



Pilomatrixomas presenting as vascular tumors on color Doppler ultrasound

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Received 8 June 2010; revised 6 July 2010; accepted 6 July 2010

Key words:

Pilomatrixoma;
Hemangioma;
Ultrasound;
Skin ultrasound;
Dermatology ultrasound;
Skin tumors ultrasound

Abstract Diagnosis of pilomatrixomas may be difficult because they can mimic other clinical conditions. Color Doppler ultrasound had been proven useful in the study of localized lesions of the skin and can both define lesion morphology and determine blood flow changes in real time, and may thus help differentiate primary from secondary vascular skin lesions. We present 3 cases of pilomatrixomas that mimic vascular lesions of the skin on physical examination. Clinical, sonographic, intraoperative, and histologic images are provided to highlight the nature of these challenging cases.

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Pilomatrixomas are cutaneous adnexal tumors, more common in the first 2 decades of life and in females. Sometimes, their diagnosis can be difficult and preoperative diagnostic accuracy in cases of pilomatrixoma is achieved in only 46% of the cases [1]. This entity may mimic other clinical conditions such as epidermal cysts, foreign body reactions, calcifications in lymph nodes, or fat necrosis [2].

Ultrasound had been useful for studying localized lesions of the skin [3]. On gray scale sonography, pilomatrixomas usually appear as target-shaped nodules with a well-defined hypoechoic rim and hyperechoic center. Scattered hyperechoic dots producing posterior acoustic shadowing that correspond

to calcium deposits may be found within the nodule nucleus [4,5]. The addition of color Doppler, with spectral curve analysis, to the usual gray scale ultrasound examination allows visualization of vascular characteristics (type and velocity of the flow), and defines its pathologic contribution [6]. Such anatomic information may be critical for determining the clinical management of localized lesions of the skin [7].

1. Setting and patients

A color Doppler ultrasound was performed in 3 patients presenting with atypical vascular lesions later confirmed as pilomatrixomas by histology. All ultrasound examinations were performed by the same physician at the Department of

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Radiology of Clinica Servet. The sonographic system consisted of a high-resolution Philips HDI 5000 ultrasound equipment (Bothell, WA) with a compact probe of variable frequency (from 7 to 15 MHz). The protocol includes sedation with chloral hydrate, in patient 4 years or younger, 30 minutes before the examination after informed consent is obtained from a parent or guardian. The study was approved by the Ethics Committee of Clinica Servet.

2. Cases

2.1. Case 1

A 2-year-old girl presented with a slow-growing, painless, erythematous, and firm swelling in the left ear lobe diagnosed as hemangioma. Further tumor growth over 3 months prompted her referral for color Doppler ultrasound examination. Sonographic imaging showed a nodular tumor of $1.6 \times 1.2 \times 1.0$ cm in the dermis and subcutaneous tissue of the pinna, with a hypoechoic rim, hyperechoic center, and multiple hyperechoic dots (suggestive of calcium deposits). Blood flow was increased within the tumor and at the periphery, with markedly enlarged vessels that represented

arteries with maximum peak systolic velocity of 26.4 cm/s. The tumor was resected, and histologic examination confirmed the diagnosis of pilomatrixoma (Fig. 1).

2.2. Case 2

A 1-year-old female infant with a 6-months history of a firm and painless mass in the right cheek associated with prominent telangiectasias was diagnosed as a hemangioma and treated with steroids. Because of a lack of therapeutic response, the patient was referred for color Doppler ultrasound examination. The imaging study showed a solid, $2.7 \times 2.7 \times 1.4$ cm mass occupying the dermis and subcutaneous tissue. The tumor had a hypoechoic rim and a hyperechoic center with multiple dots and posterior acoustic shadowing. Blood flow was increased in the surrounding dermis and superficial subcutaneous layers, with enlarged vessels representing arterial flow of maximum peak systolic velocity of 10.4 cm/s. Surgical excision was performed, and the histology confirmed the diagnosis of pilomatrixoma (Fig. 2).

2.3. Case 3

A 13-year-old girl presented with 6 months history of a rapidly growing mass under the skin of the dorsal spine. The

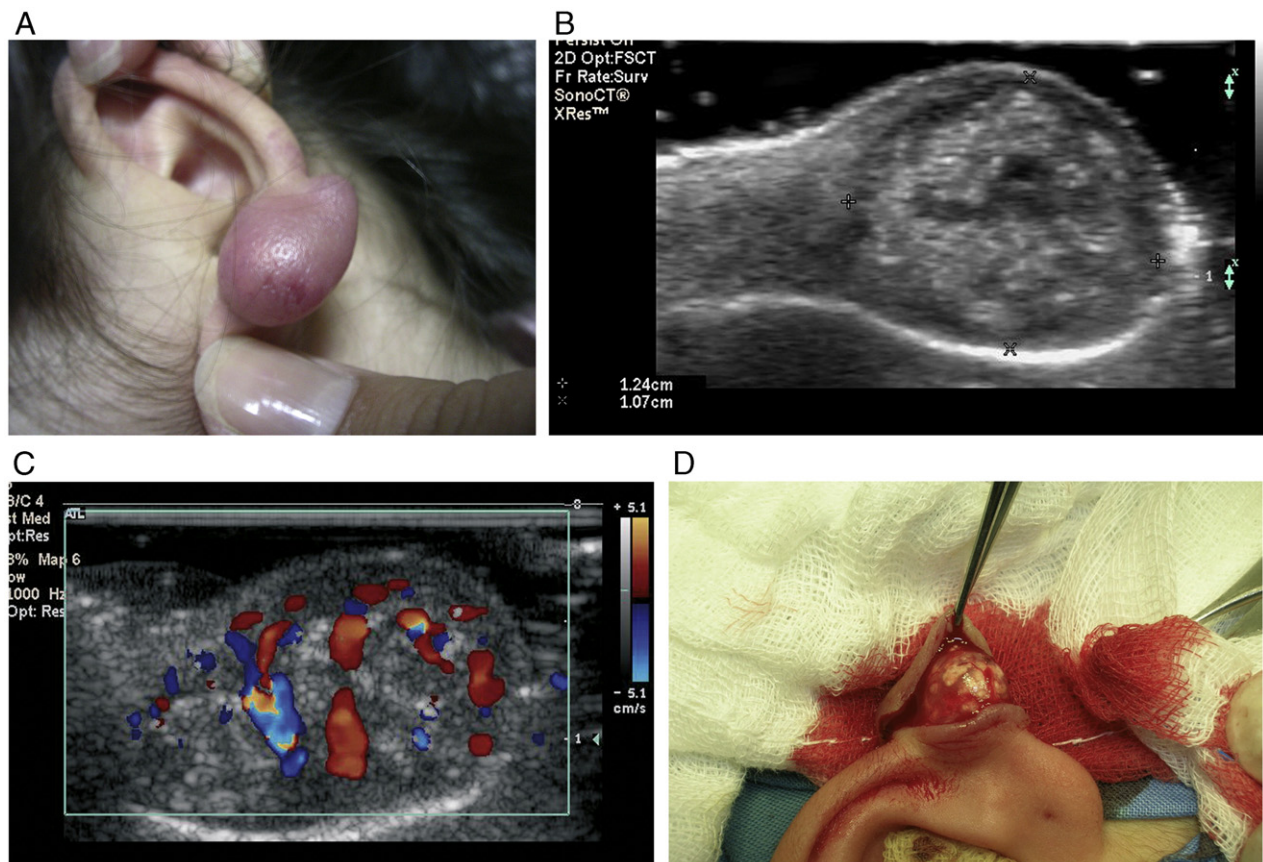


Fig. 1 Case 1. A, Erythematous swelling in the lobule of the left auricle. B, Ultrasound (longitudinal view): nodule with hypoechoic rim and hyperechoic center containing hyperechoic dots suggestive of calcium deposits. C, Color Doppler ultrasound (longitudinal view): increased blood flow within and around the nodule. D, The tumor at surgery.

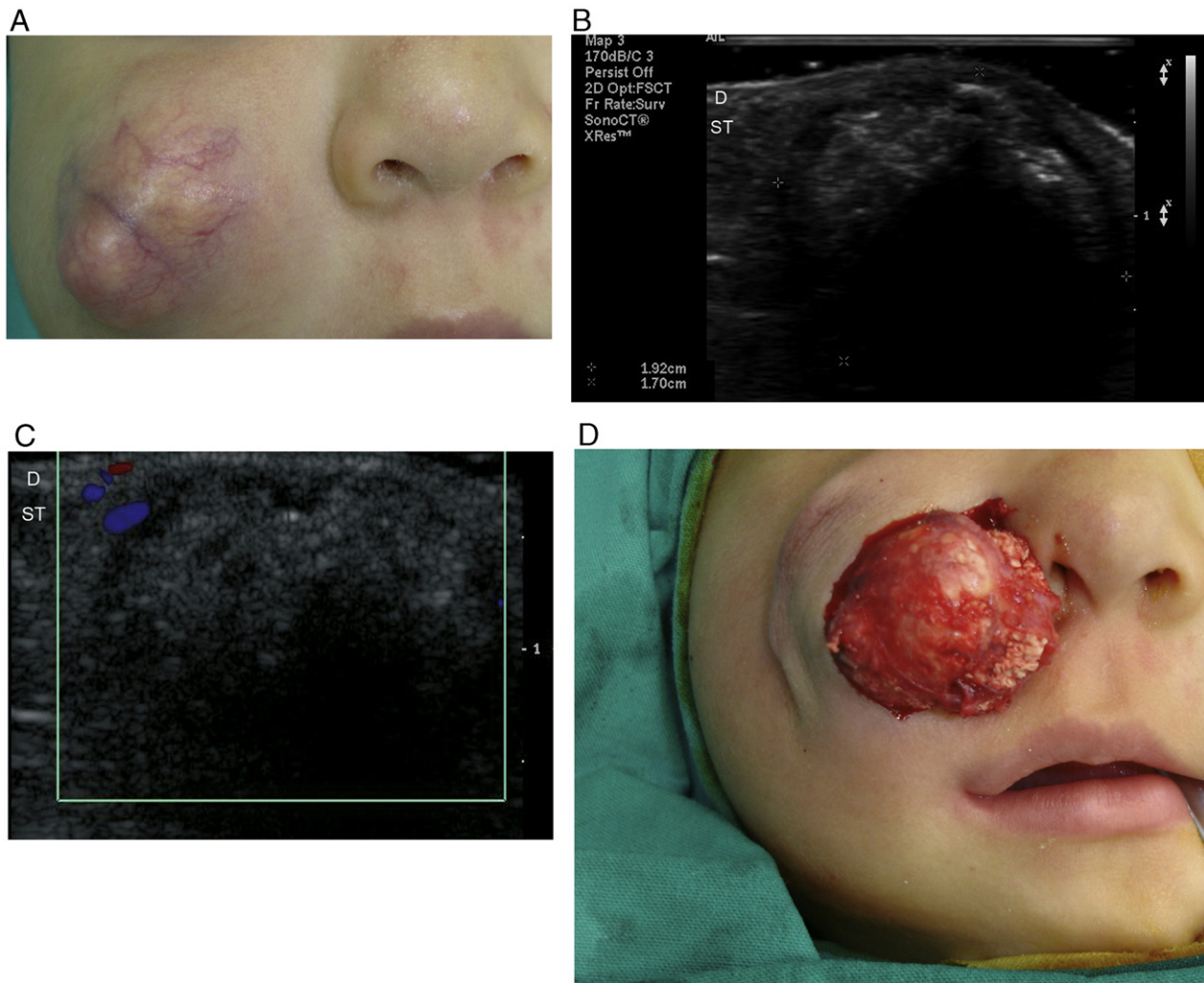


Fig. 2 Case 2. A, Mass in the right cheek. B, Ultrasound (transverse view): hyperechoic round solid lesion occupying the deeper dermis and subcutaneous tissue with hyperechoic dots suggestive of calcium deposits. C, Color Doppler ultrasound (transverse view): increased blood flow around the nodule. D, The tumor at surgery. D indicates dermis; ST, subcutaneous tissue.

tumor was clearly visible and tender on palpation. The skin was erythematous and slightly purplish, and a vascular tumor was suspected. Color Doppler sonogram showed a hollowed, $5.3 \times 5.4 \times 4.5$ -cm mass with an oval cystic cavity with thick walls. Deep within the tumor, a well-defined hypoechoic nodule ($1.7 \times 1.4 \times 1.3$ cm) was observed. Multiple septa tethered the nodule to the cavity walls, and sparse intranodular hyperechoic dots suggestive of calcium deposits were detected. Blood flow was increased throughout the nodule, with enlarged vessels representing arterial flow of maximum peak systolic velocity of 14 cm/s. Upon surgical resection, the tumor was histologically diagnosed as cystic pilomatixoma (Fig. 3).

3. Discussion

These atypical and challenging cases illustrate the importance of adding color Doppler to gray scale ultrasound to determine lesional blood flow and velocity and, if abnormal,

assess vessel location, size, and distribution to ascertain the true nature and anatomic characteristics of skin lesions. Furthermore, these cases prove that not all lesions that clinically seem to be vascular in nature (ie, hemangioma and vascular malformation) actually have a primary vascular origin.

With the combination of gray scale and color Doppler, it becomes possible to characterize blood flow patterns and isolate the vascular component, as illustrated by these very rare variants of pilomatixoma. Used alone, the gray scale ultrasound criteria for diagnosis of pilomatixomas (hypoechoic round solid lesions with hyperechoic dots suggestive of calcium deposits) would have likely missed the relevant anatomic data noted in the present cases (increased blood flow and/or cystic degeneration). The hyperechoic dots suggestive of calcium deposits are not pathognomonic for pilomatixomas having been also detected in epidermal cysts, basal cell carcinoma, and even hemangiomas [8]. Conversely, 15% to 30% of pilomatixoma cases do not show calcium on gray scale ultrasound [5].

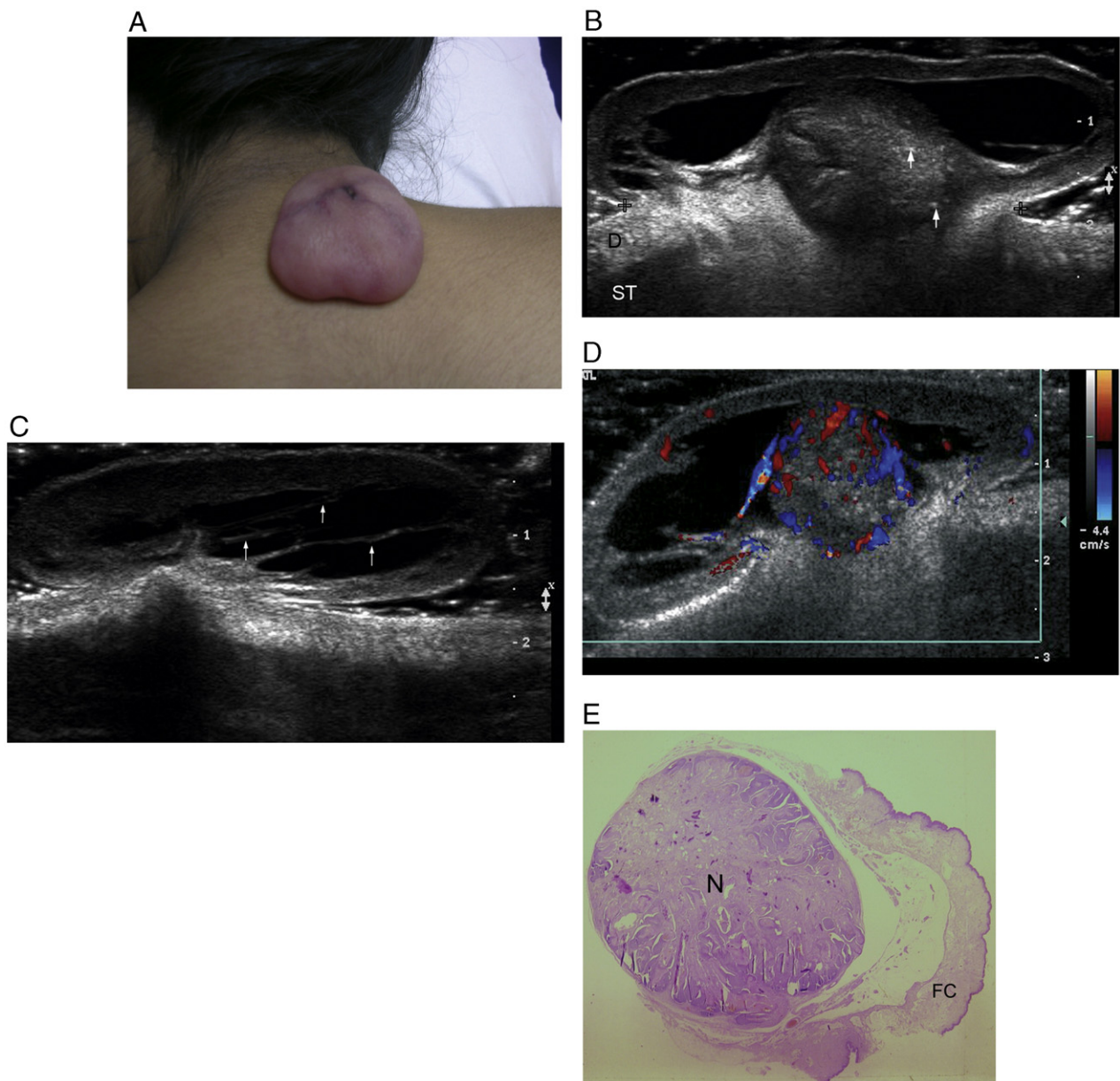


Fig. 3 Case 3. A, Erythematous swelling on the cervicothoracic spine. B, Ultrasound (transverse view): outwardly growing cystic structure with thick walls and attached hypoechoic dermal nodule; hyperechoic small dots may represent calcium deposits (arrows). C, Ultrasound (higher transverse view): thick septa coming off the cystic wall. D, Color Doppler ultrasound (transverse view): increased blood flow within the nodule. E, Photomicrograph of the surgical specimen matching the ultrasound image (hematoxylin and eosin, 2 \times ; courtesy Dr Ivo Sazunic). FC indicates fibrous cystic capsule; N, nodule.

The marked increase in blood flow on color Doppler ultrasound of pilomatrixomas differs from that of hemangiomas and vascular malformations. Pilomatrixomas are not accompanied of either venous blood flow or arteriovenous shunts that are common findings in hemangiomas [6,9]. Although the pattern of blood flow in pilomatrixoma has not been specifically described as abnormal, the present cases demonstrate that significant alterations of blood flow do occur and may actually represent a predominant manifestation. Further proof of its significance is the recorded peak systolic velocity of 24 cm/s in a pilomatrixoma vessel in one

of our cases. Because the peak systolic velocity for the posterior tibial artery is 16 ± 10 cm/s [10], it can be predicted that rupture of such a vessel could result in significant intraoperative bleeding. The pathogenesis of this hypervascularization has not been elucidated, but perhaps tumoral cell overexpression of vascular growth factors(s) and/or the corresponding receptors (s) could play a role, as has been suggested for other skin tumors [11].

Cystic variants of pilomatrixoma are rare and associated with the rapid growth suggestive of malignant tumors. It is possible that this particular presentation may be related to the

genesis of the tumor described as having infundibular-matrix cystic configuration in the early stages, and as calcified and ossified nodule without visible epithelial component in the end stage [12]. This progressive degeneration suggests episodes of microvascular bleeding that may have led to cyst formation.

Perhaps the reported low diagnostic accuracy in confirmed instances of pilomatixoma [2] can be explained by their characteristic involvement of deep cutaneous layers (dermis and subcutaneous tissue) that may make the tumors less accessible to clinical examination and its variants more likely to mimic other skin lesions.

Pilomatixomas may have a clinical pseudovascular presentation, but regardless of their appearance, color Doppler ultrasound is a useful diagnostic adjunct. Awareness of these atypical characteristics and early use of sonography may improve both diagnostic accuracy and cosmetic results, and impact on clinical management influencing the switch from a medical approach to surgical excision and thus shorten or prevent exposure of these children to the potential adverse effects of unnecessary drug administration (for e.g. steroids).

Acknowledgments

We are thankful to Drs Alicia Benavides, Carlos Misad and Filberto Guerra for their help with the histologic analysis of the biopsies.

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