

PACHYDERMODACTYLY PRESENTING AS BILATERAL PROXIMAL INTERPHALANGEAL JOINT SKIN THICKENING IN AN ADOLESCENT MALE: A CASE REPORT

Manisha Koirala and Sanket Bishokarma

Department of Dermatology, National Academy of Medical Sciences, Bir Hospital, Mahaboudha, Kathmandu, Nepal

ABSTRACT

Pachydermodactyly is a rare, benign form of superficial digital fibromatosis characterized by asymptomatic periarticular thickening of the skin surrounding the proximal interphalangeal joints. Because of its resemblance to inflammatory arthropathies, it is frequently misdiagnosed, resulting in unnecessary investigations and inappropriate treatment. An 18-year-old right-hand-dominant male presented with a four-year history of gradually progressive, painless periarticular thickening involving the proximal interphalangeal joints of both hands, most prominently affecting the index fingers. He denied joint pain, morning stiffness, functional limitation, preceding trauma, repetitive mechanical friction, or systemic symptoms. Dermatological examination revealed symmetrical, firm, non-tender fusiform thickening involving the lateral and medial aspects of both proximal interphalangeal joints, with preserved joint mobility. Laboratory investigations, including complete blood count, erythrocyte sedimentation rate, C-reactive protein, rheumatoid factor, and anti-cyclic citrullinated peptide antibody, were within normal limits. Ultrasonography demonstrated localized dermal thickening without increased vascularity or synovitis, while plain radiographs showed preserved joint spaces without erosions or other osseous abnormalities. Based on the characteristic clinical presentation and supportive laboratory and imaging findings, a diagnosis of pachydermodactyly was established. As the patient was asymptomatic and had no cosmetic or functional concerns, conservative management with reassurance and observation was advised. At three-month follow-up, the lesions remained unchanged without development of new symptoms. Pachydermodactyly should be considered in adolescents presenting with painless periarticular digital thickening and normal laboratory and imaging findings. Awareness of this uncommon condition facilitates timely diagnosis, prevents unnecessary invasive investigations and immunosuppressive therapy, and enables appropriate conservative management.

KEYWORDS

Adolescent, digital fibromatosis, finger swelling, pachydermodactyly, proximal interphalangeal joint, superficial fibromatosis

Received on: May 04, 2026

Accepted for publication: September 02, 2026

CORRESPONDING AUTHOR

Dr. Manisha Koirala
Assistant Professor,
Department of Dermatology,
National Academy of Medical Sciences, Bir Hospital,
Kathmandu, Nepal
Email: manisha1.koirala@gmail.com
Orcid No: <https://orcid.org/0009-0003-3681-6474>
DOI: <https://doi.org/10.3126/nmcj.v28i3.101265>

Cite this paper as: Koirala M and Bishokarma S. Pachydermodactyly presenting as bilateral proximal interphalangeal joint skin thickening in an adolescent male: a case report. *Nepal Med Coll J* 2026; 28: 265-70.

INTRODUCTION

Pachydermodactyly is a rare, benign form of superficial digital fibromatosis characterized by asymptomatic periarticular soft-tissue thickening involving the proximal interphalangeal (PIP) joints, predominantly affecting male adolescents and young adults.^{1,2} The term pachydermodactyly was introduced by Verbov in 1975 to describe this distinctive clinicopathological entity.⁷ Despite increasing recognition, the condition remains exceptionally uncommon. Based on the largest published compilation of cases, 139 patients had been reported in the literature up to 2022, underscoring its rarity.⁹ The condition typically presents as bilateral, symmetrical fusiform swelling affecting the lateral aspects of the second to fourth fingers without involvement of the underlying joints.^{2,3}

The exact etiopathogenesis of pachydermodactyly remains uncertain. Repetitive mechanical stimulation resulting from habitual finger rubbing, interlacing, or cracking has been implicated in many patients; however, numerous cases occur in the absence of any identifiable precipitating factor.^{2,4} Histopathologically, the lesion is characterized by thickening of the reticular dermis with increased collagen deposition, variable fibroblast proliferation, and occasional dermal mucin accumulation, with little or no inflammatory cell infiltrate.^{2,4,6}

Because of its periarticular distribution, pachydermodactyly is frequently mistaken for inflammatory arthropathies, particularly juvenile idiopathic arthritis and rheumatoid arthritis, as well as other causes of digital swelling, including knuckle pads, pachydermoperiostosis, and superficial fibromatoses.^{2,4} Unlike inflammatory arthropathies, patients with pachydermodactyly typically have normal inflammatory markers and autoimmune serology, while radiological and ultrasonographic examinations demonstrate isolated soft-tissue thickening without synovitis, erosive changes, or other structural joint abnormalities.^{2,4,6} Early recognition of these characteristic clinicoradiological findings is essential to prevent unnecessary laboratory investigations, invasive diagnostic procedures, and inappropriate immunosuppressive therapy.^{2,4,8}

We report the case of an 18-years-old male with bilateral pachydermodactyly involving the proximal interphalangeal joints of both hands, most prominently affecting the index fingers. This case highlights the importance

of a systematic clinicoradiological evaluation in establishing the diagnosis, excluding inflammatory joint disease, and facilitating appropriate conservative management.

CASE REPORT

An 18-years-old right-hand-dominant male undergraduate student presented to the Dermatology Outpatient Department with a four-year history of gradually progressive, painless thickening of the skin over the proximal interphalangeal (PIP) joints of both hands. The lesions had developed insidiously and gradually increased in size before remaining stable over the preceding two years. There was no history of pain, tenderness, erythema, pruritus, morning stiffness, restriction of finger movements, or functional impairment. The patient denied preceding trauma, repetitive mechanical friction, habitual finger rubbing, finger cracking, knuckle rubbing, finger interlacing, nail biting, or any other repetitive manual habits. He also denied fever, weight loss, fatigue, Raynaud phenomenon, or other systemic symptoms suggestive of inflammatory or connective tissue disorders. His past medical history was unremarkable, and there was no family history of similar lesions, rheumatologic disease, or connective tissue disorders.

Cutaneous examination revealed symmetrical, fusiform periarticular thickening involving the medial and lateral aspects of the PIP joints of all fingers bilaterally, with the changes being most pronounced over both index fingers (Fig. 1). The overlying skin was normal in colour and texture without erythema, hyperkeratosis, scaling, ulceration, or local warmth. On



Fig. 1: Clinical photograph demonstrating symmetrical fusiform soft-tissue thickening involving the medial and lateral aspects of the proximal interphalangeal (PIP) joints of both hands, with the changes most pronounced over the bilateral index fingers.

Table 1: Laboratory investigations performed in the patient

Investigation	Patient result	Reference range	Interpretation
Hemoglobin	14.5 g/dL	12-18 g/dL	Normal
Total leukocyte count	5,730/mm ³	4,000-11,000/mm ³	Normal
Platelet count	218 × 10 ³ /mm ³	150-400 × 10 ³ /mm ³	Normal
Erythrocyte sedimentation rate (ESR)	5 mm/hour	0-10 mm/hour	Normal
C-reactive protein (CRP)	1.2 mg/L	<5 mg/L	Normal
Rheumatoid factor (RF)	3.5 IU/mL	≤14 IU/mL	Negative
Anti-cyclic citrullinated peptide (Anti-CCP) antibody	<7 U/mL	<17 U/mL	Negative

palpation, the lesions were firm, non-tender, and non-adherent to the underlying structures.

Musculoskeletal examination demonstrated a full, painless range of motion of all affected joints without flexion deformity, instability, joint effusion, or clinical evidence of synovitis. Examination of the remaining joints and systemic examination were unremarkable.

Laboratory investigations were within normal limits (Table 1). Complete blood count, inflammatory markers, rheumatoid factor,

and anti-cyclic citrullinated peptide antibody were all within their respective reference ranges, making inflammatory arthropathy and connective tissue disease unlikely.

High-resolution ultrasonography demonstrated focal dermal thickening involving the medial and lateral aspects of the affected PIP joints without increased vascularity on colour Doppler examination (Fig. 2). The underlying extensor tendons, joint capsule, and adjacent osseous structures appeared normal, with no evidence of synovitis, joint effusion, or tendon

Table 2: Differential diagnosis of pachydermodactyly and distinguishing features

Condition	Clinical features	Laboratory findings	Imaging findings	Findings in the present case
Pachydermodactyly	Painless periarticular soft-tissue thickening of PIP joints	Normal	Dermal/soft-tissue thickening without synovitis or bony changes	Consistent
Juvenile idiopathic arthritis	Pain, morning stiffness, synovitis, reduced joint movement	Elevated ESR/CRP may be present	Synovitis, joint effusion, erosions in chronic disease	Excluded
Rheumatoid arthritis	Symmetrical inflammatory polyarthritis with joint tenderness	Positive RF and/or Anti-CCP, elevated inflammatory markers	Joint-space narrowing and erosions	Excluded
Knuckle pads	Well-defined fibrous plaques over the dorsal aspect of interphalangeal joints	Normal	Localized superficial fibrous thickening	Excluded
Pachydermoperiostosis	Digital clubbing, pachydermia, periostosis	Usually normal	Periosteal new bone formation	Excluded
Superficial fibromatoses	Localized fibrous nodules or plaques with variable distribution	Normal	Variable soft-tissue thickening	Excluded

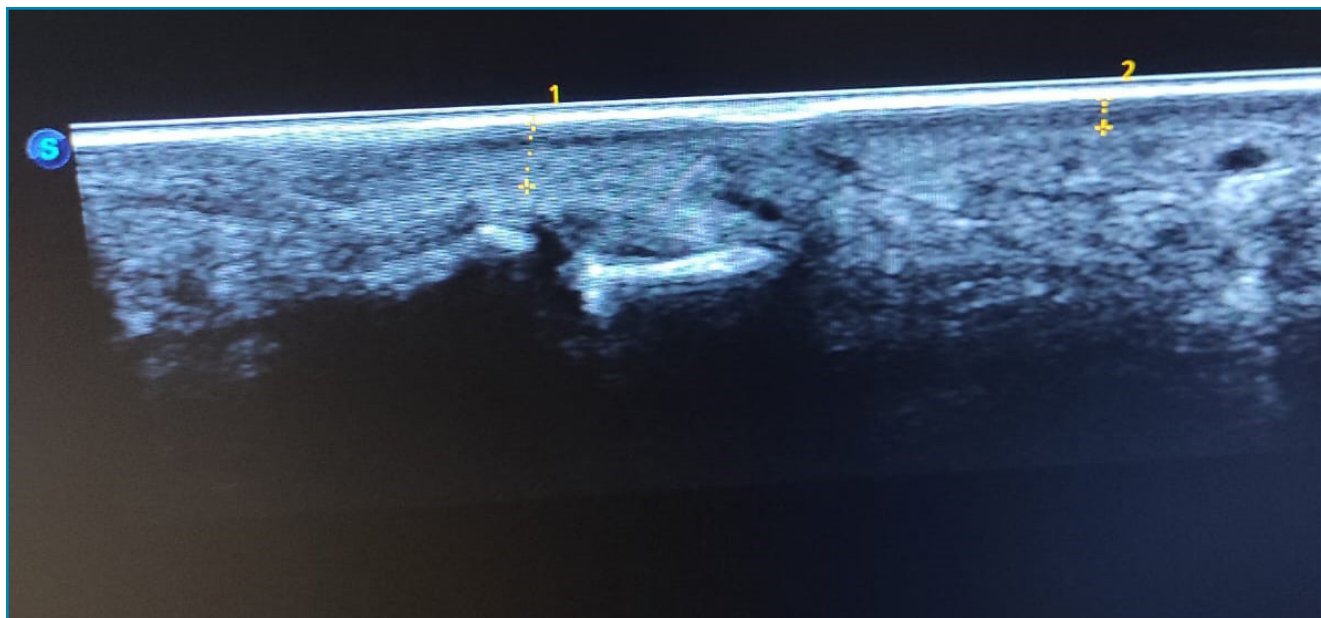


Fig. 2: Ultrasonography demonstrating focal dermal thickening involving the medial and lateral aspects of the affected proximal interphalangeal (PIP) joints.



Fig. 3: Plain radiograph (X-ray) of both hands demonstrating preserved proximal interphalangeal (PIP) joint spaces without erosions, periosteal reaction, osteophyte formation, or soft-tissue calcification

pathology. Plain radiographs of both hands demonstrated preserved joint spaces without erosions, periosteal reaction, osteophyte formation, calcification, or other osseous abnormalities (Fig. 3).

Based on the characteristic clinical presentation, normal laboratory investigations, and supportive imaging findings, inflammatory arthropathies including juvenile idiopathic arthritis and rheumatoid arthritis were considered unlikely. Other differential diagnoses, including knuckle pads, pachydermoperiostosis, and other superficial fibromatoses, were also excluded based on the

clinical morphology, distribution of lesions, and absence of characteristic radiological or systemic features. A clinical diagnosis of pachydermodactyly was established.

Skin biopsy was not performed because the diagnosis was considered sufficiently supported by the characteristic clinicrodiological findings. The patient demonstrated the typical distribution of periarticular soft-tissue thickening with preserved joint function, normal inflammatory markers, negative autoimmune serology, and absence of synovitis or structural joint abnormalities on ultrasonography and radiography. As the diagnosis was evident clinically and additional histopathological confirmation was unlikely to alter management, an invasive diagnostic procedure was considered not important.

The patient was counselled regarding the benign nature of the condition and reassured that no evidence of inflammatory joint disease was present. The patient was informed about the available management options, including observation, avoidance of repetitive mechanical trauma, intralesional corticosteroid injections, and surgical excision for selected symptomatic or cosmetically significant cases. Because the patient was completely asymptomatic, had no functional limitation or cosmetic concern, and preferred conservative management, no active intervention was undertaken. At three-month follow-up, the lesions remained clinically unchanged, with no progression, reduction in size, development of new lesions, or impairment of hand function.

DISCUSSION

Pachydermodactyly is a rare, benign form of superficial digital fibromatosis characterized by asymptomatic periarticular soft-tissue thickening, most commonly involving the proximal interphalangeal (PIP) joints of adolescent and young adult males.^{1,2} Since its recognition as a distinct clinicopathological entity, it has remained an uncommon and frequently underdiagnosed condition because its clinical presentation closely resembles inflammatory arthropathies.^{2,4} Early recognition is important, as failure to recognize this benign disorder may lead to unnecessary laboratory investigations, invasive procedures, and inappropriate treatment with immunosuppressive agents.^{2,4,8}

The precise pathogenesis of pachydermodactyly remains uncertain. Repetitive mechanical stimulation resulting from habitual finger rubbing, interlacing, cracking, or other repetitive manual activities has been proposed as a contributing factor in many patients, leading to localized fibroblast proliferation and excessive collagen deposition.^{2,4} However, not all patients report such behaviours, suggesting that repetitive trauma is not an essential prerequisite for disease development.² In the present case, the patient denied any history of repetitive mechanical trauma or habitual manipulation of the fingers, a finding that has also been documented in previously reported cases.^{3,4} Histopathologically, pachydermodactyly is characterized by thickening of the reticular dermis with increased collagen deposition, variable fibroblast proliferation, and occasional dermal mucin accumulation, with minimal or no inflammatory cell infiltrate.^{2,4,6}

Clinically, pachydermodactyly typically presents as slowly progressive, painless, symmetrical fusiform swelling involving the lateral aspects of the PIP joints, most frequently affecting the index, middle, and ring fingers.^{2,3} Functional impairment and limitation of joint movement are uncommon because the pathological process is confined to the periarticular soft tissues without involvement of the joint itself.² Our patient demonstrated the characteristic clinical phenotype, with symmetrical periarticular thickening involving the PIP joints of both hands, most prominently affecting the index fingers, while maintaining normal joint mobility and hand function.

A systematic diagnostic evaluation is essential to exclude inflammatory and structural joint disorders. Laboratory investigations are generally normal in pachydermodactyly

but are valuable in excluding inflammatory arthropathies (Table 1).² In our patient, complete blood count, erythrocyte sedimentation rate, C-reactive protein, rheumatoid factor, and anti-cyclic citrullinated peptide antibody were all within normal limits, making inflammatory arthritis unlikely. High-resolution ultrasonography demonstrated isolated dermal thickening without evidence of tendon involvement or synovitis, while plain radiographs showed preserved joint spaces without erosions, periosteal reaction, osteophytes, or soft-tissue calcification. These findings are consistent with previous reports describing imaging abnormalities confined to the periarticular soft tissues without articular destruction.^{4,6}

The principal differential diagnoses of pachydermodactyly are summarized in Table 2. The most important conditions to exclude include juvenile idiopathic arthritis, rheumatoid arthritis, knuckle pads, pachydermoperiostosis, and other superficial fibromatoses. Juvenile idiopathic arthritis and rheumatoid arthritis usually present with pain, morning stiffness, synovitis, elevated inflammatory markers, positive autoimmune serology, and progressive radiographic joint changes, features that were absent in our patient.^{2,4} Knuckle pads are well-circumscribed fibrotic plaques located over the dorsal aspect of the interphalangeal joints rather than the circumferential periarticular thickening characteristic of pachydermodactyly.² Pachydermoperiostosis is distinguished by digital clubbing, periostosis, pachydermia, and characteristic skeletal abnormalities, none of which were present in our patient.² The absence of systemic manifestations, normal laboratory investigations, characteristic ultrasonographic findings, and normal radiographic appearance collectively supported the diagnosis of pachydermodactyly while effectively excluding these important differential diagnoses.

Although histopathological examination remains the definitive diagnostic method, biopsy is not mandatory when the clinical presentation and imaging findings are characteristic.^{2,6} Histopathology primarily serves to exclude other fibromatoses or neoplastic conditions in diagnostically challenging cases. In the present patient, the characteristic bilateral periarticular distribution, absence of inflammatory features, normal laboratory investigations, and supportive imaging findings established the diagnosis with a high degree of confidence. Because histopathological confirmation would not have altered management, an

invasive diagnostic procedure was considered unnecessary.

There are no standardized treatment guidelines for pachydermodactyly because of its rarity and benign clinical course. Conservative management with patient education, reassurance, and avoidance of repetitive mechanical trauma remains the cornerstone of treatment.^{2,4} Intralesional corticosteroid injections have been reported to reduce lesion thickness in selected patients, although the evidence is limited to isolated case reports and small case series.⁴ Surgical excision has been performed primarily for cosmetic concerns or persistent symptomatic lesions but carries the risk of recurrence and unnecessary morbidity.^{2,6} As our patient was completely asymptomatic, had no functional impairment or cosmetic concern, and preferred conservative management after counselling, observation with regular follow-up was considered the most appropriate approach. At three-month follow-up, the lesions remained stable without progression or development of new symptoms.

Therefore, pachydermodactyly should be considered in adolescents and young adults presenting with painless periarticular thickening of the PIP joints and normal laboratory investigations. Careful

clinicoradiological correlation enables accurate diagnosis, distinguishes this benign condition from inflammatory arthropathies, and prevents unnecessary invasive investigations and inappropriate immunosuppressive therapy.

In conclusion, pachydermodactyly is an uncommon but important benign cause of periarticular digital thickening that may closely mimic inflammatory arthropathies. Recognition of its characteristic clinical presentation, supported by normal laboratory investigations and imaging demonstrating isolated soft-tissue involvement without joint pathology, is essential for establishing the diagnosis. Appropriate clinicoradiological evaluation can obviate the need for invasive diagnostic procedures, prevent unnecessary immunosuppressive therapy, and facilitate appropriate conservative management through patient education and reassurance.

ACKNOWLEDGEMENTS

The authors thank the patient for providing written informed consent for publication of this case report and accompanying clinical images.

Conflict of interest: None

Source of research fund: None

REFERENCES

1. Saidy E, Maalouly J, Darwish M, Cortbawi C. Pachydermodactyly: a case report and literature review. *J Orthop Case Rep* 2019; 10: 86-8. DOI: <https://doi.org/10.13107/jocr.2019.v10.i01.1650>
2. Dallos T, Oppl B, Kovács L, Zwerina J. Pachydermodactyly: a review. *Curr Rheumatol Rep* 2014; 16: 442. DOI: <https://doi.org/10.1007/s11926-014-0442-7>
3. Paravina M, Stanojević M, Jovanović D, Ljubisavljević D. Pachydermodactyly: a case report and literature review. *Serb J Dermatol Venereol* 2014; 6: 174-85. DOI: <https://doi.org/10.2478/sjdv-2014-0015>
4. Alrubaiian MT, Alharthi YH, Alfaraj S. Pachydermodactyly: an underdiagnosed condition in adolescence - a case report and literature review. *Case Rep Dermatol Med* 2025; 2025: 5560071. DOI: <https://doi.org/10.1155/crdm/5560071>
5. Nikhila PK, Santhosh P, Jensy KN, Sahadevan NV. Pachydermodactyly: a case report. *J Skin Sex Transm Dis* 2019; 1: 107-9. 10.25259/JSTD_21_2019
6. Barnes LA, Bae GH, Lewis MA, Rieger KE. Pachydermodactyly: case report including clinical and histopathologic diagnostic pitfalls. *J Cutan Pathol* 2018; 45: 949-53. DOI: <https://doi.org/10.1111/cup.13359>
7. Verbov J. Letter: Pachydermodactyly: a variant of the true knuckle pad. *Arch Dermatol* 1975; 111: 524.
8. Vázquez Fernández R, Maneiro Fernández JR, Cervantes Pérez EC, Mera Varela A. Pachydermodactyly: a systematic review. *Ir J Med Sci* 2021; 190: 1005-14. DOI: 10.1007/s11845-020-02378-1.
9. Bocutcu S, Basaran O, Batu ED. Pachydermodactyly in an adolescent boy: is it more common than we think? *Rheumatol* 2022; 61: 1298. DOI: 10.1093/rheumatology/keab453.