

## Dermatofibrosarcoma protuberans diagnosed by a single biopsy

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### Abstract

This brief report is about a 9 year old girl presenting with a 2.5 cm circular blue to violet discoloration on the anterolateral upper left thigh. The first biopsy taken revealed the diagnosis dermatofibrosarcoma protuberans. The patient underwent surgery at a specialized sarcoma center with post operative histology showing free wide resection margin. Preoperative chest x-ray showed no sign of metastasis. This brief report emphasizes the significance of the use of biopsy when cutaneous elements look suspicious and diagnosis is unclear.

### Case Report

A 9 year old, previously healthy girl, presented with a 2.5 cm circular, blue to violet, slightly elevated discoloration on the upper anterolateral left thigh (Figure 1). Two years prior to the



Figure 1. The clinical presentation of the cutaneous thickening with violet discoloration on the anterolateral upper left thigh, just before the biopsy was taken.

consultation she had a minor trauma against the area and the described cutaneous changes developed shortly after. Her general physician suspected a foreign body and conducted an ultrasound of the thigh. All findings were normal except for a 6 mm fluid accumulation. In addition, the patient's own GP tried treatment with topical steroid without success. The patient was then advised to seek further dermatological investigation.

Dermatological examination revealed the earlier described intumescens at the thigh. Except for minor tenderness when palpating the discoloration, the examination was normal. A skin biopsy was taken and histologically, the dermis was replaced by a tumor composed of interwoven bundles of rather uniform, small spindle-cells (Figure 2). Staining with CD34 showed that the tumor cells characteristically expressed CD34 (Figure 3). Thus, histology revealed the diagnosis dermatofibrosarcoma protuberans grade I.

The patient was referred to the specialized sarcoma center to the further treatment starting with a preoperative chest x-ray. The x-ray showed no signs of metastasis. The tumor was removed using wide local excision, with post-operative histology showing wide free resection margin. The patient is now set up to annual clinical controls during the next 5 years to detect any recurrence.

### Discussion

Dermatofibrosarcoma protuberans (DFSP) is a relatively rare tumor with only 0.8-5 cases per million/year.<sup>1</sup> The localization of this type of cancer is 50% on the thorax, 40% on the limbs, 10% head/neck area.<sup>1</sup> The clinical pres-

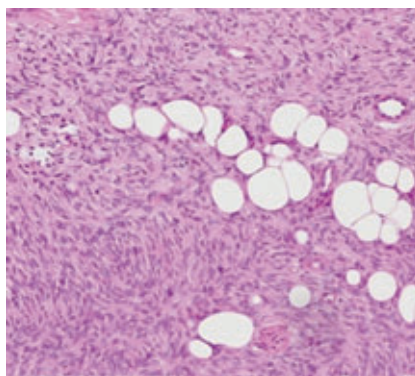


Figure 2. The dermis is replaced by a tumor composed of interwoven bundles of rather uniform, small spindle-cells. The tumor cells insinuate between fat cells in the subcutis (Hematoxylin and Eosin).

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entation of DFSPs are in the early stages plaque like areas of cutaneous thickening with violet/blue or red discoloration. When the tumor slowly enlarges it becomes raised, firm and nodular and the surrounding skin may become teleangiectatic. DFSP is low malignant and a locally aggressive growing cancer, only 1-4% disseminates.<sup>1</sup> The dissemination is hematogenic to lungs and bones.<sup>2</sup> Sentinel node biopsy is not recommended because of

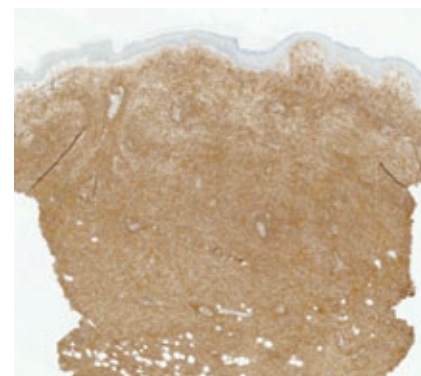


Figure 3. The tumor cells characteristically express CD34.

the seldom lymph node involvement (<1%). The mortality is high with metastasis, the majority of patients die within the 2 first years.<sup>1</sup> Without metastasis and with total surgical tumor resection, the prognosis is good with 5 year survival rate of 93-100%.<sup>1,4</sup> The recurrence rate is up to 50-75%, most of them within 3 years.<sup>1,3</sup>

The treatment for DFSP is tumor resection with pathologically negative margins. The preferred margin is 3-4 cm.<sup>1,4</sup> This is called wide resection. For the surgeon, Mohs surgery was not in consideration, since *en bloc* resection is preferred in sarcomas. Furthermore, the DFSP has infiltrating elements that are hard to detect during histology. So margins have to be large, at least some centimetres.

## Conclusions

A new kind of treatment using Imatinib, an abl-kinase inhibitor, has been approved for adult patients with unresectable, recurrent or metastatic DFSP who are not eligible for surgery. The Imatinib results on patients with locally advanced or metastatic disease show variable response, but there are cases with successful results. In particular two cases; a man with DFSP on the thigh and metastasis of the spine and a woman with metastasis in the lungs and axilla show the successful use of imatinib. They were treated with Imatinib for 4 and 3 months, respectively. A tumor size reduction of 75% was seen in the first case, permitting this patient to surgical resection. In the second case, a CT-scan 3 months after treatment start showed nearly resolution of the lung metastasis. Adjuvant radiotherapy is not commonly preferred, because of reported cases with growth of

aggressive fibrosarcomas in areas with DFSP that were treated with radiation therapy.<sup>1</sup>

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