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Darier and Ferrand Dermatofibrosarcoma with Mammary Location: About a Case

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ABSTRACT

Introduction: *Dermatofibrosarcoma is a sarcoma of intermediate degree of malignancy with strong local aggressiveness with a low but real metastatic potential. Its breast location is very rare.*

Observation: *It was a 40-year-old woman who has never been pregnant who was followed for a retro-areolar tumor of the ulcerated left breast that had been evolving for more than eight weeks. A mammary cytopract was done which was in favor of a phyllode tumor. We performed a large surgical excision on the patient and the histology of the operative specimen was in favor of a dermatofibrosarcoma of Darier and Ferrand with healthy resection margins.*

Discussion: *The mammary location of Darier and Ferrand syndrome is rare. With early diagnosis an infectious and hemorrhagic complication can be avoided whereas, wide excision improves the prognosis.*

Keywords

Darier and Ferrand Dermatofibrosarcoma, Breast, Large excision.

Introduction

Dermatofibrosarcoma (DFS) of Darrier and Ferrand is a cutaneous mesenchymal tumor with low malignancy potential, but with a characteristic tendency whose local recurrence is very high [1,2]. Local recurrence rates of 50% to 70% have been reported thanks to a lumpectomy [3,4]. DFS accounts for 1% of all soft tissue sarcomas and less than 0.1% of all malignant tumors. The annual incidence is about 0.8-4.5 / 1000000 [1].

Because of its relative resistance to chemotherapy and radiotherapy, its treatment remains exclusively surgical [5,6]. Usually, the locations are the trunk, the limbs and the cephalic extremity. Breast localization is exceptional and requires wide resection respecting the margins of the healthy excisions [7-9].

We report the case of a Darier and Ferrand tumor with breast location and we are discussing the singularity of this topography.

Observation

It is a 40-year-old who has never been pregnant with no particular health history who was received for painless ulcerated right breast mass gradually increasing in volume for more than a year without mammalian flow.

On clinical examination, we found a mass occupying the external quadrants of the right breast about 10 cm long axis with a 5 cm ulcero-necrotic zone at its center. There was no mammalian flow and the ganglionic areas were free.

Figure 1: Clinical aspect of the breast.

A compunction and a breast biopsy confirmed an adenocystosarcoma.

In front of this clinical board we performed a large excision.

The histology of the operative specimen showed fusiform, dense, monomorphic and intradermal cell proliferation under a thinned epidermis, the tumor infiltrates deeply by invading the entire dermis and underlying hypodermis in the form of trabecular spans.

Fascia

The Immunohistochemia showed an intense and diffuse CD34 positivity.

Figure 2: Macroscopic feature of the operative specimen.

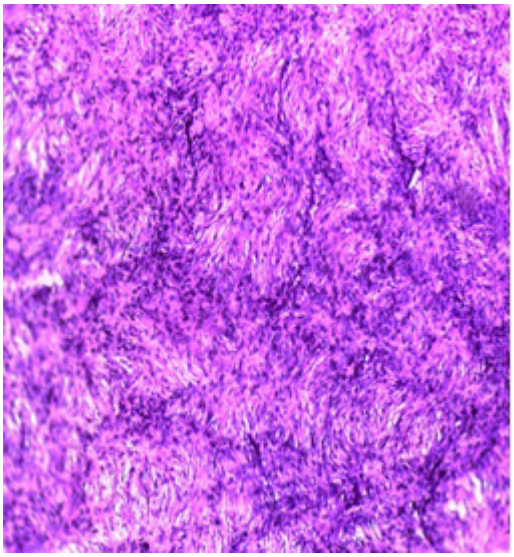

Figure 3: Microscopic aspect of the operative specimen.

Discussion

The Darier and Ferrand Dermatofibrosarcoma (DFS) is a mesenchymal cutaneous tumor with intradermal development. It is a tumor, located between the benignity pole of the very common and harmless cutaneous fibroid and the malignant pole of the true cutaneous fibrosarcoma [4,10]. Its frankly malignant transformation with metastasis is exceptional, whereas its local recurrence is very high [5,11].

DFS affects both female and male. Most often it occurs between the age of 20 and 50 [6]. The preferred seat of injury is the trunk, the root of the limbs and the cephalic extremity. On the other hand, its breast location remains rare.

Dermatofibrosarcoma can affect all the parts of the body, but its preferred seats are the trunk 50 to 60%, the limbs 20 to 30%, the head 15 to 20% [6,7]. Its mammary location is very rare [12-14].

Clinically the lesion begins with an indurated plaque, covered with skin of appearance and normal coloration, sometimes whitish, yellowish-white, pinkish, purplish or reddish, it is apparently well defined and is movable relative to the deep planes. At a later stage, the plate spreads; its surface becomes irregular and bumpy, forming after a few months or a few years, a multinodular mass, often polychrome, of variable size, hard, perfectly mobile on the deep planes.

This evolution in two stages is not constant because some forms are immediately uninodular or multinodular with secondary melting of nodules [1,3,15,16]. This typical aspect of FDS was found in our patient and its evolution may result to a painful and haemorrhagic ulceration of these nodules.

Pathologically, the tumor is in the form of dense cell proliferation, poorly limited, unencapsulated, occupying the dermis, usually entirely. The tumor sends extensions into the hypodermis, without destroying its elements, while the epidermis is respected [17-19]. The cells are elongated, spindle-shaped, with more or less abundant cytoplasm, regular oval nucleus. The mitoses are variable and rarely atypical. On the architectural plan, the cells are arranged in radiating beams (aspect "wheel radius") or swirling.

It is sometimes difficult to distinguish between DFS and other spindle-shaped tumors. Immunohistochemistry techniques make it possible, in difficult cases, to make the case for diagnosis by highlighting tumor cells in the tumor.

CD34 antigen

In fact, immunohistochemistry shows an intense and diffuse positivity of CD34 [17,18,20]. In our patient, the biopsy had previously found an adeno -cystosarcoma. Histology of the operative part and immunohistochemistry has led to the diagnosis of dermatofibrosarcoma [20].

Cytogenetic analysis is an interesting contribution in the diagnosis. With it, we found the presence of ring sunumerous chromosomes composed of sequences from chromosomes 17 and 22 or, more rarely, t (17; 22) translocations [6,21].

The treatment is difficult because of the risk of subclinical extension of the tumor, which is often the cause of local

recurrence. The wide surgical excision as we did it with our patient is the standard treatment with margins of resection of about 5 cm [22-24]. Postoperative radiotherapy and targeted therapy can be used in some of the more advanced forms and in the prevention of local recurrence [5,6]. In fact, the imatinib mesylate, which is an anti-tyrosine kinase molecule, inhibits the PDGFRβ receptor and induces clinical responses in patients with unresectable or metastatic DFS [4,10].

Rigorous clinical supervision every 6 months is necessary because of a high risk of recurrence. In our patient, we did not detect local recurrence after one year.

The prognosis of the DFS depends on its possible local malignancy. This tumor rarely metastasizes. Its high potential for recurrence despite the wide surgical excision makes it difficult to control [2,3,7].

Conclusion

DFS is a particular skin tumor because of its difficult diagnosis. Breast DFS is very rare. Immunohistochemistry and cytogenetics are essential for the diagnosis of DFS. In spite of the wide surgical excision, which is the basis of the treatment, local recurrences are not negligible.

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