A RARE CASE REPORT OF MALIGNANT PILAR TUMOUR

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Abstract:
Pilar tumor is a rare neoplasm arising from the external root sheath of the hair follicle and is most commonly observed on the scalp, these tumors are largely benign often cystic and are characterized by trichilemmal keratinisation, wide local excision has been the standard treatment of choice. Recent reports have described a rare malignant variant with an aggressive clinical course and a propensity for nodal and distant metastasis which therefore merits aggressive treatment. In this report we present a malignant pilar tumor of the scalp with multiple nodal metastasis at presentation, diagnostic and therapeutic consideration in the form of adjuvant radiotherapy are subsequently discussed. The aim of this report is to create awareness regarding this particular entity, considering its rare presentation and the need to differentiate these tumors from squamous cell carcinoma of the head and neck region.

Keyword: Proliferating pilar tumor, Trichilemmal tumor, hair Follicle tumor, scalp tumor

INTRODUCTION: Malignant Proliferating pilar tumour [MPTT] is a rare neoplasm arising from the isthmus region of the outer root sheath of the hair follicle. It is also commonly called as proliferating trichilemmal tumour [PTT]; its incidence is 0.1% of skin biopsies. There is no standardised recommendation for adjuvant treatment like chemotherapy or radiotherapy after surgery or management of surgically unresectable cases. We present a rare case of this tumour which was benign initially and later progressed to a malignant proliferating pilar variant over a period of time with multiple lymph nodes.

CASE REPORT
55 years old female patient presented with history of swelling in the right temporo-occipital region of scalp – for 30 years duration, which was about 1.5*1.5 cms, asymptomatic, static in size till about 3 years back, Swelling gradually increased in size to about 4*4 cms and was associated with pricking pain over scalp region. Surgery in the form of local excision was carried out on 28/10/2009 Histopathology report suggested a proliferative trichilemmal tumour, patient was asymptomatic for about 2 months after surgery,
again developed a swelling about 4*4 cms over the scalp in parieto-occipital region and diagnosed as local recurrence CT brain revealed a soft tissue density lesion on right parieto occipital region with no evidence of bone erosion

Patient underwent a wide local excision with flap cover on 30/1/2010. Histopathology report suggested a proliferative trichilemmal tumour. Patient was on regular follow up and asymptomatic for about 14 months. Patient later developed multiple swelling in the neck region

1. Behind the angle of mandible hard in consistency fixed and ulcerated non tender about 2 x 2cms

2. Second swelling about 7 x 7cms, nodular surface firm to hard in consistency fixed to underlying structure, ulcerated and matted Biopsy from the neck nodes revealed a malignant proliferative trichilemmal Tumour

**TREATMENT GIVEN**

Keratitis, ichthyosis and deafness syndrome [KID] (1), Birt–Hogg–Dubé syndrome [BHD] and Cowden Syndromes were ruled out as trichilemmal tumour are associated with these syndromes (2), Surgery was not possible because of fixity to underlying structures. Palliative Radiotherapy to a dose of 48Gy was given using a AP/PA portals with Co-60 Theratron phoenix machine At the end of palliative radiotherapy of 48 Gy, patient’s pain and nodal swelling had partially regressed. Only partial response could be achieved and salvage surgery of the residual disease could not be done due to extensive skin involvement and fixity to underlying structures.

**Fig-1 CT of Brain showing no intracranial extension only soft tissue involvement**

Patient is on regular follow up till now with no further progression of the disease.
CHARACTERISTICS OF TRICHELLEMMAL TUMOUR

Previous reports of trichilemmal tumour have been in frequent, comprising only about 0.1% of skin biopsies and the malignant variant even rare. Pilar tumour is a rare neoplasm arising from the hair follicle, the most common location is the scalp (90%), the other reported sites include back, chest, axilla, groin, gluteal region, and thigh, vulva, face and eye lids. The size may range from 1 to 10 cms although lesions as large as 25 cms have been reported, inflammation ulceration bleeding and or yellowish discharge may occur. A spectrum of transformation is hypothesised which begins with a benign pilar cyst proceeding to a proliferating pilar tumour and then to a malignant proliferating pilar tumour, the stimulus for the changes in these lesions is currently unknown although trauma inflammation irritation may play a role. This patient had a benign lesion for almost 30 years and then transformed into malignancy as was documented from the serial Histopathological reports which shows a gradual transformation into malignancy. This tumour lacks a distinctive histological or immunohistochemical marker to suggest a malignant transformation, increased proliferation index and DNA aneuploidy in tumour cells is an expression of a pre malignant event. It mimics squamous cell carcinoma and its biological behaviour is unpredictable. Mann et al. reported metastasis from pilar tumours and hence considered it to be a genuine neoplasm. However, metastasis from pilar tumours is rare. it has been reported after treatment of the primary. Proliferating pilar tumours have been classified into three groups; benign, locally aggressive, and malignant.

Differential diagnosis include
1. Cylindroma.
2. Dermoid cyst.
3. Squamous cell carcinoma.
5. Keratoacanthoma.

INVESTIGATION:
The best modality to determine bony invasion or erosion is CT scanning and proliferating pilar tumours are frequently found as incidental subcutaneous nodules on brain CT scan. However, for deeper tissue invasion, MRI is best which most frequently display isointensity on T1weighted images and heterogeneous signal on T2weighted images. Screening for PPT is suggested in patients with 1) keratitis-ichthyosis-deafness [KID] (8) (9) KID syndrome is a rare congenital disorder characterized by keratitis, ichthyosis, and deafness and increased follicular cancer. Surveillance in patients with KID syndrome should include screening for pilar tumors and their early removal to avoid development of malignant proliferating pilar tumors with poor prognosis. 2) Birt-Hogg-Dubé syndrome is a rare disorder that affects the skin and lungs and increases the risk of certain types of tumours.

MANAGEMENT:
Aneuploidy is common in malignant proliferative trichilemmal tumour, particularly in tumours with a high proliferative fraction, loss of CD34.
immunoreactivity is an additional feature of malignant transformation. Though limited, its values may be helpful in the diagnosis of malignant proliferative trichilemmal tumour surgery in the form of wide local excision with a 1 cm margin has been followed (11), considering the aggressive nature of the malignant variant and high rates of adjuvant loco regional as well as distant failures in previous series. Adjuvant radiotherapy is justified, although the role of radiation therapy and chemotherapy especially in the malignant variant is not established. Some centres have tried with adjuvant chemotherapy (VAC) (12) and radiotherapy to prevent local relapse although no standardised recommendation is available as of now due to non availability of randomised trials and paucity of cases.

PROGNOSIS
The prognosis is excellent with complete excision. Local recurrence and metastasis are extremely rare in benign cases, however in proven malignant cases; metastasis has been known to occur in 30% cases. (13)

ROLE OF RADIOThERAPY IN MALIG Views of comment section NANT TRICHILEMMAL TUMOUR Unlike head and neck squamous cell carcinomas which are loco regional, pilar tumours are primarily local only. Electrons to the primary and photons to the nodes is preferred in appropriate anatomical structures e.g. primary in scalp with nodes in neck. Considering the aggressive nature of the malignant variant and high rates of loco regional and distant failures in previous series adjuvant radiotherapy is justified.

CONCLUSION Since malignant proliferative trichilemmal tumour follows an aggressive course, it is essential to distinguish it from similar looking neoplasms for an appropriate therapy, with consideration for adjuvant treatment.

Bibliography


