

Supplementary Online Content

eTable 1. 2000 - 2017 case statistics of Degos' disease

Case NO.	Reported year	Gender	Age	Age at onset of cutaneous manifestations	Time to the development of systemic manifestations	Organs involved	Whether presented during the pregnant	Cutaneous histopathology	Variant	Systemic Treatment	Outcome
1	2000[1]	M	44 Yrs	42 Yrs	3 Yrs	Skin, eyes, GIT, heart	—	Ulceration, degeneration of collagen fibers in the superficial dermis, perivascular lymphocytic infiltration of the remaining skin, and annular deposition of mucin. No vasculitis or thromboemboli were seen.	Malignant	NA	Dead
2	2000[2]	F	47 Yrs	NA	NA	Skin, lungs, heart, GIT	—	Hyperkeratosis, epidermal atrophy, dermo-epidermal separation, edema, and necrosis in the papillary dermis. Fibrinoid necrosis and thrombosis were seen in the papillary dermis and in the vessels below the lesions.	Malignant	anti-inflammatory drugs	ND
3	2000[3]	M	47 Yrs	NA	NA	Skin, penile (ulceratio	—	Cutaneous tissue necrosis, surrounded by mucinous infiltration and a perivascular lymphohistiocytic infiltrate	Malignant	Aspirin, dipyridamole, cyclosporine, heparin, tacrolimus,	Dead

						n), GIT					prednisolone, azathioprine and cyclophosphamide	
4	2000[4]	M	47 Yrs	44 Yrs	3 Yrs	Skin, heart	—	Markedly thin smooth muscle of vessels with invasion of lymphoid cells.	Malignant	Aspirin, cardiac catheterization	Alive	
5	2001[5]	M	48 Yrs	48 Yrs	2 Mos	Skin, GIT	—	NA	Malignant	corticosteroids, heparin, cyclosporin, tacrolimus, aspirin, and clopidogrel	Dead	
6	2001[6]	M	52 Yrs	52 Yrs	—	Skin	—	Wedge-shaped dermal infarct with thrombosed vessels at the apex	Benign	aspirin	Alive	
7	2001[7]	M	53 Yrs	47 Yrs	—	Skin	—	Fully developed lesions exhibited a prominent lymphocytic vasculitis as evidenced by intramural lymphocytes, mural fibrin deposition, and intraluminal thrombi with a surrounding lymphocytic infiltrate containing lymphocytic nuclear debris.	Benign	pentoxifylline	Alive	
8	2001[8]	M	71 Yrs	NA	—	Skin	—	NA	Benign	pentoxifylline and aspirin	Alive	
9	2002[9]	F	32 Yrs	22 Yrs	—	Skin	Yes	Vessels adjacent and deep to the infarct show a dense perivascular infiltrate. A small arteriole deep to the apex of the wedge demonstrates endothelial hyperplasia and occlusion.	Benign	No treatment	Alive	
10	2002[10]	F	7	2 Mos	—	Skin	—	Dermal–epidermal band-like infiltrate	Benign	Aspirin and	Alive	

			Mos					together with superficial and deep perivascular mononuclear infiltrates. Dermal-hypodermal vessels showed lymphocytic infiltration of the vessel walls, fibrinoid necrosis, endothelial swelling and disruption of the intima.		dipyridamole	
11	2003[11]	F	56 Yrs	NA	NA	Skin, GIT	—	Acute stenotic endovascular proliferation with thrombosis in the absence of any appreciable inflammation, infiltrated in the walls of venules and small arteries.	Malignant	ND	Dead
12	2003[12]	F	24 Yrs	24 Yrs	1 Mo	Skin, GIT	—	Clearly demarcated necrotic and atrophic epidermis, and lymphocytes and neutrophils infiltrating in the vicinity of vessels in the dermis. Arterioles occluded by thrombi in the mid-dermis were seen in a serial section	Malignant	Nicotine patches	Alive
13	2003[13]	M	26 Yrs	13 Yrs	—	Skin	—	At the base of the area of necrobiosis, serial sections revealed an occluded vessel with a dense lymphoid cell infiltration. Marked oedema with perivascular and periadnexal lymphohistiocytic infiltrate in the reticular and middermis as well as thickening of the vessel walls with deposition of fibrinoid material.	Benign	acetylsalicylic acid and dipyridamole	Alive
14	2003[14]	M	19 Yrs	16 Yrs	3 Yrs	Skin, eyes, penile	—	ND	Malignant	steroids, dapsone, rifampicin ,cyclophos	Dead

						(ulceration), vocal cords, lungs, kidney				phamide, aspirin, dipyridamole, pentoxifylline, cyclosporine	
15	2003[15]	F	22 Yrs	NA	—	Skin	—	Wedge-shaped mucin deposits and sclerosis in the upper reticular dermis. Vacuolar interface dermatitis. Sclerosis with intact elastic fibers.	Benign	Aspirin	Alive
16	2004[16]	M	34 Yrs	14 Yrs	—	Skin	—	The epidermis is atrophic, and the dermis is hypocellular. The perivascular mononuclear cell infiltration, thrombosis, and thickening of the vessel walls with deposition of fibrinoid material.	Benign	Aspirin	Alive
17	2004[17]	F	56 Yrs	50 Yrs	6 Yrs	Skin, lungs	—	A wedge-shaped zone of necrosis, scattered necrotic keratinocytes, mild vacuolar change, dermal edema with mucin, and a sparse perivascular infiltrate.	Malignant	Dipyridamole and aspirin	Dead
18	2004[17]	F	40 Yrs	38 Yrs	—	Skin(ANA positive)	—	A wedge-shaped area of necrosis in the dermis with abundant mucin deposition. There was a sparse superficial perivascular infiltrate of lymphocytes.	Benign	Aspirin	Stable through 30 months of follow-up
19	2004[17]	F	63 Yrs	61 Yrs	—	Skin(ANA positive)	—	A wedge-shaped area of necrosis in the dermis with abundant mucin deposition, mild dermoepidermal interface change with occasional necrotic keratinocytes,	Benign	Aspirin and dipyridamole	Stable through 7 months of follow-up

								and a sparse superficial perivascular infiltrate of lymphocytes.			
20	2004[17]	F	39 Yrs	34 Yrs	—	Skin(ANA positive)	—	Incipient focal necrosis, epidermal atrophy, vacuolar interface change, dermal mucin and a superficial perivascular lymphocytic infiltrate.	Benign	Aspirin	Stable through 16 months of follow-up
21	2004[18]	F	49 Yrs	48 Yrs	1 Yr	Skin, GIT, lungs	—	Rare inflamed vessels, occlusion of vascular lumens by fibrointimal proliferation and organized fibrin thrombi, and atrophy of vessel wall with fibrin deposition.	Malignant	Dipridamole, indomethacin, and enoxaparin	Dead
22	2005[19]	F	29 Yrs	26 Yrs	26 Yrs	Skin, CNS(brain and spinal cord)	—	Deep dermis arteriolar hyalinosis with endothelial hyperplasia and perivascular lymphomonocitic infiltrate	Malignant	Antiinflammatory drugs, immunosuppressant treatments, and plasmapheresis. Antiplatelets and antidepressant drugs.	NA
23	2005[20]	M	43 Yrs	NA	NA	Skin(including palms, soles and face), penile (ulceration)CNS,	—	Hyperkeratosis, atrophy of the epidermis, superficial perivascular lymphocytes infiltration, a sprinkling of lymphocytes along The dermo-epidermal junction in conjunction with vacuolar alteration, and an occasional necrotic keratocyte in a thinned epidermis, fibrinoid necrosis in papillary dermis and hyalinization of	Malignant	Pentoxifylline, dipyridamole and immunoglobulin	Dead

						GIT		subepidermal collagen.			
24	2005[21]	M	38 Yrs	33 Yrs	5 Yrs	Skin, penile (ulceration) CNS, GIT, lungs	—	Atrophic hyperkeratotic epidermis overlying and inverted, cone-shaped area of necrosis. Blood vessels in the dermis with narrowing of the lumens and perivascular mononuclear cell infiltration.	Malignant	Aspirin, warfarin, clopidogrel, intravenous immunoglobulin and plasmapheresis.	Dead
25	2005[22]	F	22 Yrs	22 Yrs	—	Skin (coexistent with protein S deficiency)	—	A moderate lymphohistiocytic inflammatory infiltrate was present at the lateral and deeper margins of this acellular area.	Benign	No treatment (just stopped contraceptive pills)	Alive
26	2005[23]	M	33 Yrs	27 Yrs	—	Skin	—	An occluded arteriole at the base of the wedge-shaped area of sclerosis, the vessel has a discontinuous endothelial lining and is surrounded by lymphocytes and nuclear dust.	Benign	No treatment	Alive
27	2005[24]	F	24 Yrs	16 Yrs	—	Skin	—	Striking mucin deposition was noted in the papillary and reticular dermis forming an inverted wedge-like pattern with the apex pointing to the deeper portion of the biopsy	Benign	Aspirin (and stopped contraceptive pills)	Alive
28	2006[25]	M	31 Yrs	31 Yrs	6 Mos	Skin, GIT	—	ND	Malignant	Prednisolone, aspirin, pentoxifyllin, warfarin	Dead
29	2006[26]	F	58 Yrs	NA	NA	Skin, eyes (optic nerves),	—	The biopsy showed thrombosis in the small dermal vessels and intimal hyperplasia. Epidermal atrophy,	Malignant	Methylprednisolone, antiplatelet and anticoagulant therapy.	Dead

						CNS(brain and spinal cord), GIT		myxomatous degeneration, and focal necrosis in the dermis.			
30	2007[27]	F	47 Yrs	46 Yrs	—	Skin	—	Epidermal atrophy with overlying orthokeratosis and hyalinization of collagen beneath.	Benign	ND	ND
31	2007[28]	F	16 Yrs	10 Yrs	—	Skin	—	Hematoxylin-eosin stained sections reveal mild epidermal acanthosis and a lymphocytic infiltrate in the papillary dermis.	Benign	Aspirin	Alive
32	2007[29]	F	38 Yrs	37 Yrs	1 Yr	Skin, GIT	—	Epidermal atrophy and a large acellular area in the dermis in a well-developed lesion. An altered vessel below the dermal necrobiotic zone with the lumen occluded by a thrombus in a newly developed lesion.	Malignant	Warfarin, dipyridamole and lipoprostaglandin E1, IVIG	Stable through 11 months of follow-up
33	2008[30]	F	40 Yrs	38 Yrs	16 Mos	Skin, GIT	—	ND	Malignant	ND	Dead
34	2008[31]	M	60 Yrs	56 Yrs	4 Yrs	Skin, GIT, CNS, eyes(optic nerves)	—	The presence of swollen endothelial cells and lymphocyte-mediated vasculitis with fibrinoid necrosis, in absence of mucine deposits.	Malignant	Acenocoumarol, methylprednisolone, IVIG, anti-TNF α	Dead
35	2008[32]	F	2 Yrs	16 Mos	8 Mos	Skin, GIT	—	Epidermal atrophy, hyperkeratosis, necrobiotic dermal collagen, vessel	Malignant	Aspirin, dipyridamole	Alive

								thrombosis, inflammatory infiltrate, mucin deposition			
36	2008[33]	M	59 Yrs	57 Yrs	2 Yrs	Skin, GIT	—	ND	Malignant	ND	Dead
37	2008[34]	M	48 Yrs	45 Yrs	3 Yrs	Skin, GIT, CNS, heart, lungs	—	NA	Malignant	NA	Dead
38	2008[35]	F	75 Yrs	72 Yrs	3 Yrs	Skin, GIT	—	Central epidermal necrosis overlying a zone of amorphous eosinophilia in dermis surrounded by lymphocytes. Amorphous dermis eosinophilia consistent with infarction. Central small blood vessel with thickened wall with focal fibrinoid changes surrounded by lymphocytes. Increased connective tissue mucin in dermis at the edge of an infarct.	Malignant	Prednisolone (balloon dilatation for splanchnic vessel stenosis)	Stable through 12 months of follow-up
39	2009[36]	F	43 Yrs	NA	NA	Skin, GIT	—	There were fibrin-platelet thrombi in some of the dermal vessels associated with a mild perivascular lymphocytic infiltrate. Immunofluorescence studies revealed no deposition of IgG, IgM, IgA, C3, C1q or fibrinogen.	Malignant	Aspirin	Dead
40	2009[37]	M	6 Mos	6 Mos	3 Wks	Skin, CNS	—	Epidermal atrophy, wedge-shaped dermal necrosis, mucin deposition.	Malignant	Prednisolone	Dead
41	2009[38]	M	47	40 Yrs	7 Yrs	Skin, GIT	—	Atrophic and ulcerated epidermis and an	Malignant	Antiplatelet,	Dead

			Yrs					inverted wedge-shaped area of sclerosis in the underlying dermis and mucin deposition.		anticoagulant, and pentoxifyline	
42	2009[39]	F	41 Yrs	38 Yrs	3 Yrs	Skin, GIT, CNS	—	Absence of crests and atrophic epidermis. Intense perivascular inflammatory infiltrate.	Malignant	Corticosteroids, aspirin, ticlopidine	Stable through 18 months of follow-up
43	2009[40]	M	18 Yrs	14 Yrs	4 Yrs	Skin, GIT	—	Epidermal atrophy, wedge-shaped dermal necrosis, mucin deposition.	Malignant	Aspirin, Dipyridamole, methylprednisolone	Alive(but without follow-up)
44	2010[41]	F	36 Yrs	30 Yrs	—	Skin	Yes	Lobular panniculitis with lymphocytic and plasmocytic infiltrate, superficial hyalinosis, thrombus without inflammation, mucin deposition, and a wedge shaped zone of necrosis.	Benign	Prednisone, hydroxychloroquine	Alive
45	2010[42]	F	37 Yrs	36 Yrs	1 Yr	Skin, GIT	—	A raised epidermis with focal ulceration overlying a dense infiltrate of lymphocytes and thrombosis in subcutaneous blood vessels	Malignant	Methylprednisolone, IVIG, anticoagulation	Dead
46	2011[43]	M	15 Yrs	13 Yrs	2 Yrs	Skin, GIT	—	Hyperkeratosis, atrophic epidermis, basal layer hydropic degeneration in epidermis, on dermis collagen deposition and subendothelial sclerosis in arterial wall in segmental foci that caused ischemic infarct leading to atrophy of adnexal structures.	Malignant	Aspirin, Dipyridamol	Dead

47	2011[44]	F	5 Yrs	3 Mos	15 Mos	Skin, CNS, eyes	—	Central epidermal atrophy, papillary dermal sclerosis, and a sparse perivascular lymphocytic inflammatory infiltrate.	Malignant	Aspirin	ND
48	2011[45]	F	30 Yrs	30Yrs	6 Mos	Skin, GIT, pancreas, spleen	—	Wedge-shaped areas of altered dermis covered by atrophic epidermis with slight hyperkeratosis. Mucin deposition, moderate sclerosis, and collagen homogenization.	Malignant	Alemtuzumab, tacrolimus	Dead
49	2011[46]	M	41 Yrs	41 Yrs	Simultaneous	Skin, CNS, kidney	—	Narrowing and occlusion of small caliber blood vessel lumen; secondary to intimal proliferation and thrombus. The vessel wall was PAS positive.	Malignant	Methylprednisolone, hemodialysis	Alive
50	2011[47]	M	32 Yrs	32 Yrs	—	Skin(coexisted with SLE)	—	Dermal edema, mucin deposition and lymphocytic vasculitis.	Benign	Hydroxychloroquine and aspirin	Alive
51	2011[48]	F	15 Mos	Congenital	9 mos	Skin, CNS, GIT	—	Mid-dermal oedema, perivascular lymphocytic infiltration, endothelial swelling and fibrinoid necrosis of the affected vessels with loss of lumen caliber.	Malignant	Aspirin, heparin	Dead
52	2012[49]	M	42 Yrs	42 Yrs	Simultaneous	Skin, GIT, CNS	—	Ulceration with parakeratosis, abscesses, and altered collagen fibers. Perivascular lymphocytic infiltration in the upper dermis.	Malignant	Methylpredisolone, cyclophosphamide, heparin, salpogralate	Alive
53	2013[50]	M	46 Yrs	47 Yrs	Cutaneous involvement after	Skin, GIT, heart, lungs	—	A thrombotic microangiopathy along with dermal chronic microvascular changes characterized by thickened basement	Malignant	Ecuzumab, IVIG, low molecular weight heparin,	Stable through 30 months of

					systemic manifestations			membrane zones along with superficial vascular ectasia and foci of vascular dropout.			follow-up
54	2013[51]	F	64 Yrs	62 Yrs	2 Yrs	Skin, GIT	—	Wedge-shaped degeneration of dermal collagen and thrombotic vessels at the bottom of the lesion.	Malignant	Heparin, warfarin, dipyridamole	Dead
55	2013[51]	F	53 Yrs	51 Yrs	—	Skin	—	Edematous papillary dermis with inflammatory cell infiltration at the center of the lesion, perivascular lymphocytic infiltrate encroaching upon the vascular walls at the periphery of the lesion.	Benign	Prednisolone	Alive
56	2013[52]	F	42 Yrs	39 Yrs	3 Yrs	Skin, GIT, heart, bladder, CNS	—	Skin biopsy of upper arm lesion was initially read as “consistent with lupus”.	Malignant	Hydroxychloroquine, chloroquine, mycophenolate, cyclophosphamide, azathioprine, treprostinil	Stable through 3 years of follow-up
57	2013[52]	M	17 Yrs	15 Yrs	2 Yrs	Skin, GIT, bladder, CNS	—	Wedge-shaped area of infarction, epidermal atrophy with collagen degradation.	Malignant	Ecuzumab, treprostinil	Alive
58	2013[53]	M	50 Yrs	48 Yrs	2 Yrs	Skin, GIT, lungs	—	ND	Malignant	Aspirin, clopidogrel, and prednisolone	Dead
59	2014[54]	F	34 Yrs	33 Yrs(8 months duration)	—	Skin	—	Extensive epidermal necrosis with foci of re-epithelialization and superficial and deep perivascular inflammatory infiltrate in the dermis.	Benign	Aspirin	Alive

60	2014[54]	M	44 Yrs	44 Yrs(6 months duration)	—	Skin	—	An atrophic epidermis and dermal fibrosis with absence of pilosebaceous units.	Benign	Anticoagulant	Alive
61	2014[55]	M	50 Yrs	NA	—	Skin	—	An ulcerated epidermis overlying a wedge shaped area of necrosis in the dermis, with eosinophilic and densified collagen fibers and a decrease of dermal capillaries.	Benign	ND	ND
62	2014[56]	M	9 Yrs	6 Yrs	3 Yrs	Skin, CNS, GIT	—	Epidermal atrophy, hyperkeratosis, degeneration of basal cell layer, perivascular inflammatory infiltrate, mucin deposition.	Malignant	Aspirin, methylprednisolone, Immunoglobulin	Stable through 2 years of follow-up
63	2014[57]	M	14 Yrs	14 Yrs	Cutaneous involvement after systemic manifestations	CNS, skin	—	Lymphocytic vasculitis characterized by perivascular lymphocytic infiltration, mucin deposition	Malignant	Immunoglobulin, methylprednisolone, cyclophosphamide, enoxaparin	NA
64	2014[58]	F	73 Yrs	71 Yrs(18 months duration)	2 Yrs	Skin, GIT	—	Wedge-shaped necrosis in the dermis and interstitial infiltration of lymphocytes as well as extravasation of erythrocytes.	Malignant	Aspirin, Dipyridamol	Dead
65	2014[58]	F	39 Yrs	33 Yrs	—	Skin	—	Non-inflammatory endarterial thrombotic occlusion, wedge necrosis, and infarction of the dermis, can further confirm the diagnosis.	Benign	Aspirin, Dipyridamol	Alive

66	2014[59]	M	38 Yrs	37 Yrs	—	Skin	—	A wedge-shaped area of degenerated collagen with the apex extending into the dermis, which was oedematous and contained mucin deposits.	Benign	PEG-IFN	Alive
67	2015[60]	F	45 Yrs	25 Yrs	—	Skin(coexisted with Behcet's disease)	—	A wedge-shaped dermal infarct with a broad base toward the epidermis. An occluded dermal vessel is visible at the base of the dermal infarct area, and a lymphohistiocytic inflammatory infiltrate is visible at the lateral and deeper margins of the dermal area.	Benign	Aspirin, dipyridamole(prednisolone, dapsone, and colchicine were ineffective)	Alive
68	2015[61]	M	74 Yrs	NA	—	Skin	—	Dermal necrosis without inflammatory infiltration, calcification and sclerosis in papillary dermis, hyalinosis of the blood vessel wall in deep layer of the dermis.	Benign	Steroid and cyclophosphamide	Alive
69	2015[62]	F	47 Yrs	41 Yrs	7 Yrs	Skin, CNS, GIT	—	Central epidermal atrophy, papillary dermal sclerosis, and perivascular lymphocytic inflammatory cell infiltrates, in addition to vascular thrombosis.	Malignant	Aspirin, dipyridamole	Dead
70	2015[63]	F	49 Yrs	41 Yrs	8 Yrs	Skin, CNS, GIT	—	The overlying epidermis was atrophic and the wedge-shaped area of avascular necrosis was visible. Surrounded by interstitial mucin deposition in the dermis.	Malignant	Plavix, dipyridamole, heparin, salpogralate, IVIG	Dead
71	2015[64]	M	68 Yrs	68 Yrs	8 Mos	Skin, GIT	—	Wedge shaped necrosis was seen in the upper part of the dermis, with various degrees of ulceration and crust formation.	Malignant	Steroid, aspirin, dipyridamole, IVIG	Dead

72	2016[65]	F	47 Yrs	47 Yrs	5 Mos	Skin, GIT	—	Widespread area of fibrosis in the dermis subjacent to an atrophic epidermis without mucin deposition.	Malignant	Corticosteroid, aspirin, pentoxifylline, hydroxychloroquine.	Dead
73	2016[66]	M	4 Yrs	4 Yrs	Cutaneous Involvement after systemic manifestations	CNS, skin, GIT	—	Epidermal atrophy, hyperkeratosis, mucin deposition, fibrin thrombi.	Malignant	Methylprednisolone, cyclophosphamide, immunoglobulin, rituximab, Infliximab, treprostinil, eculizumab, natalizumab	Dead
74	2016[67]	F	27 Yrs	26 Yrs	1 Yr	skin, GIT	—	Hyperparakeratosis and central atrophy of the epidermis as well as thrombosed venules with hyalinized walls	Malignant	Acetylsalicylic acid, IVIG	Dead
75	2016[68]	F	55 Yrs	50 Yrs	5 Yrs	Skin, GIT, CNS, heart	—	Lichenoid dermatitis with hyperkeratosis, patchy lymphocytic infiltrate, hemosiderin deposition, increased dermal mucin, and colloid bodies in the papillary dermis.	Malignant	Eculizumab, aspirin, apixaban	Stable through 6 months of follow-up
76	2016[69]	M	56 Yrs	55 Yrs	6 Mos	Skin, GIT	—	Epidermal atrophy, thrombosis of vessel in papillary dermis along with perivascular lymphocytic infiltrate, dermal sclerosis.	Malignant	Low molecular weight heparin, pentoxifylline	Dead
77	2017[70]	F	4 Mos	Congenital	—	skin	—	Epidermal atrophy, lymphocytic interface dermatitis	Benign	Aspirin, dipyridamole	Alive
78	2017[71]	F	50 Yrs	50 Yrs	—	skin	—	Lymphohistiocytic perivascular infiltrates, lymphocytic vasculitis, thrombotic	Benign	ND	Alive

								occlusion, and abundant red blood cell extravasation			
79	2017[72]	M	4 Yrs	3 Yrs	11 Mos	Skin, CNS	—	Interface dermatitis, perivascular lymphocytary inflammation, lymphocytary vasculitis and an ischemic area in the papillary dermis with no mucin deposition.	Malignant	Eculizumab, aspirin, IVIG	Dead
80	Current case, 2017	F	30 Yrs	27 Yrs	3 Yrs	Skin, GIT, CNS	Yes	The epidermis of overlying revealed hyperkeratosis, focal atrophy, acanthosis disappeared. The junction of the epidermis and the dermis has localized vacuolar degeneration.	Malignant	Aspirin, Dalteparin, methylprednisolone, alprostadil	Dead

CNS: central nervous system; F: female; GIT: gastrointestinal tract; M: male; NA: not available; ND: none declared.

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