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PHOTOLETTER TO THE EDITOR

# Pigmented dermatofibrosarcoma protuberans in a 4-year-old girl and ultrasonographic findings

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## Abstract

Dermatofibrosarcoma protuberans (DFSP) in children is often clinically misdiagnosed as hemangioma or vascular malformation. Ultrasonography and color Doppler imaging are useful noninvasive tools for the diagnosis of skin tumors and may help distinguish DFSP from other vascular skin lesions in children. (*J Dermatol Case Rep*. 2015; 9(2): 52-54)

### Key words:

pigmented dermatofibrosarcoma protuberans, child, nodule, skin cancer, ultrasonography

## Case

DFSP is a mesenchymal tumor of intermediate-grade malignant potential. Typically it presents during early or middle adult life.[1,2] Pigmented DFSP (Bednar tumor) is a rare neoplasm, present in 1% to 5% of all cases of DFSP. Histopathologically, it is characterized as a variant of DFSP with melanin-containing cells.[3] However, the reports of DFSP in children are increasing recently. Reis-Filho JS *et al* described that pediatric pigmented DFSP may account for up to 5% of all pigmented DFSP.[3] DFSP in children is sometimes diagnosed as infantile hemangioma or vascular malformation. We describe here a case of Bednar tumor, in which ultrasonography and color Doppler imaging were helpful in the diagnosis of the tumor.

A 4-year-old girl presented with a 33 mm x 27 mm, purple-erythematous, soft, hemispherical nodule in the right buttock, surrounded by scattered, slightly red-purple macules (Fig. 1). Based on the

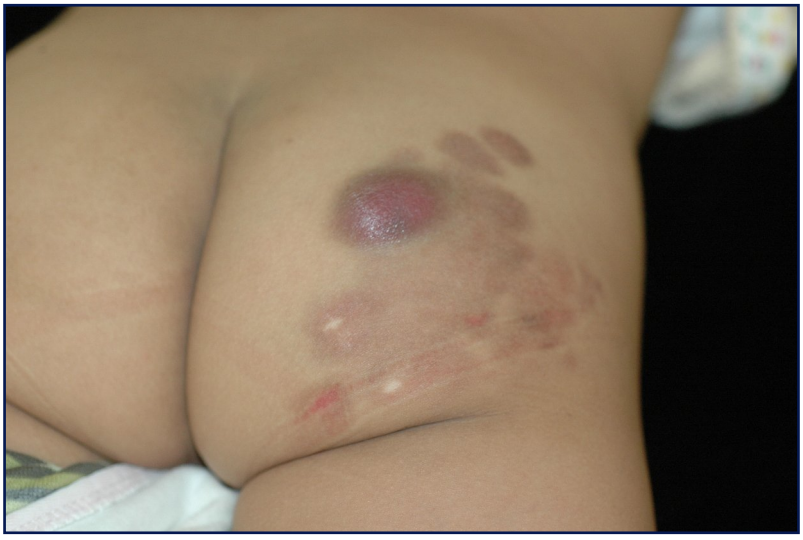

**Figure 1**

*Clinical appearance. The tumor was a 33 mm x 27 mm, purple-erythematous, hemispherical nodule on the right hip, which was soft and did not adhere to the basal tissue. Around it, slightly red-purple plaques were a scattered.*

age of the patient, hemangioma or arteriovenous malformation (AVM) were initially diagnosed. Ultrasonography showed a hypoechoic nodule in the subcutaneous layer with partial hyperechoic area (Fig. 2A). The tumor was partially invasive into the adipose tissue. Color Doppler imaging revealed increased internal vascularity without direct communication between the arterial and venous systems. (Fig. 2B). These findings suggested a malignant tumor rather than a vascular tumor.

An excisional biopsy was performed. Histopathology of the specimen demonstrated a neoplasm of spindle cell proliferation having a storiform pattern. The tumor occupied the area between the upper dermis, just beneath the epidermis and deep dermis. In the deep dermis, tumor cells spread along the connective tissue septa and partially infiltrated the subcutaneous fat in a honeycomb-like pattern (Fig. 2C,D). These spindle cells had elongated nuclei, and the ratio of mitosis was 2/10 HPF. Some melanin-containing cells were observed among the tumor cells (Fig. 2E). The spindle-shaped tumor cells were positive for CD34 and factor XIIIa, and negative for CD31, desmin, S-100, HMB45. The pigmented cells were positive for S-100 and HMB45. The tumor was diagnosed as a congenital Bednar tumor. The patient was treated with a wide deep excision having 1 cm margins. There was no evidence of recurrence 3 years after surgery.

## Discussion

Melanocytic nevi, infantile hemangioma, and AVM are relatively common congenital skin tumors. In contrast, malignant tumors of the skin are rare in children. In this case, because the patient was four years old and the tumor was a purple-erythematous nodule, initially hemangioma or AVM was diagnosed. However, ultrasonography demonstrated the irregular infiltration of the tumor into the adipose tissue with significant hypervascularity. It is difficult to diagnose only by findings of color Doppler ultrasonography, because increased intralesional vascularity is a common feature in both vascular and malignant tumors.[4] Some malignant tumors include necrotic areas with destruction of tumor vessels, and others show high vessel density. Therefore, the vascularity in

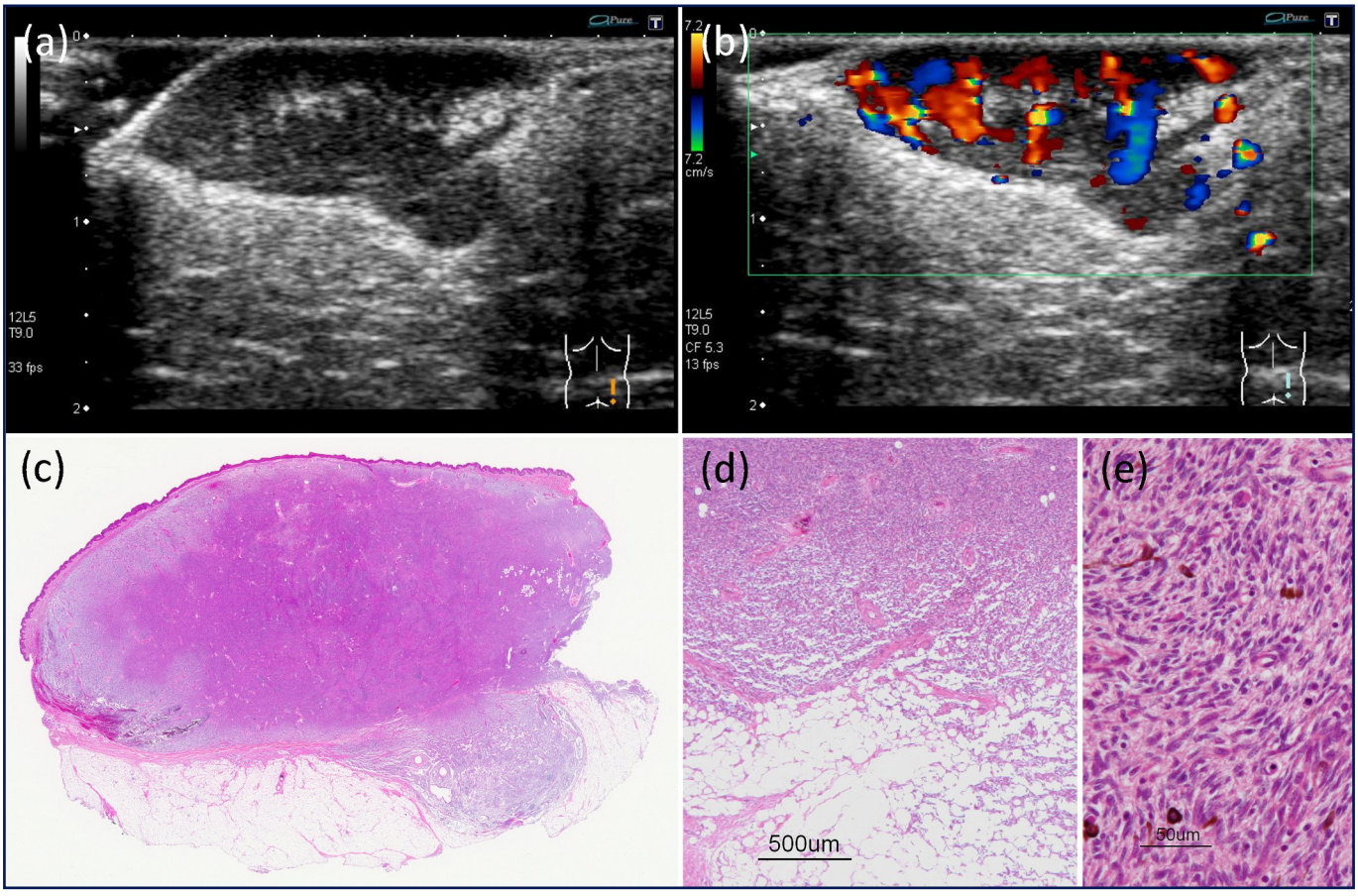


**Figure 2**

*(A-B) Ultrasound findings. (A) B-mode ultrasound showed a hypoechoic, partially mixed echoic mass lesion. The tumor was partially invading into the adipose tissue. (B) Increased internal vascularity was demonstrated.*
*(C-E) Histological findings. (C) The tumor was present between upper dermis, just beneath epidermis, and deep dermis. In the deep dermis, the tumor cells spread along the connective tissue septa, and partially infiltrated into the subcutaneous fat. The feature was similar to the ultrasound imaging, figure 2A. (D) The spindle cells extended into subcutaneous fat. (E) Several pigmented cells were observed among the spindle cells in a striform pattern.*

malignant tumors is various and not enough to clarify the differential diagnosis. The significant ultrasonographic features of a malignant tumor are infiltrated margin, scalloped shape, and size larger than 5 cm.[5] In this case, the ultrasonographic image was clearly the same shape as the microscopic feature of the excised tumor, with tumor cells infiltrating into the subcutaneous fat (Fig. 2A,C). It has also been reported that the internal echoes of DFSP are characteristically hypoechoic or mixed echogeneic and ultrasonographic findings are closely associated with the histological findings, as observed in this case. Ultrasonographic imaging is useful because it is minimally invasive and sedation is not necessary even in children. It may help distinguish DFSP from other benign tumor including hemangioma and AVM.

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