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TEST YOURSELF: ANSWER

Slowly growing mass on the left shoulder

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Diagnosis: Pilomatricoma

Pilomatricomas are relatively rare tumors, with an estimated incidence of 1.04% of all benign skin tumors [1]. This likely plays a role in their frequent confusion with other benign and malignant tumors [2]. The most common locations are the head and neck and upper extremity, respectively [3]. They typically present as a superficial, hard, slow-growing masses [3]. The majority occur within the first two decades of life [4]. They may be covered by blue, red, or normal skin, and in rare cases can ulcerate [4]. These lesions can occasionally be painful secondary to mass effect [3], although pain is not a defining presenting symptom.

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Accurate diagnosis of pilomatricomas prior to surgical excision or biopsy occurs in less than 50% of cases based on published case series [3]. Several authors have estimated that physicians outside of the field of dermatology arrive at the correct diagnosis only 3% of the time [3]. The differential diagnosis includes epidermoid cyst, hemangioma, squamous cell carcinoma, dermatofibrosarcoma, and soft tissue sarcoma.

Pilomatricomas have previously described imaging findings that aid in accurate diagnosis. A key radiographic feature of pilomatricomas across all imaging modalities is their confinement to the skin and superficial soft tissue and lack of involvement of the deeper layers [5].

Plain radiographs may show a calcified mass with regular contours [6]. Calcifications have been reported as either having an homogenous sand-like appearance or large dense foci of calcification [6].

Ultrasonography has been recommended by some to be the preferred initial imaging test [5]. It shows a hypoechoic rim and extensive acoustic shadowing owing to the calcification of the tumor [5] (Fig. 1a). The presence of vascularity on color ultrasound has also been reported by several authors; however this feature is less ubiquitous and therefore of less diagnostic value [5]. The utility of Doppler evaluation is further undercut as many lesions are extensively calcified and not amenable to such evaluation [5].

Magnetic resonance imaging (MRI) can also be requested during the diagnostic evaluation. Its characteristics include an intermediate and homogeneous signal on T1-weighted imaging, and a contrast-enhancing hyperintense rim on T2-weighted imaging [4, 7]. Furthermore, on T2 fat-suppressed contrast-enhanced images, high signal bands can be seen radiating from the low signal center to the periphery [7] (Fig. 1b, c, d).

While imaging studies are an important adjunct during the diagnostic workup, tissue sampling is often necessary for accurate diagnosis. Histologically, the tumors are well circumscribed and are composed of islands of small baseloid epithelial cells that have a high nuclear-to-cytoplasmic ratio. These cells merge with larger "ghost" or "shadow" necrotic keratinocytes that are surrounded by keratin [8, 9]. The keratinous debris can become focally calcified and elicit a prominent mono and multinucleated histiocytic reaction (Fig. 2a, b, c).

Surgical management with wide or simple resection has been advocated although wide excision offers reduced recurrence rates [8, 10]. Recurrence rates have ranged from 1.5 to 3% [3], leading some to advocate for follow-up for at least 1 year [2]. Published cases of pilomatricoma of the upper extremity have documented positive results with surgical resection [4]. Some larger tumors have required soft tissue coverage with skin grafting [4].

Although generally considered a benign tumor, malignant variants do exist. With less than 150 cases reported in the English literature as of 2014, pilomatrix carcinoma is the exceedingly rare malignant form of a pilomatricoma [10]. These tumors have a similar anatomic distribution to their benign counterparts [10]. Tumor size and local recurrence are potential risk factors related to the risk of metastatic spread [10]. In cases of pilomatrix carcinoma, initial treatment with wide versus marginal excision appears to offer no benefit when considering the likelihood of metastasis [10].

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest.

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