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Perspective

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Psychopathology down the Memory Lane: Ekbom's Syndrome(s)

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Description

Karl-Axel Ekbom, a Swedish neurologist has contributed lavishly to psychiatry. Three syndromes have been named after him. These are namely Ekbom's syndrome (s)-Delusional Parasitosis; Restless leg syndrome; and Pleurothotonus. Herein, we touch briefly on these entities.

Delusional Parasitosis describes (and is better referred to as) the delusion of infestation. It is often associated with tactile/haptic hallucination (formication) [1]. Historically, many misnomers have been coined (e.g. chronic tactile hallucinosis, Hypochondical paraphrenia, Progressive somatic schizophrenia). Similarly, Cleptoparasitosis is a form where the sufferer believes the infestation is in their dwelling. Delusional Parasitosis can be categorized into primary, secondary (functional), and organic forms [2]. In primary form, the delusion arises spontaneously as a monosymptomatic hypochondriacal delusional state (delusional disorder-somatic subtype), while in secondary forms, the delusional disorder arises in the context of another psychiatric disorder (e.g. in affective psychosis, paranoid schizophrenia, anankastic/paranoid personality disorders, neuroses). Organic causes include, inter alia, substance use (e.g. cocaine 'bugs'), nutritional deficiencies (e.g. Vitamin B12), medical conditions (e.g. Diabetes, HIV) and, organic brain syndromes (e.g. Dementia). Abnormalities in a fronto-striato-thalamo-parietal circuit have been speculated. Delusional Parasitosis has a bimodal distribution- the first peak for ages 20-30, is more likely to be secondary form and of male preponderance, whilst the second peak for ages older than 50s, more of primary form and has a female predilection by a ratio of 5:1. Interestingly, albeit debatable, a closely related condition known as Morgellons was described as a dermatopathy characterized by crawling or stinging sensation, finding fibres on skin and various rashes or sores. This entity is strongly believed to reflect delusional parasitosis. Typically recognized is 'Matchbox' sign (or 'Specimen' sign or 'Baggies' sign) where patients tend to bring a specimen of presumed pathogen to convince clinicians [3]. Three variants were characterized [4] *viz*., a shared psychosis, Folie à famille or Folie partagée; delusional infestation by proxy (belief that a child or pet is infested other than oneself); and double delusional infestation (belief that oneself and other party are infested but this is not shared by the latter). Akin to Ekbom's syndrome, Hypochondiasis circumscripta referred to intradermal dysaesthesia, tactile illusions, body fantasies like experiencing tunnels under skin and an urge to extract particles from skin leading to self-destructive behaviour (cf. Dermatitis artefacta). Delusional parasitosis should be differentiated from entomophobia-a specific phobia (of insects).

Restless Legs Syndrome (RLS) also known as Allison leg jitters; Dyslysis; Irritable legs; Wittmaack-Ekbom syndrome; Anxietas tibiarum. It is characterized by an urge to move limbs with/out sensations, improvement with activity, worsening at rest and worsening at night chiefly affecting the pre-dormitum. It may be primary (Parasomnia) or secondary to iron deficiency neurodegenerative disorders, end-stage renal disease or drugs (conceptualized as secondary akathisia) [5]. It commonly occurs de novo or gets worsening in pregnancy. When confined exclusively to pregnancy, it has been designated-Gestational Restless Leg Syndrome (gRLS). Major risk factors for RLS include increasing age, female sex, smoking, sedentary lifestyle, caffeine, alcohol consumption and family history. A link between RLS and PLMD with ADHD in children has been shown [6]. Iron and dopamine systems are purportedly incriminated [7]. A family history of RLS is very common and pedigrees in these cases suggest an autosomal-dominant transmission with high penetrance. Two forms were classically described, *viz*.; asthenia crurum paresthetica (painless) and asthenia crurum dolorosa (with unpleasant sensation). Specifiers for clinical course include a chronic-persistent RLS (defined as untreated symptoms on average occurred ≥ 2/week for past year) and intermittent RLS (<2/week for past year but more than 5 lifetime events). Arms restlessness can be rarely a presentment of RLS, although more commonly seen clinically as a reflection of RLS dopaminergic augmentation (a treatment-induced paradoxical worsening of the symptoms of Restless Legs Syndrome (RLS) that is caused by long-term dopaminergic therapy, in particular with higher doses) [8].

Pleurothotonus describes lateral flexion of trunk due to spasm in paraspinal musculature. Pisa Syndrome is a truncal dystonia with twisting and bending of upper thorax and involuntary flexion of neck and head to one side. Tilting symptoms bilaterally is known as 'Metronome Pisa Syndrome'. It has been described rarely as antipsychotic-induced EPS or by ACE it is in Alzheimer's disease, hence some form of cholinergic-dopaminergic imbalance is purported [9]. Typically older females with organic brain syndromes are more vulnerable to develop Pisa syndrome. Idiopathic Pisa syndrome is characterised by an adult-onset, segmental truncal dystonia in patients with no previous exposure to antipsychotics. It occurs rarely but shows a complete resolution with high doses of anticholinergic drugs [10].

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