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Multiple Pigmented Follicular Cyst: A Rare Case Report

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Abstract: ***Background**: Pigmented follicular cyst is a rare form of epidermal cyst that usually appears as an asymptomatic form of a single pigmented papule or nodule. Histopathologically shows the differentiation of pigmented hair shaft in an epidermal cyst. Although benign, pigmented follicular cyst can affect the quality of life of a person if the appearance of multiple and symptomatic. This is the twice case in a 5-year period (2015-2019) in Dr. RSUP. M. Djamil Padang. This disease clinically resembles multiple steatocystoma, so histopathological examination is needed to confirm. **Case report**: A 26 years old female,complained of appearing bluish and yellowish lumps that sometimes felt painful on her chest, stomach, trunk, both of leg, both of arm, which increase in number and size since ± 5 years. These lumps secreted white-yellowish mass if rupture. Working diagnosis of this patient was suspect steatocystoma with differential diagnosis with multiple follicular cyst, eruptive xanthoma. We diagnosing with multiple pigmented follicular cyst after histopathological examination showed a cyst on dermis with thin epithelial wall and lumen containing laminated keratin and pigmented hair shafts. There was no presence of sebaceous gland on PAS stain. **Discussion**: Clinically, it is difficult to distinguish multiple steatocystoma from multiple epidermal cysts. Histopathology of this patients is suitable for multiple pigmented follicular cyst, which is a rare variant of multiple epidermal cyst.*

Keywords: multiple pigmented follicular cyst, epidermal cyst, steatocystoma

1. Introduction

Epidermal cysts known by the other names as epidermoid cysts, infundibular type follicular cysts, keratin cysts, epidermal inclusion cysts or epithelial cysts. That cyst was limited by the epithelium and contains keratin. This cyst does not involve the sebaceous glands and does not contain sebum. Pigmented follicular cyst, a rare variant of epidermal cysts, was first described by Mehregan and Medenica in 1982 which mainly occurs in adult men and usually appears as a solitary pigmented papulonodule and asymptomatic in head or neck. They described seven patients with solitary pigmented papules or cysts that histopathologically described the presence of mid-dermal cysts containing the remnants of pigmented hair shaft. Ribera et al. in 1990 reported the first two cases of multiple pigmented follicular cysts, which they termed multiple pigmented terminal hair cyst.[1,2]

The pathogenesis of this variant epidermal cyst is not known with certainty, but there are several different theories about the origin of these embryonic lesions. It is can arise from ectopic skin tissue due to tissue dislocation to adjacent areas. Or is the end result of a form of teratoma monolayer derived from germ cells.[3]

Clinically, epidermal cysts are dermal or subcutaneous nodules that are mobile in the presence of a punctum in the central region. Non-traumatic lesions are usually located in the upper chest, upper back, neck and head. Traumatic lesions generally occur on both palms, both soles of the feet and both buttocks. The presence of punctum can represent a pilosebaceous unit which can produce unpleasant odors. These lesions can be skin-colored, yellowish or white. Cysts usually grow slowly and are asymptomatic, although rupture is common. Pigmented follicular cysts have a single pigmented dome-shaped or intracutaneous papulonodule with multiple pop-ups which can be seen in some patients. Pigmented follicular cysts usually occur in the age range of 20-63 years. Single pigmented follicular cysts can occur in the head or neck region, while multiple can be found in the abdomen chest and other body parts. [1,2,4]

Histopathologically, the pigmented follicular cyst is located in the mid-dermal, consists of flattened epithelium with epidermoid keratinization, and has a small opening that resembles the epidermis above. The growth of hair follicles can sometimes be found connected to the cyst wall. Characteristically, multiple pigmented hair shafts and keratin layers can be found in cyst cavities. Although pigmented follicular cysts tend to be solitary and asymptomatic, significant morbidity can occur if pigmented follicular cysts appear multiple and are symptomatic. [1,2,4]

Pigmented follicular cyst must be distinguished from other skin lesions that contain remnants of hair shaft on the inside of cysts such as dermoid cysts, steatocystomas and eruptive vellus hair cysts. Dermoid cysts are cysts located in the subcutaneous area that can usually occur from birth and are present in the head and neck region. Whereas steatocystomas and eruptive vellus hair cysts often appear as multiple lesions. All three types of cysts have the same clinical appearance, age of onset, location and hereditary pattern, but histopathologically can be distinguished.[5]

Steatocystomas occur as single (steatocystoma simplex) or multiple (steatocystoma multiplex) lesions. They tend to be a few millimeters to a centimeter in diameter and appear as cysts in the dermis that drain oily fluid if punctured. Steatocystomas are most numerous on the chest and in the axillae and groin. There are unusual facial and acral variants as well as a rare congenital linear form. Steatocystomas persist indefinitely, and they are usually asymptomatic except for cosmetic concerns. Steatocystoma multiplex is inherited as an autosomal dominant condition, and is due to mutations in the keratin 17 gene. It may occur in association with eruptive vellus hair cysts and pachyonychia congenita type 2, also caused by keratin 17 (as well as K6b) defects.Biopsy specimens of steatocystoma showed a dermal cyst lined by a thin stratified squamous epithelium, with a granular layer surmounted by a thin, irregular, corrugated eosinophilic cuticle. Small sebaceous lobules are found in or immediately adjacent to the cyst wall.[6]

Histopatology of cutaneous cysts have an epithelial lining that may be composed of stratified squamous epithelium or other forms of epithelia. Some 'cysts' have no epithelial lining at all. Approach to a cyst with stratified squamous epithelium can seen on figure.[6]

2. Case Report

We reported a 26-year-old female patient with chief complaintbluish and yellowish lumps that sometimefelt painful on the chest, stomach, trunk, both of leg, both of arm, which increase in number and size since ± 5 years ago.

Initially ± 5 years ago a bluish lump as large as corn kernels appeared on chest. The lump get bigger and secrete a white yellowish mass when rupture and leave a scar. The patient has never treated this complaint. ± 3 years ago, a bluish lump increase a number and size on stomach, trunk, both of leg, both of arm. ± 1 month ago, the bluish lump increase in number with variant size, some lumps enlarged, reddened, felt painful until they finally broke and left a blackish brown mark, patient feels disturbed with this condition. Patient was consulted with dermatologist and had mometason cream for redness lump, the complaints are reduced but the lump is still present.There was no history of hipercholesterolemia, diabetes mellitus and hypertension.

General physical examination revealed on normal limit. Dermatological state bluish, yellowish, redish papules, bluish and yellowishnoduls with varying sizes with smallest size 0,1x0,1x0.05cm, larger size 1.5x1x0,2cmon chest, stomach, trunk, both of leg, both of arm.

Histopathologic examination **(**Hematoxylin Eosin Stain) revealed hyperkeratosis, partial epidermis revealed focal atrophy on epidermis and there were mild to moderate lymphocyte and PMN cells, a cyst appears with a thin stratified squamous epithelium wall, the lumen contains a mass of keratin (laminated keratin), pigmented hair shafts and degeneration of keratin with rod-shaped organisms resembling hypae / bacterial on dermis.

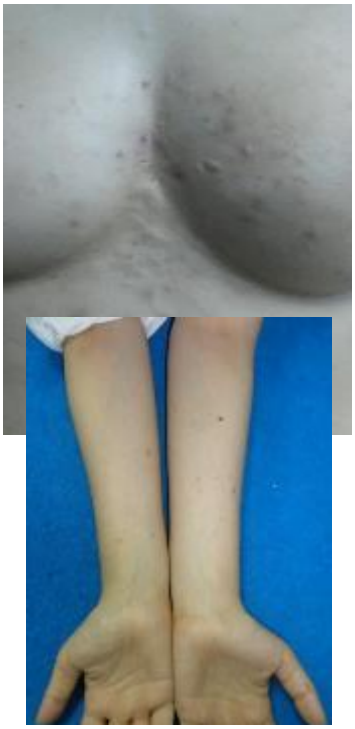


Figure 1: Bluish and yellowish papules, bluish and yellowish noduls with varying sizes a. on chest b. both of arm

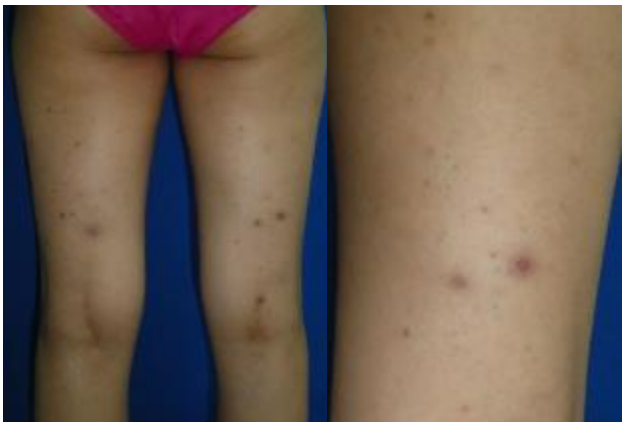


Figure 2: Bluish and redish papules, bluish and yellowish noduls with varying sizes on tight

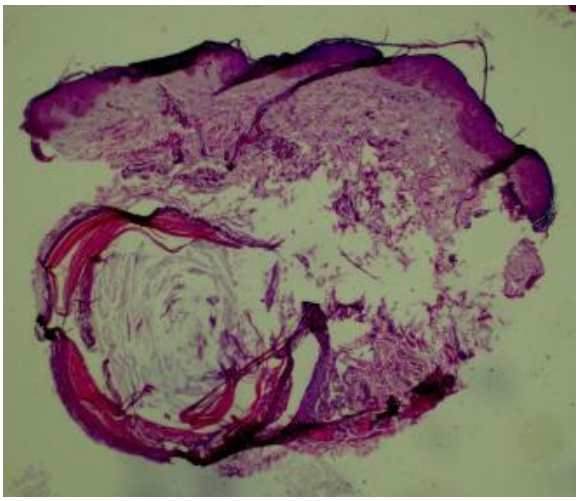


Figure 3: Skin tissue with a partially thinned epidermis, on the dermis with mild to moderate lymphocyte and PMN cells and a cyst (objective 4x)

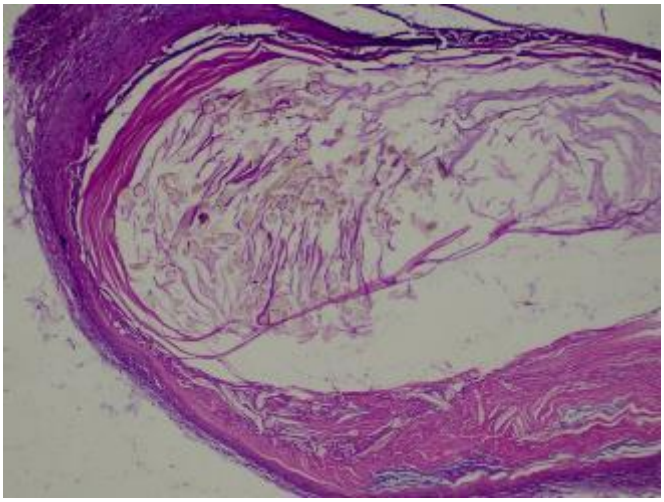


Figure 4: A cyst with stratified squamous epithelium wall and containing mass of keratin and pigmented hair shaft (objective 10x)

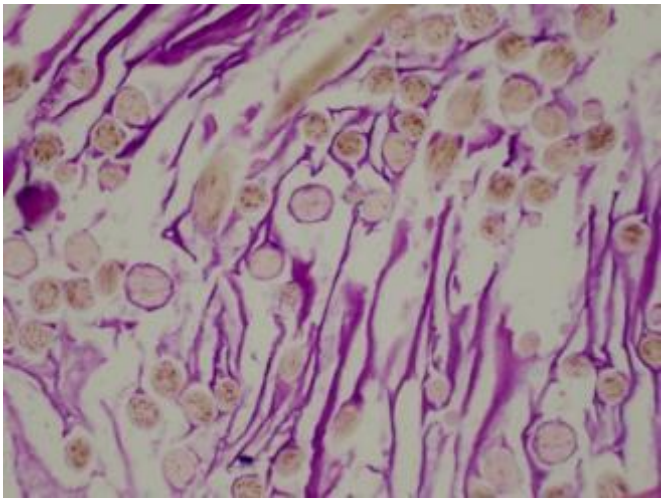


Figure 5: The structure of pigmented hair shaft fragment, mostly brown (melanin) (objective 100x)

3. Discussion

Multiple pigmented follicular cyst is one of the rare variants of multiple epidermal cyst which is characterized by bluish, yellowish papulo-nodules that can be accompanied / without clinical symptoms. In this patient from history and physical examination it was found that bluish yellowish-colored bumps appeared on the chest, stomach, trunk, both of leg, both of arm since ± 5 years ago, the patient had also tried to break the lump and a yellowish white mass came out. From the physical examination obtained: bluish-yellowish papules, bluish-yellowishnoduls with varying sizes. It was diagnosed with suspect steatocystomawith differential diagnosis multiple epidermal cyst. Then the biopsy excision on right leg of the cyst is performed on the patient and histopathological examination of the tissue and the results obtained in the form of multiple pigmented follicular cyts.

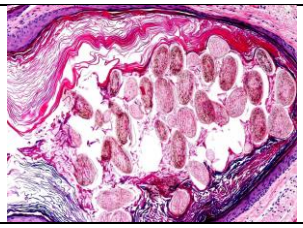
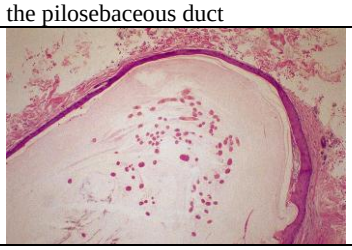
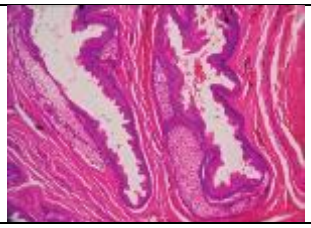
Histopatology of steatocystoma must be find the presence of sebocyte cells in the cyst wall, but after several cuts on hematoxylin eosin stain, there was no visible structure of the sebaceous glands in the cyst wall. Recommendations for did PAS stain, because cyst epithelium in steatocystoma is strongly positive with PAS because epithelium was an excretion tube for sebum.[4] PAS stain revealed cyst react weakly with PAS and there was not seen a sebocyte.

The existence of remaining pigmented hair shaft on the inner lumen cystwas characteristics of pigmented follicular cyst. There are several other epithelial cysts that contain hair shaft inside the lumen of the cyst, such as steatocystoma and eruptive vellus hair cyst. Chuang Y.dkk (Taiwan, 2004) reported one case of multiple pigmented follicular cyst in the vulva in a 62-year-old woman who had a differential diagnosis with eruptive vellus hair cyst and multiple steatocystoma Caballero L, et al (Spain, 1999) reported one case of multiple pigmented hair cysts in the head and neck region in 23-year-old men and are diagnosed differentially with eruptive vellus hair cyst.[8]To distinguish the three forms of epithelial cysts can be seen in Table 1.

Initially patients were diagnosed with eruptive xanthoma, but on histopathological examination we did not found foam cells. The characteristic histologic finding in xanthomas is the foam cell. Foam cells consist of macrophages that contain imbibed lipid within their cytoplasm. whereas in the preparation revealed a cyst lined with a cyst with stratified squamous epithelium containing laminated keratin thus concluded is an epidermoid cyst.[6]

Table 1: Differences in epithelial cysts (taken from literature no. 2,10)

	Multiple pigmented follicular cyst	*Eruptive vellus hair cyst*	*Steatosistoma multiple*
Clinical Appearance	Dome-shaped brownish-colored papules with a diameter of 1-3 cm Predilection: in the chest and abdomen Develops at puberty or young adults who appear suddenly and have no tendency for spontaneous remission	Follicular papules with a size of 1-4 mm. Often brownish, blue, yellow or red On the chest, extremities, buttocks, head and neck region Appears at the age of 4-18 years with sudden and eruptive pop-ups. Usually it is chronic and spontaneous remission can occur	Skin-colored nodules - yellowish in size from a few millimeters to centimeters Chest, back, abdomen and extremities It is inherently autosomal ominant, develops sporadically and is more common in men.
Histopatology	Cyst wall consists of flattened epithelium Sebaceous glands can be found	Constrained by flattened epithelium There is no sebaceous gland	Constrained by flattened epithelium covered by eosinophils Always accompanied by the presence of sebaceous glands

	Contains laminated keratin and pigmented terminal hair shaft Derived from the infundibulum but can also develop from the isthmus	Contains keratin beads and non-medullated, non-pigmented vellus hair Derived from the infundibulum, less often from the isthmus or rarely from the pilosebaceous duct	Contains oily material, layered keratin, non-medullated, non-pigmented lanugo hair shafts Derived from pilosebaceous ducts
			

4. Conclusion

One case report of multiple pigmented follicular cyst in a 26-year-old woman, who clinically resembles multiple steatocystoma. Histopathologically revealed a picture of multiple cysts was found with a cyst wall bounded by a flattened epithelium with a lumen containing keratin and pigmented hair shaft so that this image corresponds to multiple pigmented follicular cysts. The diagnosis is based on clinical manifestations and supported by histopathological examination.

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