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RESEARCH ARTICLE

SCROTAL PILOMATRIXOMA: A RARE CASE REVEALED BY AN HYDROCELE

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Abstract

Pilomatrixomais a cutaneous tumor, originating from the hair matrix, mostly affecting pediatric population. We report a particularly uncommon location of pilomatrixoma: the scrotal skin. A 50-year-old man, without history, has developed a hydrocele . Most often, its preoperative diagnosis is not possible, because of its clinical polymorphism. The precise diagnosis is histological, by the revealing of mummified cells. The treatment is surgical, with only purpose to perform complete removal of pathologic tissue, allowing to decrease the rate of local relapse.

Introduction:-

Pilomatrixoma is a rare, benign, slow-growing, cutaneous tumor of the hair follicle matrix.First described by Malherbe in 1880, It is also called calcifying epithelioma of Malherbe [1]. It is more common in the pediatric age group and women; however, the overall incidence is roughly one in 800 cutaneous tumors only [2]. Few retrospective studies have also reported a bimodal distribution of presentation with the first peak around the first decade of life and a second peak at the sixth decade [3]. It is commonly located over the face (preauricular, periorbital, neck, and upper trunk) [4]. It is mostly a solitary and asymptomatic lesion, whereas multiple forms have syndromic associations.

The adnexal tumoris often misdiagnosed clinically due to its resemblance with other common skin masses like sebaceous, epidermoid, or dermoid cysts. The diagnostic modality of choice is tissue biopsy,. There is no role of medical therapy as the tumor does not exhibit spontaneous regression; surgical removal is the only option by all means.

Here, we present a case of scrotal mass that was presented as a suspicious hydrocele but turned out to be a pilomatrixoma of the scrotum.

Case Presentation

We report the case of a 50-year-old man who consulted in urology for hydrocele evolving for 6 months. There was no complaint of associated fever or pain. On examination, hydrocele was noted with a little, firm, sessile mass on the lateral aspect of the scrotum with lobulated appearance (Figure 1).

Bilateral testes were normal and could be felt separately. No regional lymphadenopathy was noted. The patient was posted for surgery under spinal anesthesia. Mass was excised and sent for histopathological examination. The scrotum was closed in layers with absorbable sutures. The patient was discharged in clinically stable condition on the next day.

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The histopathological report was suggestive of a pilomatrixoma of the scrotum described as a benign proliferation made of solid nests of basaloid cells undergoing abrupt trichilemmal-type keratinizationwithpresence ofmummified shadow cells (ghost cells)

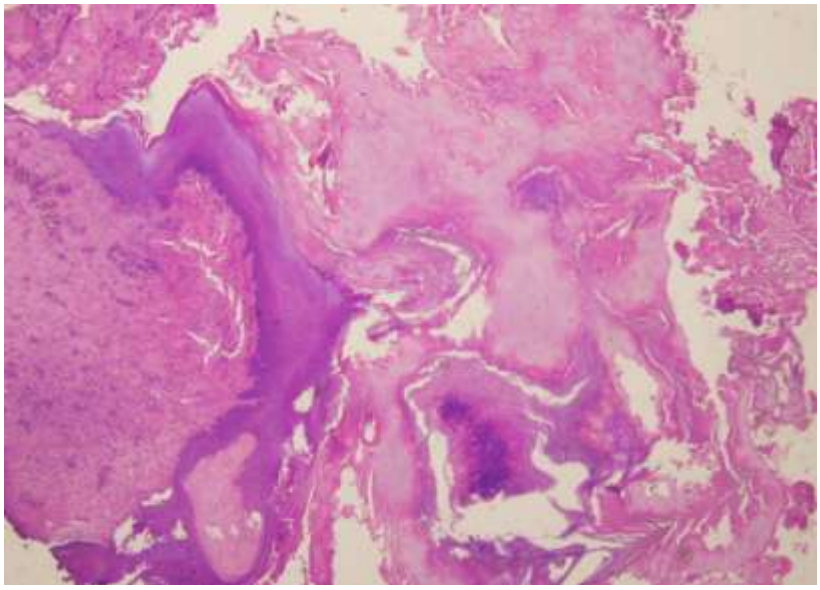

Figure 1:- Low power view of pilomatrixoma(HE staining x 20).

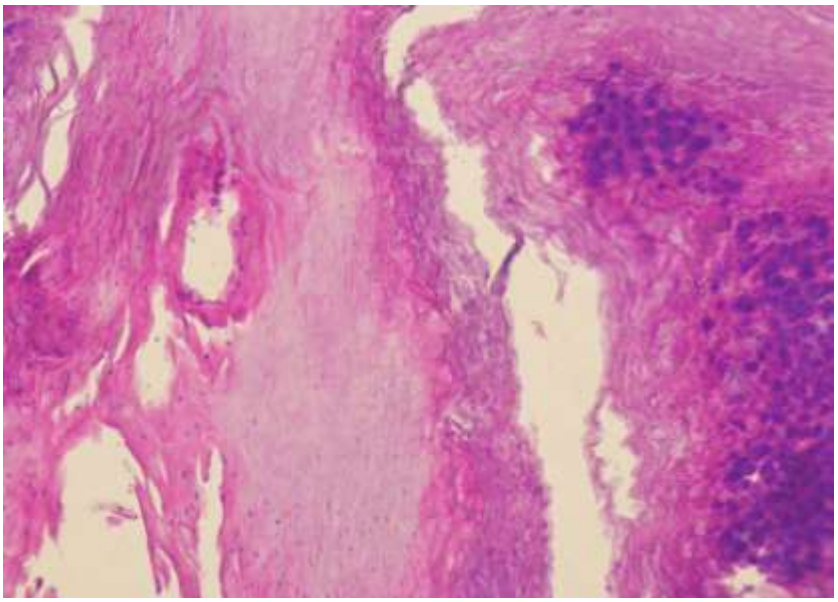

Figure 2:- Intermediate power view showing nucleated basophil cells surrounding ghost cells. Characteristic of pilomatrixoma. (HE staining x40).

Discussion:-

Pilomatrixoma commonly is a single, nodular, painless mass, adhered to the underlying skin but not extending into the deeper planes [5]. Based on appearance, a pilomatrixoma lesion can be classified into five clinical types: protruding mass, pigmented, mixed type, ulcerated, and keloid-like. The mass type is a predominant presentation in the available literature [6]. Four stages of the histomorphological pattern were explained by Kaddu et al. as early, fully developed, early regressive, and late regressive stages [7].

The beta-catenin gene (CTNNB1), which plays a vital role in the development of hair follicles, is mutated in up to 75% of cases. The presence of proto-oncogene B-cell lymphoma antigen 2 (BCL2) has also been associated with pilomatrixoma. Multiple forms have been observed to be associated with familial conditions such as Gardner syndrome, myotonic dystrophy, sarcoidosis, Steinert'sdisease, or Turner syndrome [8]. The risk of malignancy is rare; however, locally aggressive pilomatrix carcinoma with distant metastasis has been reported in studies before [9,10]. Differential diagnoses are sebaceous cyst, epidermal cyst, dermoid cyst, sebaceous adenoma or carcinoma, capillary hemangioma, squamous cell tumor, etc. [11].

Imaging modalities like ultrasound (USG), CT, and MRI scans have all been used to support the diagnosis. USG is acceptable enough in the majority of the cases with a positive predictive value of 95.5% since it is quick, cheap, and easily available. The definitive diagnosis of pilomatrixomais, however, made by histopathological examination [6]. On histology, the tumor is well-defined, comprising of peripheral basaloid cells proliferation and centrally located structureless eosinophilic cells, which lack nuclei called shadow cells or ghost cells typical of trichilemmal keratinization. Areas of calcification can be seen within the region of shadow cells [12].

The treatment of pilomatrixoma is essentially complete surgical resection, with 5 to 10 mm of safety margins. With a well-performed surgical procedure, the rate of recurrence is low at around 1.5% [13]. Mohs micrographic surgery is now being used to ensure complete margin-free excision, especially in cases of suspected malignancy [14].

Conclusions:-

Pilomatrixoma of the scrotum is a very rare entity. It is often misdiagnosed due to high clinical polymorphism. An ultrasound examination of a suspicious scrotal mass should be done to support the preoperative diagnosis. Histopathological evidence is confirmatory and the treatment is complete surgical excision with extremely low chances of recurrence.

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