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TEST YOURSELF: ANSWER

Middle-aged female with palpable swelling over the abdominal wall

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Answer

Dermatofibrosarcoma protuberans

Discussion

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing, locally aggressive spindle cell tumor of the dermis [1–3]. DFSP arises in the dermis and presents as a multinodular mass, which can later infiltrate into deeper soft tissues [1]. Although it can occur at any age, it is commonly diagnosed in the second to fifth decades [1, 3]. It is most commonly found in the trunk, the extremities and head-neck being lesser frequent sites [1]. However owing to its dermal origin, it can occur anywhere on the body. Even though it has a low malignant potential, the chances of local recurrence are high [3]. More than 90% cases are associated with chromosomal translocation involving 17q22; 22q13 [3]. The resultant gene product binds to the constitutively expressed PDGF receptor stimulates the growth of DFSP cells [3].

Clinically, it presents as a very slowly growing reddish-brown or light blue plaque-like lesion with intracutaneous and subcutaneous extensions but without epidermal invasion [2, 3]. Occasionally it can also present as a reddish, firm lesion with irregular borders or multinodular appearance [3]. Definitive diagnosis is by biopsy which shows diffuse infiltration of the dermal layer and the subcutaneous fat by densely packed spindle-shaped cells arranged in typical cartwheel or storiform pattern (Fig. 5) which are CD34-positive (Fig. 6) [3, 4].

Owing to the typical clinical appearance and superficial location of the tumors, imaging is not usually performed for evaluation. MRI is recommended to evaluate local extension and preoperative planning of larger, atypical and recurrent tumors [1, 4]. CT can be used for evaluation if bony involvement is suspected, while USG can be used for detection of smaller lesions and for guided biopsies [4]. On USG these tumors are seen as well-circumscribed round-to-oval heterogeneously hypoechoic lesions with moderate peripheral vascularity [5]. Lee et al. found intratumoral tiny echogenic foci (<0.5 mm) without an accompanying comet tail artifact and posterior enhancement in all their cases [5]. On CT scans these are seen as well-defined isodense nodular soft tissue masses in the skin and subcutaneous fat with characteristic protrusion from the skin and moderate homogeneous post contrast enhancement [2, 6]. They appear homogeneously iso-hypointense on T1W images (Fig. 1) and hyperintense on T2 (Fig. 2) with homogeneous post-contrast enhancement (Fig. 4). Restricted diffusion (Fig. 3) is documented to be a feature of the lesion in a few studies, which is of particular importance in order to distinguish residual/recurrent tumors from scar tissue in the postoperative setting [7]. Few authors have reported early arterial enhancement in the lesions [8]; however, our case did not show significant early arterial enhancement but rather showed homogeneous enhancement in the venous phase (Fig. 4).

To conclude, although DFSP is diagnosed by its characteristic clinical manifestations, radiologists must be familiar with the typical imaging features of this uncommon neoplastic pathology and include it in the differential diagnosis of dermal lesions.

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Compliance with ethical standards

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