Botryomycosis with compressive brachial plexus neuropathy: A rare tropical disease with dermato-neurological association

Dear Editor,

Botryomycosis is a rare chronic granulomatous bacterial skin infection characterised by nodules, abscesses and ulcers on the hands, feet, genitals and head.[1] It usually results from the inoculation and subsequent persistent infection of injured skin by bacteria such as *Staphylococcus aureus*, *Pseudomonas aeruginosa or Escherchia Coli*.[1] It is also known as granular bacteriosis, actinobacillosis or bacterial pseudomycosis.[2] We report a patient with cutaneous botryomycosis who had compressive brachial plexus neuropathy as a result of the nodules in her axilla. We highlight this case due to its rarity and to stress upon this rarely reported complication that can result from untreated botryomycosis.

A 50-year-old female patient, who was an agricultural labourer, presented to the dermatology out-patient department with multiple painful lesions on the back exuding serous discharge since 6 months. The condition started as a single lesion on the back, which ruptured with serous discharge. Later, she developed multiple lesions extending into the bilateral axillae. The lesions were associated with pain, which exacerbated in the past one month. She also developed severe radiating pain over the left upper limb. There was no history of evening rise in temperature or weight loss. General examination revealed pallor; there was no generalised lymphadenopathy. There was mild ulnar clawing of the left upper limb and impaired touch sensation on the medial aspect of the forearm. Cutaneous examination revealed diffuse soft tissue swelling over the upper back studded with numerous sinuses, exuding serous discharge [Figure 1]. There were sinuses in the bilateral axillae too, with puckering of the skin [Figure 2]. The complete haemogram was within normal limits except for a haemoglobin of 4.8 g/dL. Peripheral smear showed microcytic hypochromic anaemia. The erythrocyte sedimentation rate was 80 mm/hour. Mantoux test was negative. Gram stain and Ziehl Nielsen stain revealed only pus cells and no organisms were seen. Pus and tissue culture sensitivity showed *Staphylococcus aureus* sensitive to cloxacillin and co-trimoxazole. Culture for acid-fast bacilli and fungus showed no growth. Serology for human immunodeficiency virus was negative. Skin biopsy revealed epidermal hyperplasia with dermis showing fibrocollagenous stroma densely infiltrated by neutrophils forming abscesses at places [Figure 3]. Periodic acid Schiff, Grocott's methenamine silver and gram stains were non-contributory. Magnetic resonance imaging showed multiple linear hyperintense inflammatory sinuses and edema of the subcutaneous and muscle plane of the nape of the neck and upper back. It also showed multiple discrete enlarged upper and lower cervical lymph nodes, causing a mass effect on the brachial plexus bilaterally [Figure 4]. A neurology opinion was sought and it was opined that the patient had left

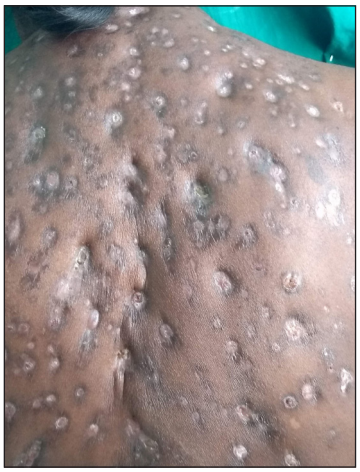

Figure 1: Multiple puckered sinuses on the back, few discharging serous liquid.

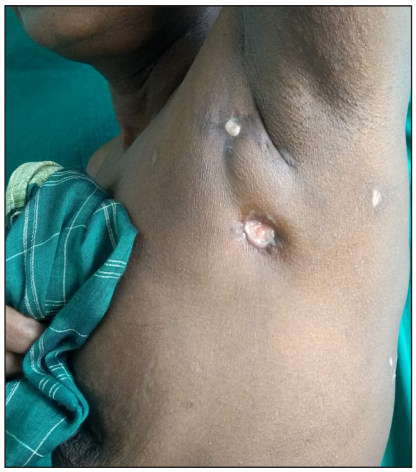

Figure 2: Left axilla showing discharging sinuses.

How to cite this article: Mohanan S, Vijaikumar M., Carounanidy U, Banushree C. S. Botryomycosis with compressive brachial plexus neuropathy: A rare tropical disease with dermato-neurological association. Indian J Dermatol Venereol Leprol. 2023;90:99–102. doi: 10.25259/IJDVL_1005_2022

Received: November, 2022 **Accepted:** January, 2023 **EPub Ahead of Print:** April, 2023 **Published:** December, 2023

DOI: 10.25259/IJDVL_1005_2022 **PMID:** 37317767

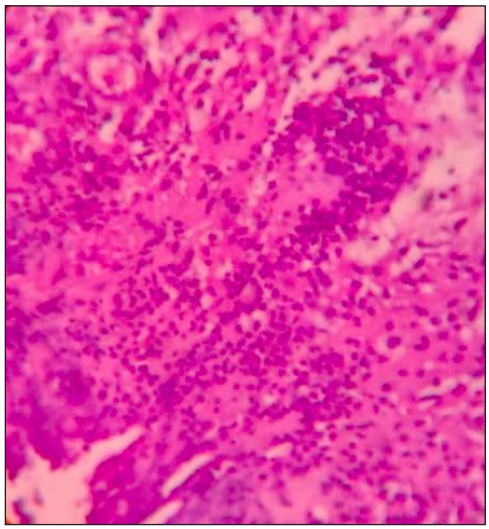

Figure 3: Histopathology showing the neutrophil collection in the dermis (H&E,100x).

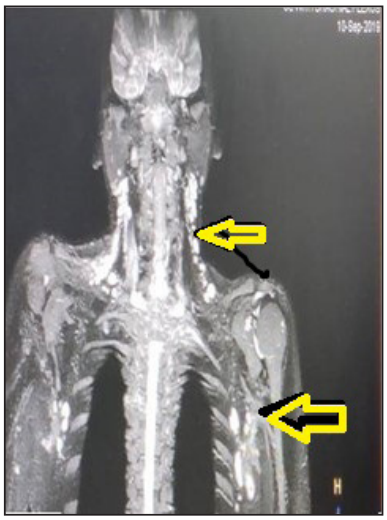

Figure 4a: Short tau inversion recovery coronal images of the neck and thorax multiple enlarged ovoid hyperintense lymph nodes, (yellow arrow) with some of them showing matting and abscess formation in the bilateral axillary region compressing the brachial plexus (yellow arrow).

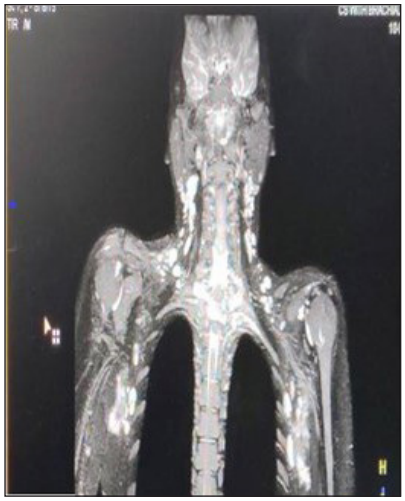

Figure 4b: Short tau inversion recovery coronal images of the neck and thorax multiple enlarged ovoid hyperintense lymph nodes, with some of them showing matting and abscess formation in the bilateral axillary region.

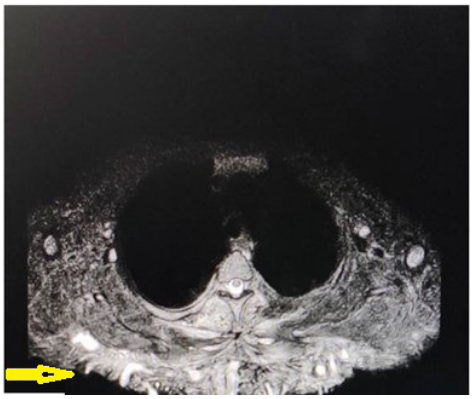

Figure 4c: Magnetic resonance imaging: Multiple linear serpiginous hyperintense sinus tracts (yellow arrow) and abscess formation noted in the posterior upper back, involving the subcutaneous and superficial muscle plane.

brachial preganglionic plexopathy. Nerve conduction studies showed that the compound motor action potential (CMAP) of the left axillary, left suprascapular, left musculocutaneous and left radial nerve were reduced by more than 50% compared to the healthy right side. The CMAP amplitude of the left median and ulnar nerve was reduced with normal latency and conduction velocity. The CMAP of the right median and ulnar nerves and the sensory nerve action potential of bilateral median and ulnar nerves had normal amplitude, latency and conduction velocity, confirming the plexopathy. The patient was started on intravenous cloxacillin 2 gm 6 hourly for 10 days. The sinuses started healing and the pain over the left upper limb started improving [Figure 5]. After discharge, the patient was asked to continue taking the tablet co-trimoxazole (Trimethoprim 160 mg/sulphamethoxazole 800 mg) twice a day. The pain completely disappeared in 3 weeks and all the sinuses started healing. The patient was later lost to follow-up during the coronavirus disease 2019 pandemic.

The first case of botryomycosis was reported by Bollinger in 1870.[3] In 1884, Rivolta proposed the term 'botryomycosis', from the Greek term 'botrys', meaning bunch of grapes, because of the characteristic groups of granules that resemble grapes and mycosis, because he thought the disease was caused by a fungus.[3] The major associated predisposing factors are skin trauma, postoperative complications, diabetes mellitus, liver disorders, treatment with steroids, alcoholism and cystic fibrosis.[3] Less common factors are malnutrition, glomerulonephritis, acquired immune deficiency syndrome, bronchial asthma and follicular mucinosis.[3] Our patient probably had malnutrition in view of her microcytic hypochromic anemia. Botryomycosis can be cutaneous or visceral.

Table 1: Differential diagnoses of soft tissue swelling with sinuses

Feature	Botryomycosis	Actinomycosis	Actinomycetoma	Eumycetoma	Nocardiosis
Agent	*Staph aureus* (commonly)	*Actinomyces israelii*	*Actinomadura* *Nocardia* species	*Madurella* mycetomatis	*Nocardia* species
Site	Extremity	Jaw, lungs, pelvis, gastrointestinal tract	Extremities	Extremities	Extremities, chest wall
Duration of evolution	Chronic	Chronic	Faster progress, more destruction, Early bone invasion[8]	Slower progress, less destruction, late bone invasion[8]	Acute to chronic
Histology	Grains with bacteria, cells, debris	Sulfur grains gram-positive filaments; nonacid fast	Grains with thin filaments	Grains with thicker filaments	Gram-positive partially acid-fast filaments
Culture media	Ordinary culture media	Anaerobic, brain heart infusion agar	Subculture: Half-strength cornmeal agar. Brain heart infusion agar	Subculture: Half-strength cornmeal agar. Saboroaud's agar	Ordinary media takes 2 weeks[7] Saborauds glucose agar without antibiotics
Treatment	Surgery with antibiotics	Penicillin G Ampicillin	Streptomycin or amikacin with co-trimoxazole	Surgical excision with antifungals	Sulfonamide

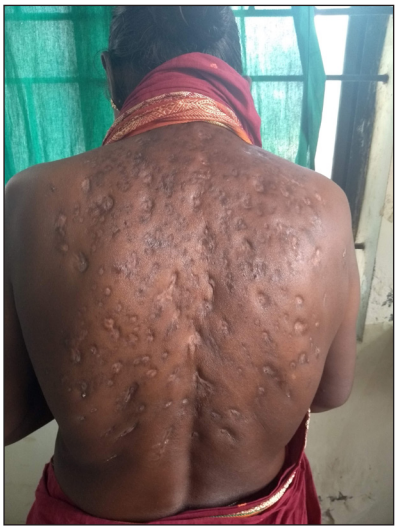

Figure 5: Healed sinuses forming puckered scars on the back.

Cutaneous botryomycosis occurs in the exposed parts of the body. Our patient developed cutaneous botryomycosis on her back. She also had swelling of soft tissues and enlarged axillary lymph nodes, causing compressive neuropathy of the brachial plexus. There are multiple reports of actinomycosis and mycetoma causing cord compression;[4,5] however, we could not find a previous report of such a complication caused by botryomycosis. The differences in the differential diagnoses of soft tissue swelling with sinuses are summarised in Table 1. Usually, botryomycosis is caused by *staphylococcus aureus*, but *Escherichia coli* and *Pseudomonas aeruginosa* have been described as causative organisms.[2] In our patient, too, materials from the pus swab grew *staphylococcus aureus*. Diagnosis usually depends on histopathological demonstration of bacteria. In our patient, since other cultures did not grow acid-fast bacterium or fungus, we narrowed our diagnosis to botryomycosis. In a 2018 review of 43 cases by Bailey *et al*.,[6] they observed that most patients responded to cephalosporins or co-trimoxazole; however, 15 patients required surgical debridement. Our patient, too, responded to co-trimoxazole. There seem to be quite a few reports from India[6], most probably due to increased reporting of these rare conditions. To conclude, botryomycosis continues to be reported from India and can be rarely complicated by compressive neuropathy, as in our patient. Early diagnosis and treatment can reduce the need for surgical debridement.

Acknowledgements

We gratefully acknowledge the contribution of Dr. S. Shrinuvasan, Associate Professor, dept of Radiology, SLIMS, Puducherry, for his contribution of MRI images of the patient

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Saritha Mohanan, Vijaikumar M.[1], Udayashankar Carounanidy, Banushree C. S.[2]

Department of Dermatology,
Indira Gandhi Medical College and Research Institute,
[1]Department of Skin and STD,
Indira Gandhi Government General Hospital and
Post Graduate Institute, Puducherry
[2]Department of Pathology,
Indira Gandhi Medical College and Research Institute,
Puducherry, India.

Corresponding author:
Dr. Vijaikumar M,

Department of Skin and STD,
Indira Gandhi Government General Hospital and
Post Graduate Institute,
Puducherry, India.
vijaiderm@gmail.com

References

1. Mechow N, Göppner D, Franke I, Kolesnik M, Bonnekoh B, Gollnick HP, *et al*. Cutaneous botryomycosis diagnosed long after an arm injury. J Am Acad Dermatol 2014;71:e155–6.
2. Bender-Saebelkampf S, Franke I, Alter M, Tueting T, Gaffal E. Cutaneous and pulmonary botryomycosis. Lancet Infect Dis 2019;19:670.
3. Bonifaz A, Carrasco E. Botryomycosis. Int J Dermatol 1996;35:381–8.
4. Oruçkaptan HH, Senmevsim O, Söylemezoğlu F, Ozgen T. Cervical actinomycosis causing spinal cord compression and multisegmental root failure: case report and review of the literature. Neurosurgery 1998;43:937–40.
5. Hussein A, Ahmed AEM, Fahal AH, Mahmoud A, Sidig A, Awad K, *et al*. Cervical cord compression secondary to mycetoma infection. Sudan J Public Health 2007;2:112–5.
6. Bailey P, Raybould JE, Sastry S, Bearman G. Cutaneous botryomycosis and staphylococcus aureus: diagnosis, management, and a systemic literature review. Curr Treat Options Infect Dis 2018;10:347–61.
7. Sommer LL, Reboli AC, Heymann WR. Bacterial diseases. In: Bolognia JL, Schaffer JV,Cerroni L.eds. Dermatology. 4th ed, China. Elsevier. 2018.p 1259–93.
8. Relhan V, Mahajan K, Agarwal P, Garg VK. Mycetoma: an update. Indian J Dermatol 2017;62:332–40.

PLACK syndrome associated with alopecia areata and a novel homozygous base pair insertion in exon 18 of CAST gene

Dear Editor,

Peeling skin syndromes (PSS) are rare autosomal recessive genodermatoses characterised by superficial painless shedding of stratum corneum.[1,2] Traupe classified PSS into Type A and B in which the former presents without skin blistering and the latter is a form of exfoliative ichthyosis.[3] Lin *et al* in 2015 first reported four cases with generalised peeling, leukonychia, acral punctate keratoses, cheilitis, knuckle pads and proposed an acronym PLACK syndrome.[2]

An 11-year-old boy born to nonconsanguinous parents presented with a history of recurrent bullae and peeling of skin subsiding with hyperpigmentation since 3 months of age. Peeling of skin with or without bullae developed spontaneously or following trauma. It was generalised till 7 years of age and later remained confined to extremities. The child also had recurrent episodes of patchy hair loss in the scalp. The child had no significant systemic illnesses. There was no history of similar lesions in the family.

Clinical examination revealed generalised xerosis, peeling of skin over dorsae of hands and feet, forearm, elbows, knees and lower legs, punctate palmoplantar keratoderma, hyperkeratotic papules over knuckles and dorsae of feet and multiple hyperkeratotic follicular papules over both lower limbs. Though all nails showed proximal leukonychia and distal onycholysis, partial nail dystrophy was seen only on toe nails [Figures 1a, 1b, 1c and 1d]. A single patch of non scarring alopecia over the vertex of scalp was noted and the dermoscopy of this patch revealed yellow dots, black dots, short vellus hair and exclamation mark sign [Figures 2a and 2b].

Skin biopsy from peeled area showed subcorneal blister and from the hyperkeratotic papule showed hyperkeratosis and acanthosis [Figures 3a and 3b].

Genomic DNA was isolated from the patient's blood sample (Qiagen). The specific exon regions from CAST gene were PCR amplified and sequenced using Sanger sequencer (ABI 3500, Thermo Scientific). The chromatogram was analysed for variants but all the previously reported mutations were absent in our patient.

Further whole exome sequencing and subsequent analysis of genomic DNA was performed (MedGenome Labs Ltd).A homozygous single base pair insertion in exon 18 of the CAST gene (chr5:g.96748570_96748571insT; Depth: 33x) encoding calpastatin that resulted in a frameshift mutation and premature truncation of the protein at codon 441 (p.Glu441Ter; ENST00000510756.5) was detected. As the patient had alopecia areata we also looked for the variant of PTPN22 (Protein tyrosine phosphatase non-receptor type 22) encoding Lyp phosphatase (Lyp R620W) and this was detected while doing whole exome sequencing.

How to cite this article: Vidya AS, Khader A, Devi K, Archana GA, Reeshma J, Reshma NJ. PLACK syndrome associated with alopecia areata and a novel homozygous base pair insertion in exon 18 of CAST gene. Indian J Dermatol Venereol Leprol. 2023;90:102–5. doi: 10.25259/IJDVL_1138_2021

Received: December, 2021 **Accepted:** March, 2023 **EPub Ahead of Print:** May, 2023 **Published:** December, 2023

DOI: 10.25259/IJDVL_1138_2021 **PMID:** 37317743