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Direttore Nicola Picardi

Acellular dermal matrix for breast reconstruction after surgery for giant dermatofibrosarcoma protuberans

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Antonio Zanghì, Maria Di Vita, Andrea Cavallaro, Marcello Majorana, Sebastiano Bentivegna, Francesco Cardì, Alessandro Cappellani

Department of Surgery and Medical Surgical Specialties, General and Breast Surgery Unit AOU Policlinico "Vittorio Emanuele II", University of Catania, Italy

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Acellular dermal matrix for breast reconstruction after surgery for giant dermatofibrosarcoma protuberans

INTRODUCTION: *Dermatofibrosarcoma protuberans (DFSP) is a rare neoplasm that exceptionally can affect the breast, always originating from skin and dermis, so imposing large sacrifice of skin. Only few cases have been reported of intraparenchymal DFS. We describe a unique case of giant intraparenchymal DFS that required removal of all the gland and reconstructive surgery.*
CASE REPORT: *A 34 years old woman presents with a quickly growing breast mass, about 12 cm, with radiological features suggestive for giant fibroadenoma or mesenchymal neoplasm. The histology showed a Dermatofibrosarcoma protuberans infiltrating the surrounding parenchyma. The need for radicalization imposed, due to the size of the tumour, a nipple sparing mastectomy. The reconstruction has been performed using a new type of Acellular Dermal Matrix (ADM) mesh to wrap the prosthesis that has been placed and fixed over the great pectoral muscle.*
RESULTS: *The final histologic report showed that the residual parenchyma and the skin removed were free from neoplastic infiltration. The patient is free from recurrence at 24 months from the surgery and the cosmetic result is excellent.*
DISCUSSION AND CONCLUSIONS: *The treatment of DFSP should be aimed to prevent local recurrence, that are usually located in the scar or very close to it. Large size DFS can impose even mastectomy. If skin is not compromised like in this case, a nipple sparing mastectomy is suitable and the one time reconstruction with ADM wrapping of the prosthesis and fixation over the muscle can help to spare time, avoid complications and pain medication and reach excellent cosmetic resu.*

KEY WORDS: Acellular Dermal Matrix (ADM) mesh, Protuberans, Breast neoplasms, Dermatofibrosarcoma, Mastectomy, Nipple sparing, reconstructive surgery

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a malignant breast tumor that originates from the deeper layers of the dermis and the subcutaneous tissue. Not uncommon to the trunk and extremities, it is a rather exceptional finding in the breast where, nevertheless, involves skin and dermis.
We present an exceptional case in which a young woman with an unusually giant breast DFSP, not involving the skin or subcutaneous tissue. It was so performed a nipple sparing mastectomy and a new technique of one step muscle-sparing reconstruction, using a biological mesh in ADM of new conception.

The Clinical Case

A 34-year-old woman presents to our outpatient office complaining rapid growing of the right breast volume,

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Correspondence to: Maria Di Vita, University of Catania, Department of Surgery and Medical Surgical Specialties, General and Breast Surgery Unit AOU Policlinico "Vittorio Emanuele II", via Santa Sofia 78, 95123, Catania Italy (e-mail:Maria.d.divita@gmail.com)

supported by a large neoplasm, about 12 cm in diameter, occupying the upper quadrants, which at mammography presented as a homogeneous opacity with polycyclic contours. The US showed characters very alike to a giant fibroadenoma.
The age, the distance from the last pregnancy (7 years) and rapid growth, did suggest a phyllodes tumor or a mesenchymal neoplasm.
It is so proceeded to surgical excision of what resulted to be three coalescing nodules, sized respectively 7 cm, 6 and 5,5 maximum diameter, and of the pseudo-capsule that surrounded them. Sent the specimen for frozen section, it was obtained in the first instance a pathology report of spindle cell neoplasm, richly cellular, with a low mitotic index.
The final report was DFSP. The neoplasm in some places infiltrated the surrounding breast parenchyma, imposing a re-excision. It was so decided a mastectomy. This consisted of a nipple-sparing mastectomy, through bat-wing skin incision that enclosed the primary wound edges, corresponding to the tract of skin closest to the neoplasm. An immediate reconstruction has been performed with placement in the subcutaneous of a double chamber prosthesis, completely wrapped in a mesh Acellular Dermal Matrix (Braxon®) pre-moulded and shaped to perfectly accommodate the implant. A suction drainage has been placed under the prosthesis and removed three days after surgery, when the serum collected was less than thirty ml per day. Stitches have been removed 12 days after surgery. No seroma has been observed.

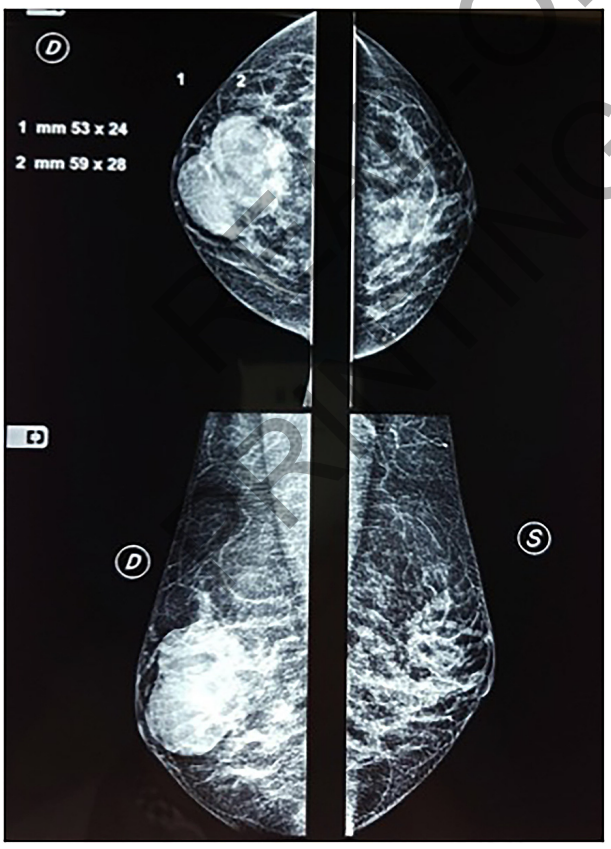

Fig. 1: RX-bilateral mammography.

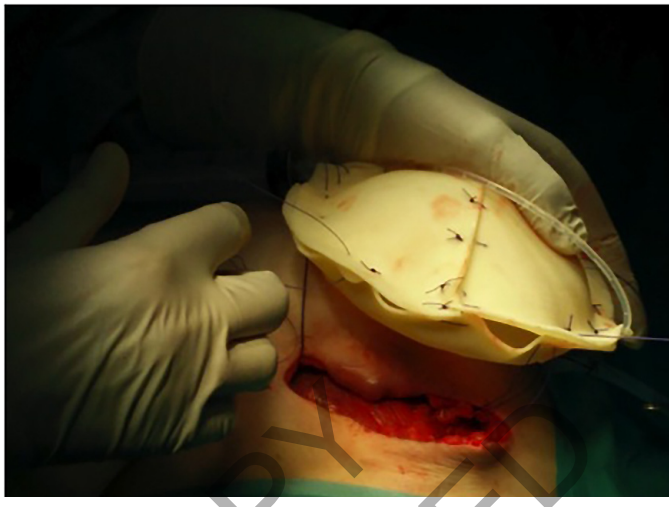

Fig. 2: a double room prosthesis completely wrapped in a mesh ADM (Braxon®) - packaging. Sutures are passed through the pectoralis major muscle so to fix the prosthesis and prevent a dislocation.

Results

The postoperative course was uneventful and totally pain-free. The patient did not complain any discomfort or impairment of motility and the cosmetic result was characterized by symmetry and natural ptosis and softness.
The final histological examination showed the absence of neoplastic elements in the residual parenchyma and in the skin removed.
At 18 months examination scar was well healed and the size, shape and softness of reconstructed breast no different from the natural one.

Discussion

Known from 1924, when Darier and Ferrand formerly described it, DFSP is a rare neoplasm that arises from deeper layer of skin, dermis and subcutaneous tissue [1].
Though the tumour is most frequently located in the trunk and upper limbs, localization at the breast is very rare. The literature in fact reports only about 40 cases located in the breast [2], most of them originating from the skin or dermis and visible as bluish, reddish or brownish mass, sometimes multinodular or very large. In some cases the lesion clinically looks as a true breast mass, but it is still clear the dermal or subcutaneous location. Only 4 cases in the literature presented as iraparenchymal and none of them reached the size we observed in this case [3-5].

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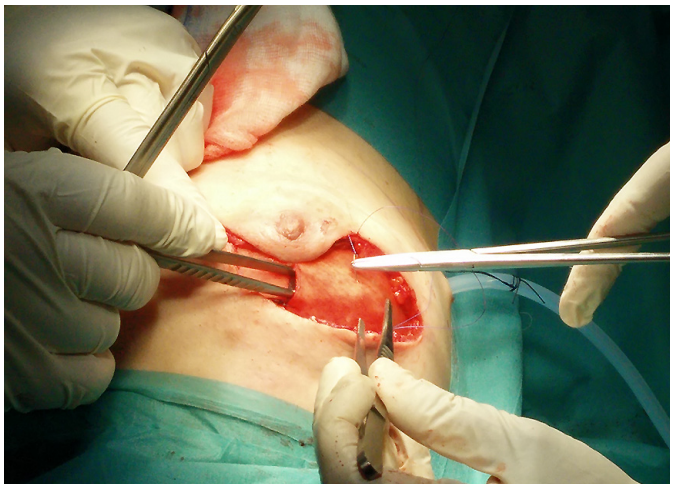

Fig. 3: The mesh in ADM is anchored to the subcutaneous, before the synthesis of the skin incision.

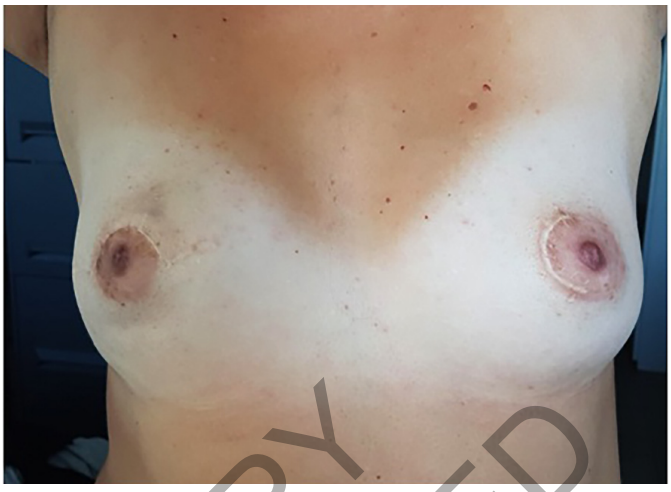

Fig. 5 Cosmetic results at 24 months

The preferred ages are those from twenty to fifty, but in the breast it has been described even in children (2 years old)[6] or older people (102 yrs)[7].
The tumour can remain for long time as a stable fibrous plaque and the rapidly grow up in larger nodules [8] characterized by low malignancy. Nevertheless literature has showed multiple cases of local recurrences and even ability to metastasize [7,9,10].
The high risk of local recurrence imposes the excision of the skin that is primitively involved by the tumour and of all surrounding tissues [2]. DFSP in fact easily infiltrates even subcutaneous fat, fascia and muscles. This ability is the principal cause of early recurrences, usually seen within three years from the primary excision, though even later recurrences have been described [10].
Nevertheless, in our case skin was not involved, as proved by histology, and the mass had the characteristics of mobility that could have lead to preoperatively consider the diagnosis of giant fibroadenoma. The deep location, inside the gland, and the infiltration of the surrounding parenchyma imposed a radicalization. NCCN Guidelines for Surgical treatment of DFSP recommend 2-4 cm of margins, and Parker recommended for DFSP larger than 2 cm at least 2.5 cm of margins, through fascia and periostium [11]. Dealing with breast and with a giant neoplasm, the only option was the mastectomy and the relative distance from surface encouraged us to spare the skin with the exception of the tract more close to the neoplasm and that had been incised to access the mass. A bat-wing incision looked fit to achieve this result. But along with mastectomy we had to perform an immediate reconstruction.
The usual reconstruction would require the sub-muscular placement of an expander. This technique is not free from discomfort and complications of various types. The expander needs to be gradually filled requiring repeated outpatient accesses. A post-operative and post expansion pain is usual, requiring medication, and upper limb functional deficits are possible.
For some time, in order to ensure optimal coverage to the expander, it is introduced in the surgical practice the use of mesh Acellular Dermal Matrix (ADM), allowing expanding the housing of the prosthesis and avoiding contact between this and the subcutaneous tissue [12].
Experience with these methods has convinced us to test a new type of ADM. The result at 24 months from surgery is completely satisfactory. Now evidences are increasingly confirming the good performance of this kind of reconstruction [13].

Conclusions

This exceptional case has so consented to test a new method of reconstruction that allowed to spare operative time, to avoid multiple outpatient accesses to fill the expander and a second surgical time to change expander with definitive prosthesis, and last to prevent complications and motility defects. After two years follow up the patient is free from recurrence with excellent cosmetic results

Riassunto

BACKGROUND: Il dermatofibrosarcoma protuberans (DFSP) è una neoplasia rara che interessa solo eccezionalmente la mammella, originano sempre dalla cute e dal derma, imponendo quindi un ampio sacrificio del rivestimento cutaneo. In letteratura sono stati descritti fino ad ora solo pochi casi di DFSP intraparenchimali. Il caso qui descritto è l'unico, a nostra conoscenza, di DFSP gigante intraparenchimale, tale da richiedere una mastectomia nipple sparing con ricostruzione immediata.

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Caso Clinico: Una donna di 34 anni si presenta all'osservazione con una neoformazione della mammella destra accresciutasi rapidamente fino a raggiungere un diametro massimo di 12 cm. Il quadro radiologico era suggestivo per un fibroadenoma gigante o per una neoplasia mesenchimale. Il reperto istologico, dopo asportazione della massa con tessuto circostante dimostrò un Dermatofibrosarcoma protuberans che infiltrava il parenchima circostante, per cui si impose una radicalizzazione dell'intervento con esecuzione di una nipple sparing mastectomy. La ricostruzione è stata eseguita in un tempo utilizzando una protesi a doppia camera avvolta in una mesh in ADM di nuova concezione per avvolgere la protesi e fissarla in sede prepettorale.
Risultato: L'istologia ha mostrato che il parenchima residuo e la cute asportata erano indenni da infiltrazione neoplastica. Il risultato cosmetico sia immediato che a distanza di 24 mesi è ottimale e la paziente è esente da recidiva.
Discussione e Conclusioni: Il Dermatofibroasrcoma protuberans, per quanto eccezionale nella mammmella, dovrebbe comunque essere tenuto in considerazione nella diagnosi differenziale delle masse mammarie, anche quando il rapporto con la cute ed il derma non sia evidente. Il trattamento deve essere mirato a prevenire le recidive, che si presentano di solito a livello della cicatrice o in sua prossimità. I DFSP di grandi dimensioni possono rchiedere l'esecuzione di una mastectomia. Se la cute non è compromessa, come in questo caso, una nipple sparing mastectomy rappresenta l'intervento di scelta. La ricostruzione in un tempo con apposizione di protesi in sede prepettorale, grazie al rivestimento in ADM, permette di risparmiare tempo, sedute operatorie, complicanze, dolore post-operatorio, deficit funzionali, e raggiungere un eccellente risultato estetico e funzionale con soddisfazione della paziente.

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