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CASE REPORT

A RARE CASE OF PILOMATRIXOMA

Rajshekhar Patil[1], Vishal Kadeli[2], Palla Abhishek Reddy[3]

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ABSTRACT: Pilomatrixoma is a benign neoplasm derived from hair follicle matrix cells. They usually present in 1st decade of life and most commonly occur in head and neck region. Here we present an interesting case of a 6 year old girl with pilomatrixoma bilaterally over the arms.
KEYWORDS: Pilomatrixoma, Calcifying epithelial tumor of Malherbe

INTRODUCTION: Pilomatrixoma is an uncommon and harmless skin lesion derived from hair matrix cells.It is also called 'pilomatricoma' and sometimes known as 'calcifying epithelioma of Malherbe'.Calcifying epithelioma was originally described in 1880 by Malherbe and Chenanlais as a neoplasm of sebaceous glands. The term pilomatrixoma was introduced based on further studies performed in 1961 by Forbis and Helwig who demonstrated that the cells differentiated in the direction of cortical cells of the hair follicle[1, 2, 3].Pilomatricoma is most often diagnosed in young children. The female to male ratio is around 3:2.It arises as a single skin coloured or purplish lesion on the head and neck region but rarely it may occur at other sites. It is characterized by calcification within the lesion, which makes it feel hard and bony and often results in an angulated shape (the 'tent' sign) [4].

CASE REPORT: Six year old female patient presented to our department with history of swelling in both arms. Swelling was first noticed in right arm 1year back which of size of a peanut and gradually increased to present size. There was also a swelling on left arm noticed 1 month back. There was no history of fever, pain in the swellings. On examination there was a non tender swelling in right arm about 3*2cm in size, hard in consistency, mobile with irregular surface. Another swelling was seen in left arm, measuring 2*1cm nontender, mobile and hard in consistency with irregular surface. X-ray of both arms showed no bony involvement. Both swellings were excised. Grossly they were well circumscribed and pearly white histopathology of which showed it to be pilomatrixoma (figure 1).The histology is presented in figures 2 and 3.

DISCUSSION: The pilomatricoma tumor commonly occurs in the head and neck regions of children[5].The diagnosis of pilomatrixoma can be made clinically if the characteristics of the tumor are known. Diagnostic tests and Imaging studies are often unnecessary in the work up of a superficial benign lesion like pilomatrixoma however tests are done to exclude malignancy or to determine the depth of lesion. Fink and Berkowitz found ultrasound to be helpful in children[6].

The diagnosis of pilomatricoma can be made clinically. Danielson-Coheson et al[7] said that the preoperative diagnosis might be improved by being aware of the fact that pilomatrixoma was a harmless benign skin tumour in children and patients usually present with a solitary nodule that has been slowly growing over several months or years. Patients are usually asymptomatic but some report pain during episodes of inflammation or ulceration. Pilomatricoma lesions slide freely over

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the underlying area. Graham and Merwin described the 'tent sign' elicited by stretching the skin over the pilomatrixoma to feel the irregular surface of the mass. There is no associated lymphadenopathy and a blue discoloration may be seen[8].Yoshimura etal[9]suggested that the diagnosis of pilomatrixoma should be suspected when the mass is adherent to skin but not fixed to underlying tissue. Pilomatrixoma generally presents with subcutaneous red to blue mass that is fairly well circumscribed, freely movable and firm to gritty on palpation[10]. Clinical features as documented by Duran et al[11]and later also by Perez and Nicholson[12]should arise clinical suspicion and they include onset in childhood or early adulthood, average size of 10 mm or less, consistency ranging from firm to cystic, moderate pattern of growth, pink to purple hue with sub-epithelial yellowish tinge, and intact overlying skin with telangiectatic vessels.

Clinical differential diagnosis includes epidermoid cysts, dermoid cyst, sebaceous adenoma or carcinoma, juvenile xanthogranuloma, capillary hemangioma, chalazion, and rhabdomyosarcoma[13, 10, 12]Although they grow slowly;they occasionally demonstrate rapid growth and may resemble keratoacanthoma[14].

A rare malignant counterpart, pilomatrix carcinoma, has been described, and approximately 90 cases have been reported in the literature. It is locally aggressive and can recur. In several cases, it has demonstrated metastases. Many key features are similar between these benign and malignant counterparts; the primary differentiating characteristics include a high mitotic rate with atypical mitoses, central necrosis, infiltration of the skin and soft tissue, and invasion of blood and lymphatic vessels[18, 19].

Histopathologic examination reveals the tumor to be grossly well circumscribed and firm to gritty in consistency (figure 1). Microscopic examination shows numerous islands of epithelial cells with characteristic arrangement of basophilic cells in the periphery and shadow cells in the center (figure 2). As the tumor matures a number of basophilic cells lose their nuclei and become shadow cells (figure 3). Calcification is seen in 75% of the cases. Sheets of intensely eosinophilic keratinous material is seen within necrotic areas, and this may induce a foreign body giant cell reaction.[15, 10, 16] Histopathologic differential diagnosis include basal or squamous cell epitheliomas as well as a variety of skin and subcutaneous cysts[17].

As performed in this case, management of pilomatrixomas typically involves complete surgical excision. Lesions on the extremities may be left untreated unless they become large or symptomatic, however in many cases these are excised for definitive diagnosis. If the tumor adheres to the dermis, the overlying skin may be excised. The recurrence rate is low, ranging from 0 to 3 percent[20].

CONCLUSION: Although a very rare tumour and often misdiagnosed as epidermal or dermoid cyst, it has its distinctive clinical and unique histological features which differentiates it. Since spontaneous regression never occurs, cosmetic problems and reports of its malignant transformation demand its complete excision.

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Gross specimen after excision (2 in no.)

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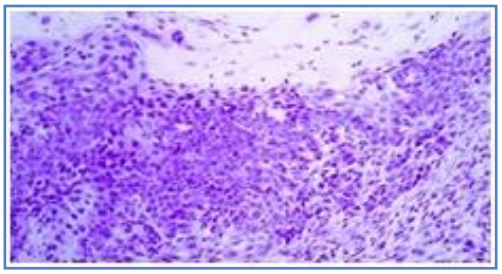

Fig. 2

Features which are helpful in making the diagnosis include asymmetrypoor circumscription, presence of several markedly sized and variably shaped basaloid aggregations of tumor cells continuity of basaloid cells with epidermis, extensive areas of necrosis en masse, infiltrative growth pattern, presence of ulceration.

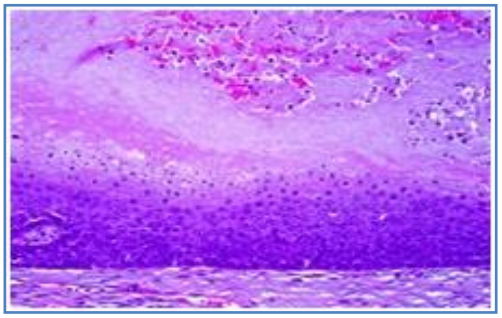

Fig. 3

The transformation of basaloid cells into shadow cells is associated with loss of nuclei.

AUTHORS:

1. Rajshekhar Patil
2. Vishal Kadeli
3. Palla Abhishek Reddy

PARTICULARS OF CONTRIBUTORS:

1. Associate Professor, Department of General Surgery, Basweshwar Teaching and General Hospital, Gulbarga.
2. 3rd Year Resident, Department of General Surgery, Basweshwar Teaching and General Hospital, Gulbarga.
3. 1stYear Resident, Department of General Surgery, Basweshwar Teaching and General Hospital, Gulbarga.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr.Rajshekhar Patil,
Block No. 56, Swastik Nagar,
Bilgundi Layout, Sedam Road,
Gulbarga – 585105.
Email-drrjsp@gmail.com

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