

of atypia in the histopathology of the lesions in our patient excluded this diagnosis.

Treatment can be provided to patients with symptomatic lesions or for cosmetic reasons. Numerous treatments with satisfactory results have been described in the literature, including topical retinoids, cryotherapy, electrocoagulation, radiofrequency, carbon dioxide laser,² surgical excision, oral tetracyclines, and photodynamic therapy.^{3,9} Given the benign nature of the disease and the absence of any cosmetic issue, our patient was not a candidate for treatment.

In conclusion, milia en plaque is a rare but easily diagnosed disease. No cause is detected in the majority of cases. In our case, the recent history of exposure to radiotherapy, with a plausible temporal relationship, would suggest a causal relationship.

References

1. Hubler WR Jr, Rudolph AH, Kelleher RM. Milia en plaque. *Cutis*. 1978;22:67-70.
2. Tenna S, Filoni A, Pagliarello C, Paradisi M, Persichetti P. Eyelid milia en plaque: A treatment challenge with a new CO₂ fractional laser. *Dermatol Ther*. 2014;27:65-7.
3. Muñoz-Martínez R, Santamarina-Albertos A, Sanz-Muñoz C, Miranda-Romero A. Quistes miliares múltiples agrupados. *Actas Dermosifiliogr*. 2013;104:638-40.
4. Martín-Ezquerro G, Molinero-Caturla J, Umbert-Millet P. Quistes miliares en placa en las extremidades. Posible toxicodermia

- por 6-mercaptopurina. *Actas Dermosifiliogr*. 2004;95:449-50.
5. Quist SR, Franke I, Bonnekoh B, Gollnick HP. White papules around the ears: a quiz. Milia en plaque. *Acta DermVenereol*. 2010;90:445-7.
 6. Beutler BD, Cohen PR. Cryotherapy-induced milia en plaque: Case report and literature review. *Dermatol Online J*. 2014;21, pii: 13030/qt4dw7k4nk.
 7. Lee A, Griffiths WAD. Multiple milia due to radiotherapy. *J Dermatol Treatment*. 2002;13:147-9.
 8. Ronchese F. Cicatricial comedos and milia. *Arch Dermatol Syphilol*. 1950;61:498-500.
 9. Carbia SG, Malah V, Glorio R, Ruiz Beguerie J, Devés A, Vogel J, et al. Milia en placa en varias y sucesivas áreas. *Med Cutan Iber Lat Am*. 2005;33:183-5.
 10. Song JC, Hughes MH, Ramolia PB, Wells MJ. Milia-like papules on upper trunk, neck, and left forehead. *J Am Acad Dermatol*. 2011;65:467-9.

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Why Is Mohs Micrographic Surgery Underused in the Treatment of Dermatofibrosarcoma Protuberans in Children?☆



¿Por qué la cirugía micrográfica de Mohs está infrutilizada en el tratamiento del dermatofibrosarcoma protuberans infantil?

To the Editor:

Dermatofibrosarcoma protuberans (DFSP) is a slow-growing, intermediate-grade fibrohistiocytic tumor characterized by high rates of local recurrence but low metastatic potential. Few cases of congenital forms have been reported, but this is probably because early lesions can go unnoticed or be confused with other entities. There is evidence that recurrence rates are lower with Mohs micrographic surgery (MMS) than

with conventional surgery with wide margins.¹ Nonetheless, in 3 recently published pediatric series, wide excision was used in the vast majority of patients (41/43).²⁻⁴ We present a case of congenital giant-cell fibroblastoma, a histologic subtype of DFSP, which was diagnosed and treated in an 8-year-old boy. In this report, we highlight the advantages of MMS and the use of negative-pressure wound therapy (NPWT) in children with DFSP.

The patient was an 8-year-old boy with no relevant past history who was seen for a lesion in the right axilla that had grown and become progressively harder. The lesion, which was initially flat and reddish in color, had been present since birth. Examination revealed an indurated, mobile plaque measuring 4 × 2.5 cm. The plaque was pink but had a purple, slightly elevated area. The regional lymph nodes were not enlarged. Biopsy showed a poorly delimited tumorous lesion composed of spindle and oval cells that occupied practically the entire dermis and extended down into the subcutaneous tissue. Pseudovascular spaces surrounded by multinucleated giant cells were also visible. The spindle cells expressed CD34 and were negative for S-100 protein and smooth muscle actin (Fig. 1). A diagnosis of giant cell fibroblastoma was established, and extracutaneous invasion was ruled out by magnetic resonance imaging. The tumor was excised under general anesthesia. The technique chosen was slow MMS (Breuninger technique), with excision of 1-cm margins in the first stage (Fig. 2). This technique consists of the en

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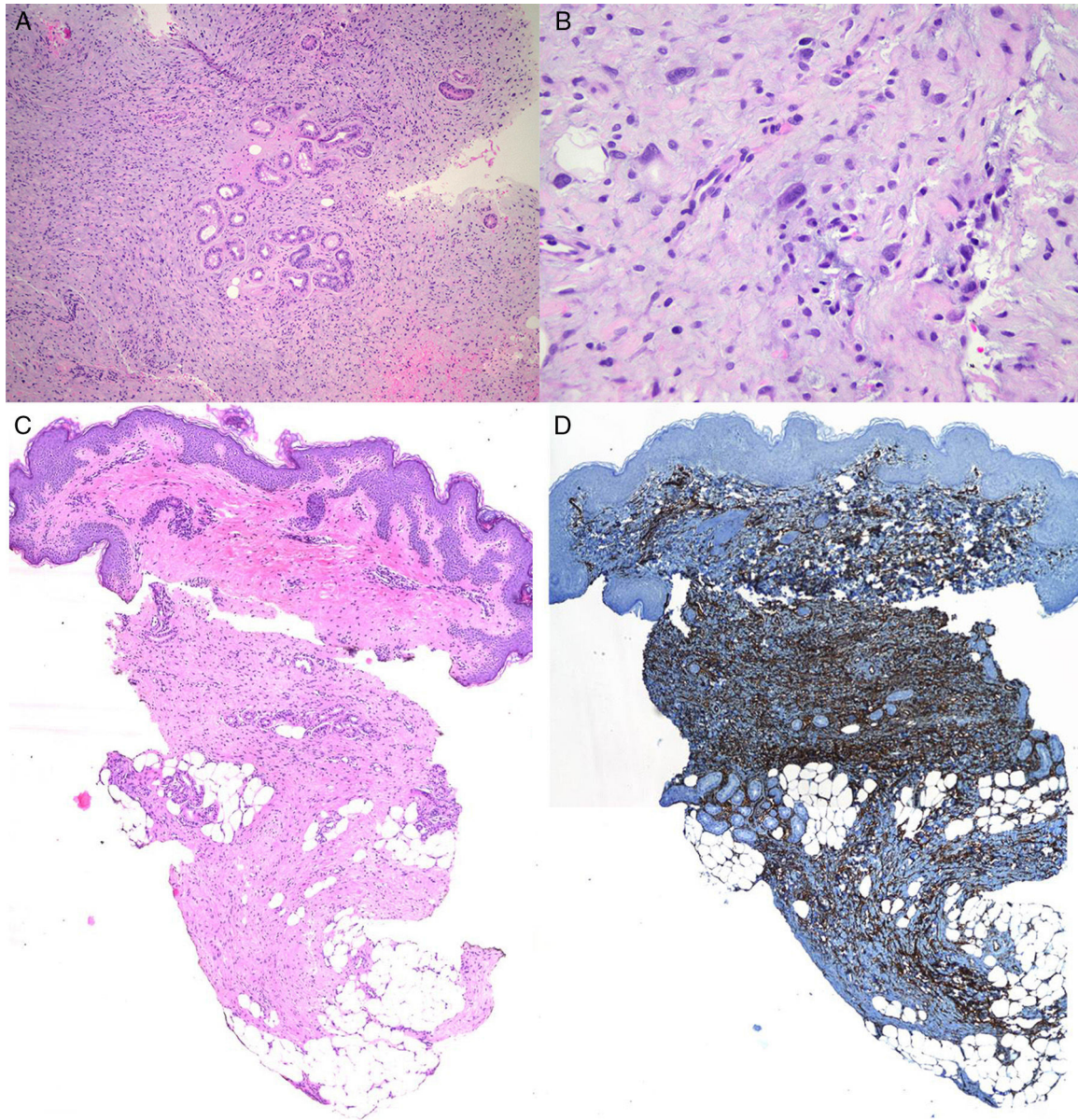


Figure 1 A. Proliferation of spindle cells arranged in fascicles (original magnification, hematoxylin-eosin $\times 100$). B, Irregular pseudovascular spaces surrounded by multinucleated giant cells (original magnification, hematoxylin-eosin $\times 100$). C, Involvement of full thickness of the dermis with extension into the subcutaneous tissue (original magnification, hematoxylin-eosin $\times 20$). D, Spindle cells expressing CD34 ($\times 20$).

bloc resection of the tumor at a 90° angle to the surface and the achievement of a uniform wound bed. In our case, the depth of excision reached the fascia. The resected specimen was fixed in formol and embedded in paraffin. Because the slides with samples from the deep and peripheral margins were to be reviewed at a later date, it was decided to cover the surgical defect with a NPWT system with a negative continuous pressure of 125 mmHg (Kinetic Concepts Inc.) (Fig. 3 A-C). On day 9, with histologic confirmation of tumor-free margins, the surgical defect was reconstructed using a Limberg flap over the granulation tissue formed during NPWT; no specific preparation was required

(Fig. 3D). There have been no recurrences in 4 years of follow-up.

DFSP is a rare tumor in children, with an estimated incidence of 1 case per 1 000 000 population under the age of 20 years.⁵ Its prevalence could, however, be underestimated as time to diagnosis ranges from 5 to 15 years. Congenital forms of DFSP are even rarer. In 1 series of 152 cases of childhood DFSP, for example, only 20 tumors were congenital.⁶

Childhood and adult forms are similar in terms of immunohistochemical characteristics, the translocation of genetic material between chromosomes 17 and 22, and the association with the *COL1A1-PDGFB* fusion gene. Congenital

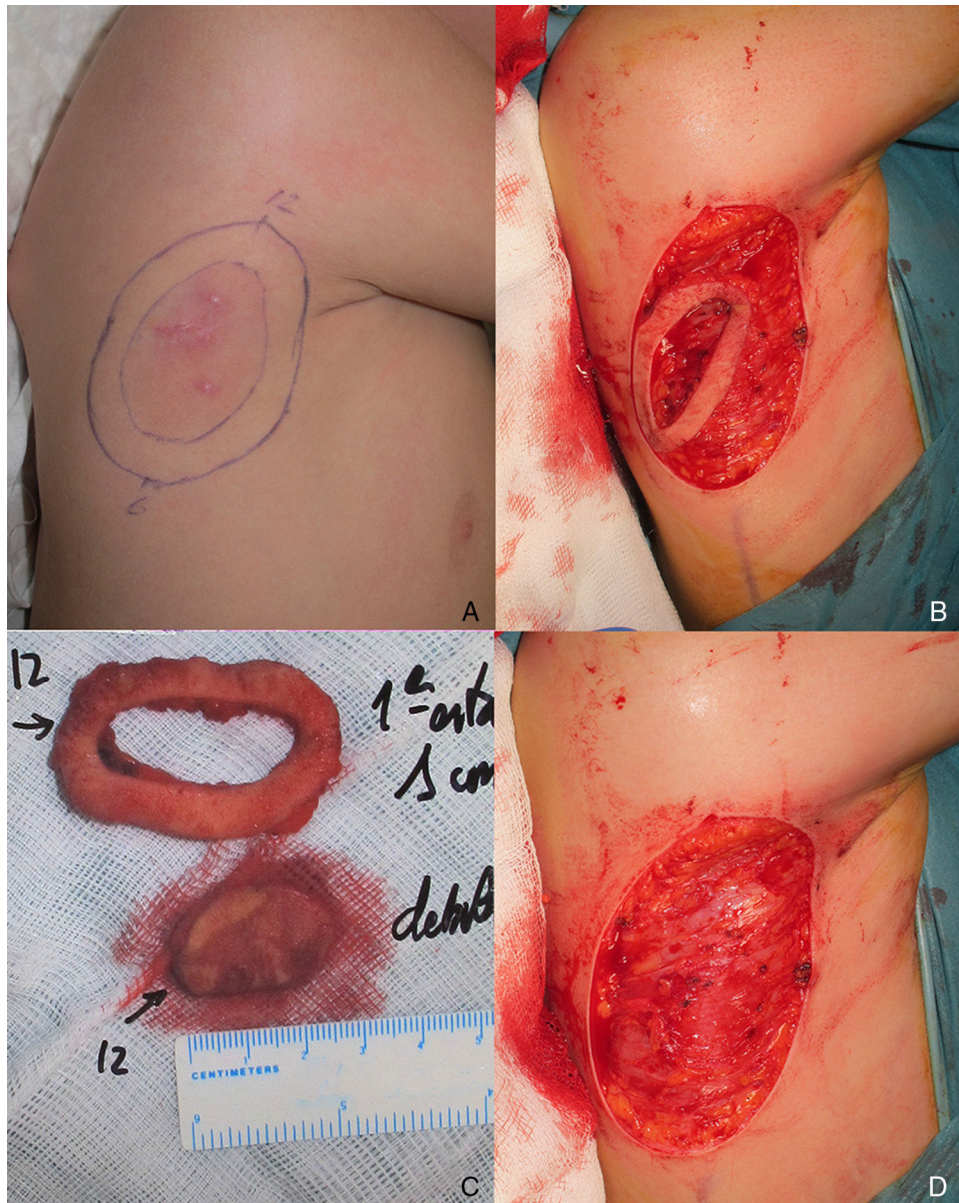


Figure 2 A, Schematic of specimen. B, Ring-shaped excision of a 1-cm margin of healthy skin around the debulking specimen. C, Specimen from first slow MMS stage embedded in paraffin for examination. D, Final surgical defect measuring 6 × 4.5 cm.

variants of DFSP present more frequently as a stain or an atrophic plaque than as a tumorous lesion. The differential diagnosis is broad and includes vascular anomalies, aplasia cutis congenita, atrophoderma, myofibromatosis, childhood fibromatosis, and fibrosarcomas.

Compared with adults, in children, tumors are more frequently located on the lower extremities and at acral sites. Other differences in children include lower recurrence rates, very few cases of metastasis, and little use of MMS.⁶

The treatment of choice for DFSP is surgery,⁷ with imatinib⁸ and radiation therapy reserved for recurrent, unresectable, or metastatic tumors. In a report of 74 patients, mostly adults, with DFSP, and a review of the literature, Serra-Guillen et al.¹ concluded that MMS was associated with considerably fewer recurrences than

conventional surgery with wide margins, and also achieved greater preservation of healthy tissue. The most common MMS technique used is slow MMS, which requires coverage of the surgical defect with biosynthetic skin substitutes or other dressings. NPWT, however, has been proposed as an alternative to skin substitutes for temporary wound coverage.⁹ This system reduces the need for analgesia and sedation and also requires less frequent wound dressing. In our case, it additionally allowed the patient to walk around and move his arm. Portable vacuum systems can be used on an outpatient basis, but because of the geographic distribution of our community (our hospital is on an island), we were not able to use a portable NPWT system.

In 2014, there were 3 reports of childhood DFSP in hospitals in Taiwan,² Canada,³ and Spain.⁴ The series included,



Figure 3 Negative pressure wound therapy (NPWT) system. A, Coverage of skin defect with foam. B, Placement of adhesive film over the foam to seal the system. C, Placement of vacuum tube connected to NPWT system. D, Reconstruction of surgical defect with Limberg flap.

respectively, 13, 17, and 13 patients under the age of 18 years, but surprisingly, MMS was used in just 2 of these 43 patients. If MMS has been shown to achieve better therapeutic, functional, and cosmetic results, why is it underused in children with DFSP? Finally, we would like to stress the importance of working closely with multidisciplinary teams to ensure that children with DFSP are diagnosed and treated correctly.

References

1. Serra-Guillén C, Llobart B, Nagore E, Guillén C, Requena C, Traves V, et al. Mohs micrographic surgery in dermatofibrosarcoma protuberans allows tumour clearance with smaller margins and greater preservation of healthy tissue compared with conventional surgery: A study of 74 primary cases. *Br J Dermatol*. 2015;172:1303–7.
2. Tsai YJ, Lin PY, Chew KY, Chiang YC. Dermatofibrosarcoma protuberans in children and adolescents: Clinical presentation, histology, treatment, and review of the literature. *J Plast Reconstr Aesthet Surg*. 2014;67:1222–9.
3. Posso-de Los Rios CJ, Lara-Corrales I, Ho N. Dermatofibrosarcoma protuberans in pediatric patients: A report of 17 cases. *J Cutan Med Surg*. 2014;18:180–5.
4. Valdivielso-Ramos M, Torrelo A, Campos M, Feito M, Gamo R, Rodriguez-Peralto JL. Pediatric dermatofibrosarcoma protuberans in Madrid, Spain: Multi-institutional outcomes. *Pediatr Dermatol*. 2014;31:676–82.
5. Kornik RI, Muchard LK, Teng JM. Dermatofibrosarcoma protuberans in children: An update on the diagnosis and treatment. *Pediatr Dermatol*. 2012;29:707–12.
6. Valdivielso-Ramos M, Hernanz JM. Dermatofibrosarcoma protuberans in childhood. *Actas Dermosifiliogr*. 2012;103:863–73.
7. Dermatofibrosarcoma protuberans. NCCN Guidelines for treatment of cancer by site. [access 19 Mar 2015]. Available from: www.nccn.org
8. Gooskens SL, Oranje AP, van Adrichem LN, de Waard-van der Spek FB, den Hollander JC, van den Ven CP, et al. Imatinib mesylate for children with dermatofibrosarcoma protuberans. *Pediatr Blood Cancer*. 2010;55:369–73.
9. Barysch M, Weibel L, Neuhaus K, Subotic U, Schärer L, Donghi D, et al. Dermatofibrosarcoma protuberans in childhood

treated with slow Mohs micrographic surgery. *Pediatr Dermatol.* 2013;30:462–8.

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Abdominal Wall Angiosarcoma in a Patient With Drug-Induced Immunosuppression Following Renal Transplantation[☆]



Angiosarcoma de pared abdominal en paciente con inmunosupresión farmacológica por trasplante renal

To the Editor:

An 80-year-old woman with a history of hypertension and type 2 diabetes mellitus had been under treatment with ciclosporin (75 mg/d) and mycophenolate (1500 mg/d) since a 2004 kidney transplant for chronic renal failure. An infiltrating ductal carcinoma led to left mastectomy and axillary clearance in 2006 without adjuvant chemotherapy or radiotherapy. She was referred to the dermatologist for a lesion that had appeared on her abdomen 2 months earlier. The only symptom was a pulling sensation. The lesion formed a large reddish-purple bruise-like plaque covering almost the entire lower half of the abdomen. Nodular structures were also present (Fig. 1). No signs of adenopathy, ascites, or other cutaneous lesions were found. Histology revealed neoplastic endothelial cells forming channels that dissected collagen fibers. The cells exhibited atypia with loss of nuclear-cytoplasmic ratio, pleomorphism, and abundant mitoses (Fig. 2). Immunohistochemistry was positive for CD31 (Fig. 3) and CD34 markers in tumor cells and negative for human herpes virus 8. A diagnosis of angiosarcoma was established. A computed tomography scan of the abdomen revealed dermal tumors infiltrating the subcutaneous tissue as far as the abdominal wall below the umbilicus and mild ascites possibly related to peritoneal carcinomatosis. The patient was referred to the oncology department, where she declined the recommended chemotherapy. One month after diagnosis, she was admitted. Her general condition had declined, she had edema in the lower extremities and evident ascites. Paracentesis confirmed peritoneal infiltration of ascitic fluid secondary to the angiosarcoma. She died 2 weeks later from kidney failure.

Angiosarcoma, or malignant angioendothelioma, is an uncommon tumor of endothelial cells that accounts for only 2% of all sarcomas.¹ It is an aggressive tumor with a 5-year

survival rate of 50% to 60%.¹ Mean survival after diagnosis ranges from 18 to 28 months.² Three types of angiosarcoma are traditionally described. The first is idiopathic cutaneous angiosarcoma, which appears on the head and scalp of older patients.³ The second and third are angiosarcoma secondary to chronic lymphedema (Stewart-Treves syndrome) and a type that appears in skin exposed to radiotherapy. This tumor generally presents as a lesion that resembles bruising, with erythematous-violaceous nodules. The various types



Figure 1 Large bruise-like reddish-purple plaque covering nearly the entire lower half of the abdomen.

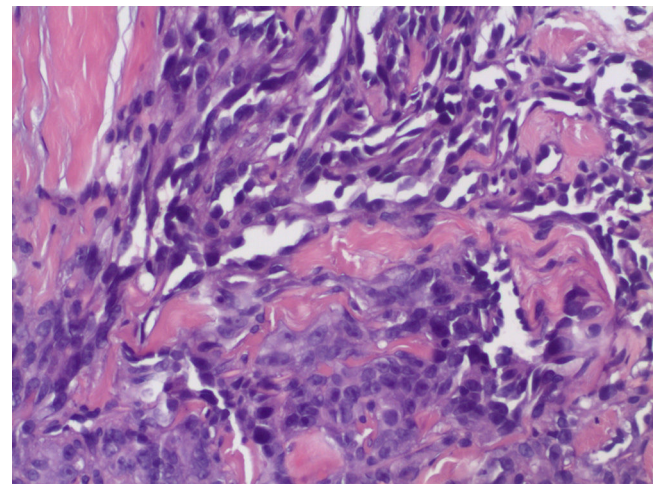


Figure 2 Anomalous proliferation of blood vessels forming structures that resemble channels (hematoxylin-eosin, original magnification $\times 200$).

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