

Testicular metastasis of dermatofibrosarcoma protuberans: A case report (an ultrasonography approach)

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Dermatofibrosarcoma protuberans (DFSP) is a highly recurrent low-grade soft tissue sarcoma, which is usually located on the trunk. In a review of the literature there were several reports of patients with dermatofibrosarcoma with histologically documented metastasis. In all of these, there was local recurrence and subsequent metastasis to local lymph nodes and hematogenous spread to the lungs. We present a case of dermatofibrosarcoma protuberans first time reported in the literature with metastasis to the testis evaluated by Colour Doppler sonography.

KEY WORDS: *Dermatofibrosarcoma; Testicular Neoplasms; Neoplasm Metastasis; Ultrasonography, Doppler, Color*

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a relatively common soft tissue neoplasm with low-grade to intermediate malignancy. Although metastasis is rarely seen, DFSP is a locally aggressive tumor with a marked tendency to recur. In the literature review there were several reports of patients with dermatofibrosarcoma and histologically documented metastasis. In all of these, there was local recurrence and subsequent metastasis to local lymph nodes and hematogenous spread to the lungs.

We present a case of DFSP first time reported in the literature, with metastasis to the left testis evaluated by Colour Doppler sonography.

CASE REPORT

A 34-year-old man presented with a lesion on his left zygomatic bone, measuring 5.5x4.5 cm. According to his clinical history it was documented that he had been operated seven times in the last 17 years for DFSP on the upper back, near the right shoulder. The latest operation was performed a year ago and the histology report stated the diagnosis of DFSP but the resection margins were not clear. A biopsy of the lesion confirmed the diagnosis of recurrence of the DFSP. A year later he presented complaining for

cough and a painless swelling of the left testis. A plain chest radiograph was normal but in view of the rare occurrence of hematogenous spread to the lungs a CT scan of the thorax and abdomen were performed. CT scan of the chest showed multiple angiocentric pulmonary nodules ranging in the size from 4 mm to 7 mm and several nodules in the subcutaneous tissue of the posterior chest wall. CT scan of the abdomen was normal. Because of the findings of the left testis swelling, an ultrasound examination of the testicles was ordered. The echo-examination revealed a solid heterogeneous mass measuring 27x25 mm with ill-defined margins. Inside the mass the presence of echogenic dense bands of fibrous tissue and foci casting was noticed (Figure 1).

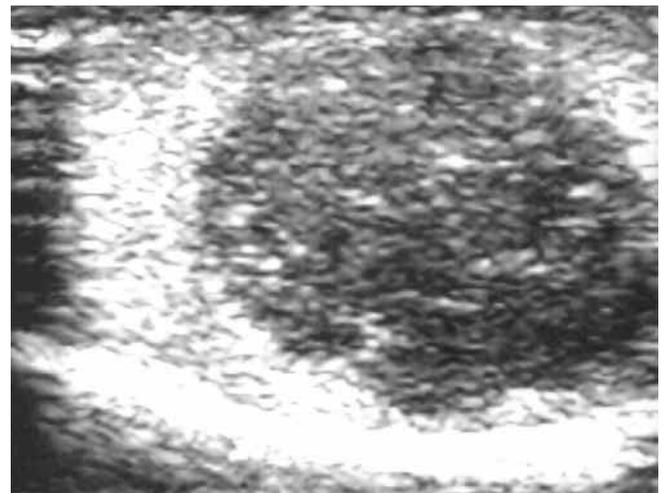


Figure 1. Sonogram of the left testis reveals a heterogenous, hypoechoic, solid mass measuring 27X25 mm with ill-defined margins. The mass contains peripheral and internal echogenic bands

Colour Doppler sonography showed medium grade colour flow within the lesion with increased resistive index in comparison with

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normal intratesticular vessels (Figure 2).

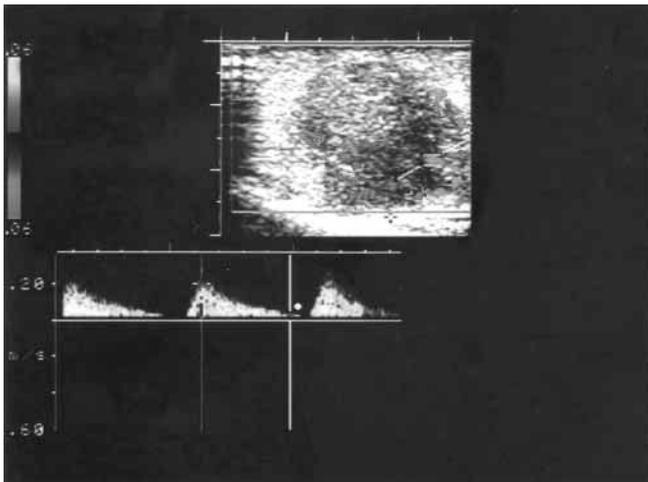


Figure 2. Colour Doppler sonogram analysis demonstrates increased resistive index arterial flow within hypoechoic testicular tumor

An excision of the left testis was performed and histology report confirmed the diagnosis of a meta-lesion from DFSP (Figure 3). After six months the patient developed brain metastases and shortly after he passed away.

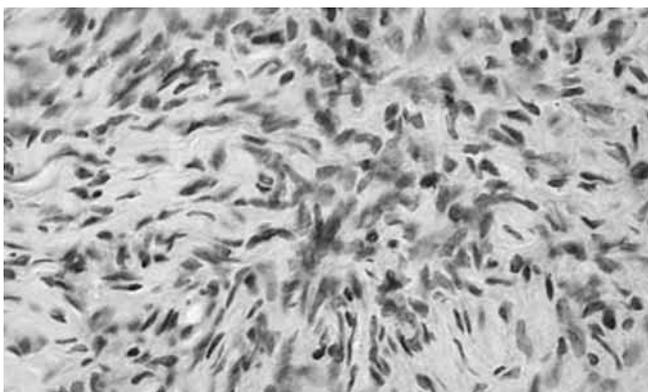


Figure 3. High power histological image of the lesion with cellular storiform appearance.

DISCUSSION

DFSP constitutes less than 0.1% of all malignant neoplasms. Incidence has been estimated between 0.8-5 cases per 1 million population per year and usually occurs in adults aged 20-25 years with almost equal sex distribution. It is most often presented as a large indurated plaque several centimeters in size and it is composed of firm, irregular nodules varying in colour from flesh to reddish brown. DFSP is a very slowly growing tumor and is most commonly seen on the trunk, followed by the proximal extremities. It is characterized by its aggressive local recurrence rate of 26%. Despite the local invasiveness, DFSP rarely metastasizes. The development risk of metastatic disease is only 5%. The most common site of distant metastasis are the lungs occurring via hematogenous spread and the regional lymph nodes.

The pre-mentioned patient had multiple local recurrence and was found to have multiple metastases throughout both lungs and also bone metastases. No lymphatic metastases were identified. The patient also developed a lesion of the left testis. The age of the patient and the clinical appearance indicated the presence of a primary tumor of the testis, specifically a seminoma. But the sonographic appearance of the mass with the medium grade vascularization and the presence of echogenic bands of fibrous tissue was favorable for a non-seminoma tumor or a metastatic lesion from the primary tumor. The histology study established the diagnosis of metastasis from DFSP. This case highlights the rare incidence of metastasising dermatofibrosarcoma protuberans to the testis and also the difficulty of establishing diagnosis only from ultrasonography findings. In the interpretation of the sonographic findings with colour Doppler sonography, the clinical history of the patient has to be taken into account.

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