

Supplementary Table 1 Brief description of eligible studies included in the systematic review

Ref.	Year	Study	No. of cases	Imaging results	Histologic features	immunohistochemical	Cutaneous manifestations	Rheumatologic manifestations	Other symptoms/diseases	Systemic treatments
Suri <i>et al</i>	2020	USA	1	Not reported	Diffuse dermal fibrosis with prominent fibroblastic proliferation	Spindled fibroblasts demonstrated positive for CD34; S100 and SOX10 was negative; Factor XIIIa, and Verhoeff-Van Gieson stain revealed the loss of dermal elastin fibrils	A single 0.8 cm × 0.6 cm indurated, flesh-colored to pink nodule on the dorsal surface of left hand	Joint pain in the right hand and bilateral knees	No	Not reported

Cai et al ^[6]	2020	UK	1	X-rays were negative; MRI demonstrated Dermal proliferation of bland spindled cells with thickened collagen bundles; Elastic fibers were absent thickening over DIP joints	Positive CD86, usually negative factor CD34 SMA	for but for XIIIa, and	Skin-colored nodules were present over bilateral metacarpals, proximal, and distal phalanges with fixed finger flexion deformity; papules in the inframammary	Symmetrical polyarthritis, proximal myopathy, Raynaud phenomenon	Scoliosis and benign ovarian cysts	Prednisolone, hydroxychloroquine and methotrexate 15 mg weekly
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region

Motegi <i>et al</i>	2018	Japan	MRI showed the synovitis and synovial hyperplasia of the joints and wrist in both hands	Nodule in the left hand revealed a prominent dermal fibrosis	Cells in fibrotic lesion were positive for vimentin, but not for CD34, CD68, SMA and S100 protein	Multiple reddish firm nodules located on the finger joints, palms and wrists of bilateral hands	Arthralgia in the shoulder, knee and hip joints	No	30 mg/d oral prednisolone; methotrexate (MTX, 8 mg/wk)
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Zou et al	2015	C	hi	1	na	<p>Periarticular osteopenia without joint destruction involving the ITP, MCP-geal, shoulder and hip joints</p> <p>Thickened and disorganized collagenous fibers with marked proliferation of spindle cells</p>	<p>Spindle cells were positive for vimentin, and negative for SMA, desmin and CD68. mononuclear histocytes and multinucleated giant cells were positive for CD68, but not for CD1a, CD4 or CD8; absence of elastic fibers</p> <p>Numerous flesh coloured erythematous, firm, smooth, flat, freely mobile papules and nodules, 2-5 mm in diameter and nontender to touch</p>	<p>Joint contractures without any redness or swelling involving all fingers</p>	<p>No</p>	<p>Oral thalidomide 100 mg/d; methotrexate</p>
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								Distal			
				X-rays				sclerodact			
				showed				ylitis,			
				a carpal				several	Painful		
				synoviti				firm flesh-	joints and	Diffuse	
				s and	A dermal	Positive for	colored	synovitis of	myalgia	Intensive	
				flexor	fibrosis	SMA, desmin	skin	the wrists;	and	physical	
Courtie	201	Fr	1	tenosyn	with an	and vimentin;	nodules of	forefingers	fatigue;	therapy (3	
<i>s et al</i>	4	an		ovitis of	increased	lack of elastic	5 mm	palpable	Raynaud	times/wk);	
		ce		II to V	number of	fibers	diameter	retracted	phenome	methotrex	
				fingers	fibroblasts		on the	flexor	non	ate	
				without			backside of	tendons			
				any			the hands				
				erosion			and				
							fingers				
					Thickened	Positive for	Skin	Painful			
				Not	collagen	SMA and	thickening	joints			
Trotta	201	Ita	1	reporte	fibers	vimentin	involve	(wrists,	No	Not	
<i>et al</i>	2	ly		d	occasionall	; negative for	forearm,	PIP,DIP and		reported	
					y arranged	CD34,CD68	arm and	MCP joints)			

				in a			trunk		
				'whorling'					
				pattern					
				Spindle			Firm skin-		
				cell			colored		Initiated
			X-rays	proliferatio			cutaneous		oral
			of the	n with	Positive	for	nodules (8		methylnisolone
		Po	hands	irregular	CD34	and	to 20 mm	No joint or	(32 mg/d),
Cabral	201	rt	and	collagen	vimentin	and	in size)	systemic	followed
<i>et al</i>	3	ug	joint	deposition	negative	for	located at	complaints	by
		al	MRI	and a	SMA,S100	and	the back of	no	methotrex
			were	marked	CD668		the hands,		ate (20
			normal	decrease in			over the		mg/wk)
				elastic			second		
				fibers			and third		
							MCP joints		

Jyoti Ranjan PARID A et al	201 2	In di a	1	<p>Radiogr aph of the hands demon- strated severe periartic- ular osteope- nia without any erosion</p> <p>Proliferatio n of spindle- shaped fibroblastic stromal cells and thick collagen bundles</p> <p>Myofibroblasts was positive for vimentin but negative for desmin and SMA</p>	<p>Multiple superficial, non- tender, firm, and freely mobile cutaneous nodules about 2-10 mm in size on the dorsum of both elbows and proximal and distal interphala- ngeal</p>	<p>Arthritis involved bilateral shoulder, elbow, wrist, MCP, PIP, DIP, knee, ankle and MTP</p>	No	Not reported
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joints

Jurado <i>et al</i>	2012	USA	4	Not reported	fibroblastic proliferation; Elastic fibers were absent in all cases	SMA in one of 3 cases in a myofibroblast-like pattern; other stains were negative	Four patients displayed cutaneous nodules and arthralgias	Flexion contractures /decreased motion ; Raynaud phenomenon and erosive joint disease	an upper respiratory tract infection	Methotrexate; interferon-alfa; ribavirin; and methylprednisolone
Ji <i>et al</i>	2011	China	1	Marginal bone erosions, joint space narrowing	Myofibroblast-like proliferation and thickened collagen	Vimentin and SMA were positive	Cutaneous nodules located on the par-articular sites of	Limited motion and stiffness of many joints, involving bilateral	no	IFN- α along with leflunomide (20mg per day)

			ng and fibers		hands,	hands,	
			periartic		ankles,	wrists,	
			ular		elbows	elbows,	
			osteopo		and	shoulders,	
			rosis in		shoulders	feet, and	
			DIP,			even	
			PIP,			cervical	
			MCP			joints	
			joints				
			and				
			wrists of				
			bilateral				
			hands,				
			The	Spindle-	Stiffness of	Polyarthritis	
			hand	cell	hand and	involved the	Prednisone
Watan		Ja	and foot	(fibroblast)	foot joints,	wrists, and	,
abe et	201	pa 1	showed	proliferatio	with small	DIP, no	methotrex
al	0	n	erosion	n; collagen	subcutane	metacarpop	ate
			in the	fibers were	ous	halangeal,	
			DIP and	thickened	nodules	and	

			PIP joints				metatarsal joints		
Kluger <i>et al</i>	2010	France	1	Not reported	Proliferation of SMA and spindle or stellate-shaped cells and thickened collagen fibers	CD68 positive, as well; CD34, CD163, FXIIIa, PS100, EMA and desmin were negative	Shiny, smooth, firm, mobile, flesh-colored to purplish papules and nodules, mainly on the hands	Joint stiffness and painful disabling nodules of the elbows, hands, ankles and feet	Prednisolone, 20 mg/d; methotrexate 20 mg/m ² /week
Romiti <i>et al</i>	2009	Brazil	1	Tenosynovitis of the flexors, synoviti	Sclerodermoid	Not reported	Cutaneous nodules	Joint stiffness and painful	Prednisone (0.4 mg/kg/d)

				s of					
				MCP					
				joints					
				Tenosyn					
				ovitis of	Proliferatio				
				the n of					
				flexors, spindle-					
				synoviti shaped			Firm,		
				s of the cells	Positive for	tender,	Symmetric		Oral
				second focally	nuclear and	pink-	polyarthritis		prednisone
Juliane		Br		and arranged	SMA; but not	colored	involving		(1
Aline	200	az	1	third in a	for CD1a,	nodules	the hands, no		mg/kg/d);
Paupit	9	il		(MCPs), storiform	CD34, CD68,	that were	wrists,		methotrex
<i>z et al</i>				margina pattern,	factor XIIIa or	0.2–1.5 cm	knees and		ate (25
				l erosion thickened	S-100 proteins	in	ankles		mg/wk)
				of the collagen		diameter			
				hand of fibers and					
				the dermal					
				third fibrosis					
				MCPs					

Marco <i>et al</i>	200 9	Br az il	1	<p>Erosions A and proliferatio periartic n of ular spindle- osteope shaped nia on cells the DIP focally and PIP arranged joints of in a the storiform second pattern, to the thickened fifth collagen fingers fibers and of both dermal hands fibrosis</p>	<p>Positive for anti-SMA; but CD1a, CD34, CD68 or S-100, anti-DNA, anti-Sm, anti- RNP,anti- SCL70, anti-Jo- 1, anti-SSA, anti-SSB,RF were negative</p>	<p>Firm, smooth, pink to skin- colored nodules that were adherent to the epidermis</p>	<p>Polyarthralg ia and symmetrical polyarthriti of the wrists, MCP joints, proximal and distal interphalan geal joints were noted</p>	<p>Sclerodac tyly, Raynaud' s phenome non</p>	<p>Prednisone (0.4 mg/kg/ d) and after 3 mo</p>
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Réty <i>et al</i>	2007	France	1	Both hands showed bilateral erosion of the DIP and PIP joints, of the right trapezium and acroostolysis of the third phalanx of the right-	A proliferation of the plump, uniform, spindle-shaped cells, mixed with abundant collagen and numerous small vessels	RF, anti-DNA, anti-Sm, anti-RNP, ANCA were negative	Painless subcutaneous nodules, 5-15 mm in diameter and slightly erythematous on the back of both hands and on the right elbow	Polyarthralgia involved the PIP and MCP joints, wrists, shoulders, elbows, hips, knees and temporomandibular joints, with morning stiffness lasting 2 hours	20 mg/wk of methotrexate
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			hand						
			second						
			finger						
			X-rays				Firm,		
			of the				nontender,	Polyarthralg	
			hands				skin-	ia, later	
			showed	Fibroblasti			colored	involving	A
			a single	c			nodules	bilateral PIP	combinatio
	So		erosion,	proliferatio			(5-10 mm	and DIP	n of
	ut		MRI	n,			in size)	joints;	a
Toit, et	200	h	showed	thickened	RF, and ANCA		across the	painfully	Sclerodac
al	6	Af	further	collagen,	were negative		extensor	swollen left	tyly
		ric	erosions	and			surfaces of	knee;	oral
		a	as well	dermal			her PIP	progressive	prednisolo
			as soft	fibrosis			and DIP	stiffening of	ne
			tissue				joints as	both hands	
			and				well as her		

Joyce M. Lee <i>et al</i>	2002	America	1	X-rays showed soft tissue swelling	Dermal fibrosis and interspersed histiocytes and lymphocytes	RF, anti-DNA, and ANCA were negative	Multiple, skin-to-pink-colored, firm papules were noted on the periungual areas, hands, feet, and nose	Polyarthritides and skin lesions; swelling of the PIP joints, wrists, elbows, ankles, and knee	No	Methotrexate and corticosteroids
Laura colonna <i>et al</i>	2002	Italy	1	X-rays of both hands and feet showed MCP, PIP, DIP	An increased number of spindle and stellate fibroblast-	RF, anti-DNA, anti-Scl-70, anti-RNP, anti-Ro, anti-Jo-1, and ANCA were negative	15-20 pink to skin-colored, solid, cutaneous nodules, 5-20 mm	No joint or systemic complaints	No	Oral prednisone, 25 mg/d, and hydroxychloroquine, 200 mg/d

			subcorti	like	cells		in			
			cal	with			diameter			
			erosions	monomorp			on elbows,			
				hous			the volar			
				nuclei and			tenar			
				small			aspect of			
				nucleoli			the hands;			
							the dorsal			
							surface of			
							the hands,			
							fingers			
							and knees			
			X-rays	Fibroblasti			Nodules			
			destruct	c			on the	Arthralgia		
B.		Fr	ive and	proliferatio	Negative		MCP and	and stiffness		Colchicine
Chkirat	200	an	erosive	n,	results	for	PIP,	of both	no	and
<i>et al</i>	1	ce	lesions	thickened	ANA and		elbows	wrists, and		rehabilitati
			on both	collagen,	RF		and tibia	then of the		on
			wrists	and				big toes		
			and on	dermal						

				the PIP fibrosis					
				of the					
				second					
				and					
				third					
				fingers					
				and the					
				big toes					
				Well-	An			Firm and	
				circums	expanded			flesh-	
				cribed	papillary			colored	
				margina	dermis,			skin	His elbows,
				l	with			nodules	shoulders,
Masso		A		erosions	uniform	Desmin and a-		and	knees, and Bouts of Prednisone
n C et	199	us	1	in the	plump	SMA are		measured	ankles also exudative (0.4
al	7	tr		MTP	spindle	positively		3-5 mm.	became tonsillitis mg/kg/d)
		ali		and ITP	cells			located	painful and
		a		joints of	arranged			over the	stiff
				both	haphazard			extensor	
				feet	ly			surface of	

Olivier VIITEC OQ <i>et al</i>	1996	France	1	Erosions and periartic ular osteo nia the proxima l and distal ITP joints	A dermal fibrosis with an increased number of fibroblasts	Negative results ANA and RF	for	the index fingers Stiffness of hand and foot joints, with small subcutane ous nodules	No joint or systemic complaints	A febrile eruption and inflamma tory polyarthr algia	Methotrex ate dosage 7.5 mg/wk
Matthe w H <i>et al</i>	1995	America	2	Marked erosive changes	Interlacing fascicles of plump spindle cells and fibroblastic	Negative results ANA and RF	for	Small pink nodules, 2 mm to 2 cm in diameter, overlying	Pain and swelling in his elbows, wrists, and hands	no	Prednisone ; methotrex ate

L.S.OS TLERE <i>et al</i>	1994	U ni te d 1 Ki ng do m	negative	proliferatio n Increased numbers of stellate fibroblasts and thickened poorly stained collagen fibers	CRP, ESR, anti-ANA, anti-Scl-70, anti-RNP, anti- Ro, anti-Jo-1, anti-SSB/SSA and RF were negative.	the involved joints and volar aspects of his palms and fingers Numerous pink-to- flesh- colored nodules ranging from 3 to 10 mm in diameter on the dorsum of the fingers	Pain, swelling and stiffness of the hands and finger	Raynaud' s phenome non	A- interferon 3 million units/wk; prednisolo ne 30 mg/d; methotrex ate
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Ph . L A C O U R et al	1993	France	An homoge neous deminer alizatio n of shoulde rs, knees, and hands, but no erosions	An increase in the number of fibroblasts and marked dermal fibrosis	An increase in fibronectin and tenascin deposition	and hands extending to the wrist Cutaneous nodules appeared on the dorsal aspects of the hands and the extensor surfaces of the forearms	Arthritis of the left knee; bilateral arthralgia	Sclerodac tyly	prednisone (1 mg/kg/d)
R.J. Hernan dez et	1989	Am eri	X-rays marked destruct	An expanded papillary	RF and Lyme serology were negative	Multiple skin nodules	Pain and stiffness in his wrists	Raynaud' s phenome	Prednisone ; methotrex

<i>al</i>	ca		ive	dermis,		over	his	and knees	na	ate
			changes	with a		right				
			involvin	uniform,		elbow,				
			g	plump,		knees,				
			carpus,	spindle		dorsum of				
			MTP ,	cell		the hands				
			PIP and	population		and				
			DIP	against a		fingertips,				
			joints	backgroun		skin				
				d matrix of		lesions				
				collagen		were				
						papulo-				
						nodular				
						and				
						reddish.				
Juliane	Br		MRI	Spindle-	Fibroblast-like	Severe		Sclerodactyl		
Aline	201	az	showed	shaped	cells found	cutaneous	y	and		Methotrex
Paupit	2	ili	solid	cells	that they were	nodules	contracture	No		ate 25 mg
<i>z et al</i>		an	nodular	focally	positive for	located	of the			week
			in the	arranged	nuclear and	mostly	fingers and			

dorsal in a cytoplasmic b- over the toes
subcutaneous storiform catenin extensor
tissue pattern, surface of
thickened the hands
collagen and toes
fibers and
dermal
fibrosis

ANAs, antinuclear antibodies; ANCA, anti-neutrophil cytoplasmic antibodies; RF, rheumatoid factor; CRP, C reactive protein; SMA, Smooth muscle actin; EMA, Epithelial membrane antigen; RA, rheumatoid arthritis; PsA, psoriatic arthritis; NS, nodular scleroderma; MCRH, multicentric reticulohistiocytosis; FR, fibroblastic rheumatism; DIP, distal interphalangeal joint; PIP, proximal interphalangeal joint; MCP, metacarpophalangeal joints; SMA, Smooth muscle actin; ESR, erythrocyte sediment rate

