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Maxillofacial Menace in Morbid Syndromes: Review Article

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ABSTRACT

Syndrome literally means running together and it refers to collection of signs, to groups of symptoms, and to mixed assortments of signs and symptoms.The term refers to a group of manifestations when cause is poorlyunderstood.Some use the term syndrome for multiple anomalies of genetic origin. It is of paramount significance that the clinical features of syndromes especially those relating to maxillofacial and oral manifestations have a promising role to play in diagnosis.

Key words: Maxillofacial manifestations, oral manifestations, syndromes.

INTRODUCTION

The wordsyndromeis derived from Greek language and it literally means running together**.** The term refers to a group of manifestations when cause is poorly understood.[1]Face is the ferociously affected aesthetic aspect of a syndrome among the multitude of major associated pathologies. The following syndromes are a part of such prominent pathologies portraying the pathetic face in the phase of facial manifestations focusing fervently on cosmetic considerations in such situations apart from the overall health as a whole.

Caffey - silverman syndrome(infantile cortical hyperostosis,smyth's syndrome)

It was first described in 1930 by Roske, however the clinical and roentgenographic features was brought to attention by Caffey, Silverman and Smith in 1945. It is an autosomal dominant condition and its etiology has been suggested that it was caused by a congenital anomaly of the vessels supplying the periosteum of the involved bone, the hypoxia effecting a focal necrosis of the overlying soft tissuesand resulting in new periosteal bone formation. The average onset of this syndrome is 9 weeks. It is characterized by – Bilateral swelling over the mandible or other bones, Roentgenographic evidence of new bone formation in this area, Hyperirritability and Mild fever.

Systemic manifestations

Face

Symmetrical swelling over the face located over the body and ramus of the mandible.

Skeletal system

The most frequently affected bone is mandible. Others involved are clavicle, tibia, and ulna. The new periosteal bone formation appears mostly during 9th week and undergoes resolution by around 9 months; however roentgenographic evidence may persist for many years.

Oral manifestations

Involvement of the mandible was formerly thought to be necessary for diagnosis of the condition, but analysis of a large series of cases has revealed that this is not so. Nevertheless, swelling of the jaws is the most common presenting sign.[2-5]

Hutchinson-gilford syndrome(Progeria)

It was first described by Hutchinson & Gilford. It is a combination of Dwarfism, immaturity and Pseudo senility. Because of a peculiar form of hyper metabolism, persons with this affliction succumb to old age and die of coronary disease during their middle teens.

Systemic manifestations

Face & Appearance:It is disproportionately small giving the head a hydrocephalic appearance. The ears are small without lobules. The nose is beaked giving a bird facies. Eyebrows and occasionally eyelashes are lost. Scalp hair is lost and replaced by a downyfuzz, giving a newly hatched bird appearance. The chest is narrow and the abdomen protuberant.

Oral manifestations

Poor middle-face development and mandible hyperplasia are constant features. The mandible angle is155 degree (normal 120 degree).Because the jaw is small, the teeth, usually of normal size, are crowded. . In most cases, eruption of the teeth has been delayed and the deciduous, dentition is often retained. The teeth are stained yellowish brown with microscopic evidence of senile papal changes. The palate has also been found to be high. The submandibular glands are often prominent and in some, cases, may stimulate a double chin.[6-8]

Parry-romberg syndrome(progressive hemifacial atrophy)

This syndrome was first described by Parry & Romberg in 1825 & 1846 respectively.It is an autosomal dominant condition. It consists of slowly progressive atrophy of thesoft tissues of essentially half the face, accompanied most often by contralateralepilepsy, by trigeminal neuralgia & by changes in hair and eyes.Occasionally there may be associated atrophy of half the body. The suggestedetiologyis the irritation in the peripheral trophic sympathetic system.

Systemic manifestations

Face

Asymmetry noted.

Oral manifestations

Except for atrophy of half the tongue, oral involvement appears to be rare.Roentgenographic study of the jaws revealed that the body and ramus of the mandiblewere shorter on the involved side and that there was a delay in development of theangle.In a few cases the teeth on the involved side were retarded in eruption .[9, 10]

Treacher collins' syndrome

(Mandibulo facial Dysostosis, France schetti- Zwahlen-klein Syndrome, Bilateral Facial Agenesis)

This syndrome was first described by Treacher Collins and Franchestti in 1846 and 1940 respectively. It is an autosomal dominant condition.The suggested etiology is incorrect development of blood relay (from the remains of first aortic arch to stapedial artery to external carotid artery)

Manifestations

Face

Downward sloping palpebral fissures, depressed cheek bones, deformed pinna, receding chin, large fish like mouth, tongue shaped process of hair that extends towards cheek.

Oral manifestations

The mandible is almost always hypoplastic. Roentgenographic studies have shown that the angle is more obtuse than normal and that the ramus may be deficient. The undersurface of the body of the mandible is often pronouncedly concave. The palate is noted to be high or cleft in over 40 percent of the patients. Macrostomias, ie., Failure of fusion of the maxillary and mandible processes, may be unilateral or bilateral. Its exact frequency of occurrence has not been estimated, but it is not very common. Microstomia has also been seen. Because of the poor development of the maxilla and the frequency of high or cleft palate, dental malocclusion is frequent. The teeth may be widely separated,hypoplastic, displaced, or associated with open bite. Aphasia of the parotid gland and macroglossia were also reported. [11-13]

CONCLUSION

Sometimes to appreciate, understand and value the goodness, a comparison with contrary is needed. In other words the true value of health in relation to appearance and function is felt when pathology causes loss of normal morphology and functions. Syndromes are one such pathology which arises from different etiologies resulting in mild to fatal consequences both with respect to morphology and function. Thus though nothing is completely perfect yet every creation of Almighty is beautiful in its own unique way. Ultimately, every individual yearns to see their face in good health both in terms of appearance and as part of overall function of the body.

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