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PATIENT CHARACTERISTICS, LOCATIONS AND HISTOPATHOLOGICAL FEATURES OF STEATOCYSTOMA MULTIPLEX CASES

Hilal BALTA[1], İlknur ÇALIK[1], Şenay ERDOĞAN DURMUŞ[2], Sevilay ÖZMEN[3]*, Ahmet Erkan BİLİCİ[4]

[1]*Fırat University, Faculty of Medicine, Department of Pathology, 23200, Elazığ, Türkiye*
[2]*Prof Dr Cemil Taşçıoğlu City Hospital, Department of Pathology, 34384, İstanbul, Türkiye*
[3]*Atatürk University Faculty of Medicine, Department of Pathology, 25240, Erzurum, Türkiye*
[4]*Erzurum Region Training and Research Hospital, Department of Pathology, 25240, Erzurum, Türkiye*

Abstract: Steatocystoma multiplex (SM) is a rare benign genetic disorder consisted of multiple cystic with little or no nail and hair involvement. Steatocystoma is a hamartomatous malformation of the pilosebaceous unit consisting of dermal cysts filled with a sebum-like material. To examine the patient characteristics, locations, presentations and histopathological features of SM in a group of patients. We analyzed 9 patients retrospectively that histopathologically diagnosed as SM. Demographic data collected from pathology reports. Patients' mean age was 43.7 years (range: 15-69 years), with a male to female ratio of 7:2. The average diameter of the lesion at presentation was 13.2 mm (range: 2–30 mm), although 4 cases were less than or equal to 5 mm in size. The lesions located in neck, conjuctiva, face and frontal region. The most common clinical presentation was a painless skin lesion which was noticed incidentally. Histopathological examination of skin showed presence of cystic formation with sebaceous gland on its wall in the dermis and typical stratified squamous epithelium that showed wavy shaped stratum corneum, refractile and strong eosinophilic infiltrated in the cystic wall. SMs are benign lesions more frequent in male patients and especially located in head and neck region. These lesions may be misdiagnosed or underreported.

Keywords: Steatocystoma multiplex, Histopathology, Hamartomatous, Pilosebaceous unit

***Corresponding author:** Atatürk University Faculty of Medicine, Department of Pathology, 25240, Erzurum, Türkiye
E mail: ertekozmen@gmail.com (S. ÖZMEN)

Hilal BALTA https://orcid.org/0000-0003-3745-9694
İlknur ÇALIK https://orcid.org/0000-0003-2160-7453
Şenay ERDOĞAN DURMUŞ https://orcid.org/0000-0003-3388-9312
Sevilay ÖZMEN https://orcid.org/0000-0002-1973-6101
Ahmet Erkan BİLİCİ https://orcid.org/0000-0003-4458-0791

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1. Introduction

Steatocystoma multiplex (SM) is a rare benign genetic disorder consisted of multiple cystic with little or no nail and hair involvement (Gordon et al, 2013; Pietrzak at al., 2015). It is a hamartomatous malformation of the pilosebaceous unit consisting of dermal cysts filled with a sebum-like material (Cho et al., 2002; Surej et al., 2014; Krishnegowda et al., 2015; Santana et al., 2016). Mutations in the Keratin 17 gene are considered in its pathogenesis (Cho et al., 2002; Gordon et al., 2013; Surej et al., 2014; Pietrzak et al., 2015; Krishnegowda et al., 2015; Kromann et al., 2015; Santana et al., 2016). SM lesions are typically located in areas with sebaceous follicles, although atypical presentations involving sites lacking sebaceous follicles have exceptionally been described (Kromann et al., 2015). Steatocystomas can ocur anywhere and have been reported on the oral mucosa and in the subdermal tissue (Kaiser et al., 2016). The aim of this study is to examine the patient characteristics, locations, presentations and histopathological features of SM lesions in Erzurum, Türkiye with a group of patients to increase the knowledge about these lesions for pathologist and clinicians.

2. Materials and Methods

We reviewed 9 patients that diagnosed as SM histopathologically in Pathology Department of Erzurum Training and Research Hospital between 2009-2017, retrospectively. The data of patients' characteristics such as age, gender, site of lesions, lesions characteristics collected from patients' pathology reports. Histopathological analysis done with light microscope and hematoxylen and eosin (H&E) staining sections. Descriptive statistics for the evaluation of results have shown in the form of mean, the nominal variables have shown as the number of cases and percentages (%).

3. Results

Patients' mean age was 43.7 years (age range: 15-69 years). 7 of 9 patients were males (77.8%) and 2 of them

females (22.2%) The average diameter of the lesion at presentation was 13.2 mm (range: 2–30 mm), although 4 patients' lesions ≤5 mm. The most common location of lesions is face followed by neck, frontal region and conjuctiva (Table 1). The most common clinical presentation was a painless skin lesion which was noticed incidentally.

Histopathological examination showed presence of cystic formation with sebaceous gland on its wall in the dermis and typical stratified squamous epithelium that showed wavy shaped stratum corneum, refractile and strong eosinophilic infiltrate in the cystic wall (Figure 1 and 2).

Table 1. Locations of cases

Location	n	%
Face	4	44,45
Neck	2	22,22
Frontal region	2	22,22
Conjuctiva	1	11,11
Total	9	100

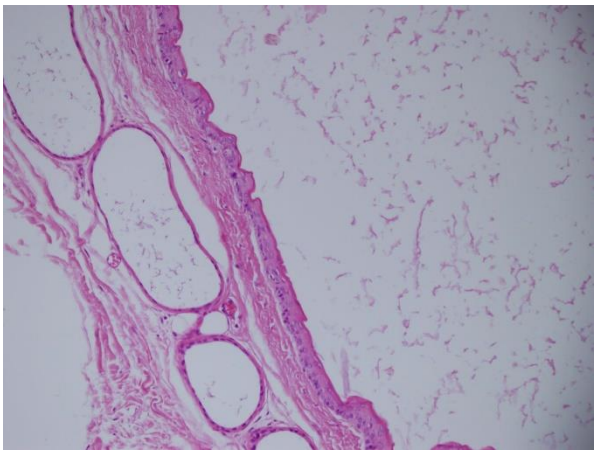

Figure 1. There is a cyst lined by a thin, squamous epithelium with a crenulated surface and the sebaceous gland lobule within the cyst lining (H&Ex200).

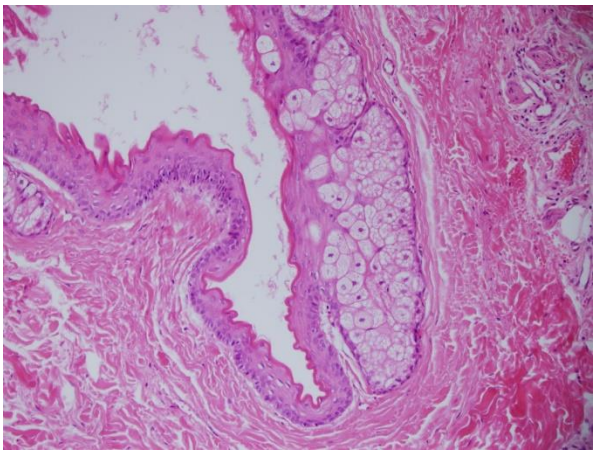

Figure 2. Sebaceous glands, granular layer loss and eosinophilic stratum corneum were seen on the wall of the cystic spaces (H&Ex200).

4. Discussion

SM was first described by Jamieson in 1873, and the term was coined by Pringle in 1899. It is an unusual benign disorder that characterized by the development of numerous sebum containing dermal cysts. It is an autosomal dominant inherited disorder, but most cases are sporadic (Cho et al., 2002; Varshney et al., 2011; Gordon et al., 2013; Surej et al., 2014; Pietrzak et al., 2015).

Lesions are most commonly found on the chest and arms (proximal extremities). Although other sites are also described such as; axillae, neck, abdomen, scalp, vulva and inguinal region (common in women) (Cho et al., 2002; Varshney et al., 2011; Gordon et al., 2013; Surej et al., 2014; Pietrzak et al., 2015; Kromann et al., 2015). However, they can be occur anywhere and have been reported on the oral mucosa and in the subdermal tissue (Kaiser et al., 2016). The largest case series of SM is a review of 64 patients for demographic, clinical, and histopathologic information. The study reported that the majority of cases were sporadic and that an average age of onset was 26 years (range 4 to 64 years-old). The most common location of lesions was on the arms (35%), chest (29%), axillae (20%), and neck (23%). Lesions were found on the legs in only 12.5% and buttocks in only 3.1% of patients (Cho et al., 2002) .In our study the age range was 15-69 years in a mean of 43.7. Locations of our cases were face 4 (44.45%), neck 2 (22.22%), frontal region 2 (22.22%), conjuctiva 1 (11.11%). Contrary to the literature no lesion was observed in the extremities.

Even though the disease usually starts in adolescence or early adulthood, it may also develop at the age of 1 year as well as in the elderly (Varshney et al., 2011; Surej et al., 2014; Krishnegowda et al., 2015, Waldemer-Streyer et al., 2017). In our study the youngest patient was 15 year old. Although there is no sex or racial predilection, according to some authors the condition is more common in males (Krishnegowda et al., 2015; Sawatari et al., 2017). Similarly, in our study there was a male predominance.

SM lesions' sizes ranging from a few mm to several cm (Varshney et al., 2011; Surej et al., 2014; Kaiser et al., 2016; Alina et al., 2017; Sharma et al., 2018). In our cases mean of lesions size was 13.2 mm. The size of 4 lesions were ≤5 mm.

The lesions are usually asymptomatic excepting the suppurative variant which the lesions become inflamed and suppurative after minor trauma (Kamra et al., 2013; Gordon et al., 2013; Santana et al., 2016; Waldemer-Streyer et al., 2017). Cystic lesions are usually multiple, small, soft, movable, yellow to skin-colored (Surej et al., 2014; Santana et al., 2016; Kaiser et al., 2016; Waldemer-Streyer et al., 2017). In macroscopic examination SM cysts are smooth and round ranging in size from a few mm to several cm (Kaiser et al., 2016; Waldemer-Streyer et al., 2017).

Histopathologically SM lesions are multiple cysts with little or no nail and hair involvement examination

(Pietrzak et al., 2015). The differential diagnoses include epidermal and epidermoid cysts, eruptive vellus hair cysts, neurofibromatosis, lipomatosis, milia, sebaceous hyperplasia, sebaceous adenomas, and xanthomatosis. The term “sebaceous cyst” is often used interchangeably with epidermal cyst, which is essentially a misnomer. In fact, SM is the only cyst with a true sebaceous origin. On histopathology, SM shows flattened sebaceous lobules located close to the cystic wall (Sharma et al., 2018).

The prevalence of these lesions is not currently known which is likely due to the fact that they are rarely reported (Kaiser et al., 2016).

Treatments for SM have included cryosurgery, aspiration, and time-consuming and scarring surgical excisions (Gordon et al., 2013). All of our patients were surgically excised.

5. Conclusion

In conclusion, SMs are benign lesions more frequent in male patients and especially located in head and neck region. These lesions may be misdiagnosed or underreported. We think that our work may contribute to the literature because it is the first patient series work in the East Anatolia region in Türkiye.

Author Contributions

Concept: H.B. (20%), İ.Ç. (20%), Ş.E.D. (20%), S.Ö. (20%), and A.E.B. (20%), Design: H.B. (20%), İ.Ç. (20%), Ş.E.D. (20%), S.Ö. (20%), and A.E.B. (20%), Supervision: H.B. (20%), İ.Ç. (20%), Ş.E.D. (20%), S.Ö. (20%), and A.E.B. (20%), Data collection and/or processing: H.B. (20%), İ.Ç. (20%), Ş.E.D. (20%), S.Ö. (20%), and A.E.B. (20%), Data analysis and/or interpretation: H.B. (20%), İ.Ç. (20%), Ş.E.D. (20%), S.Ö. (20%), and A.E.B. (20%), Literature search: H.B. (20%), İ.Ç. (20%), Ş.E.D. (20%), S.Ö. (20%), and A.E.B. (20%), Writing: H.B. (20%), İ.Ç. (20%), Ş.E.D. (20%), S.Ö. (20%), and A.E.B. (20%), Critical review: H.B. (20%), İ.Ç. (20%), Ş.E.D. (20%), S.Ö. (20%), and A.E.B. (20%), Submission and revision H.B. (20%), İ.Ç. (20%), Ş.E.D. (20%), S.Ö. (20%), and A.E.B. (20%). All authors reviewed and approved final version of the manuscript.

Conflict of Interest

The authors declared that there is no conflict of interest.

Ethical Approval/Informed Consent

Research was conducted in line with the Declaration of Helsinki and Good Clinical Practice. Ethical permission was obtained from the Erzurum Region Training and Research Hospital Clinical Research Ethics Committee (Number: 2022/07-75; Date: 06.06.2022).

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