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Childhood Eyelid Pilomatricoma Mimicking Recurrent Chalazion

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Abstract: Pilomatricoma is a rare benign subcutaneous childhood tumor first described in 1880. It has no gender predilection. The exact etiology of this disease is still unknown. However, association with mutation in the CTNNB1 gene was reported. The most frequent location is the neck, although the upper eyelid is an uncommon site of involvement. The diagnosis of pilomatricoma is clinically challenging, and is based on histopathologic examination, after complete surgical excision, which is the radical treatment. It shows foreign body reaction that surrounds nucleated basophilic cells and islands of enucleated calcified ghost cells. The tumor may have diverse clinical presentations and aspects. It commonly manifests as an asymptomatic, solitary subcutaneous mass. Some clinical signs are specific such as the tend sign and the teeter-totter sign. Squamous and basal cell carcinoma are the main clinical differential diagnoses, and should be routinely ruled out. Radiologic investigations (ultrasonography and Magnetic resonance imaging: MRI) may enhance diagnosis accuracy in atypical cases. Ultrasonography shows hyperechoic and heterogeneous mass. MRI is more specific for diagnosis showing homogeneous well-defined mass on T1-weighted scans and high signal intensity on T2-weighted images. Pilomatricoma of the upper eyelid may be misdiagnosed and treated as recurrent chalazion. Recurrence and malignant transformation of pilomatricoma are rare after complete surgical excision. This case report is about a 14-year-old healthy boy who exhibited large pilomatricoma in the upper eyelid initially misdiagnosed and mistreated as chalazion.

Keywords: Pilomatricoma, Eyelid, Chalazion, Childhood, Surgery

1. Introduction

Pilomatricoma is a common benign childhood tumor, which manifests on the neck and cheeks [1]. This disorder was first described by Malherbe in 1880 [2], and is the second most common superficial lump excised in children [3]. Pilomatricoma may uncommonly involve the eyelids [4]. The definite diagnosis is somewhat challenging, and is based on histopathology after complete surgical excision [5].

This is an unusual case of eyelid pilomatricoma in a healthy boy misdiagnosed as chalazion.

2. Case Report

A 14-year-old otherwise healthy boy, with a 6 months history of surgical excision of a chalazion involving the right upper eyelid, presented with a gradually increasing and painless swelling lesion in the chalazion excision site. On examination, there was a 10 mm x 7 mm x 6 mm, purple, single, large, ovoid, well-circumscribed, firm, superficial and mobile mass, located on the outer portion of the right upper eyelid (Figure 1).

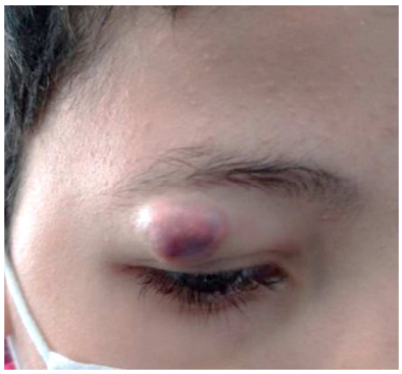


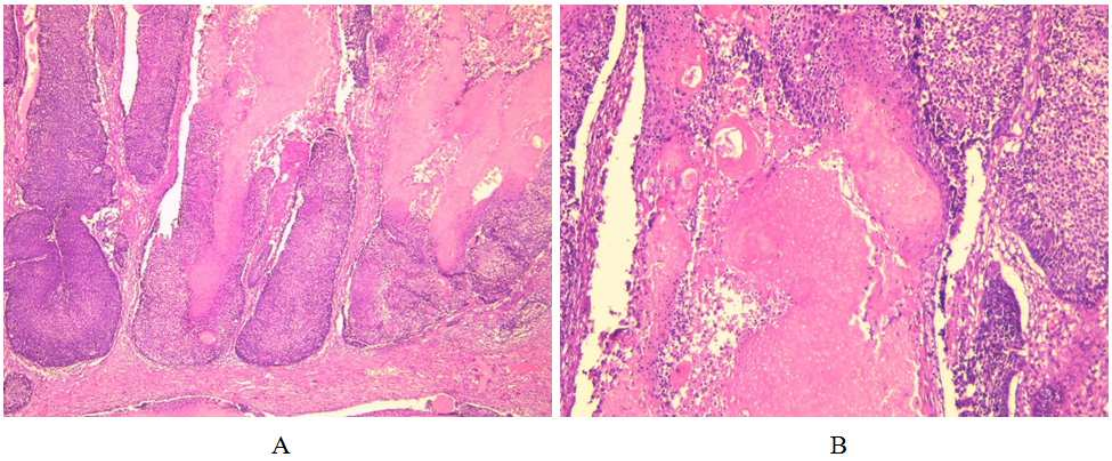
Figure 1. *Color photograph of the right eye shows a purple large, ovoid, well-circumscribed, superficial mass located on the outer portion of the upper eyelid.*

Visual acuity was 20/20 in both eyes, and no afferent pupillary defect was noted. The ocular motility was full. Slit-lamp examination showed normal anterior segment bilaterally, and funduscopy was unremarkable. Systemic examination was normal.

Ultrasonography showed subcutaneous ovoid mass with heterogeneous echostructure tissue and mixed blood supply.

Total surgical excision was performed and was uneventful. Histopathological examination revealed nucleated basophilic cells in the periphery, and islands of enucleated calcified ghost cells surrounded by foreign body reaction (Figure 2). A final diagnosis of pilomatricoma was finally made.

After a follow-up period of 6 months, there was no evidence of local recurrence (Figure 3).



A B

Figure 2. *Photomicrograph of excised tumor (A: Hematoxylin Eosin X40; B: Hematoxylin Eosin x 100) shows multilobulated tumor with basophilic cells and ghost or mummified cells which are eosinophilic and enucleated. Note the high mitotic index of the tumor.*

Figure 3. *Color Photograph of the right upper eyelid, 6 months after surgery shows no evidence of local recurrence.*

3. Discussion

Pilomatricoma is a benign skin tumor growing from cells in the hair follicle and is also known as pilomatrixoma or calcifying epithelioma of Malherbe [6, 7]. It may occur at any age, most often in children and young adultes [1, 8, 9]. No gender predilection was reported [10]. There is a particular diagnostic challenge to clincians due to the diverse presentations of the tumor. Classically, pilomatricoma usually manifests as an asymptomatic subcutaneous nodule [5, 11]. In this patient pilomatricoma presented as unique well-delimited subcutaneous nodule. The size of these lesions ranges from 0.5 to 3 cm [5]. Pilomatricomas are usually solitary nodules, but multiple lesions have been also reported [1]. This clinical form is reported in patients with multisystemic disorders, including Gardner syndrome, Rubinstein-Taybi syndrome, myotonic dystrophy, Kabuki syndrome and Trisomy 9 [9]. The most common affected sites are the head and the neck [1]. Periorbital region is involved in 17% of cases [12]. As in this patient, eyelid pilomatricoma may be misdiagnosed clinically as chalazion [4, 13]. The lesion for this patient was misdiagnosed and mistreated as chalazion. The recurrence of the lesion 6 months later, and pathological examination showed that it was initially an eyelid pilomatricoma. The tumor generally manifests as subcutaneous red to blue mass that is well-circumscribed, mobile, and firm or gritty to palpation [14]. The tend sign is pathognomonic of this lesion, stretching the skin, over the tumor, makes its elongation [4, 11]. In addition, pressing on one edge of the lesion causes elevation of the opposite side like a teeter-totter. Both signs are helpful clinical features for diagnosis [11]. The diagnosis is rarely

made clinically. It may be related to the fact that most clinicians are not familiar with this rare eyelid tumor [1]. There are many clinical differential diagnoses such as pyogenic granuloma, arteriovenous malformation, epidermal inclusion cyst, soft tissue sarcomas, cutaneous lymphoma, and matrical carcinoma [9]. Chalazion is not a common differential diagnosis [15]. All these conditions should be routinely ruled out in eyelid tumors mimicking pilomatricoma.

Radiologic investigations (ultrasonography and MRI) may enhance diagnosis accuracy in atypical cases. Ultrasonography, may enhance the exact preoperative diagnosis from 33 to 76% [15**,** 3, 1]. It shows well-defined, ovoid, hyperechoic heterogeneous mass with internal calcifications, vascularity and posterior shadowing [6, 14, 16], such features were also noted in this patient. Magnetic resonance imaging (MRI) shows well-defined mass with intermediate homogeneous signal intensity on T1-weighted scans and high signal intensity on T2 – weighted images [14].

Pilomatricomas do not spontaneously regress [6]. Surgical excision with complete resection and clear margins is necessary to final diagnosis and successful treatment [3, 14]. No recurrence was noted after surgical management in this patient. Incomplete excision has been associated with high recurrence rates [15]. When excised, these lesions are often larger than they appear on the surface [4]. Typical histopathologic findings show cells in dermal nodules with nucleated basophilic cells on the periphery and enucleated shadow cells with calcifications in the center [17], with a high mitotic index [9]. However, immunohistochemical stains are not necessary for the diagnosis [15]. Mutations associated, include beta-catenin and bcl-2, which were high level expression for the basophilic cells in pilomatricomas [15]. An association with a mutation in the CTNNB1 gene was previously reported [9]. The exact etiology of the disease remains controversial. Some authors speculate that repeated skin trauma may induce an inflammatory response that leads to an overgrowth of hair matrix [6]. In this patient, no evidence of ocular trauma was reported.

Proliferating pilomatricoma is a rare variant [1**,** 9]. It was previously reported in 69 cases [18], and the presence of multiple tumors has been associated with myotonic dystrophy [3, 19]. This serious form is asymmetrical, rapidly growing, vascular and infiltrating tumor [9, 20]. It occurs more frequently in males and elderly patients [9, 20]. There are 4 reported deaths as a consequence of metastatic pilomatricomas. The most frequent sites are lungs, bones and lymphatics [20]. In the literature, there is fewer than 10 cases reported of pilomatricoma with extensive osseous metaplasia [21]. There is new described histopathologic entity reported as an intermediate form between pilomatricoma and pilomatrical carcinoma [22].

After complete surgical exision of pilomatricoma, the outcome is usually good with no evidence of local recurrence or metastasis [23]. The frequent benign nature is reassuring for both patients and clinicians [24], but pilomatricoma should be routinely considered in the differential diagnosis of eyelid skin tumors [25, 26]. Recently, there are more reports of malignant and borderline forms of pilomatricoma, which require an early diagnosis and adequate management, to prevent transformation and extension. Squamous and basal cell carcinomas, kertoacanthoma, epidermoïd cyst and metastasis are the main clinical differential diagnosis, and should be systematically ruled out [20].

4. Conclusion

Pilomatricoma of the eyelid is a rare benign tumor, which may occur in young children, and may be easily misdiagnosed as chalazion. The ophthalmologist should include pilomatricoma in the differential diagnosis of eyelid tumors in children, especially in the case of chalazion. The clinical diagnosis is somewhat challenging, and requires a high index of suspicion. Some clinical features are specific such as the tend sign and the teeter-totter sign.

Radiologic investigations (ultrasonography and MRI) may enhance diagnosis accuracy in atypical cases. Recently, there are more reports of malignant and borderline forms, which require early diagnosis and appropriate management to prevent transformation and extension. Complete surgical excision and pathological examination are the clue for the definite diagnosis with a good long-term outcome.

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