. Rarely, the proliferating trichilemmal cyst showed malignant degeneration resulting in local tissue invasion and distal metastasis.<sup>2</sup> Usually, the trichilemmal cyst presents as asymptomatic or mildly painful, smooth, mobile, firm nodule, mainly over scalp, while the proliferating trichilemmal cysts are usually larger and multilobulated.<sup>5</sup> In malignant transformation of

proliferating trichilemmal cyst, the lesions may ulcerate and bleed on touch.

More than 90% of trichilemmal cyst occur in scalp and occasionally other hair bearing sites maybe affected like neck, trunk, limbs, back, nose, vulva or groin.<sup>6</sup>

Although, the trichilemmal cysts are considered as biologically benign, however, these cysts maybe locally aggressive and malignant degeneration can occur resulting in direct invasion to adjacent structures, regional lymphadenopathy and distant metastasis.<sup>7</sup>

The aim of the study was to radiological and pathological evaluation of trichilemmal cysts of scalp.

## MATERIALS AND METHODS

A hospital-based cross-sectional retrospective study was conducted after approval from the institutional ethics review committee. The study group comprised of 20 patients presenting to the Departments of Radiodiagnosis, Radiotherapy, General Surgery, Plastic Surgery and Dermatology in a tertiary care hospital from July 2015 to August 2017.

**Patient Selection-** We included both outpatients and inpatients of both sexes presenting with various scalp lesions in whom both cross sectional imaging and histopathological examinations were done. Patients without cross-sectional imaging study were excluded from the study. Informed consent was obtained from patient, parents/guardian before undergoing CT or MRI scan. CT scan was done in 17 patients and MRI were done in 3 patients.

**CT Protocols-** Siemens Somatom Spirit Dual Slice CT Scanner (Siemens Healthcare, Erlangen, Germany) was used in this study. Patients were scanned in supine position. Non-contrast CT scans of brain were obtained followed by contrast CT scan were obtained after IV injection of 1mL/kg body weight of iodinated contrast agents. Scanning parameters used were spiral mode with slice thickness of 5mm and collimation 5 x 2.5 mm, pitch: 1.4; kVp: 120; mAs: 80. Multiplanar reformatted images were obtained. Recon parameters included slice thickness of 3 mm and recon increment of 1.5 mm.

**MRI Protocols-** MRI scans were done in Siemens Avanto 1.5 Tesla B15 machine (Siemens Medical Systems, Erlangen, Germany). MRI of brain was obtained using axial T1WI, T2WI, FLAIR (fluid attenuated inversion recovery), SWI (susceptibility weighted images), DWI (diffusion weighted images) sequences using 5 mm slice thickness. Sagittal T1WI and coronal T2WI sequences were obtained with 4-5 mm slice thickness. Postcontrast fat suppressed T1W images in all three planes were obtained with 5 mm

slice thickness after IV administration of 0.1mmol/kg gadolinium.

Axial T2WI images were obtained with TE: 80-95, TR:3800-4000, FLAIR weighted images with TE:90-95, TR:9000 and TI:2500 and T1W images with TE:7-9 and TR: 450-500. Sagittal T1W images were obtained with TE: 7-9, TR: 450-500 and FOV of 230. Coronal T2W images were obtained with TE:80-95, TR:3800-4000 and FOV of 230.

**Evaluation-** Twenty patients of variable sizes scalp lesions were examined in this study for the appearance, shape, location, components, calcifications, nearby bony calvarial erosion or intracranial extension. Histopathological confirmation was done in all patients.

**Histopathological Evaluation-** The paraffin embedded Haematoxylin and Eosin stained histopathology slides were reviewed.

**Statistical Analysis-** Data were presented in terms of percentage, mean and standard deviation. Calculations were done using SPSS programs (Statistical Package for the Social Science version 16. SPSS Inc. Chicago, USA).

## RESULTS

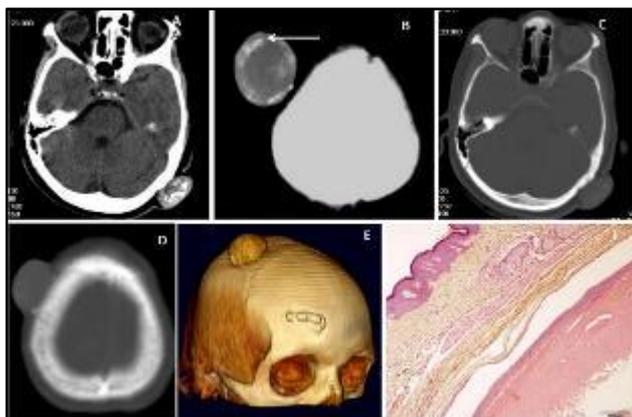
The mean age of patient with trichilemmal cyst was 47.9 yrs.  $\pm$  1.5(SD) with male:female ratio of 1:3. The mean size of the largest dimension of trichilemmal cyst was 3.6cm  $\pm$ 1.8 (SD). Out of 20 patients 17 patients (85%) presented with only scalp swelling (Figure 1, 2 and 3) and 3 patients (15%) were with scalp swellings, ulcerations and bleeds on touch (Figure 5 and 6). In 20 patients, total 51 trichilemmal cysts were detected out of which 46 (90.2%) were benign trichilemmal cysts (Figure 1-3) and 5(9.8%) showed malignant transformation (Figure 4-6). The mean duration of presence of trichilemmal cyst was 5.9 yrs.  $\pm$  3.2 (SD). CT scan were done in 17 patients (85%) and MRI scan were done in 3 patients (15%). Fifteen patients (75%) showed calcifications in the trichilemmal cysts (Figure 1-3). Out of 5 malignant trichilemmal cysts 3 patients (15%) showed bony calvarial erosion and 2 patients (10%) showed intracranial extensions. In all 20 patients, trichilemmal cysts were excised and in 3 patients' excision were done followed by radiotherapy. Recurrence of disease process was noted in 8 lesions (15.6%) after excision. The clinical, radiological and histological findings were summarised in Table 1.

Statistical significance with 'p' value of 0.003 was noted between the size of largest dimension of trichilemmal cyst and their histopathology. No statistical significance was noted between the duration of swelling and their histopathology with p value of 0.61.

Number of Patients	Age	Sex	Clinical Presentation	Duration of Swelling(yrs.)	Total Number of Lesions	Calcifications	Bony Calvarial Erosion	Size of Largest Dimension of Lesion(cm)	HPE Findings	Treatment	Follow up
1	60	F	1.0	13	8	+	-	5.0	Benign	Excision	5 months RD
2	45	M	1.0	6	3	-	-	1.4	Benign	Excision	3 months NR
3	22	M	1.0	2	1	+	-	5.1	Benign	Excision	6 months NR
4	10	F	1.0	3	1	-	-	2.7	Benign	Excision	2 months NR
5	55	F	1.0	6	4	+	-	2.1	Benign	Excision	5 months NR
6	57	F	1.0	4	1	+	-	2.0	Benign	Excision	3 months NR
7	40	M	1.0	7	1	+	-	3.6	Benign	Excision	9 months NR
8	37	F	1.0	4	1	+	-	1.6	Benign	Excision	11 months NR
9	59	F	1.0	7	1	+	-	2.3	Benign	Excision	15 months NR
10	38	F	1.0	3	1	+	-	2.7	Benign	Excision	10 months NR
11	63	F	1.0	15	5	+	-	3.4	Benign	Excision	3 months RD
12	35	F	1.0	4	3	+	-	1.4	Benign	Excision	4 months NR
13	68	F	1.0	5	2	-	-	6.3	Benign	Excision	6 months RD
14	46	M	1.0	3	1	+	-	2.1	Benign	Excision	2 months NR
15	58	M	1.0	4	1	+	-	2.8	Benign	Excision	6 months NR
16	36	F	2.0	7	5	-	+	7.8	Malignant	Excision and LND	3 months RD
17	60	F	1.0	5	2	+	+	3.9	Malignant	Excision and LND	8 months RD
18	55	F	1.0	7	2	-	+	4.6	Malignant	Excision	5 months RD
19	62	F	2.0	6	5	+	-	6.0	Malignant	Excision	7 months RD
20	52	F	2.0	8	3	+	-	5.8	Malignant	Excision and LND	2 months RD

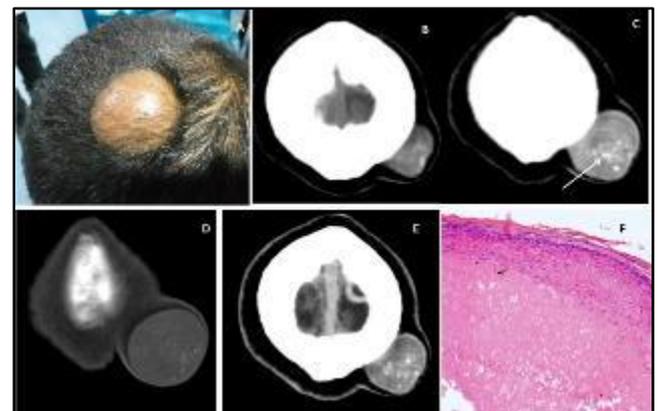
**Table 1. Summary of Trichilemmal Cysts of Scalp in 20 Patients**

LND-Lymph node dissection, NR- No recurrence, RD- Recurrence of disease.



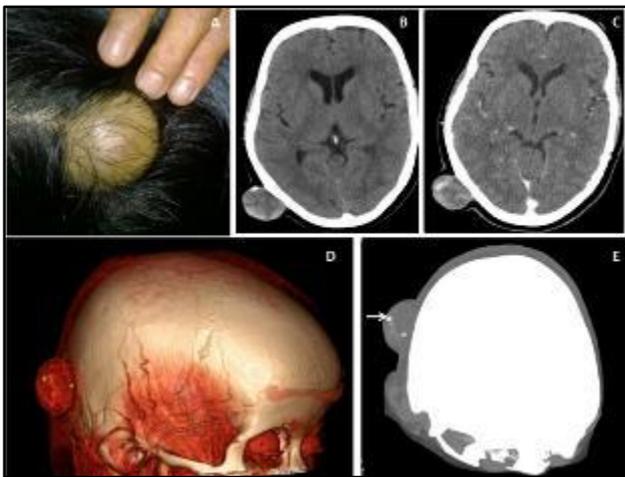
**Figure 1**

60 years old women presented with multiple asymptomatic scalp swellings from last 13 years. Non-contrast axial (A and B) and bone algorithm (C and D) CT images showed variable sizes oval to plaque like scalp lesions in left postauricular region and right parietal scalp with irregular chunky and curvilinear calcifications(white arrow in image B) within with intact underlying bony calvaria. Surface Shaded Display (SSD) image (E) showed the bump like lesion in right parietal scalp and plaque like lesion in right frontal scalp. HPE image (F) of trichilemmal cyst lined by stratified squamous epithelium showing trichilemmal type of keratinisation (H and E 4X).



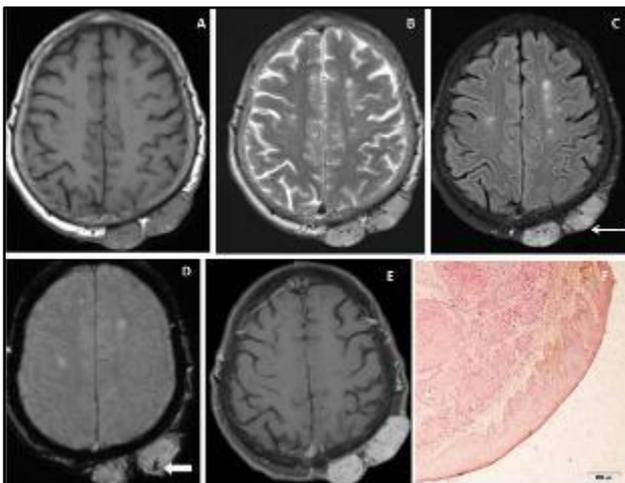
**Figure 2**

40 years male patient presented with solitary left parietal scalp swelling from last 7 years. Photograph of patient showed oval-shaped swelling in left parietal scalp (image A). Noncontrast axial (B and C) and bony algorithm (D) CT images showed left parietal mixed density lesion with chunky calcifications (white arrow) with intact underlying bony calvaria. Axial contrast CT (E) image showed heterogenous enhancement of the lesion. HPE image (F) of proliferating trichilemmal cyst lined by benign squamous cells showing abrupt keratinisation (H and E 10X).



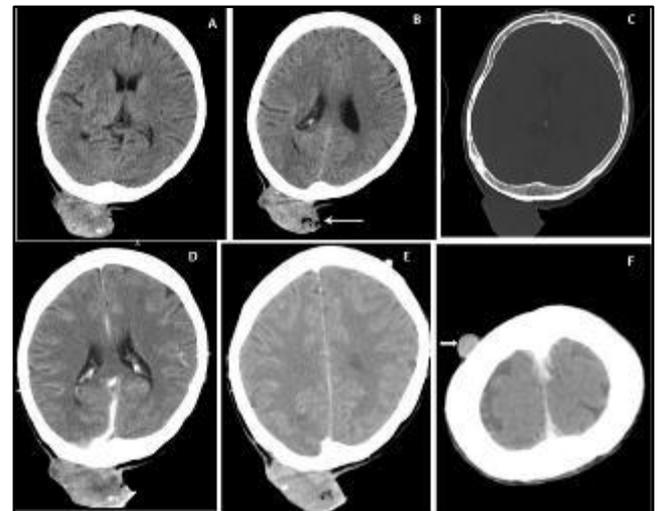
**Figure 3**

38 years female presented with right occipital scalp swelling for last 3 years. Photograph of patient showed oval-shaped swelling in right occipital scalp (image A). Non-contrast and contrast axial (B and C) CT images showed right occipital scalp mixed density lesion with fine nodular calcifications, which showed mild heterogenous enhancement with smaller nonenhancing cystic areas. Surface Shaded Display (SSD) image (D) showed the bump in right occipital scalp. Reconstructed maximum intensity projection (MIP) image (E) showed nodular bright calcifications within the scalp swelling (white arrow).



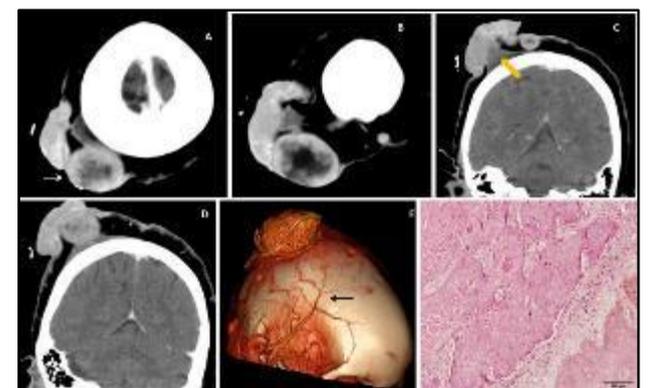
**Figure 4**

Axial T1W, T2W and FLAIR images (A-C) showed T1W isointense and T2W is to slight hyperintense lobulated lesions in left parietal scalp (white arrow). Axial Susceptibility Weighted Image (SWI) showed nodular blooming (white block arrow) within the scalp lesion representing calcifications (D). Axial post gadolinium image (E) showed moderate homogenous enhancement of the nodular scalp lesions. HPE images (F) of malignant trichilemmal cyst showing lobules of atypical squamous cells invading the surrounding tissue (H and E 4X).



**Figure 5**

62 years women presented with multiple scalp swellings with ulcerated right parietal scalp swelling. Non-contrast axial CT images (A and B) showed heterogenous appearing right occipital scalp lesion with surface ulcerations and air foci within (white arrow). Bone algorithm image (C) showed intact underlying bony calvaria. Axial post contrast (D, E and F) showed heterogenous enhancement of the scalp lesion with eccentric necrosis. Another smaller nodular enhancing right parietal scalp lesion also noted (white block arrow).



**Figure 6**

52 years female presented with a larger right parietal scalp for last 8 years with recent ulcerations and overlying raw areas, which bleed on touch. Post-contrast CT axial (A and B) and coronal (C and D) images showed moderate heterogeneously enhancing larger multilobulated right parietal scalp swelling (white arrow) with less enhancing centrally located stem (yellow block arrow). Surface Shaded Display (SSD) image (E) showed the bump like lesion in right parietal scalp with multiple engorged meningeal arteries supplying the lesion (black arrow). HPE image (F) showed malignant transformation of proliferative trichilemmal cyst with lobules of atypical squamous cells invading the surrounding tissue (H and E 10X).

## DISCUSSION

The trichilemmal cyst or proliferating trichilemmal cysts usually present with solitary scalp lesion, however, multiple scalp sites can occur at times.<sup>8</sup> Both trichilemmal cyst and proliferating trichilemmal cyst shows trichilemmal type of keratinisation and sometime can occur together. The proliferating trichilemmal cyst may arise from preexisting trichilemmal cyst either due to repeated trauma or inflammation. Coexistence of benign and malignant areas within the same scalp lesion representing that PTT arises from a preexisting trichilemmal cyst.<sup>9</sup>

Takata et al suggest loss of p53 tumour suppressor gene responsible for transformation of PTT to malignant PTT.<sup>10</sup>

The common differential diagnosis of trichilemmal cysts of scalp includes epidermal inclusion cyst, dermoid cyst, pilomatrixoma, acne keloidalis nuchae, lipoma, sebaceous cyst, cylindroma, etc. Uncommon differential diagnosis of trichilemmal cyst includes haemangioma, meningocele, atretic encephalocele, slow flow vascular malformation of scalp, sinus pericranii, hibernoma and liposarcoma. The common differential diagnosis of proliferating trichilemmal cyst or malignant trichilemmal cyst include squamous cell carcinoma, plexiform neurofibroma and uncommonly metastasis, basal cell carcinoma, lymphoma and melanoma.<sup>11,12</sup>

Kim H J et al<sup>7</sup> reported that trichilemmal cyst or proliferating trichilemmal cyst can appear either as cystic or solid mass on cross-sectional imaging. On CT scan solitary lesion appears as focal bump like elevation of scalp with intact underlying bony calvaria. Variable pattern of intralesional calcifications like nodular, plaque like chunky and linear calcifications can be encountered. On Magnetic Resonance Imaging (MRI) the trichilemmal cyst showed homogenous isointense to slight hypointense signal intensities on T1W images and homogenous to heterogenous hyperintense signal intensities on T2W images.<sup>13</sup> Susceptibility Weighted Images (SWI) showed the blooming within the lesion with bright signal intensities on phase images representing intralesional calcifications. Proliferating Pilar Tumour (PTT) showed heterogenous signal intensities with multiple locations and larger sizes of lesion. PTT on post gadolinium images showed heterogenous enhancement of the lesion with variable areas of enhancing solid or nonenhancing cystic components.<sup>13</sup> However, the malignant transformation of PTT shows heterogenous T2W signal intensities with more central or eccentric T2W hyperintensities because of intralesional necrosis. It also exhibit poorly defined margins, nearby tissue plane infiltrations, bony calvarial erosion or even intracranial extensions.<sup>7</sup> Regional necrotising lymphadenopathy usually exhibit metastatic nodal disease.

Rapid enlargement of a longstanding scalp lesion with surface ulceration and histopathological evidence of abnormal mitosis, marked cellular pleomorphism, irregular infiltrative margins suggest malignant transformation of trichilemmal cyst or proliferating trichilemmal cyst.<sup>14,15,16</sup>

In high-resolution USG, the trichilemmal cyst appear as dermal and subcutaneous tissue rounded or oval shaped lesion. It is usually anechoic and may shows internal echoes or debris from keratin. Nodular, chunky, curvilinear or plaque like calcifications can be detected. With use of Micropure technique intralesional calcifications are well detectable. The proliferating trichilemmal cyst or malignant trichilemmal cyst shows hypoechoic to heteroechoic appearance of scalp lesion on USG without appreciable cystic component. Nearby local infiltrations, intratumoural vascularities and underlying bony calvarial erosions can be detectable.<sup>17,18,19</sup>

High resolution ultrasonography with colour Doppler also helps to differentiate from lipoma, haemangioma, slow flow vascular malformation, sinus pericranii, meningocele or even encephalocele from trichilemmal cyst. But, USG has less sensitivity in identifying the actual local invasiveness or bony calvarial erosion associated with malignant trichilemmal cyst.

In our study, sample of 20 patients with 51 trichilemmal cyst or PTT, 5 lesions (9.8%) showed malignant changes.

Histopathologically, the trichilemmal cyst reveals a well circumscribed cyst with trichilemmal type of keratinised layer with intervening granular layer.<sup>11,20</sup> The epithelial cells of the trichilemmal cyst proliferates from periphery to centre of the cyst.<sup>11</sup>

The Proliferating Pilar Tumour (PTT) reveals bands of proliferating epithelial cells that may surround cystic areas or are interconnected and separated by fibrous stroma.<sup>11</sup> Sometime malignant trichilemmal cyst in histopathologically difficult to differentiate from squamous cell carcinoma.<sup>21</sup> Malignant proliferating trichilemmal cyst has tendency to recur and metastasise more frequently than squamous cell carcinoma.<sup>22</sup> Hence, histological identification of trichilemmal type of keratinisation is of utmost important.

Trichilemmal cyst are often easily enucleated,<sup>12</sup> while proliferating trichilemmal cysts requires wide local excision to prevent recurrence.<sup>2</sup> In malignant transformation of proliferating trichilemmal cyst, more aggressive treatment should be considered with wide local tumoural excision, lymph nodal dissection, radiotherapy or chemotherapy.<sup>23,24,25,26</sup>

In our study, sample we encountered recurrence of disease process after local excision in 8 lesions (15.6%).

Literatures reported recurrences of trichilemmal cysts or PTT after diagnosis and treatment ranged from within 6 months to more than 10 years.<sup>23,27</sup> The proliferating trichilemmal cysts may exhibit local aggressiveness with local tissue plane invasion, bony calvarial erosion and even intracranial extensions, which may create considerable morbidity and or even mortality.<sup>24,25,28</sup>

More than 30 cases of malignant transformation of proliferative trichilemmal cysts have been reported in literature including more than 12 cases of metastatic diseases.<sup>7,11,29,30</sup> In our study, sample of 20 patients with 51 lesions 5 lesions (9.8%) shows malignant

transformation and associated lymph nodal metastasis in 3 patients.

In literature, the metastatic diseases associated with proliferating trichilemmal cyst have been reported as early as at initial presentation or as late as 10 years.<sup>11,26,31</sup>

### CONCLUSION

The discrete trichilemmal cyst, larger multilobulated Proliferating Trichilemmal Cyst (PTT) and more irregular indistinct margin heterogenous appearing invasive scalp lesion of Malignant Proliferating Trichilemmal Cyst (MPTT) having various clinical course and behaviour. The benign trichilemmal cyst and proliferating trichilemmal cystic lesions whose cross sectional imaging findings and even histological findings may not correlate with their clinical behaviour. Hence, close clinical follow up and careful histological examination is necessary to exclude focal malignant transformations in clinically benign looking trichilemmal cysts or proliferating trichilemmal cysts. Even close follow up is necessary after excision to look for disease recurrence.

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