

Case Report

RARE DIAGNOSIS OF A PROLIFERATING PILAR TUMOUR IN A FACIAL HAIRLINE CYST

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ABSTRACT

Sebaceous cyst, also known as an epidermoid cyst, is a subepidermal nodule filled with keratin and it is a benign capsulated tumor. It is often located on the scalp region, face, neck, and trunk; but can be found elsewhere such as the scrotum, genitalia, fingers, and buccal mucosa. Proliferating Pilar Tumors (PPT) are rare tumors. It is derived from the external root sheath of the hair follicle. These tumors are like irregular subcutaneous nodules and often appear on the scalp. This case report was about a 59 years old woman who came to the hospital following excision of a frontal lump elsewhere, with a sebaceous cyst as the initial diagnosis. From the histopathologic examination, grossly there was a whitish and greyish lump with a soft outer surface. Microscopically, there were malignancy signs with areas with keratinization. The tumor formed a solid pattern of enlarged cells with moderate to marked nuclear pleomorphism with vesicular nuclei, prominent nucleoli, and abundant pale eosinophilic to clear cytoplasm. There was also much free keratinous debris noted and numerous foci of calcification identified within the tumor. Mitotic figures with abnormal forms were frequently seen. The final diagnosis after the histopathological examination was Proliferating Pilar Tumour with focal malignancies. In conclusion, facial hairline tumor differentially diagnosed as a sebaceous cyst turned out to be a rare Proliferating Pilar Tumor (PPT). Following histopathological confirmation, the patient was referred for further management by a specialist team.

Keywords: Sebaceous cyst; proliferating pilar tumor; rare tumor; disease

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Hii j ni j tu

1. Sebaceous cyst or epidermoid cyst is a benign capsulated tumour on the scalp region, face, neck, and trunk that subepidermal nodule filled with keratin.
2. Proliferating Pilar Tumours (PPT) are rare tumours was found in hair follicle.
3. Sebaceous cyst diagnose on facial hairline tumour turned out to be a rare Proliferating Pilar Tumour (PPT).

INTRODUCTION

Sebaceous cyst or epidermoid cyst is a benign cyst derived from infundibulum epithelium from the hair follicle. This cyst can grow in every part of the body, grow slowly, and remain present for years. To diagnose this cyst, it needs a histopathological examination. To treat these patients, doctors may do observations, complete excision or incision, drainage of infected cyst, and steroid injection (Naftali et al. 2018). This cyst can happen at any age, from birth until 72 years of age. However, it often happens between 15 years and 35 years (Puranik et al. 2016). Malignancy transformation is so rare, but there is a possibility of it.

The malignancies of this cyst can be squamous cell carcinoma, basal cell carcinoma, and merkel cell carcinoma (Wollina et al. 2018).



Figure 1. Malignancies of *Sebaceous cyst*

Proliferating Pilar Tumour (PPT) is a very rare tumour, and there are less than 100 cases reported. This tumour can be classified as benign, low- and high-grade malignant tumour because of their different significant biologic activities (Gulati et al. 2011). Because of the rarity of these cases, there are no guidelines available for the management of these tumours. The standard treatment has been still wide local excision (Siddha et al. 2007). Here, this case was initially diagnosed by *sebaceous cyst*, but after the first excision and biopsy, the histopathological examination showed that it was a proliferating pilar tumour with focal malignancies. This will be treated again with re-excision of residual tissue in the area that was previously operated.

CASE REPORT

Female patient, 59 years old, attended Universiti Kebangsaan Malaysia Hospital referral by other hospital. A small lump was found 1.5 cm in diameter in frontal region of the facial hairline. Clinical examination showed a fixed lesion, well-defined and soft on palpation. No cervical lymph nodes palpable (group I – V and facial/ parotid/ occipital all negative). The patient admitted that there was no pain felt in the lump as well. The patient denied having suffered any trauma or having undergone any surgical procedure in that region. She said that the lesion began to develop like 20 years ago but was slow in progression and just decided to go to an expert recently, because she thought that it might be dangerous and this had no symptoms either. The patient had bronchial asthma in her medical history. She had undergone an excision biopsy for the lump in the previous hospital. After the excision, there was a scar in her frontal region 2 x 0.5 cm in size, 8 cm above the right brow, and not palpable.

The histopathology examination received grossly a piece of whitish grayish lump weighing 1.0 g measuring 17 x 16 x 9 mm. The outer surface was smooth. Cut section showed a homogenous whitish yellowish cut surface. Sections microscopically showed a well-circumscribed mass surrounded by thin fibrous capsule composed of islands and lobular proliferation of squamous epithelial cells in areas showing microcystic formation exhibiting glassy eosinophilic cytomorphology. Areas with keratinization were noted. The basement membrane appeared hyalinized and thickened. In areas, the tumor formed a solid pattern having enlarged cells with moderate to marked nuclear pleomorphism with vesicular nuclei, prominent nucleoli and abundant pale eosinophilic to clear cytoplasm. There were also many free *keratinous debris* noted. Numerous *foci* of calcification were identified within the tumor. Mitotic figures were frequently seen; showing abnormal form.

No necrosis present. It was diagnosed as a frontal *sebaceous cyst* at first, but after histopathology examination, it was diagnosed as a proliferating pilar tumor with focal malignant features. Because of this histopathology examination, the patient was referred to Universiti Kebangsaan Malaysia Medical Centre for the next treatment.

When operated in another hospital before coming to Universiti Kebangsaan Malaysia Medical Centre, there was no marginal resection when operated before, because it was possibly just diagnosed as a benign *sebaceous cyst*, so that the next treatment was an additional surgery, and it was a re-excision of the residual tissue of the area that has been previously operated to prevent the presence of the residue of cancer cells in that area, because the patient had been diagnosed by malignant tumor.

DISCUSSION

Epidermoid Cyst often occurs in the head and neck region, for example the scalp (34%) and neck (18%) often occurred in the areas (Nicollas et al. 2000). This benign cyst grows very slowly and usually occurs without any symptoms, but inflammation may happen and cause pain (Nicollas et al. 2000, Ohta et al. 2012, Weedon & Strutton 2010). This cyst was so rare to become a malignancy, but there were some cases of malignancies, such as Basal Cell Carcinoma, Bowen disease, Squamous Cell Carcinoma, and Mycosis Fungoides (Debaize et al. 2002). This cyst can happen in any age and often in the third and fourth decade of life (Sunil et al. 2014).

There was an English literature that showed a case of malignancy in 1968, when McDonald, in his analysis of 637 epidermal and sebaceous cysts, found malignancy only in seven cases (1.1%). There were just eight cases of basal cell carcinomas and only one was a squamous cell carcinoma. There was no metastasis. The treatment was local excision and it was satisfactory (Arianayagam & Javalakshmi 1987).

Proliferating Pilar Tumor (PPT) was first described by Wilson-Jones in 1966 named as "proliferating epidermoid cyst". After that, there were so many names to name this disease. There were proliferating trichilemmal cyst, hydatidiform keratinous cyst, giant hair matrix tumor, invasive hair matrix tumor, and trich chlamydia carcinoma. It showed that there were many interpretations about the biological activity of this tumor (Javid et al. 2020).

Proliferating Pilar Tumor (PPT) often occurs in elderly woman patients. There were 76 cases that divided PPT into three groups based on the degree of stromal invasion and the level of cytological atypia—benign, low- and high-grade malignant. A study indicated that there were 20 cases of low-grade malignant tumor and occurred in age 64.1 with men to female ratio of 1:2. The tumor location often occurred in the head and neck in 80% cases. It also often happens in the scalp (Network 2004). Most of the cases had benign lesions before it was operated (Gulati et al. 2011). Proliferating Pilar Tumors usually attack patients with excess hair than patients with bald scalp. These pilar tumors grew from the increasing of epithelial proliferation within pilar or *sebaceous cyst*. Patients often gave a history that they have a long remaining cyst in the same area (Siddha et al. 2007).

This reported case was diagnosed as a sebaceous cyst after physical examination in another hospital before referral. After the excision biopsy, histopathological examination was done and showed focal malignancies features found in this case like areas with keratinization, enlarged cells with moderate to marked nuclear pleomorphism with vesicular nuclei, prominent nucleoli and abundant pale eosinophilic to clear cytoplasm. There were also many free *keratinous debris* noted. Numerous *foci* of calcification were identified within the tumor. Mitotic figures were frequently seen; they showed abnormal form. This leads to malignancy of proliferating pilar tumor that is very rare to be found.

In another case report based on Budrukkar et al. 2007, there was a histological examination result that was almost the same. There was a keratinized nodule. Nucleus showed hyperchromatic and pleomorphic with the present of multinucleated giant cells. There were mitotic figures that showed a malignancy of this tumor. There was also a distinct area, resembling a typical benign pilar tumor. It was composed of interlacing nodules of small peripheral cells that were matured into large central cells with central keratinization, so that it was diagnosed as a proliferating pilar tumor in the histological examination.

Sebaceous cyst grew from *infundibulum follicular* because of the plugging of keratin. Proliferating pilar tumor is a very rare tumor that appear from the external root of hair follicle. Both of them are derived from hair follicle with different etiologies, so that misdiagnosis might have happened, because from the appearance itself, this case looked like a *sebaceous cyst* without histopathological examination, because it is often diagnosed by just a physical examination.

This patient was referred from another hospital to Universiti Kebangsaan Malaysia Medical Centre because of malignancies found in histopathological examination. When operated in another hospital before coming to Universiti Kebangsaan Malaysia Medical Centre, there was no marginal resection or surgical margin, because it was just probably diagnosed as a benign *sebaceous cyst*. Surgical resection margin status is important to malignant lesions surgery.

Surgical resection margin is an area around the infected tissue that will take together with the tumor itself. This area can possibly have healthy cells or cancer cells, so that the surgeons must take the infected tumor cells together with the area around it called the surgical margin. In some hospitals, doctors want 2 millimeters (mm) or more of normal tissue around the cancer and the outer edge of the removed tissue, but it may vary in different hospitals to be a clear margin. Tumour free margins are the success key to the goal of treatment of tumor (Breastcancer.org, 2019).

A margin was called positive if an invasive tumor was cut by the surgical blade, but if the tumor was close and not transected, it can be considered as a negative for tumor by National Surgical Adjuvant Breast and Bowel Project (NSABP) B-06 study. A positive margin shows that there was a presence of tumor (Emmadi & Wiley 2012). In this case, because there was no margin resection in this patient, there was a re-excision of the residual tissue in the area that was previously operated in the previous hospital to prevent any residual cancer cells infecting the area.

After the patients underwent the surgery, there was a not-so-big wound after surgery, and there must be a dressing after surgery. The dressings should make a good environment for healing. The dressing should be painless and helps patients return to normal function (Imran et al. 2018). After that, a scar was formed in that area, because there was a wound healing after the surgery had been done. Wound healing is a natural process in human body to restore injured tissue. There are 4 phases of wound healing, namely hemostasis, inflammation, proliferation, and remodeling of the wound (Imran et al. 2011).

The patient's scar was a result of remodeling. Patients should take care of their wound after surgery even after the skin is healed, because these postoperative patients need to visit the doctors to focus on prevention and treatment of the scars. Before undergoing an operation, the patient needs to be aware of excessive scars formation. Sometimes, it can become a hypertrophic

scar and if the hypertrophic scar does not improve after six months, it will become a keloid. Keloid needs a variety of methods to treat. Keloid needs combination therapies to reduce it, because there are some of those that are effective to reduce it, and there are some are not effective. It is the same as for treating wounds, a combination treatment may be needed to heal the wound (Chik et al. 2016, Imran et al. 2011, Son & Harijan 2014).

This patient had a very rare malignant tumour, although the cyst had already been taken, but there will still be a re-excision for the residual tissue, because it turned out to be a malignancy and there was no margin resection. For malignancy, the patient must be thinking about it seriously and should be allowed to decide what happens to their bodies, so that the patient can recover soon (Muhammed et al. 2017). Pathologists in previous hospitals should need a certain diagnosis confirmation, because this case was a very rare case. The pathologists should coordinate with other hospitals to confirm this rare diagnosis by referring the patient to Universiti Kebangsaan Malaysia Medical Centre.

With this very rare case, it is hoped that pathologists can work together and communicate well to resolve this case. In addition, cooperation between surgeons and pathologists is needed for properly diagnosing this case, so that successful treatment can be achieved.

Strength and limitation

The case report provides a detailed description of a rare type of tumor, Proliferating Pilar Tumor (PPT), which can help increase awareness of this condition among healthcare professionals. The report highlights the importance of accurate diagnosis and histopathological examination in cases where a benign lesion such as a sebaceous cyst may be suspected but turns out to be malignant. The report can provide useful information for future case studies and research. The report does not provide information about the patient's medical history or potential risk factors for developing PPT, which may impact the prognosis and management of the condition. The report does not provide information about the patient's outcome or response to further management by a specialist team, which may be useful for future studies and patient care.

CONCLUSION

A 59 years old woman who was first diagnosed as epidermoid cyst after surgery and biopsy were done, the histopathological examination showed that there was proliferative pilar tumour with focal malignancy features. There will be future management by Universiti Kebangsaan Malaysia Medical Centre. This patient would have a re-excision of residual tissue in

the area that was previously operated, because there was no surgical margin before. It will prevent patients from having residual cancer cells in that area.

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Conflict of interest

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Author contribution

All authors contributed conceptual, study design, and analysis data. FHI was write the manuscript and final validation to r udlksh.

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