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Case Report

Post-traumatic pilomatricoma presenting as rapidly enlarging pediatric scalp mass ☆

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ABSTRACT

Pilomatricomas (PM) are benign neoplasms that arise from hair follicle matrix cells. They are one of the most frequently excised pediatric skin masses. A typical clinical presentation is a slow growing, painless, firm, superficial mass in the head and neck of a child. We present a rare presentation of PM. A preschool aged girl presented with an enlarging scalp mass following a history of minor blunt trauma to the area. Two months prior, the child sustained a ground level fall and subsequently developed a painless "bump" at the site of injury on the scalp. The "bump" was initially the size of a "mosquito bite" and enlarged to the size of a "ping pong ball". Given the rapid progression and history of trauma, there was a broad differential diagnosis. An extensive workup including sonography, computed tomography, MRI, and biopsy were performed. The final pathologic diagnosis was confirmed as pilomatricoma. An atypical presentation of PM in a child can mimic a wide variety of pathology. Our case demonstrates unusual features of rapid enlargement and preceding trauma. With the widespread use of sonography for evaluation of superficial lesions, the radiologist may be the first to suspect the diagnosis of PM. Knowledge of the typical imaging findings in PM can be valuable, especially in more unusual cases.

Introduction

Pilomatricomas (also termed pilomatrixoma) are typically benign, superficial neoplasms of hair follicle matrix cells [1]. Pilomatricomas (PM) were first described by Malherbe in 1880 and were termed "calcifying epithelioma of Malherbe" [2]. The etiology of this tumor is unknown [3]. A majority of PM's present in the pediatric population, commonly in patients younger than 20 years old with a mean age of 8.9 years in 1 study [4]. A slight female predominance has been reported [1,3].

One study found that PM is the second most excised pediatric skin mass after epidermoid cyst, making up 10% of all superficial masses evaluated by pathologists [4,5]. PM's pre-

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dominantly occur in the head and neck as a solitary mass, but can occur in any hair follicle bearing skin surface [1,4]. Other common locations include the upper extremities and trunk. Multiple lesions are uncommon but are reported in 2%-10% of cases [3].

Physical exam typically demonstrates a solitary, slow-growing, painless, firm, oval, mobile mass. There may be an overlying reddish-blue discoloration [1,3,4]. PM's typically measure between 1-2 cm in the greatest dimension [3]. The majority range from 0.4 to 20.0 cm in size [1]. When PM exceeds 5.0 cm, they can be classified as a "Giant Pilomatricoma." [5].

Surgical excision of the lesion is almost always curative and confirms the diagnosis [1]. Local recurrence and complications are uncommon. Malignant PM's can occur and are termed pilomatrix carcinomas [3]. This entity is extremely rare and most often seen in adults in the sixth decade of life [4].

We present an atypical case of a 5-year-old female who sustained mild blunt trauma to the scalp. She then developed a rapidly enlarging superficial mass at the area of injury. The differential diagnosis was broad due to the presentation. After multimodal imaging, surgical excision with histopathologic examination yielded a final diagnosis of PM.

When the presentation is unusual and typical imaging findings of PM are present, the diagnosis may be able to be suspected preoperatively by the radiologist. As a result, it may be possible to reduce uncertainty and avoid diagnostic delay in similar clinical situations.

Case report

A 5-year-old female presented to clinic with a 2-month history of an enlarging, scalp mass at a previous injury site. The child sustained a witnessed ground level fall with minor blunt trauma to the scalp. Mother of child endorsed minor pain and focal swelling. One week later, the mother noticed a "mosquito-bite" sized, flesh-colored "bump" at the injury site, which continued to enlarge to the size of a "ping pong ball". The patient denied fever, pain, itching, drainage, vision, or hearing changes.

Physical examination at that time revealed a well-appearing young female with a 3.0 × 3.0 cm oval-shaped, circumscribed mass at the left frontotemporal scalp. The mass was erythematous, firm, nonmobile, and painless Figure 1.

Unenhanced computed tomography of the head was obtained to rule out traumatic etiology such as the rare possibility of an undiagnosed skull fracture and subsequent leptomeningeal cyst. Computed tomography demonstrated a circumscribed solid superficial mass with internal calcification. There was no underlying bony involvement, osseous destruction, or fracture. No fat stranding or surrounding inflammatory changes were present Figure 2. Ultrasound demonstrated a heterogeneous solid mass with a peripheral hypoechoic rim and central Doppler flow Figure 3.

Magnetic resonance imaging (MRI) showed a heterogenous mass with patchy and reticulated foci of increased signal on T2 weighted sequences. The mass demonstrated both peripheral enhancement and irregular internal reticular enhancement. A few internal foci of susceptibility-weighted blooming artifact were present, suggesting calcification or blood product Figure 4. The mass did not restrict diffusion Figure 5.

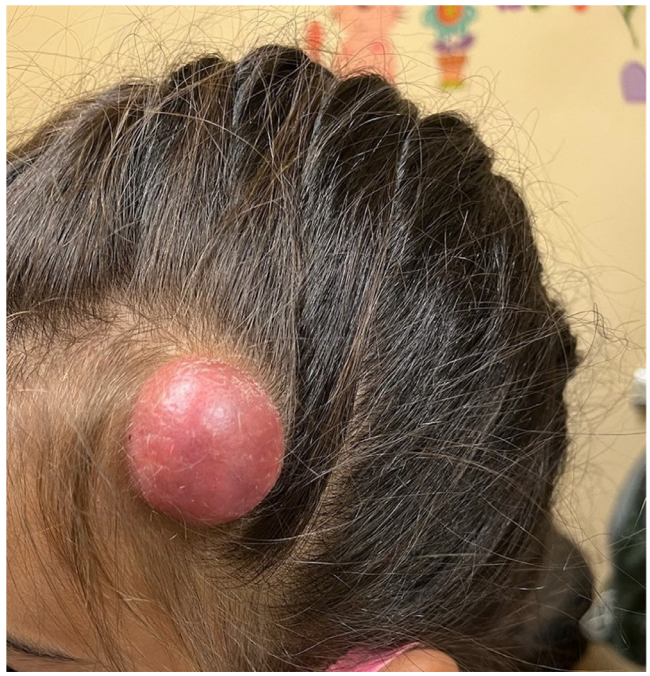

Fig. 1 – Clinical photo of left frontotemporal superficial mass on initial presentation, approximately 2 months postinjury. The mass was firm, painless, and mobile with an overlying reddish discoloration.

Ultimately, core needle biopsy demonstrated epithelium composed of follicular matrix cells and aggregates of shadow cells. These keratinized, anucleate, eosinophilic cells with distinct cell borders along with the presence of follicular matrix cells led to the diagnosis of a Pilomatricoma. Surgical excision by both pediatric and plastic surgery with final pathologic review confirmed the diagnosis. At her 3-week follow-up, the surgical site had healed as expected without complication.

Discussion

Our case describes a challenging clinical presentation of PM in a female child. Preceding trauma and rapid enlargement are unusual for PM and are likely to raise clinical concern for significant traumatic injury or malignant neoplasm.

In our case, there was a well-documented relationship between the patient's trauma and subsequent development of PM. A few cases in the literature describe a potentially similar relationship. One review described 21 patients with some preceding form of blunt force trauma prior to developing a PM at the site of injury [1]. Additional sources of trauma reported include insect bite, dog scratch, blunt head trauma, intramuscular injection, and vaccination [1,5–7]. Anetodermic Pilomatricoma, a rare variant of PM, is characterized by a bullous ap-

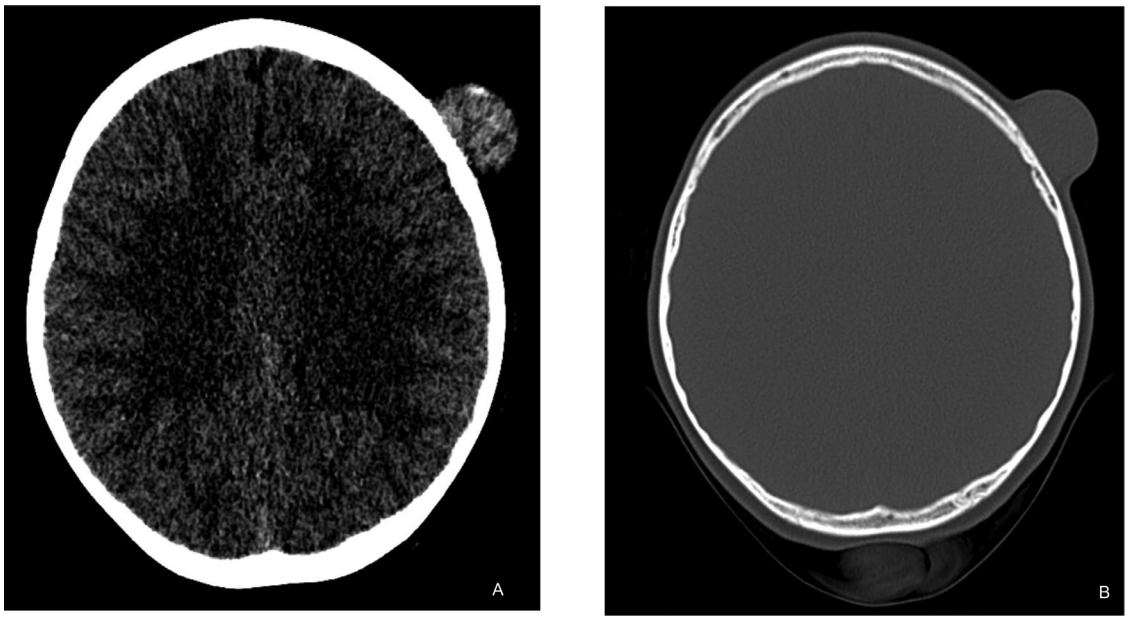

Fig. 2 – (A) Axial noncontrast CT image of the head demonstrates a marginated, heterogeneous, soft tissue density with a small peripheral focus of internal calcification and (B) There is no underlying fracture or osseous involvement. The mass is confined to the subcutaneous fat and skin.

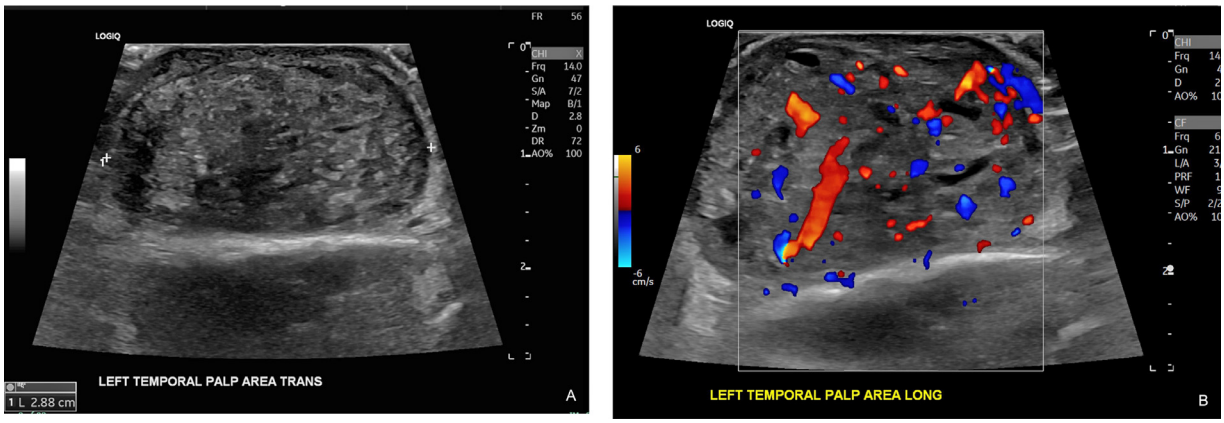

Fig. 3 – (A) Longitudinal grayscale sonographic image of the mass reveals a faint peripheral, hypoechoic rim with an internal heterogeneous appearance. There are a few internal hyperechoic foci with posterior shadowing, likely reflective of calcification and (B) Longitudinal Doppler image reveals prominent internal flow.

pearance and has a more definitive association with mechanical trauma [8]. While the role of trauma in PM development is ultimately unclear, further investigation may be warranted.

The rapid enlargement of the mass in our case was of significant clinical concern. Sudden enlargement of any mass can be concerning for a malignant neoplasm and is not typically present in PM [1,3,4]. A similar case in the literature describes an 8-year-old child with a rapidly growing shoulder mass, mimicking a malignancy. Multimodality imaging, tumor board evaluation, and biopsy were performed as part of the evaluation, which ultimately yielded a PM [9].

PM is a notoriously elusive preoperative diagnosis [1,3,4]. Prior studies have established that staff and pediatric fellowship trained radiologists are not consistently familiar with this entity, with the correct diagnosis being made in 12% [4] in 1 study and 15% in another [3]. The difficulty in diagnosing PM preoperatively is not exclusive to radiologists. The preoperative diagnosis for PM has been previously demonstrated to be less than 50% in multiple studies [4]. The lack of diagnostic certainty from both clinicians and radiologists can lead to additional invasive workup.

Although not always definitive, the typical imaging findings of PM may be helpful in reducing diagnostic uncertainty, especially in unusual clinical scenarios. The top differential considerations in the pediatrics population include inclusion cysts (both epidermal and dermoid), trichilemmal cysts, cal-

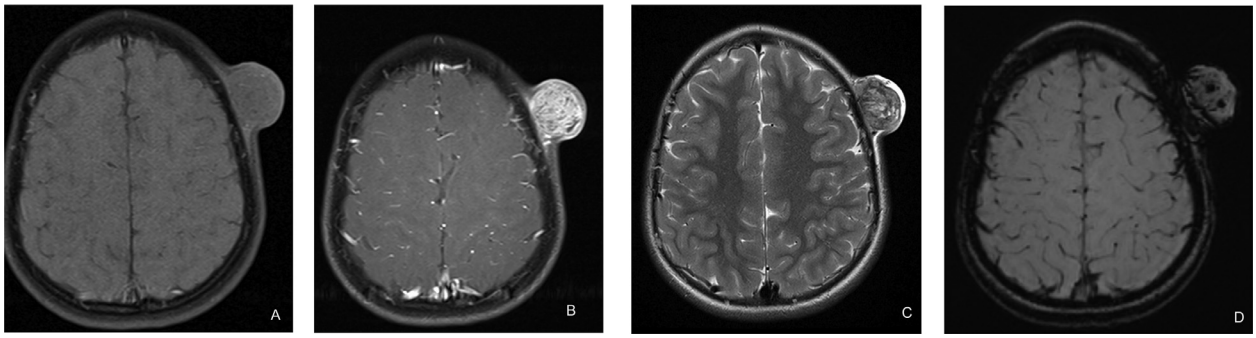

Fig. 4 – (A, B) Axial T1 Fat saturation pre and postcontrast sequences. The mass is uniformly T1 isointense with rim enhancement and internal patchy/reticular enhancement and (C) Axial T2 weighted sequence. The mass has a heterogeneous T2 signal with scattered internal areas of T2 hyperintensity (D) Axial susceptibility weighted sequence. Two central foci of susceptibility are present within the mass suggesting internal calcification or blood product.

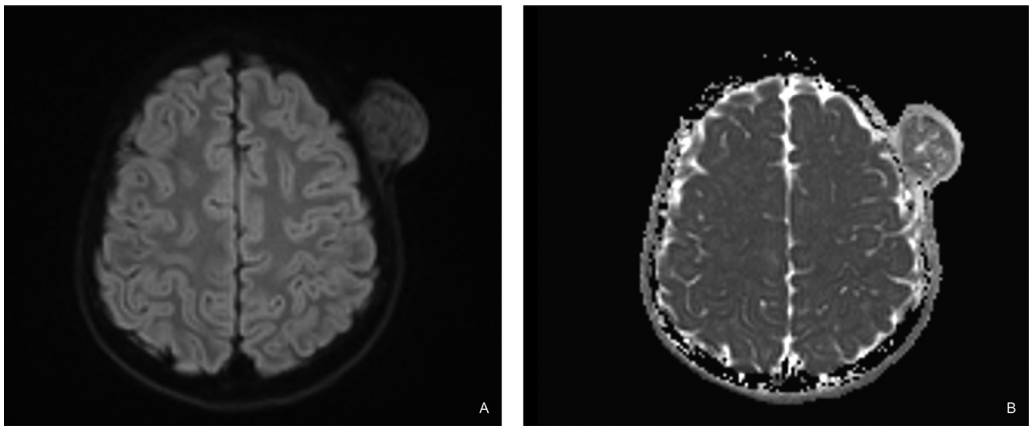

Fig. 5 – (A) and (B) Axial diffusion weighted image (DWI) with associated apparent diffusion coefficient (ADC) map. The mass does not demonstrate restricted diffusion.

cified lymphadenopathy, vascular tumors, and malignant soft tissue tumors, among others [3,4,10–12].

Although the imaging appearance of PM's can vary, the typical findings of a PM would be an ovoid, circumscribed, heterogeneous, noncompressible mass confined to the subcutaneous fat/skin with a connective tissue rim. The margins of the lesion are typically well preserved. Calcifications and internal vascularity are common but variably present. When identified, they can be potential distinguishing features. When MRI is obtained, PM's should demonstrate heterogeneous enhancement and no abnormal restricted diffusion.

A high frequency transducer should be utilized for sonographic evaluation [11,13]. It should be noted that the sonographic appearance of PM can vary, as demonstrated by the Solvetti classification system, which describes 5 distinct appearances of PM [10,13].

However, a common ultrasound finding reported is the presence of a peripheral hypoechoic rim [3,4,10]. One study demonstrates this finding to be present in 82% of cases [4], but other studies have suggested this finding is less common [10]. The hypoechoic rim is thought to correspond to the connective tissue capsule seen pathologically [4]. Internal reticulations are also commonly observed on ultrasound [4].

PM commonly demonstrates internal and/or peripheral vascularity, but it is not always present. US Doppler evaluation may not be feasible due to intralesional calcifications. When vascularity is present, the lesion may mimic an aggressive neoplasm. The presence of vascularity would argue against more common etiologies such as inclusion cysts or trichilemmal cysts, neither of which should demonstrate internal or peripheral vascularity unless inflamed [4,12].

Calcifications are another common finding in PM and can be identified on multiple modalities. Some studies show up to 80.6% of PM contain calcifications of some variety [4]. The degree of calcification can vary in PM with some lesions demonstrating no calcifications at all and others being completely calcified [4,13]. A completely calcified lesion may be specific and could clinch the diagnosis of PM [4,13].

The presence of calcifications could aid in distinguishing between PM from dermoid cysts, in which calcifications

are not typically observed [4]. Additionally, calcifications can help distinguish PM from trichilemmal cysts, where echogenic foci secondary to calcifications were reported in only 24% of cases and were described as more "lumplike" in nature [12]. Trichilemmal cysts are also typically less dense than their surrounding tissues and are characterized by posterior enhancement [12]. A calcified PM could potentially mimic a calcified lymph node. In these cases, the morphology of the lesion and identification of a fatty hilum is key [4].

Cross sectional imaging may be helpful by confirming that the mass is confined to the subcutaneous fat and is closely related to the dermis [4]. Fat stranding, edema, or inflammatory change would be unusual findings for PM and may suggest an alternative diagnosis or superimposed infection [4]. PM will have heterogeneous enhancement on MRI and should not restrict diffusion, distinguishing from epidermal inclusion cysts [4]. Fat would not be expected to be seen within a PM, potentially differentiating between a dermoid or a lymph node [4]. Finally, cross sectional imaging may be helpful when the clinical situation is unusual or traumatic etiologies are on the differential, as in our case.

The atypical presentation of PM can mimic aggressive pathology to include a malignant neoplasm or sequela of significant trauma. Our case identifies that preceding trauma and rapid enlargement can occur. Since these patients are likely to be evaluated with imaging, the radiologist may be best suited to initially suspect the diagnosis. Familiarity with the typical imaging findings and a high clinical suspicion are key for the radiologist to guide appropriate management.

Patient consent

Written informed consent to publish this case and the use of anonymized clinical and radiologic material was obtained from the patient's legal guardian.

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