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A Case Report Of Abdominal Wall Pilomatricoma

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Introduction

Pilomatrixoma is a benign neoplasm. It originates from the matrix cells of hair follicles. It is also called as pilomatricoma or calcifying epithelioma of Malherbe. It is typically found in the upper extremities and head and neck region. It is rarely occurs elsewhere in the body.
Nearly 60% of tumours are found in individuals who are less than 20 years of age.
In literature the largest case series was done with 346 pilomatricomas of which 15.3 percent were observed in the upper extremity. It is rare to find pilomatricomas over the abdomen, and we present one such case.

Case Report

A 76 year old male patient came to the tertiary hospital with complaints of mass over the abdomen for the past 6 months. Patient also gives history of pain for the past 3 months, with no aggravating or relieving factors. He doesn't give history of rapid increase in the size of the swelling. He gives no history of trauma, weight loss, fatigue or chills. Patient is a known hypertensive for the past 40 years and a known diabetic for the past 6 years, on regular medications for both.
Patient also gives surgical history of coronary bypass done 12 years back and right sided carotid endarterectomy in 2020. Patient also gives history of allergy to sulpha drugs. On physical examination, he is a moderately built and well nourished male. A midline scar on the chest (post CABG) and a right carotid endarterectomy scars were noted and were healthy. A swelling of size 2 x 2 cms was found over the left lumbar region. The swelling was globular in shape, well defined, with smooth surface, erythematous and with no active discharge. On palpation, the swelling did not have local rise in temperature, non-tender and mobile. Patient underwent excision and biopsy under local anesthesia. The specimen was sent for histopathology, reported as a - epidermis with focal pseudoepithalomatous hyperplasia, an underlying well defined dermal neoplasm composed of large islands and sheets of basaloid cells with scanty cytoplasm, with abrupt transition to ghost cells enclosing sheets of keratin. The intervening fibrocollagemnous stroma shows lymphocytic collections admixed with histiocytes and giant cells focally forming vague granulomas. Thus confirming, Benign Skin Adnexal Tumour-Pilomatricoma

Discussion

Pilomatricoma is a benign appendageal tumour. It arises from hair follicle matrix cells. It is also known as calcifying epithelioma of Malherbe. It is generally considered a rare tumour but it's occurrence has increased in recent times. Its known to occur more commonly in children than in adults.

The lesion was initially described by Malherbe and Chenantais in the year 1880. They identified it as a calcifying epithelioma although it was said to arise from sebaceous glands.9 The term pilomatrixoma was coined in a journal by Forbis and Helwig in 1961 in order to indicate the histological source.

In a study consisting of 10 pilomatricoma lesions, the results of immunostaining were strongly positive for BCL2 among all the patients. BCL2 is a proto-oncogene. It plays a role in the suppression of apoptosis in both benign and malignant tumours. This data suggests that faulty suppression of apoptosis is a contributory factor in the pathogeneis. Recently researchers have proved that the proliferating cells of pilomatricomas show distinct staining with antibodies that directed against LEF-1. LEF-1 is a marker for hair matrix cells. There is evidence to indicates that the S100 proteins, which is a family of cytosolic proteins that bind to calcium, and have a wide range of both intracellular and extracellular functions by regulating not only calcium balance, but also, cellular apoptosis, cellular migration, cellular proliferation and differentiation, energy metabolism and inflammation; can be used as a biomarker for the diagnosis of pilomatrixoma.

These data provide biochemical of morphological evidence that these tumours are derived from hair matrix cells. Moreover, researchers have demonstrated that at least 75% of people with pilomatricoma are associated with a mutation in the gene CTNNB1. These data directly indicate that beta-catenin/LEF misregulation as the major cause for hair matrix tumorigenesis in humans beings.

These lesions are more commonly found in head and neck. They are also found in upper limbs. These lesions have a predilection for children, young adults and women. The fact that this was found in an elderly male over the abdomen makes it a rare presentation.

Its malignant variant pilomatrix carcinoma is rare. In literature there have been no more than 90 cases that were documented. When it is present, its locally aggressive and known to recur. Metastasis has been documented. The key differentiating characteristics of the malignant variant are a high mitotic rate with atypical mitosis, central necrosis, skin and soft tissue infiltration, blood vessel and lymphatic infiltration.

Pilomatricoma is commonly presented as a solitary, firm subcutaneous nodule. It is a slowly progressive tumour that might grow over a period of months or years as it was in this case. Some patients may complain of pain, that is common during periods of inflammation or ulceration.
In a series consisting of 346 cases, 32 % of patients reported pain and tenderness.
In most cases, the skin overlying the swelling is normal in colour and texture. But sometimes the patient may present with tent sign, in which, there is flattening of the complete surface or only a portion of the tumour with an angulation, so that it represents a side of the tent. This is often noticed only when the skin is stretched. This occurs because the tumour is attached to the overlying epidermis and is pathognomonic sign. When pressing on one edger of the skin the opposite edge protrudes from the skin ans this sign is called teeter-totter sign. It is associated with bluish or reddish discolouration that occurs because of neo-vascularization. Pilomatricomas are commonly solitary tumours and multiple pilomatricomas are usually associated with certain genetic disorders. These are. Gardner syndrome, xeroderma pigmentosum, myotonic dystrophy and basal nevus syndrome.

Pilomatricomas are also commonly misdiagnosed and the confirmation is usually done on histopathology.
Wells et al discovered that in a series of 51 histologically proven pilomatricomas, 94 % of cases were incorrectly diagnosed. It was also proven that 57% of preoperative diagnosis was incorrect.
In the latest series consisting of 346 cases of pilomatricomas, it was discovered that only 28.9% of cases had a preoperative diagnosis that was consistent with the pathological diagnosis. Kumaran et al., in a retrospective review consisting of 78 excised pilomatricomas, 46% had accurate preoperative clinical diagnosis.
Preoperative diagnosis that is incorrect diagnosed includes- sebaceous cysts, dermoid cysts, epidermoid cysts, foreign bodies, masses that are unidentified, nonspecified cysts.

The histopathological feature of pilomatricoma is that of a well defined tumour that is most often surrounded by a capsule of connective tissue. Mostly it is located in the subcutaneous or dermal layer. It is made up of islands of epithelial cells.These islands of epithelial cells are made up of varying degrees of uniform basaloid matrical cells and it also shows cystic changes. As the tumour matures, the basaloid cells show central degeneration, which due to central unstained areas is characterised by presence of anucleated ghost or shadow cells. Other characteristic findings include foreign body giant cells, keratin debris and central calcification. 70 to 85 % of cases have been found to have calcification.

Since the tumour is small, superficial and well-circumcised, diagnostic imaging is usually not done. In MRI a non-enhancing lesion with small areas of signal dropout is usually seen which is consistent with calcification.
CT may show a subcutaneous lesion that is sharply demarcated and has soft tissue density, plus or minus calcification.
MRI may show a rim-enhancing lesion that has minor areas of signal dropout that is signifies calcifications. Ultrasound shows a mass that is well-defined consisting of central foci that is echogenic and a peripheral rim that is hypoechoic or a mass that is entirely echogenic that has a strong acoustic shadow in the subcutaneous layer.

Wang et al. did a review of multiple cases reports and series and noted that fine needle aspiration cytology incorrectly diagnosed 45% of cases of pilomatricoma.
The two sepecific findings for pilomatricoma are basaloid cells and ghost cells, which are quite precisely diagnosed by fine needle aspiration cytology.
The management of pilomatricoma is excision and biopsy. The rate of recurrence is low, from the range of 0 to 3%.

Conclusion

Pilomatricomas more commonly occur in the upper limb and, head and neck region. They also have a predilection for children, women and young adults. We present a case where pilomatricoma was diagnosed in an elderly 79 year old male patient over the anterior abdominal wall.

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