INTRODUCTION :-

Fahr Disease is a rare degenerative neurological disorder characterized by the presence of abnormal calcium deposition and associated cell loss in the areas of brain that control movement, including basal ganglia and cerebral cortex. The condition was first described by Fahr in 1930. According to reports in medical literature, Fahr Disease is often familial. It is believed to have autosomal dominant inheritance but a few cases have been reported to have autosomal recessive inheritance and even some sporadic cases have been reported in literature. The association between the abnormal phenotypes and abnormal genes remain unclear despite the recent mapping to chromosome 14q of a susceptible locus for Fahr Disease.

CASE REPORT :-

A 36 year old woman presented with progressive deterioration of mentality and motor functions, extrapyramidal signs, dysarthria, ataxia, athetosis. Plain radiographs of the skull demonstrated irregular calcifications in bilateral fronto-parietal region of skull. A contrast enhanced CT scan head was done in Spiral CT scanner wipro GE which revealed bilaterally symmetrical non-enhancing hyperdense (HU + 234) lesions s/o calcification, involving globus pallidus, putamen, caudate nucleus, internal capsule, thalami, dentate nucleus, cerebellum and subcortical white matter (fig:-1&2). Blood
chemistry revealed normal serum levels of calcium, phosphorus and alkaline phosphatase. The CT Scan findings when correlated with typical clinical history and normal blood chemistry was suggestive of Fahr Disease.

DISCUSSION :-

Fahr Disease or familial idiopathic basal ganglia calcification is characterized by bilateral basal ganglia calcification. The most common site of calcification is the globus pallidus. However additional areas of calcification are putamen, caudate nucleus, internal capsule, dentate nucleus, thalamus, cerebellum and cerebral white matter. The calcium deposits occur in the extracellular and extravascular space often surrounding the capillaries. It is not clear whether the calcification in Fahr Disease is a metastatic deposition, secondary to local disruption of blood brain barrier, or is due to disorder of neuronal calcium metabolism.

Typically the age at onset of clinical symptoms is 30 to 60 years. The clinical evolution is that of a degenerative disorder, rather than a developmental disorder. The core clinical features are dysarthria, extrapyramidal signs, ataxia, progressive deterioration of mentality, loss of motor accomplishments, symmetrical spastic paralysis, athetosis, eye impairment due to optic atrophy.

There is neither a cure for Fahr Disease, nor a standard course of treatment. The prognosis is variable and hard to predict.

REFERENCES:-