

# Treatment Outcome Evaluation of Multiple Familial Trichoepithelioma Cases Treated with Electrosurgery and Curettage: Review of Four Cases

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**Abstract:** ***Background:** Multiple familial trichoepithelioma (MFT) is benign tumor arising from hair follicle, characterized by skin-colored papules usually located on the forehead, nose, cheek and chin. The management of MFT is challenging due to the recurrence and complication. This case series hence intended to evaluate the treatment outcome of four MFT cases treated with electrosurgery and curettage in Dermatology and Venereology Department of Dr. M. Djamil Hospital. **Case report:** Four cases of MFT in 20-year-old male patient, 13-year-old girl patient, 9-year-old girl patient and 16-year-old girl patient were reported. All patient developed smooth, shiny and skin colored-papules on the forehead, nose, cheek and chin. Lesions were distributed predominantly in the central face. Positive family history of similar presentation were presented by all patients. Histopathological examination revealed the diagnostic criteria of MFT. Patients were diagnosed as MFT and treated with electrosurgery and curettage. Treatment outcome evaluation described that first and fourth patient felt satisfied after treatment due to significant reduction of the lesion although new lesions still persist. While, second and third patient felt less satisfied after the treatment, because there were recurrence and new lesions. Only first patient had atrophic surgical scarring and hyperpigmentation patches as complication. **Discussion:** Electrosurgery and curettage had possibility of partial removal resulting the persistence or recurrence lesion and the appearance of complications such as scar. Due to the complication of non-pharmacological procedure like electrosurgery and curettage, pharmacotherapy has been also investigated for MFT treatment.*

**Keywords:** electrosurgery and curettage, treatment outcome, recurrence, scarring.

## 1. Introduction

Trichoepithelioma is a benign tumor of the pilosebaceous unit that originates from the hair follicles. Trichoepithelioma can be divided into 3 subgroups including multiple familial trichoepithelioma (MFT), solitary non-hereditary trichoepithelioma and desmoplastic trichoepithelioma.<sup>1</sup>

The MFT is an uncommon autosomal dominant disease skin tumor, associated with mutations on chromosome 9p21 or in cylindromatosis tumor suppressor gen (CYLD) located on chromosome 16q12-q13. The multiple familial trichoepithelioma (MFT) is characterized by asymptomatic, multiple skin-colored papules and nodules located around the nasolabial area, nose, forehead, upper lip and occasionally the scalp, neck and upper trunk. The multiple familial trichoepithelioma (MFT) can undergo the transformation into malignant neoplasms, such as trichoblastic carcinoma or basal cell carcinoma (BCC).<sup>1,2</sup>

The MFT involve the face in 80% of cases, leading to significant psychological morbidity due to cosmetic appearance. Surgical excision is considered as the effective treatment, but it impractical where multiple lesions are evident. Other treatment modalities have been reported, such as electrosurgery, cryosurgery, dermabrasion, erbium:Yag and carbon dioxide laser, with variable result. Plausible complication of treatment include partial removal resulting persistence or recurrence and scarring.<sup>1,2</sup>

Herein we evaluate the treatment outcome of four cases of MFT treated with electrosurgery and curettage in

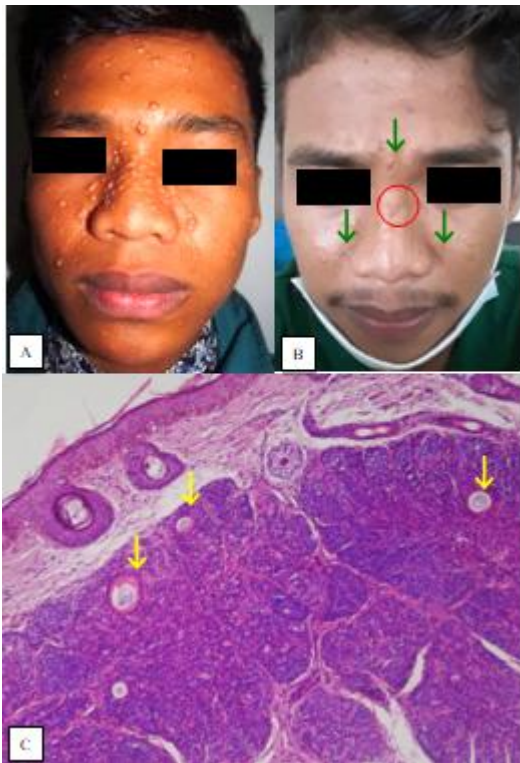
Dermatology and Venereologi Department of Dr. M. Djamil Hospital Padang.

## 2. Case Report

### Case 1

A 20-year-old male patient was admitted to the outpatient clinic with a 10-years history of multiple skin colored, firm, asymptomatic papules in the forehead, nose, both of cheek and chin. The lesion had started as only a few papules on his nose during childhood. The papules gradually increased in number and appeared in the forehead, both of cheek and chin with various size. There was no lesion found in the other part of the body. There was no history of seizure or other systemic abnormalities. His little brother had the similar lesion in the face.

General state was in normal limit. Dermatologic examination showed multiple, smooth, shiny and flesh-colored papules in the forehead, nose, nasolabial fold, both of cheek and chin. The center of some lesions was slightly depressed and umbilicated. The lesions were distributed predominantly in the central face. Skin biopsy was performed and histopathological examination revealed tumoral formation consisting of mainly basaloid cells that made peripheral palisading of nuclei with horn cyst. Patient was diagnosed as MFT and treated with electrosurgery and curettage.



**Figure 1:** (A) Before electrosurgery and curettage procedure, there were multiple flesh-colored papules on the forehead, nose, both of cheek and chin. B. Two years after the treatment, there were surgical atrophic scar (green arrow) and hyperpigmented patches (red circle). C. Histopathological examination, a tumoral formation consisting of mainly basaloid cells that made peripheral palisading of nuclei with horn cyst (yellow arrow). H&E, 40x.

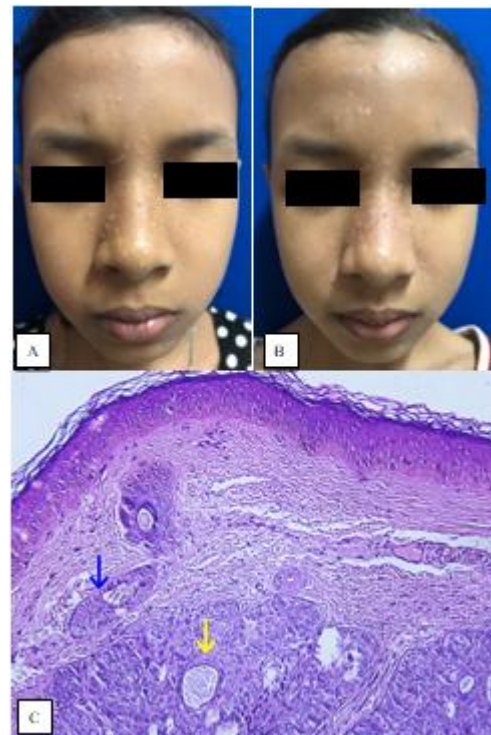
About 2 years later, there were no recurrence lesion, but there were some new lesion in forehead, nose and both of cheek with a smaller size. Atrophic surgical scarring and hyperpigmentation patches was found as complication after the treatment.

### Case 2

A 13-year-old girl patient was admitted to outpatient clinic with multiple skin colored, smooth, shiny and asymptomatic papules in the forehead, nose, both of cheek and chin, increased in number since 3 years prior to visiting the hospital. The parent noticed that the first lesions (approximately 5 skin-colored papules) appeared on the nose. Over the time, the lesions increased in number and appeared on the forehead, cheeks and chin. The skin lesions were a source of great embarrassment on the part of the patient, limited her social interactions and affected her school performance. There was no lesion in the other part of the body. Patient had no history of systemic disease and seizure. Grandmother, mother and aunty had the same lesions.

General examination was in normal limit. Dermatologic examination revealed multiple, smooth, shiny, skin-colored papules on the forehead, nose, nasolabial fold, both of cheek and chin. Most of the papules were distributed in the central face. Histopathological examination showed multiple tumoral formation consisting of mainly basaloid cells that

made peripheral palisading of nuclei with horn cyst. Patient was diagnosed as MFT and treated with electrosurgery and curettage.



**Figure 2:** (A) Before electrosurgery and curettage procedure, there were multiple flesh-colored papules on the forehead, nose, both of cheek and chin. B. One month after the treatment, some of skin-colored papules reappeared in the same location. C. Histopathological examination, multiple basaloid cells with peripheral palisading (blue arrow) and keratin-filled cyst horn cyst (yellow arrow). H&E, 40x.

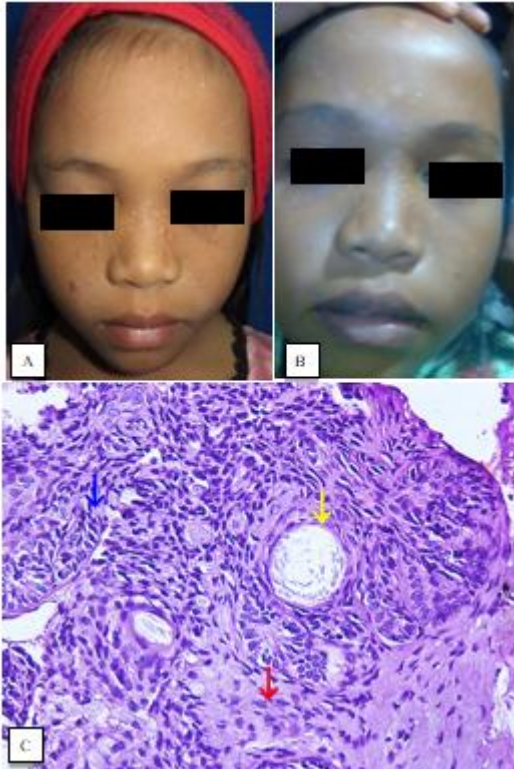
The surgical wound improved after 4 weeks, but there were several papules reappeared in the same location. The parent felt less satisfied with the treatment result.

### Case 3

A 9-year-old girl patient was admitted to outpatient clinic with multiple skin colored, firm, smooth, shiny, asymptomatic papules in the forehead, nose, both of cheek and chin, increased in number since 4 years prior to visiting the hospital. The parent said that the first lesions (approximately 5 skin-colored papules) appeared on the nose. The papules gradually increased in number and appeared on the forehead, cheeks and chin. Sometime patient peeled off the papules and left the erosion. There was no lesion in the other part of the body. Patient had no history of any systemic disease and seizure. Her aunty had the same lesions like patient.

The general examination was in normal limit. Dermatology examination revealed multiple smooth skin-colored, shiny papules on the forehead, nose, nasolabial fold, both of cheek and chin. Most of papules were distributed in the central face. There erosion on the right cheek due to the peeling of papule by her self. Histopathological examination showed multiple tumoral formation consisting of mainly basaloid cells that made peripheral palisading of nuclei with horn cyst and papillary mesenchymal bodies.

Patient was diagnosed as MFT and treated with electrosurgery and curettage. After one year, there were recurrence and new lesions. The parent did not report about scarring and hypo or hyperpigmentation due to the treatment. The parent felt less satisfied for result of the treatment.



**Figure 3:** **A.** Before electrosurgery and curettage procedure, there were multiple flesh-colored papules on the forehead, nose, both of cheek and chin. **B.** One year after treatment, some of skin-colored papules reappeared in the same location and there were new lesion in the other side. **C.** Histopathological examination, multiple basaloid cells with peripheral palisading (blue arrow), keratin-filled cyst horn cyst (yellow arrow) and papillary mesenchymal bodies (red arrow). H&E, 100x

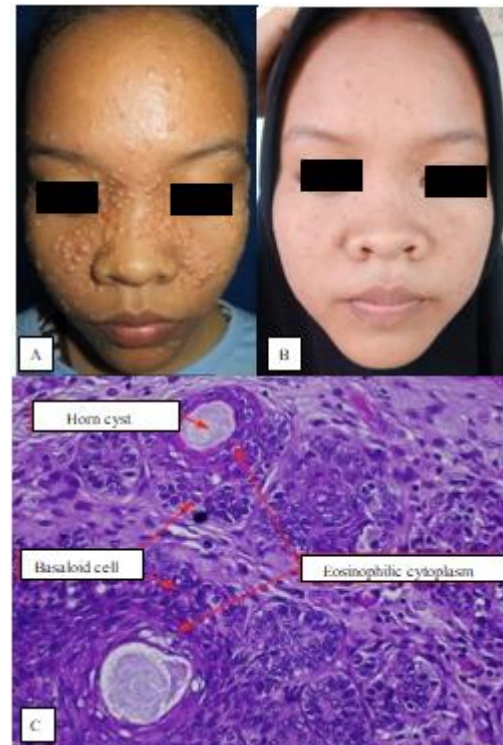
#### Case 4

A 16-year-old girl patient was admitted to the outpatient clinic with multiple skin-colored, firm, asymptomatic papules and nodules in the forehead, nose, both of cheek and chin, increased in number within the last 10 years. The first lesion (4 skin colored papules) appeared around the nose. Over the time, the papules increased in number and affected almost the entire of face. Patient reported that there was no papule in the other part of body. Although the papules were asymptomatic, they were cosmetically unacceptable to the patient. There was no associated systemic disease. Her uncle and aunty had the same lesions like the patient.

Approximately 6 months prior to visiting the hospital, patient came to the Dermatologist in the local hospital. Electrosurgery procedure had been done to the patient. But several weeks after the procedure, the same lesion reappeared in the same location. Then patient got topical medication applied once a day at night for 1 month. But there was no improvement, the lesion was still in the same size.

There was no abnormality in the general state. Dermatology state revealed multiple smooth skin-colored, shiny papules and nodules on the forehead, nose, nasolabial fold, both of cheek and chin. The papules are distributed predominantly in the central face. Histopathological examination showed basaloid cells with eosinophilic cytoplasm around the horn cyst.

Patient was diagnosed as MFT and treated with electrosurgery and curettage. One year later, there was a significant reduction of the lesion and recurrence and new lesions still persist. Patient did not report about scarring and hypo or hyperpigmentation. The patient felt satisfied after the treatment.



**Figure 3:** Before electrosurgery and curettage procedure, there were multiple flesh-colored papules and nodules on the forehead, nose, both of cheek and chin. **B.** One year after the treatment, some of skin-colored papules reappeared in the same location and there were new lesion in the other side but in smaller size and number. **C.** Histopathological examination, there were multiple basaloid cells with peripheral palisading and keratin-filled cyst horn cyst. (H&E, 100x)

### 3. Discussion

Multiple familial trichoepithelioma is an autosomal dominant inherited genodermatosis, associated with mutations on chromosome 9p21 or in cylindromatosis tumor suppressor gene (CYLD), located on chromosome 16q12-q13. The disease is characterized by the progressive development of trichoepitheliomas and affected more than one family member. Patients with MFT typically begin developing tumors during childhood or adolescence. The tumors present as skin-colored papules or nodules on the face, located preferentially on the nasolabial, upper lips and cheeks, but may also occur on the neck, scalp, or trunk.

They may grow larger and increase in number over time. It can be disfiguring enough to cause depression and contribute to other psychological problems leading to losing self-esteem and confidence. Most of the patients seek treatment for cosmetic reasons.<sup>1</sup>

This case series reported four cases of MFT that had the same onset and clinical manifestation. The patients presented the skin-colored, smooth and shiny papules and nodules in the forehead, nose, both of cheek and chin with various size. The lesions were distributed predominantly in the central face. Papules were not presented in the other site of body, such as scalp, neck and trunk. The patients noticed that the first lesions appeared in childhood and over the time gradually increased in number. There was no history of associated systemic disease in all of patient. This clinical manifestation are typical for diagnosis of MFT.

Multiple familial trichoepithelioma affected more than one family member. All of this cases had positive family history of similar presentation. Genetic studies can be used to detect the abnormalities in band 9p21 or in the CYLD gene. Due to the limitation of facility, the genetic studies can not be performed for all cases.

Defenitive diagnosis of MFT was established based on histopathological examination. Histologically, MFT are dermal tumors with focal continuity with the overlying epidermis. They are composed of islands of uniform basaloid cells, sometimes showing peripheral palisading. Moreover, there are usually branching nests of basaloid cells. Small keratinous cysts lined by stratified squamous epithelium are quite common. Histopathologic features of trichoepithelioma also include the presence of papillary mesenchimal bodies.<sup>3</sup> Excisional biopsy and histopathological examination had been performed for all patients. All patients had histopathological features of trichoepithelioma and there was no sign of malignancy.

All patients in this case series had the similar medical history, physical examination, dermoscopy and histopathological examination. Multiple familial trichoepithelioma diagnostic criteria were met for all patients. Therefore, all patient were diagnosed as MFT.

Most of MFT patient seek the tretament for cosmetic reason. The are various of modalities treatment for MFT, but there is no ideal treatment to get a good result. Surgical excision is considered as the effective treatment, but it is not feasible for the multiple lesions. Other treatment modalities have been reported, such as electrosurgery, cryosurgery, dermabrasion, erbium:Yag and carbon dioxide laser, with

variable result. Plausible complication of treatment include partial removal (may result in persistence or recurrence) and scarring.<sup>1,2</sup>

All of the patients were treated with electrosurgery and curettage. Electrosurgery is used to cause deeper tissue destruction and hemostasis with minimal carbonization. Curettage was indicated to removing the tumor by scraping it away with a curette. The deep destruction provided by electrocoagulation results in scarring. The possibility of scarring, however, should be noted when discussing therapeutic alternatives with the patient.<sup>4</sup>Shaffelburg M et al (Canada, 1998) reported the successful treatment of two patients with electrosurgery. In comparison to carbon dioxide laser, this modality yields identical cosmetic results yet is less expensive and considerably more time efficient.<sup>5</sup>Sulistiyowati SA et al. (Indonesia, 2013) reported a case of MFT treated with combination of electrosurgery and dermabrasion. These modalities are effective, less bleeding, shorter time and relatively inexpensive.<sup>6</sup>

Treatment outcome evaluation described that all patient had new lesion in the different location. The second, third and fourth patient had several recurrent lesions after electrosurgery and curettage procedure, but in smaller size. Only the first patient who did not has recurrent lesions. The possible causative of recurrent lesions is partial removal of the tumor resulting persistence or recurrence lesions. The prevention of partial removal had been done for all patient by performed the procedure until reached the end point, characterized by the capillary bleeding.

Complication of electrosurgery and curettage was only found in the first patient. The first patient developed atrophic post-operative scarring. Atrophic post-operative scarring occurring after surgical procedures is a common cosmetic problem for patients. Atrophic scars, which present as topographical depressions, result when dermal collagen and connective tissue production during the physiologic wound-healing process inadequately compensate for the tissue loss present after injury. Wound tension, individual variations in wound healing and scar contraction are all factors that contribute to the creation of a depressed, atrophic scar. With varying success, numerous ablative, nonablative, and fractional devices have been used to stimulate neocollagenesis and dermal remodeling in an attempt to improve the appearance of atrophic scars.<sup>7,8</sup>

Due to the complication of non-pharmacological procedure, like electrosurgery and curettage, pharmacotherapy has been also investigated for MFT treatment.

**Table 1:** The various pharmacotherapy treatment for MFT from several literature

Author	Year	Treatment modalities	Dose, route and duration of administration	Efficacy
Urquhart et al <sup>1</sup>	2005	Imiquimod cream in combination with tretinoin gel 1%	3 times a week and then, twice daily topically (imiquimod) along with once a day topically (tretinoin) for 3 years.	About 80% improvement
Fisher et al <sup>1</sup>	2006	Aspirin in combination with adalimumab	325 mg twice a day orally (aspirin) along with 40 mg every other week for the first 2 months and thereafter, 40 mg every week as subcutaneous injection (adalimumab) for 8 months 5	Remarkable degree of improvement
Alessiet al <sup>1</sup>	2009	Imiquimod cream 5% Dose,	5 to 7 times per week topically for 32 weeks	Partial clinical response
Tu JH et al <sup>9</sup>	2017	1% sirolimus cream	Twice a day for one month after CO <sub>2</sub> laser	Prevent rapid recurrence, no

				need for additional CO <sub>2</sub> laser ablation.
Baur V et al <sup>10</sup>	2018	Vismodegib	150 mg/daily, peroral	Significant decrease of lesion in size and number
Dreyfus et al <sup>11</sup>	2019	Topical rapamycin 1%	Twice daily for 7-12 months	Interruption the prorsion of the lesions
Shimizu <sup>12</sup>	2020	0,2% sirolimus cream	Twice a day for 3 month	Nodule dramatically decrease in child

#### 4. Conclusion

The management of MFT is challenging due to the recurrence and complication. Electrosurgery and curettage had possibility of partial removal resulting the persistence or recurrence lesion and the appearance of complications such as scar. Due to the complication of non-pharmacological procedure like electrosurgery and curettage, pharmacotherapy has been also investigated for MFT treatment.

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#### Author Profile



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