

Nicolau Syndrome Presenting as an Ischemic Upper Limb - Management & Functional Outcome

Dr. Radhika Yadav¹, Dr. Rupali Rastogi², Dr. Ruchi Tyagi³, Dr. Shashank Mishra⁴

¹Junior Resident, Department of General Surgery, Netaji Subhash Chandra Bose Subharti Medical College, Swami Vivekanand Subharti University, Meerut, Uttar Pradesh, India

²Junior Resident, Department of General Surgery, Netaji Subhash Chandra Bose Subharti Medical College, Swami Vivekanand Subharti University, Meerut, Uttar Pradesh, India

³Professor, Department of Physiology, Netaji Subhash Chandra Bose Subharti Medical College, Swami Vivekanand Subharti University, Meerut, Uttar Pradesh, India

⁴Professor, Department of General Surgery, Netaji Subhash Chandra Bose Subharti Medical College, Swami Vivekanand Subharti University, Meerut Uttar Pradesh, India

Abstract: Nicolau syndrome condition also known as *embolia cutis medicamentosa*, could be an uncommon complication characterized by tissue ischemia & gangrene that occurs after the infusion of drugs. The precise pathogenesis is questionable, but there are a few theories, counting coordinate harm to the end supply route and cytotoxic impacts of the mediate. Extreme torment within the quick post - injection period and purplish discoloration of the skin with a reticulate pigmentary design is characteristic of this disorder. In adverse conditions, it may cause tissue rot and compartment disorder. Determination is basically clinical, and there's no standard treatment for the illness. In this, we present a case with the conclusion of Nicolau Syndrome displaying with appendage ischemia due to diclofenac infusion; which could be an uncommon event.

Keywords: Nicolau syndrome, Tissue Ischemia, Gangrene, Diclofenac infusion, Appendage Ischemia

1. Introduction

Nicolau Syndrome may be an iatrogenic disorder, which was initially reported by Freudenthal and Nicolau in 1924 and 1925, respectively in patients treated with bismuth salts for syphilis. Nicolau syndrome is also known as livedoid dermatitis (LLD) or embolism cutis medicamentosa (ECM) and has been reported with administration of various other drugs. Nicolau syndrome is a rare complication caused by intra - muscular injection of various medications (1). It may also occur due to subcutaneous, intravenous injection. Beginning as pain and pallor over site of injection it may rapidly transform into livedoid reticular, haemorrhagic patches, and hence to ulcers and skin necrosis. The necrosis in the injection site of skin and sometimes muscle is a characteristic feature of this syndrome. Pathology is although multifactorial but pre - dominantly involves intense vasospasm with extensive perivascular inflammatory infiltrates and vascular thrombosis. The development of intense vasospasm taking after intravenous or around the vein infusion is the foremost broadly acknowledged theory in its pathogenesis [2]. Histologic findings include thrombosis of vessels in the reticular dermis either as a primary or secondary event. Treatment is mainly supportive and symptomatic.

2. Case Report

A 17 year old male from Meerut, NCR presented to us in Subharti medical college, Meerut with complaints of severe

pain in right upper limb reaching up to the right axilla and discolouration till the level of right mid - arm following history suggestive of intra arterial injection of diclofenac sodium by an unregistered medical practitioner ("jhola chaap") in a village 36 hours back.

Local examination: On inspection, the overlying skin of right hand, forearm and lower midarm was hyperaemic, tense and shiny. On palpation, severe tenderness was present even on light touch with restriction of movement on right elbow joint and right wrist. Patient was unable to move his fingers of right hand. Whole of the right upper limb up to mid arm was swollen and reddish brown in colour with petechial haemorrhages all over the limb, more prominent towards ventral aspect. Right side ulnar artery and radial artery were not palpable; very feeble right brachial pulsations were present.

Laboratory tests; showed Leukocytosis, increased Lactate Dehydrogenase 630 U/l (normal 200 - 420 U/l) with myoglobinuria.

Management: Due to worsening of condition and concern for compartment syndrome with absent peripheral pulses; patient underwent fasciotomy and debridement of necrotic tissue with interval split skin grafting. Following fasciotomy, he had improvement in pain control, skin, and neurovascular findings. He was discharged to outpatient rehabilitation, and his hand function improved and achieved full range of movement of right upper limb.

**Figure 1****Figure 2****Figure 3 (a)****Figure 3 (b)****Figure 3 (c)****Figure 4 (a)****Figure 4 (b)**

Figure 1: Nicolau syndrome presenting as limb ischemia; **Figure 2:** Fasciotomy extending from mid upper arm till thenar eminence; **Figure 3a, 3b, 3c:** necrotic tissue debridement; **Figure 4a, 4b:** Post split skin grafting with full range of movement of limb.

3. Discussion

Nicolau disorder, too known as embolia cutis medicamentosa (ECM), is characterized as an iatrogenic disorder taking after intramuscular infusions. In any case, cases with Nicolau disorder after subcutaneous, intravenous, or intraarticular infusion have been as of late detailed within the writing 3– 5.

In spite of the fact that the pathogenesis of Nicolau disorder isn't completely caught on, direct vascular harm, perivascular signs of inflammation, and vascular contraction after an infusion are thought to be the main reason 6. In expansion, it has been recommended that pharmacological properties of an individual medicate may play a part within the pathogenesis 6.

In Nicolau disorder, taking after the infusion of the clinically active agent, erythematous, ecchymosed, and reticular injuries show up within the infusion location with extreme torment. Progressive ischemic necrosis with sharp edges in a

livedoid design comes up later. Lesions frequently mend leaving atrophic scars 8.

Nicolau disorder has no authoritative treatment. Within the early period, the most objective of treatment is to avoid the advancement of necrosis. Hence, pentoxifylline, hyperbaric oxygen, intravenous alprostadil, and heparin, which strengthen the vasculature, can be utilized 4. Intralesional steroid infusion can too be viable by decreasing irritation. Surgical debridement ought to be performed within the case of necrosis 4. Systemic anti - microbials ought to be utilized in case of secondary infection 4. Contraction and deformity advancement are among late complications, and surgical treatment can be required in these cases. Nicolau disorder is unprecedented with appropriate infusion strategies – aspirating just before infusing medicine has been suggested as a strategy of anticipating this disorder 1

The current case of NS is special since the quiet created compartment disorder of his right upper appendage (lower arm) at his infusion location. The arrangement of

compartment disorder depends on different components. Palmer and Mercer portrayed front tibial compartment syndrome taking after femoral supply route perfusion in a arrangement of four patients. They hypothesized that the created compartment disorders might due to direct ischemia and harm to the femoral supply routes. Essential muscle corruption and venous harm or impediment may too contribute to improvement of compartment disorder (11).

Within the current case the appendage ischemia might be delivered by a plausible intra - arterial infusion that had created embolic hindrance of the little and medium vascular supply in its course or a likely perineural or periarterial infusion had created intense broad vasospasm, compromised microcirculation and the advancement of compartment syndrome.

4. Conclusion

Intravenous injections should be given only by a trained personnel and government policies should be directed to demotivate and ban unregistered medical practitioners at primary health care level or proper training is required to avoid these complications and limb loss, if not timely managed at tertiary care level.

References

- [1] Mott SE, Peña ZG, Spain RI, White KP, Ehst BD. Nicolau syndrome and localized panniculitis: A report of dual diagnoses with an emphasis on morphea profunda - like changes following injection with glatiramer acetate. *J Cutan Pathol*.2016; 43: 1056–61. [PubMed] [Google Scholar]
- [2] Anıl M, Çakmak B, Bal A, et al.: Nicolau Syndrome in Two Children Following Penicillin Injection: Case Report. *Türkiye Klinikleri J Pediatr*.2010; 19 (2): 144–7. Reference Source [Google Scholar]
- [3] Sonntag M, Hodzic - Avdagic N, Bruch - Gerharz D, et al.: [Embolia cutis medicamentosa after subcutaneous injection of pegylated interferon - alpha]. *Hautarzt*.2005; 56 (10): 968–9.10.1007/s00105 - 005 - 1026 - 2 [PubMed] [CrossRef] [Google Scholar]
- [4] Geukens J, Rabe E, Bieber T: Embolia cutis medicamentosa of the foot after sclerotherapy. *Eur J Dermatol*.1999; 9 (2): 132–3. [PubMed] [Google Scholar]
- [5] Cherasse A, Kahn MF, Mistrih R, et al.: Nicolau's syndrome after local glucocorticoid injection. *Joint Bone Spine*.2003; 70 (5): 390–2.10.1016/S1297 - 319X (03) 00137 - 4 [PubMed] [CrossRef] [Google Scholar]
- [6] Dadacı M, Altuntas Z, Ince B, et al.: Nicolau syndrome after intramuscular injection of non - steroidal anti - inflammatory drugs (NSAID). *Bosn J Basic Med Sci*.2015; 15 (1): 57–60.10.17305/bjbms.2015.1.190 [PMC free article] [PubMed] [CrossRef] [Google Scholar]
- [7] Ezzedine K, Vadoud - Seyedi J, Heenen M: Nicolau syndrome following diclofenac administration. *Br J Dermatol*.2004; 150 (2): 385–7.10.1111/j.1365 - 2133.2004.05808. x [PubMed] [CrossRef] [Google Scholar]
- [8] Lee MW, Kim KJ, Choi JH, et al.: A case of embolia cutis medicamentosa. *J Dermatol*.2003; 30 (12): 927–928.10.1111/j.1346 - 8138.2003. tb00351. x [PubMed] [CrossRef] [Google Scholar]
- [9] Corazza M, Capozzi O, Virgili A: Five cases of livedo - like dermatitis (Nicolau's syndrome) due to bismuth salts and various other non - steroidal anti - inflammatory drugs. *J Eur Acad Dermatol Venereol*.2001; 15 (6): 585–8.10.1046/j.1468 - 3083.2001.00320. x [PubMed] [CrossRef] [Google Scholar]
- [10] Nischal KC, Basavaraj HB, Swaroop MR, et al.: Nicolau syndrome: an iatrogenic cutaneous necrosis. *J Cutan Aesthet Surg*.2009; 2 (2): 92–95.10.4103/0974 - 2077.58523 [PMC free article] [PubMed] [CrossRef] [Google Scholar]
- [11] Palmer BV, Mercer JL. Anterior tibial compartment syndrome following femoral artery perfusion. *Thorax*.1973; 28 (4): 492–4. [PMC free article] [PubMed] [Google Scholar]