Article published online: 2022-11-28

Giant Sternal Pilomatricoma: An Unusual Tumor at an Uncommon Site

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J Health Allied Sci[NU] 2023;13:440–442.

Abstract

Pilomatricoma, pilomatrixoma, or calcified epithelioma of Malherbe is an uncommon, benign, isolated tumor of hair matrix primarily seen in children and young adults. The most common location is the head and neck region. It usually manifests as a small nodular mass. Here we report a rare case of giant pilomatricoma over the sternum in a 60-year-old male patient with an unusual clinical and radiological presentation mimicking a calcified sebaceous cyst.

Keywords
- pilomatricoma
- sebaceous cyst
- ghost cell
- tumor
- histopathology

Introduction

Pilomatricoma is an unusual skin adnexal tumor that arises from a hair matrix derived from the epidermal primitive basal cells, first described in 1880 by Malherbe and Chenantais.[1] They manifest as small, firm asymptomatic nodular masses with a good prognosis. It occurs in the head (periauricular, periorbital, frontal, and temporal) and neck region.[2,3] But some unusual locations have also been documented in the literature: arm, upper back, cymba conchae(ear), thigh, breast, sternum, and lower leg.[1–5] Usual lesions are less than 3 cm in size. According to the literature, pilomatricoma with the most significant size documented was 34×21 cm.[6] They are usually solitary, but multiple lesions associated with Turner's syndrome and myotonic dystrophy can be seen. Here we are reporting a rare case of giant pilomatricoma of the chest wall, causing a diagnostic dilemma.

Case Report

A 60-year-old male patient presented with swelling over the sternum, gradually increasing in size for 1 year. There has been a sudden increase in the size in the last 2 months (►**Fig. 1A**). The swelling was polypoid with an ulcerated surface and firm to hard in consistency. Clinically the diagnosis of the calcified sebaceous cyst was made. The MRI showed a well-defined subcutaneous lesion with thin internal septations. Findings were in favor of benign epidermoid cyst with secondary infection. Chest wall involvement was not seen. Hence, wide excision was done, and the mass was $9 \times 7.5 \times 6$ cm in size with a cut surface showing a gray–white lesion with hemorrhage (►**Fig. 1B**). Microscopy showed the dermis with a large well-circumscribed neoplasm composed of islands of peripheral basaloid cells, abruptly transitioning into squamoid cells. Focal shadow cells and calcification were also seen (►**Fig. 2A**, **2B**, **2C**). Surrounding the epithelial islands, there was a mixed inflammatory infiltrate composed of histiocytes, foreign body giant cells (►**Fig. 2D**), lymphocytes, and plasma cells. No atypia or malignancy was noted. Histological features suggested benign pilomatricoma. The patient was followed up for 1 year. No evidence of recurrence or signs of malignancy was seen.

article published online
November 28, 2022

DOI https://doi.org/10.1055/s-0042-1758035.
ISSN 2582-4287.

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

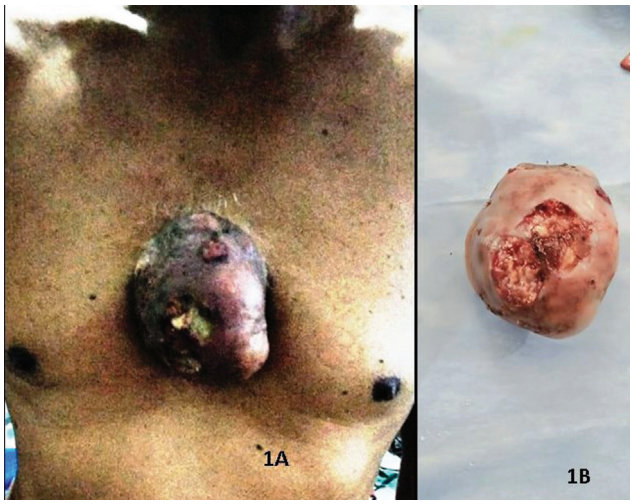

Fig. 1 (**A**) Clinical image showing polypoid growth with stalk over the sternum having an ulcerated surface. (**B**) Excised sternal mass with ulcerated surface.

Discussion

Pilomatricoma is a rare, benign hair matrix tumor. The clinical presentations have been noted from asymptomatic to painful tender nodules. A few lesions have presented as bulla, discoloration, erythematous lesion, giant lesion with ulceration, and intermittent bleeding.[1,2] These are slow-growing lesions but may show a sudden increase in size, as seen in our case. Many of them will have dystrophic calcification and present as hard calcified nodules. Due to varied clinical presentations, it is often misdiagnosed as benign cysts to tumors. Hence, the differential diagnosis of sebaceous cyst, dermoid cyst, retention cyst, abscess, benign vascular tumors, and adnexal tumors trichoepithelioma, trichoblastoma, basal cell carcinoma and trichoblastic carcinoma should considered.[1–5] Secondary features such as inflammation and soft tissue changes in a pilomatricoma mimic many mentioned lesions. Hence an imaging workup is useful with unusual presentations. The role of imaging modalities in such cases is controversial. MRI on the T1 image shows an isointense lesion in 67% of patients and hyperintense to hypointense in 76.2% in T2. Increased uptake of fluorodeoxyglucose is seen in the positron emission tomography scan. Calcification is detected in 81% by computed tomography, whereas USG identifies internal echogenic foci in most cases.[3]

Fine needle aspiration cytology can also be utilized to detect pilomatricoma. The typical central pale nuclear zone and the presence of shadow cells are distinct cytological features but may not be present in all cases. The background may show calcium deposits, multinucleated giant cells, basaloid, and squamous cells. Literature shows many pilomatricoma cases are diagnosed as epidermal cysts, giant cell-rich tumors, and fibrohistiocytic lesions.[5–7] Some cases may be confused with malignant lesions. This was due to high cellularity and primitive-looking cells. The cells have a high nuclear–cytoplasmic ratio and prominent nucleoli. Mitotic figures can be seen. Inflammatory background with the debris is noted, mimicking tumor necrosis.[8,9]

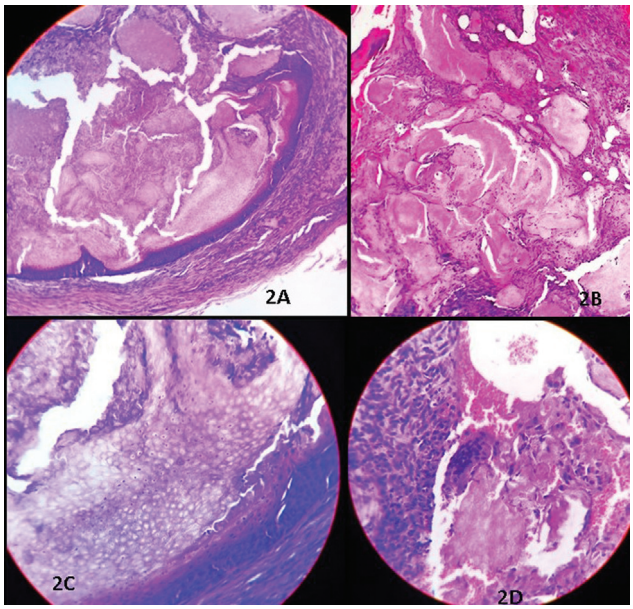

Fig. 2 (**A, B**) Histopathology showing lesion with islands of peripheral basaloid cells, abruptly transitioning into squamoid cells (H&E, X400). (**C**) Histopathology showing lesion with basaloid cells and ghost cells (H&E, X400). (**D**) Histopathology showing lesion with foreign body giant cell (H&E, X400).

Definite diagnosis is only by histopathology. It shows solid nests composed of basaloid cells and ghost cells. There will be abrupt keratinization with the typical feature of "ghost" or "shadow" cells. Calcification, ossification, and foreign body reactions are commonly seen. Pigmentation and extramedullary hematopoiesis can be seen in rare cases. Some cases show β-catenin gene mutation. It can be aggressive, indicating atypia, recurrence, and a tendency to invade. Malignant transformation (pilomatrix carcinoma) is rare and shows infiltrating border, atypical features, necrosis, and atypical mitosis.[10] In our case, histology showed classical features of pilomatricoma.

The treatment of choice for pilomatricoma is local excision.[11] Wide excision is done in suspected malignancy or history of recurrence or diagnostic dilemma, assuring safety margins. In our case, it was managed with wide excision. These tumors rarely recur unless with malignant transformation. In our case, no evidence or features of malignancy was detected after 1 year of follow-up.

Conclusion

A giant pilomatricoma is often misdiagnosed due to its size. There are very few documented cases occurring in the sternum. When accompanied by an unusual location and presentation like ours, it can be easily confused with any other subcutaneous lesion. Hence, a mandatory workup is needed before excision, keeping a rare lesion such as pilomatricoma as one of the differential diagnoses considering the chances of pilomatrical carcinoma.

Conflict of Interest
None declared.

References

1 Marzouki A, Chbani B, Bennani A, Lahrach K, Boutayeb F. Giant pilomatrixoma of the arm: an unusual presentation (a case report). J Saudi Soc Dermatol Surg 2013;17(01): 33–35

2 Lee SY, Ryu YJ, Her Y, Sung KY. Pilomatricoma: an unusual presentation. Ann Dermatol 2014;26(06):791–792

3 Inagaki A, Yamashita Y, Mori Y, Takeuchi E, Asaoka K, Murakami S. An unusual tumour of the ear: pilomatricoma in a middle-aged woman. Case Rep Otolaryngol 2019;2019:4975216

4 Marino MA, Ascenti G, Cardia R, Ieni A, Colonna MR. Pilomatrixoma of the right thigh: Sonographic-pathologic correlation in a young man. Radiol Case Rep 2019;15(03):230–233

5 DeRosa DC, Lin-Hurtubise K. Pilomatricoma: an unusual dermatologic neoplasm. Hawaii J Med Public Health 2012;71(10):282–286

6 Beattie G, Tai C, Pinar Karakas S, Cham E, Idowu O, Kim S. Colossal pilomatrixoma. Ann R Coll Surg Engl 2018;100(02):e38–e40

7 Bansal C, Handa U, Mohan H. Fine needle aspiration cytology of pilomatrixoma. J Cytol 2011;28(01):1–6

8 Thapliyal N, Joshi U, Vaibhav G, Sayana A, Srivastava AK, Jha RS. Pilomatricoma mimicking small round cell tumor on fine needle aspiration cytology: a case report. Acta Cytol 2008;52(05):627–630

9 Gupta M, Gupta V, Kumar R, Jhajj K. Pilomatrixoma: a diagnostic pitfall in fine-needle aspiration cytology. J Evol Med Dent Sci 2014;13(03):1691–1697

10 Bilings DS. Tumour and Tumour like conditions of the skin. Rosai and Ackerman's Surgical Pathology 11th ed. Philadelphia: Elsevier; 2018:67–68

11 Harsha MS. E RG, Jakkula S. Benign proliferative pilomatricoma over sternum: a rare case. Int Surg J 2018;5(07):2657–2659