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RESEARCH ARTICLE

"ORAL MYIASIS IN RETT SYNDROME: A RARE ENTITY"

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Abstract

Oral Myiasis is a rare disease caused by larvae of certain dipteran flies often associated with poor oral hygiene, alcoholism, senility, suppurating lesions and severe halitosis. Here we are presenting a case of parental negligence of a 15 year old mentally retarded girl with oral myiasis which on subsequent radiographic examination and clinical correlation was diagnosed to be a case of Rett syndrome. Rett syndrome is a neuro-developmental disorder that affects girls almost exclusively with initial normal development. These patients are often initially misdiagnosed as idiopathic mental retardation, cerebral palsy, or autism. Treatment consisted of manual removal of maggots, with the help of forceps and turpentine oil, surgical debridement of wound followed by broad-spectrum antibiotics. This case gives a glimpse of social stigma regarding undiagnosed mentally retarded children and parental negligence leading to such a miserable condition of the patient.

INTRODUCTION

The word Myiasis is derived from Greek "Mya" meaning fly and "isasis" meaning disease.[1] Hope. F.W. in 1840 was the first person who coined the term "Myiasis" to describe a human disease which originated from dipterous larvae. Common etiological factor for Myiasis is dipterous larvae that breed on dead or living tissues of host, liquid body substances and ingested food.[2] The incidence of disease is higher in developing tropical and subtropical countries and less frequent in western countries.

The larvae responsible for the disease prefer to grow in warm and humid environment; therefore myiasis is preferably seen in summer months. Myiasis have been classified in two categories, Primary myiasis caused by larvae feeding on living tissue and commonly seen in animals, and rarely seen in humans. Secondary myiasis which is caused by flies that feed on dead tissues, this is more common variant and seen in patients with necrotic lesions.[3] According to sites involved in the body the myiasis can be oral, ocular, cutaneous, nasopharyngeal and intestinal and urogenital.[4]

Rett syndrome, originally reported to affect only girls, was described by Andreas Rett in Vienna as "Cerebral atrophy associated with hyperammonemia". The latter attribute is no longer considered an essential part of the syndrome. The clinical features have been described as similar to those of a uniform and progressive encephalopathy[5-7]. It is characterized by

normal development till 6 months to 48 months of age followed by gradual loss of purposeful hand movements and development of characteristic, stereotypical hand movements; loss of previously acquired speech; psychomotor retardation; ataxia; truncal apraxia; deceleration of head circumference; and autistic symptoms. A characteristic hand – wringing or hand – washing stereotypies develop. Expressive and receptive language skills become severely impaired and are associated with marked mental retardation. A loss of social interaction skills is frequently observed during the preschool years, but social interest often increases later. Originally, the condition was thought to be confined only to girls, but boys with this disorder or quite close to it have been described.[8] The estimated prevalence ranges from 1 in 10,000 to 1 in 22,000.[9]

So, Here we are presenting a case report of parental negligence of a 15 year old mentally retarded girl with oral myiasis which on subsequent radiographic examination and clinical correlation found to be a case of Rett syndrome.

Case Report :

A developmentally disabled 15-year-old girl patient presented to us as a case of Oral Myiasis secondary to dento-alveolar abscess of the anterior maxilla. She was accompanied by a female attender who according to her was a caretaker in a local orphanage where this patient was left by her parents around 5-years back.

On initial inspection patient looked severely underdeveloped according to her age, her face showed ape like features, she was not able to stand on her feet and was walking using all her four limbs, patient also did not have any communication skills and was continuously making incomprehensive sounds, her fingers were continuously in her mouth, care taker of the patient informed us that the patient was crying and that there was severe bad smell coming from her mouth following fall of her upper front tooth.

As the patient was adequately cooperative it allowed us to perform an initial local examination of her oral cavity , which revealed a large burrowing ulcerative wound in respect to anterior maxilla which showed clear infestation with maggots. Her upper lip and anterior palate was swollen with adjacent teeth displaced, there was pus discharge present from the wound. Meanwhile patients parents arrived as they were contacted by the orphanage, but it was clearly noticeable that the patient was being roughly handled by her parents (father specially), patient also started becoming increasingly agitated and kept running away from them. Patient was referred for CT scans to the medical college where she became uncooperative and so was referred to the department of paediatrics for opinion regarding sedating the patient, later her scans were performed under sedation which showed involvement of anterior maxilla but the scans also showed an interesting finding of severe cerebro-atrophic changes in her brain. Correlating the clinical & radiological findings a conclusion was drawn by the paediatric department that the patient was suffering from a rare Rett's syndrome, which was subsequently informed and discussed with us.

Patients parents were informed about the condition and the treatment required for the same, they were also informed regarding the syndrome patient was suffering from and the reason behind her abnormal appearance and cognitive skills. Patients father was not showing any interest in her treatment and was very uncooperative. Patients mother requested to continue the treatment anyways.

Patient was admitted and started with intravenous antibiotics & analgesics, with proper consents patient was successfully treated using turpentine oil and thorough debridement under sedation. Patient was discharged after 3 days with oral antibiotics and analgesics. Despite clear instructions patient and her parents never returned for follow up.

PHOTOGRAPHS

Figure 1: Myiasis of anterior maxilla in Rett syndrome patient

Figure 2: Destruction of bone and loss of teeth due to larvae in anterior maxilla.

Figure 3: Removal of larvae after application of turpentine oil in the lesion.

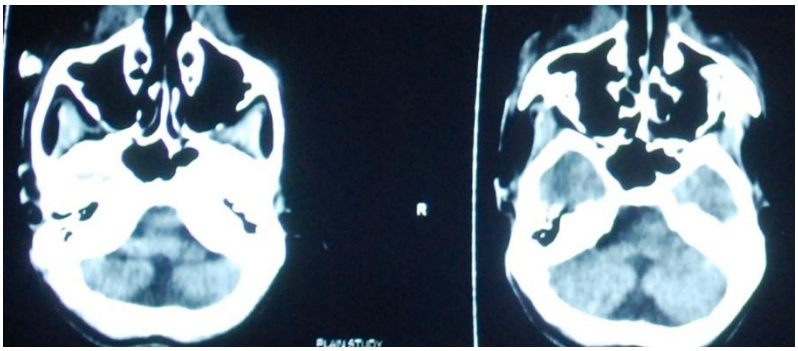

Figure 4: Brain atrophic changes in CT scans

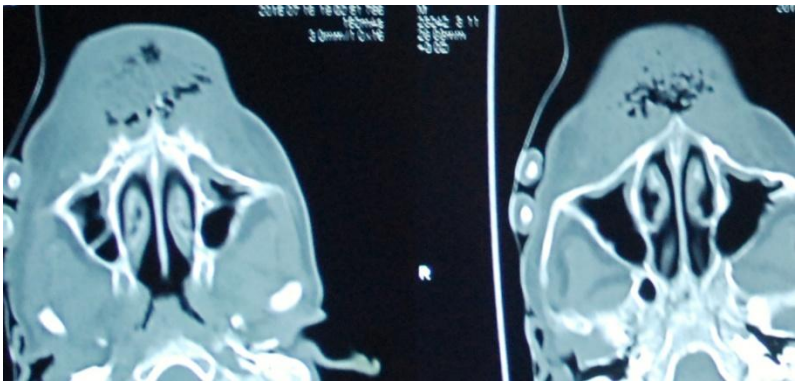

Figure 5: CT scan showing maxillary destruction due to Myiasis

Figure 6: Larvae as obtained from patient's oral cavity

Discussion:-

The term myiasis was first proposed by Hope in 1840 [10]. In Hindu mythology myiasis was considered as "God's punishment for sinners" [11].

It can be clinically classified as (a) dermal and sub dermal myiasis, (b) facial cavity myiasis, (c) wound or traumatic myiasis, (d) gastrointestinal myiasis, (e) vaginal myiasis and (f) generalized myiasis.

Primary myiasis caused by biophagous larvae (feed on living tissue), is common in cattle (called bicheiras) and is rare in humans.Secondary myiasis is caused by necrobiophagous flies (feed on dead tissue). This is a more common type and attack patients with necrotic cavity lesion. [12,13]

Myiasis of oral cavity is caused by flies of the order Diptera, larva of which develop in decaying tissue. *Musca nebulo* is the commonest Indian housefly. They are seen in abundance in human dwellings and are very active during the rainy season.

Patient reported here was mentally challenged, hence could not protect herself against flies. She was more susceptible as she was living in an orphanage with poor family care. This along with the presence of a recently exfoliated incisor provided as a fresh wound which lead to the infestation of the flies causing Myiasis.

Diagnosis of Myiasis can be made by observing the larvae as it surfaces periodically. Definitive diagnosis of exact species of fly responsible for infestation can be made only by rearing of larvae on natural or synthetic medium until they grow to adult fly stage. Ultrasound can also be used as diagnostic tool.

It is likely that poor oral hygiene, lip incompetence and lack of awareness added as contributory factors for infestation of larvae in this case [17]. The risk factors for oral Myiasis include suppurative lesions, trauma in the face, mouth-breathers, extraction wounds, alcoholics and abandoned person. [14,15,16]

Rett syndrome is characterized by profound cognitive impairment, problems with communication, stereotypic hand movements, and pervasive growth failure that follow a normal period of development during the first six to 18 months of life[17]. Diagnosis of RS is based on clinical criteria, as only 70-80% of patients with typical RS phenotype have mutations in the RS gene[18]. It is a genetic disorder transmitted in an X linked dominant fashion, 99% of the cases are sporadic, and familial recurrence is seen in less than 1%. Mutations in the MECP2 gene, located in the region Xq28, have been identified in 70-80% of patients [19]. Oral manifestations as mentioned in the literature are as shown in the table:

Oral Manifestations	*Oral Digital Habits*
• Abnormal chewing	• Handbiting
• Bruxism	• Drooling, spitting
• Hypersalivation	• Biting or rubbing of the dorsum of the hand against the teeth and lips
• Micrognathia	
• High arched palate	• Stereotypic quick mouthing of objects using both hands
• Narrow maxillary arch	• Digital-hand sucking, licking, or wetting with saliva between stereotyic hand-washing episodes
• Tongue protrusion	

(Adapted from Adkins 1986; Budden 1986; Hagberg 1986; Hanks 1986; Holms 1986; Iwata 1986; Moser 1986; Naidu 1986; Olsson and Rett 1987; Opitz 1986; Philippart 1986; Zappella 1986)

This case report shows us how parents of children with special needs and low socioeconomic background may lead them to take drastic steps such as abandoning the child.

As in most published reports, the burden of caring for a child with disability was borne primarily by the mother (Heller et al, 1997; Peshawaria et al, 1998). Reports of the effect of gender on parental stress have been variable in Indian studies. Padencheri et al (2011) reported that marital intimacy is more impaired when the child with disability is female.

India is a diverse country with regional subcultures where a female child is traditionally considered more of a burden than a male child (Gupta, 1987). A female child with disability is likely to be considered even more burdensome, raising the spectre of neglect and abuse. This was the probable cause why the child was left in an orphanage despite having a family with other siblings. Which also became more apparent when the patients father was asked about his other children, and he told us that he had other children, daughters who were "beautiful". Such is the dilemma of the society which makes a parent to abandon his own child.

CONCLUSION:-

Rett syndrome is a recently discovered, progressive neurological disorder that occurs exclusively in females early in life. Females with this rare and often misdiagnosed clinical entity have unusual but distinctive oral/digital habits. Dentists aware of Rett syndrome and its distinct manifestations can aid in early diagnosis and treatment of those afflicted with the disorder.

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