Case report

Huge pilomatrixomas of the scalp: A case report

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\textbf{ABSTRACT}

Pilomatrixoma is a rare benign skin tumor differentiating toward hair matrix cells usually encountered in the head and neck region. It is most frequently appearing in the first and second decades of life. Histopathological examination is essential to make definitive diagnosis. Herein, we present an atypical case of multiple pilomatrixomas. A 69-year-old man with multiple voluminous masses over the scalp. Among the three lesions, one was clinically suspicious for malignancy, it measured 17 cm and was ulcerated in places. Histopathology confirmed the diagnosis of pilomatrixoma. The tumors were removed surgically with free margins. Otolaryngologist should be familiar with this benign tumor when evaluating soft-tissue mass in the head and neck region.

1. Introduction

Skin adnexal tumors are a heterogeneous group of neoplasms for which the diagnosis may be challenging. In an effort to simplify their classification, adnexal neoplasms into 3 groups: sebaceous, sweat gland-derived, and follicular [1].

Pilomatrixoma is a rare benign skin tumor differentiating toward hair matrix. It occurs as a firm nodule, most often on the face. Most occur in children and adolescents, but they can rarely occur in elderly patients [2]. We present a rare case of multiples pilomatrixomas in a 69-year-old patient. The clinical presentation, investigation and management are discussed.

This study has been reported in accordance with the SCARE criteria [3].

2. Case presentation

A sixty-nine year old male, farmer by profession, presented in our outpatient department with a 12-year history of multiple scalp masses. His past medical history was significant for diabetes mellitus. He stated that he did not consult beforehand, for fear of surgery.

Clinical examination revealed 3 subcutaneous bulky, rounded, firm masses of varying size on the parietal region. The largest one measured $17 \times 9$ cm and was ulcerated in places. Regional lymphadenopathy was absent (Fig. 1).

Head computed tomography (CT) showed 3 round, soft-tissue density lesions with calcifications.

They was no evidence of osteolysis (Fig. 2).

The initial differential included malignant causes of scalp masses such as basal cell carcinoma, squamous cell carcinoma, metastasis and malignant adnexal tumors.

The patient underwent an incisional biopsy of the mass under local anesthesia of the largest mass.

Microscopically it consisted of multiple circumscribed islands of ghost cells and basaloid cells. Areas of ossification were noted. The histopathological pattern was consistent with a pilomatrixoma. The patient was scheduled for surgery.

Under general anesthesia, the lesions were infiltrated with 2% lidocaine containing 1:10,000 norepinephrine. an elliptical incision was made over the parietal scalp masses. Skin was excised as the tumors were removed en bloc (Fig. 3). Using bipolar electrocautery adequate hemostasis was achieved. The whole surgical intervention was performed by a senior head and neck surgeon. The 2 small wounds were closed by direct suture without tension. The largest one was partially closed and daily dressing were performed until complete healing by secondary intention. (Fig. 4). Post-operative follow up was uneventful.

The patient was reevaluated at 3 months intervals by physical examination in our outpatient clinic. He showed no signs of recurrence.

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3. Discussion

Pilomatrixoma or pilomatricoma, also known as calcifying epithelioma of Malherbe is a benign skin tumor of the hair follicle [4]. This pathology was first described by Malherbe and Chenantais in 1880 [5].

Pilomatrixoma is a rare disease, studies report the incidence to be between 0.001% and 0.0031% of all dermatohistopathologic materials submitted for examination [6]. Its peak incidence is in the first and second decades, but it can occur at any age. It is slightly more common in women than men (ratio 1.15:1) [2].

The exact etiology and pathogenesis of pilomatrixoma are still unknown. Some studies have suggested that pilomatrixoma and pilomatrix carcinoma have mutations in the CTNNB1 gene, which encodes beta-catenin. This mutation leads to an upregulation of intracytoplasmic
Reactive inflammatory foreign body giant cells, signifying a granuloma. The transitional cells, are located between basaloid and ghost dead cells that show a central unstained area that corresponds to the lost densely staining nuclei with scant cytoplasm. The ghost cells, represent calcification and posterior acoustic shadowing located in the skin [2].

In the literature, ultrasound is the commonest imaging modality used for aid diagnosis. Pilomatrixomas appear as well-defined, ovoid, heterogeneous, hypoechoic masses, with internal echogenic foci; sign of calcification and posterior acoustic shadowing located in the skin [2]. CT scans can also be helpful in the diagnosis. Typical features of pilomatrixoma include a well-defined subcutaneous mass with mild to moderate enhancement and varying amounts of calcification [10]. This is consistent with our findings.

CT findings in three patients, J. Comput. Assist. Tomogr. 24 (2) (2000 Mar-Apr) 332-335, https://doi.org/10.1097/00004728-200003000-00028 (PMID: 10752903).

4. Conclusion

Pilomatrixoma is a rare benign skin neoplasm differentiating toward hair matrix cells. It is more common in children and young adults. Complete surgical excision with clear margins remains the treatment of choice. Multiple pilomatrixomas should raise the suspicion of an underlying etiology.

Declaration of competing interest

The authors of this article have no conflict or competing interests. All of the authors approved the final version of the manuscript.

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

References

[1] E.H. Fulton, J.R. Kaley, J.M. Gardiner, Skin adnexal tumors in plain language: a practical approach for the general surgical pathologist, Arch. Pathol. Lab. Med. 143 (7) (2019 Jul) 852-851, https://doi.org/10.5858/arpa.2018-0199-A4 (Epub 2019 Jan 14. PMID: 30638401).

[2] C.D. Jones, W. Ho, B.F. Robertson, E. Gunn, S. Morley, Pilomatrixoma: a comprehensive review of the literature, Am. J. Dermatopathol. 40 (9) (2018 Sep) 631-641, https://doi.org/10.1097/DAD.0000000000001118 (PMID: 30191102).

[3] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus surgical Case Report (SCARE) guidelines, Int. J. Surg. 84 (2020) 226-230.

[4] K.O. Attaad, N.A. Obaidat, D. Ghazarian, Skin adnexal neoplasms-part 1: an approach to tumours of the pilosebaceous unit, J. Clin. Pathol. 60 (2) (2007 Feb) 129-144, https://doi.org/10.1136/jcp.2006.040337 (Epub 2006 Aug 1. PMID: 16882096).

[5] N.J. Aberne, D.A. Fitzpatrick, D. Gibbons, et al., Pilomatrix carcinoma 315. Presenting as an extra axial mass: clinicopathological features, Diagn. Pathol. 3 (2008) 47.

[6] M.A. Marino, G. Ascenti, R. Cardia, A. Ieni, M.R. Colonna, Pilomatrixoma of the right thigh: Sonographic-pathologic correlation in a young man, Radiol. Case Rep. vol. 15 (3) (2019 Dec 25) 230-233, https://doi.org/10.1016/j.radcr.2019.11.007 (PMID: 32071653; PMCID: PMC7010961).

[7] N.C. Demirkhan, B. Dir, O. Erdem, et al., Immunohistochemical expression of beta-catenin, E-cadherin, cyclin D1 and c-myc in benign trichogenic tumors, J. Cutan. Pathol. 34 (2007) 467-473.

[8] J.L. Graham, C.F. Merwin, The tent sign of pilomatricoma, Cutis. 22 (1978) 577-580 [PubMed] [Google Scholar].

[9] A. Vance, W.H. Seitz Jr., Pilomatrixoma of the upper arm in an orthopaedic clinic, J. Shoulder Elb. Surg. 21 (8) (2012 Aug), https://doi.org/10.1016/j.jse.2012.01.005 (e12-5). (Epub 2012 May 8. PMID: 22572401).

[10] K.H. Lee, H.J. Kim, C.H. Suh, Pilomatrixoma in the head and neck: CT findings in three patients, J. Comput. Assist. Tomogr. 24 (2) (2000 Mar-Apr) 332-335, https://doi.org/10.1097/00004728-200003000-00028 (PMID: 10752903).

[11] T.K. Simon Cypel, V. Vijayasekaran, G.R. Somers, R.M. Zuker, Pilomatrixoma: experience of the hospital for sick children, Can. J. Plast. Surg. 15 (3) (2007 Fall) 159-161. PMID: 19554149; PMCID: PMC2687500.

[12] D. Kwon, K. Grekov, M. Krishnan, et al., Characteristics of pilomatricoma- omen in children: a review of 137 patients, Int. J. Pediatr. Otorhinolaryngol. 78 (2014) 1237-1341.

[13] S.F. Hassan, E. Stephens, S.C. Fallon, et al., Characterizing pilomatrixomas in children: a single institution experience, J. Pediatr. Surg. 48 (2013) 1551–1556.