Pilomatrixoma On The Lateral Canthus
E Copcu

Citation

Abstract
We report a 4-year-old girl with a painless lump located on the left lateral canthus and infero-orbital region. The lesion on the left lateral canthus was 8 X 8 mm in diameters. An initial diagnosis of dermoid cyst was made. Excision was performed and the specimen was diagnosed histopathological as "pilomatrixoma". Pilomatrixoma of the eyelid is often misdiagnosed clinically. The most frequent site is in the head and neck with more than 75 per cent of the pilomatrixomas being located on the scalp, face, neck or arms. Although pilomatrixomas located on the periorbital region were reported in the literature, as far as we are aware, lateral canthus has not been previously reported.

Pilomatrixoma (also called pilomatrixoma or benign calcifying epithelioma of Malherbe) is a rare, benign tumour originating from the matrix of the hair root. In 1880, Malherbe and Chenantais, first described pilomatrixomas, then thought to arise from sebaceous glands, and called calcifying epitheliomas of Malherbe. Lever and Griesemer suggested that the origin of the tumor was hair matrix cells. The term pilomatrixoma was first used by Forbiss and Helwig in 1961, thus avoiding the word epithelioma, which carries the connotation of malignancy. The term was later corrected to pilomatrixoma, to be more etymologically correct. Since that time, multiple reports have established that pilomatrixoma typically involves the head and neck, and the majority occurs in children. Many different anatomical sites were presented in the literature but according to our knowledge our report is the first which presented lateral canthus located pilomatrixoma.

CASE REPORT
4 year-old girl presented our Plastic and Reconstructive Surgery Department with a painless lump located on the left lateral canthus. According to the history from her parents, this lump had appeared four months ago. She had no symptoms.

On examination, it measured 8 mm by 8 mm, had a red-white discoloration, and was hard and painful to touch (Figure 1). Her ocular examination was otherwise normal. She had no other signs and symptoms. Results of her routine laboratory test were normal. An initial diagnosis of dermoid cyst was made. She was operated under general anesthesia. Lumb was totally removed. Excision was quite easy. Defect was closed with 5/0 prolene.

Figure 1
Figure 1: Pre-operative view of the patient.

Histopathology showed a pseudoencapsulated mass composed mainly of eosinophilic acellular material in which ghost cells were prominent with considerable calcification. At the periphery there were focal areas of basaloid cells with little cytoplasm. Histological diagnosis revealed pilomatrixoma (Figure 2). There was no early or late complication.

Patient was followed six months. There was no recurrence.
DISCUSSION

Pilomatrixoma typically presents as a firm, non-tender, subcutaneous nodule, and adherent to the skin but not fixed to underlying tissue. Growth is usually slow and may occur over a period of months to years. The vast majority of head and neck masses occurring in children are of either inflammatory: infectious or congenital origin. Neoplastic processes are uncommon. Pilomatricoma can present at any age, it demonstrates bimodal peak presentation during the first and sixth decades of life, however 40% of cases occur in patients younger than 10 years of age and 60% of cases occur within the first two decades of life. Female preponderance has been reported with a male:female ratio of 2:3.

It is more common in women and the majority of cases are Caucasians. The most frequent site is in the head and neck with more than 75 per cent of the pilomatrixomas being located on the scalp, face, neck or arms. The parotid region being one of the more common sites. No cases have been reported on the palms or soles, perhaps because of the lack of hair-bearing skin in the areas. The diagnosis of pilomatrixoma is usually suspected based on palpation of a superficial, rock-hard mass and confirmed by histopathologic examination. Histologically, pilomatrixoma is a deep subepidermal tumor consisting of irregular islands of epithelial cells. The epithelial cells are organized in a characteristic biphasic architectural pattern with keratinized ghost cells in the center surrounded by variable amounts of peripheral basaloid cells. The basaloid cells exhibit deeply staining basophilic nuclei, which often contain small nucleoli. The keratinized ghost cells in the center have lost their nucleus and thus have a central unstained area. Ghost cells represent the attempts of the immature basaloid cells to manufacture hair; however, hair shaft formation is absent because differentiation in pilomatrixoma is toward the hair matrix. Neither presents with irregular nodules on the skin as pilomatrixoma does.

Radiologic imaging is of little diagnostic value for pilomatrixoma. The relatively superficial location of these tumors makes routine radiographic imaging unnecessary. Multiple or recurring tumors may be found in association with Gardner syndrome, myotonic dystrophy, sarcoidosis or Turner's syndrome. Familial pilomatrixoma and multiple familial pilomatrixoma are even rarer. In the vast majority of cases, pilomatrixoma is a solitary neoplasm, however, multiple tumors occurring synchronously account for 2-3.5% of reported cases.

The differential diagnosis of pilomatrixoma is varied. Firstly, pilomatrixoma should be differentiated from epidermal and dermoid cysts. Epidermal cysts are firm, round, and mobile, and they have normal overlying skin. Dermoid cysts are firmly attached to underlying tissue and are often found in children. Other entities to be considered in the differential diagnosis include: inclusion cysts, giant cell tumour, foreign body reaction, eccrine spiradenoma, osteoma cutis, trichilemmal cyst, basal cell carcinoma, and hydrocystoma. The treatment of choice and standard therapy for benign pilomatrixoma is complete surgical excision. Occasionally, overlying skin will need to be excised secondary to tumor adherence to the dermis. Pilomatrixoma of the eyelid is often misdiagnosed clinically. But there are characteristic features of the lesion that can help clinicians differentiate it from other tumours seen in children. This tumor is commonly misdiagnosed preoperatively (75%) when evaluation is based on clinical evidence alone, and should always be considered in the differential diagnosis with other eyelid lesions. Location of the periorbital region is not unusual for this tumour; although involvement of eyebrow, lower and upper eyelid
were reported in the periorbital region, as far as we are aware, our case is the first with pilomatricoma located on the lateral canthus.

CORRESPONDENCE TO

Eray COPCU, MD Plastic and Reconstructive Surgery
Department Adnan Menderes University, Medical Faculty, Aydin, 09100 TURKEY E-Mail: copcu@lycos.com Phone: +90.535.7368430 Fax: +90.256.2120146

References

Author Information

Eray Copcu, M.D.
Assistant Professor, Plastic and Reconstructive Surgery Department, Medical Faculty, Adnan Menderes University