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ONLINE CASE REPORT

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Colossal pilomatrixoma

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ABSTRACT

We describe the largest reported case of pilomatrixoma in the literature. While pilomatrixomas typically present as small soft-tissue nodules of the head, neck and upper extremities, they can also present as much larger masses in atypical locations. When they present in their usual size, pilomatrixomas have typical imaging features and can be correctly diagnosed with imaging studies before histological confirmation. Their clinical and imaging diagnosis become challenging when they are very large, as in our case. A giant pilomatrixoma should also be considered for paediatric patients presenting with a large subcutaneous soft-tissue mass.

KEYWORDS
Pilomatrixoma–Pediatric

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Introduction

Pilomatrixoma (pilomatricoma, epithelioma calcificans of Malherbe; Online Mendelian Inheritance in Man entry 132600) is a benign solid tumour derived from the hair follicle matrix. It was first described by Malherbe and Chenantais in 1880.[1] The current nomenclature was proposed by Forbis and Helwig in 1961.[2] It typically presents in the head, neck and upper extremities during the first two decades of life.[3] Lesions often range in size from 0.5 cm to 3.0 cm. When the lesion exceeds 5 cm in diameter, it is described as a giant pilomatrixoma. The largest previously described giant pilomatrixoma measured 24 cm × 21 cm × 9 cm in a 52-year-old man.[4] It is the second most common nodule encountered in the paediatric age group although it can be found in all ages, with a bimodal distribution in people under 30 years and in the sixth to seventh decade of life.[2,4] Pilomatrixoma is often a solitary lesion but multiple lesions can occur and may be associated with myotonic dystrophy, Turner's syndrome, trisomy 9 and Sotos syndrome.[3] This report describes a large pilomatrixoma resected from an otherwise healthy paediatric patient. It measured 34 cm × 21 cm × 17 cm.

Case history

A 17-year-old boy was initially evaluated in a surgery clinic for a large upper back mass (Fig 1). According to both mother and patient, a small mass had been present for 10 years. It had increased in size significantly over the preceding year and had developed intermittent bleeding according to the patient. The patient did not inform his parents about the increase in size of the mass over the years. Computed tomography was ordered, which showed a large pedunculated and centrally calcified soft-tissue mass arising from the right posterior back in proximity to the right trapezius muscle, with large feeding vessels (Fig 2). Imaging did not show deep bony or muscle invasion. The boy was

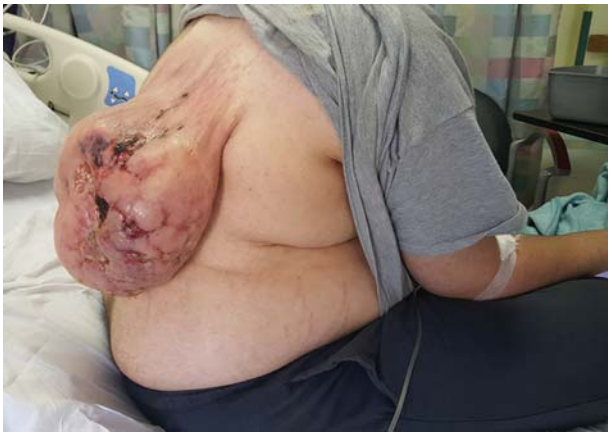

Figure 1 A large mass is noted on the right upper back of the patient

scheduled for an elective resection. However, he presented to the emergency department prior to elective operation with worsening haemorrhage from the surface of the mass. An examination of the haemorrhagic area showed necrotic skin breakdown and serous drainage. He was admitted for intravenous antibiotics and further preoperative magnetic resonance imaging (MRI). The scan showed a large exophytic pedunculated complex soft-tissue mass with central calcification. It appeared to extend into the trapezius muscle and large feeding vessels were noted arising from the axillary and paraspinal arteries with multiple vessels traversing through the mass (Fig 3).

The patient was taken to the operating room for resection. The mass and the overlying skin were completely excised. The tumour did not extend into the trapezius muscle. A primary multilayer closure was performed with Jackson-Pratt drain placement to prevent seroma formation. One week later, the wound was noted to be healing well and the surgical drain was removed. At his subsequent clinic visit, the wound had become infected and partially separated. A second operation was needed for wound debridement and closure.

Histological sections demonstrated a sharply circumscribed neoplasm composed of keratinised 'ghost cells' with focal basaloid cells with a high nuclear to cystoplasmic ratio (mitotically active without atypia) and no necrosis. The mass was surrounded by an extensive oedematous subcutaneous-type tissue with fibrosis, lobules of capillary proliferation, lymphatic dilatation and patchy lymphocytic inflammation (Fig 4). The mass weighed 3222 g (Fig 5).

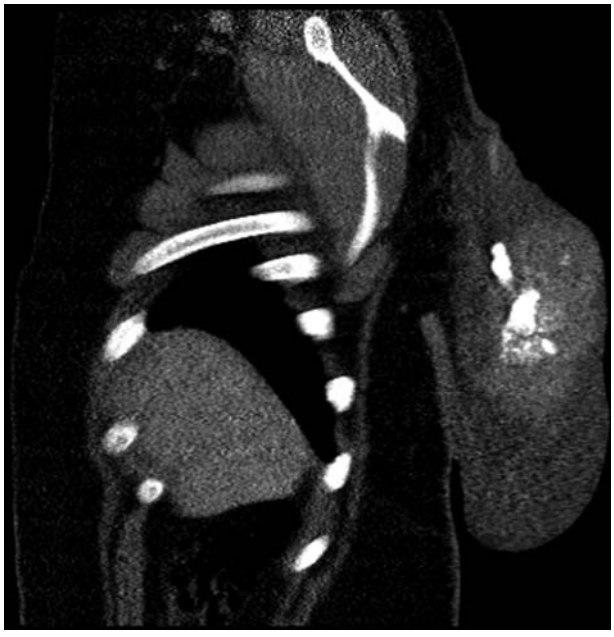

Figure 2 A sagittal computed tomographic view of the patient shows the back mass with internal calcification

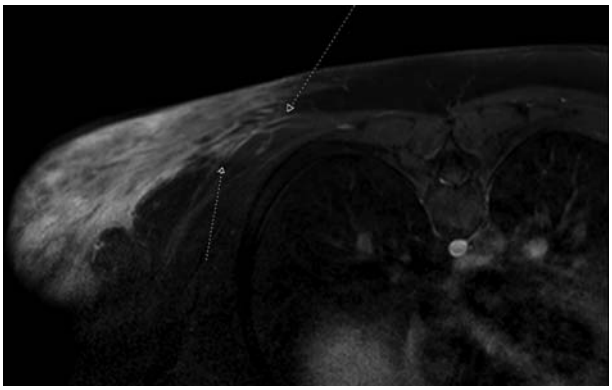

Figure 3 Magnetic resonance imaging shows the feeding vessels, indicated by the arrows, going into the back mass

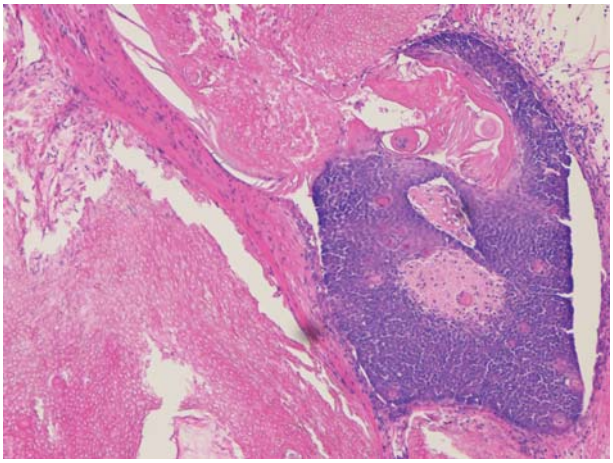

Figure 4 Haematoxylin and eosin stain slide of pilomatrixoma is shown (10 times)

Discussion

The case presented is the largest pilomatrixoma documented to date. Its size contributed to its diagnostic difficulty. Pilomatrixoma rarely exceed 3 cm and commonly affect the head and neck regions. It is unusual to present on the trunk. Interestingly, the largest reported prior case was also found on the posterior thorax.[4] Typically, it is a slow growing tumour but rapid expansion with haemorrhage has been described.[5] In our patient, the rapid growth was probably due to the expansion of the oedematous, extensive soft-tissue surrounding the pilomatrixoma. We suspect that the giant pilomatrixoma triggered a pronounced reactive inflammatory process, as supported histopathologically by the presence of dense perilesional inflammatory hyperplastic soft tissue changes. The subcutaneous location, central calcification, heterogeneous signal intensity and post-

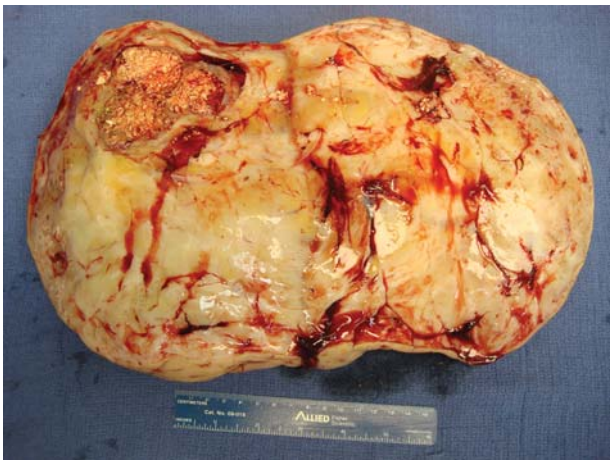

Figure 5 In this cross-section of the tumour, a calcified portion of the giant pilomatrixoma is seen in the upper left. The entire tumour is surrounded by extensive oedematous subcutaneous tissue. A 15-cm ruler is placed near the mass as a comparison

contrast enhancement of our patient's lesion were typical of a pilomatrixoma. The extensive soft tissue around the pilomatrixoma contributed to the total size of the mass and led to a clinical impression of soft tissue sarcoma. Differential diagnostic considerations are shown in Table 1.

Accurate diagnosis of pilomatrixoma can be made with imaging when they present with typical size and location. On ultrasound, the lesion appears as round or ovoid well-defined hyperechoic or heterogeneous solid nodules within the subcutaneous soft tissues. Demonstration of hypoechogenicity, heterogeneity, internal echogenic foci (calcifications) in scattered-dot pattern and hypoechoic rim with or without posterior shadowing is highly suspicious for pilomatrixoma with specificity and positive predictability of 95% and 92%, respectively.[6] Recent advent of high-frequency transducers (12–15 MHz) has allowed employment of real-time spatial compound ultrasound imaging (RTSCI) in examination of the skin and subcutaneous tissue lesions. RTSCI is a method of combining several overlapping scans of an object from different view angles into a compound image thus minimizing artefact.[7] Dermoscopy (examination of the skin using skin surface microscopy) is an additional modality that can aid in the diagnosis of pilomatrixomas. The presence of irregular white structures and streaks with polymorphous vascular structures (reddish homogenous areas, hairpin vessels or linear-irregular vessels) are the most characteristic features of pilomatrixoma on dermoscopy.[8]

Histopathology provides the definitive diagnosis for pilomatrixoma. Pilomatrixomas are located in the dermis or subcutaneous fat layer and are generally well circumscribed and multilobulated. They begin as an infundibular matrix cyst with maturation into a calcified and ossified dermal nodule containing two characteristic cell types – peripheral basaloid cells and central eosinophilic shadow cells (enucleated basaloid cells filled with eosinophilic keratin, also known as ghost cells). Calcification predominates within the ghost cell milieu. Inflammatory infiltrate with multinucleated giant cells, dystrophic ossification and melanin pigmentation are also often present.[3,7,8]

Table 1 Differential diagnoses for soft-tissue masses.

Benign	Malignant
Congenital haemangioma*	Basal cell carcinoma
Dermatofibroma	Cutaneous lymphoma
Epidermoid cyst	Cutaneous metastases
Leiomyoma	Dermatofibrosarcoma protuberans*
Lipoma*	Leiomyosarcoma*
Myofibroblastic tumour*	Malignant histiocytoma
Plexiform neurofibroma*	Malignant melanoma
Pilomatrixoma	Synovial cell sarcoma
	Squamous cell carcinoma

*Considered for this case based on radiographic features

Treatment of choice for pilomatrixomas is surgical excision with histologic confirmation. As this case demonstrates, pilomatrixomas at times can pose a diagnostic challenge; that is, when large size and extensive inflammatory hyperplastic soft-tissue changes are present. In such instances, adjunct imaging modalities such as ultrasound, dermoscopy, computed tomography or MRI may aid in preoperative diagnosis and surgical planning.

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