

Multifocal type of pilomatrixoma

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Abstract

Pilomatrixoma is a benign skin neoplasm that arises from hair follicle matrix cells. The skin lesion occurs usually as a solitary tumor and the multifocal types are very rare. Skin changes can be described as a firm to hard, non-painful, oval-shaped tumor that is covered by normal skin. It commonly occurs on a scalp, face, neck and rarely back and extremities. Complete surgical excision with the proper margin is the treatment of choice, what guaranteed the radical therapy of pilomatrixoma.

In this paper case of 16-years-old male patient with many solid tumors in subcutaneous tissue on both arms will be reported. The first skin lesion appeared on the left arm 6 years ago. Clinically the disturbance was diagnosed as an atheroma, and it was excised. One year after surgical procedure the patient observed the appearance of new nodules on both arms. In the therapy surgical excision was performed with histopathological examination of the tissues. Histopathological test has proved the clinical diagnosis of pilomatrixoma.

The case of multifocal pilomatrixoma, which is rarely diagnosed and described in professional literature, will be presented.

Key words: pilomatrixoma, multifocal localization, children, neoplasm.

Introduction

Pilomatrixoma (*Malherbe and Chenantais, Forbis and Helwig*), also known as a calcifying epithelioma, is a benign skin neoplasm that arises from hair follicle matrix cells [1-3]. It may occur at any age, ranging from children to adults (but rather rarely) [4]. This benign skin neoplasm occurs most often in cases of patients at the age of 20 and younger [5]. There are two main peaks of appearance of pilomatrixoma depending on the age of the patients: below 20 and 50 years of age [5].

This tumor occurs more often in case of women, due to reporting literature the female: male ratio is 2, 4:1 [6], 3:1 [4] or in another reports 2:1 [7].

The skin lesion occurs usually as a solitary tumor and the multifocal types are very rare [1]. In some cases pilomatrixoma could coexisted with systemic abnormalities: myotonic dystrophy [8-13], myotonic dystrophy within AIDS [14], internal anomalies in Gardner [15,16], Turner's [17] and Rubinstein-Taybi syndrome [18], that is why the patient with this neoplasm should be carefully examined towards these abnormalities. Skin changes characterized as a firm to hard, non-painful, oval-shaped tumor that is covered by normal skin. The diameter was ranged from several millimeters to several centimeters. The most common localization is the scalp, face, neck and rarely back and extremities [6,19-22]. Complete surgical excision with the proper margin is the treatment of choice, which guaranteed the radical therapy of pilomatrixoma [1,20].

Case report

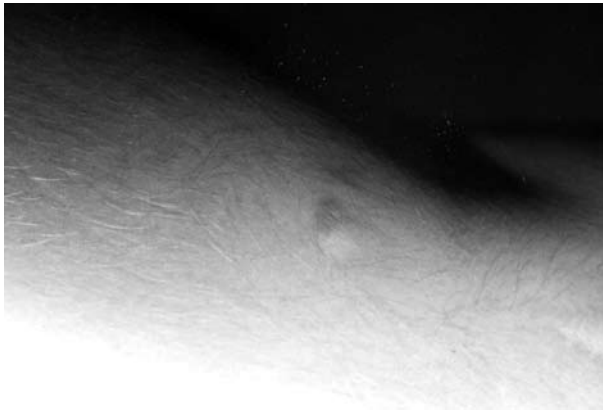
In the year 2002 a 16-years-old male patient was admitted to the Dermatological Outpatient Clinic in Katowice because of recurrence of the two skin lesions. Clinically in dermatological examination three asymptomatic, firm, solid tumors in subcutaneous tissues on both arms were proved. The first skin lesion appeared on the left arm 6 years ago. Clinically

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Figure 1. Typical skin lesion on the arm, with cicatrix after surgical excision



the disturbance was pre-diagnosed as an atheroma, which was excised by a surgeon in the ambulatory at the patient's living area. One year after surgical procedure the patient observed the appearance of new nodules on the left arm and one new on the skin of the right arm. Because of this, the patient came to The Department of Dermatology of Silesian Medical University in Katowice, where pilomatrixoma was recognized.

In dermatological examination three skin lesion were described as a well-circumscribed, firm nodules, oval-shaped, varied in diameter from 0.5 to 1.0 cm, localized on both arms (*Fig. 1*). There was slight pink discoloration of the overlying skin.

The patient has undergone surgical procedure in topical anesthesia with 0,5% solution of xylocaine. Three lesions were excised totally with the healthy margin of the skin with adherent and overlying skin.

In the therapy surgical excision was performed with histopathological examination of the nodules (*Fig. 2*), which showed masses of mummified shadow squamous epithelial cells, focally, with rows of basophilic cells resembling the hair matrix. The surrounding fibrous connective tissue showed prominent resorption including numerous, multinucleated, foreign body type giant cells. No features of osseous metaplasia or calcifications were found. The lesion was diagnosed as pilomatrixoma.

After surgical procedure the patient was treated by neurologist because of peripheral inflammation of the left facial nerve with total improvement.

In additional examination no systemic abnormalities were found. In ophthalmologic consultation normal state and function were described.

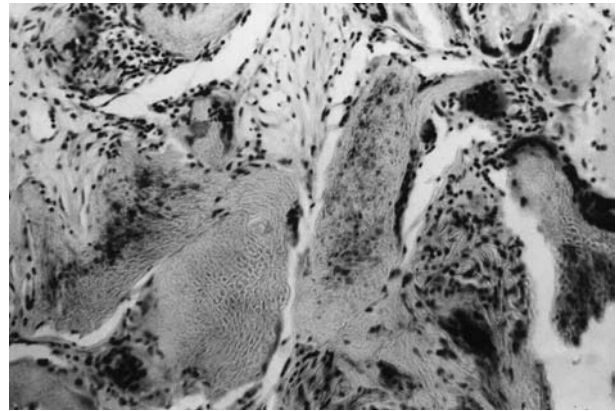
The follow-up period was 4 years and no recurrences were found.

Discussion

The presented case of multifocal pilomatrixoma is a rarely diagnosed and described in the professional literature.

The appearance of this neoplasm is almost asymptomatic.

Figure 2. Histopathological examination of excised nodule (haematoxyline-eosine stain, mag.150x)



In some cases the lesion is associated with pain, inflammation and ulceration [20].

Multiple occurrences of pilomatrixoma is rarely reported in the literature [7,8,11,14,15,23-26] and it is assessed to 3.5% of cases [24]. Mostly appearance of this tumor is associated with familial occurrence [8,11,21,27]. Pilomatrixoma is a well-known pathognomic sign of myotonic dystrophy [8-13]. Pujol, at al. [15] reports that multifocal pilomatrixoma coexists with adenomatous colonic polyps, osteoma of the mandible and ocular-pigmented retinal macules as changes in patients with recognized Gardner syndrome. In the case of our patient no familial and gastrointestinal disturbances were observed. Another rare clinical types of calcifying epithelioma of Mahlerbe are: bulbous form [3,28] and perforating type [29-31]. Bertazzoni at al. [32] reported pilomatrixoma with perilesional anetoderma caused by inflammatory processes and lack of elastic fibers.

These neoplasm-involved areas include the scalp, head and the upper extremities [3]. The most affected skin regions are face [4,20], scalp [20,22,33], neck [4,22,33], chest [33] and upper limbs [22,33]. The head, especially the cheek ad preauricular and parotid region are the most common sites – in about 50% [7]. Over 25-30% of present lesions are localized on the skin of the upper limbs.

Most typical clinical picture of pilomatrixoma is occurrence of solitary, small, firm nodule, covered with normal skin, varying in size from 5 to 30 mm [6]. The skin lesion is usually less than 3 cm [4,5]. Pilomatrixomas of atypical large size has been termed giant pilomatrixomas [24,31]. Although pilomatrixoma is a benign skin tumor, in the literature there are reports due to occurrence of pilomatrixoma carcinoma [4,25,34-36]. The surgeons should be aware of the various types of pilomatrixoma with rare occurrence of malignancy [19]. Pilomatrixoma carcinomas usually lead to the metastases to the lungs [34-36], liver [34,36] regional lymph nodes [4,34,35] and brain, heart, pancreas, kidney, adrenal gland, gastric mucosa, skin and bones [35].

Although the pilomatrixoma has its typical clinical picture in many cases the diagnosis is incorrect. In reports of Wells at al. [33] the referring diagnosis was improper in 94% of cases, and the preoperative recognition in 57% was misdiagnosed.

The best-known therapy of the pilomatixoma is total surgical excision including adherent skin [6,20].

Histopathological picture of pilomatixoma depends on the stage of the tumor development. There is prevalence of living epithelial elements in early stages, and retrogressive changes in older ones, leading to the formation of foci of mummified epithelium with shadow cells, calcifications, and reactive resorptive response in the connective tissue. The periphery of the basophilic epithelium resembling hair matrix contains viable cells, white central parts undergo mummification. In 15-25% of cases there is osseous metaplasia within the tumor or in its vicinity [37,38].

Conclusions

Despite the typical clinical picture and benign character of pilomatixoma, the recognition of this dermatological entity may lead to misdiagnosing. Clinically in most cases the lesion occurs like a solitary nodule, but doctors should remember about rare, but existing, multiple localization. Complete surgical excision, including the overlying skin is the treatment of choice.

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