

further evaluation. In cases in which the CT scan findings are indefinite or clinical suspicion of a pancreatic injury remains, further investigation with magnetic resonance cholangiopancreatography (MRCP) may be used. Although injury of the main pancreatic duct may be strongly suggested upon initial CT scanning, MRCP can demonstrate clear delineation of the duct and its integrity. Accurate recognition of major pancreatic injury is essential because delay in diagnosis and associated vascular injuries are largely responsible for the high morbidity and mortality. In majority of the cases of blunt pancreatic injury, surgical intervention is not required. If exploratory laparotomy is carried out, ductal damage may be visualized directly. Pancreatic resection is usually the most suitable treatment if CT scan or ERCP show that the duct has been damaged or transected. Minor or isolated pancreatic injury recovers well. Severe injuries have poor prognosis due to frequent association with other injuries. Pancreatic injuries continue to be an ordeal for trauma surgeons. The relatively infrequent incidence, the complexity in making an apt diagnosis and high morbidity and mortality, justify the unease these injuries provoke. The management of blunt pancreatic injuries has been controversial, with some suggesting selective observation and others advocating immediate exploration to prevent the delay-induced escalation in morbidity and death^{10, 11}. The situation may be further complicated by the presence of associated major visceral injuries in these patients.

CONCLUSION

Blunt abdominal trauma with isolated rupture of the pancreas is a rare injury. The key criterion for deciding on a treatment option is whether the main pancreatic duct is damaged or not. The value of CT scans, in

this regard, is limited by low sensitivity. Although endoscopic retrograde pancreatography or magnetic resonance imaging pancreatography is ideal for diagnosing an injury to the pancreatic duct, neither is available on an emergency basis in most hospitals. Early laparotomy appears to be the best option for patients with lesions to the pancreatic duct given that delayed surgical treatment is accompanied by increased morbidity and mortality. Also, late consequences, such as pancreatic cysts, that necessitate further therapy are likely to develop. However, if the pancreatic duct is not involved, mere drainage and a watch-and-wait strategy are usually sufficient.

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Progeria: A Case Report with Review of Literature.

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Abstract: Progeria is rare, severe genetic condition of premature ageing occurring at an early age. The incidence is one in 8 million births. Progeria is also known as Hutchinson Gilford syndrome.

INTRODUCTION

Progeria is a rare premature aging syndrome², characterized by retarded growth – dwarfism, abnormal onset of scleroderma. The classic form is known as Hutchinson Gilford progeria syndrome (HGPS).

Progeria was first described in 1886 by Jonathan Hutchinson and also by Hastings Gilford, so named as HGPS. It occurs sporadically in autosomal inheritance. Males are affected more than females 1.5:1. Caucasians are more susceptible 97%¹.

The clinical presentation is classical, conventional radiological & biochemical investigations will confirm the diagnosis. Affected children are normal at birth and growth retardation starts in 2nd year, skin becomes thin & shiny in some areas but lax & wrinkled in other areas.

We present a rare case of progeria with all the classical physical & radiological findings.

CASE REPORT

Twelve year old male child presented with progressive history of coarsening of skin, failure to thrive & with dwarfism. The child developed alopecia for the past few years.

This child of para-1 of non-consanguineous marriage, had an uneventful antenatal & prenatally history. The child was apparently normal till 1 year of age & thereafter started noticing abnormal features.

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General examination showed the child was short statured and malnourished; eyes appeared prominent, 35 inches in height and 14 kg in weight with patent anterior Fontanilla, beaked nose and crowded irregularly erupted teeth with micrognathia. (Fig-1).

Small thorax and small thin limbs. Total alopecia with prominent veins noted on the scalp, there were numerous brownish coarse thickened patches specially on the dorsum of hands & shoulder. (Fig 1.2) Terminal finger of the hand and foot are stubby. (fig 4)

Based on the history and clinical findings a provisional diagnosis of progeria was made, biochemical investigations were normal.

Skeletal survey of the child confirmed the diagnosis.

Radio graph of the skull reveals gross sutural diastasis with wormian bones, small mandible with small ascending Ramus with infantile obtuse angle with crowded irregularly erupted teeth. Fig 1

Radiograph of the chest showed sloping slender ribs with thinning of both ends. The clavicles



Fig. 1: photo-skull showing engorged vein & beaked nose & hypognathia. x-ray skull showing wide fontanelle with multiple wormian bones & hypoplastic obtuse angle ascending rami of the mandible.

Fig. 2: Photograph of both hands showing stubby terminal phalanges. X ray of the hand showing acroosteolysis.

were small and hypoplastic, long bones showed narrowing at the proximal diaphyseal end with an area of sclerosis at proximal end. Fig 3 Radiograph of the spine showed fish mouth vertebra, radiograph of the foot and hand showed resorption of terminal phalanges - acrosteolysis. Fig 4 The bone age was corresponding to chronological age. The radiological findings confirms the diagnosis of progeria.



Fig 3: x ray of the lower limb bone showing the long slender bones.

Photo of limbs showing loss of subcutaneous fat, engorged veins and sclerodermat changes.



Fig 4: Photo of both legs showing stubby terminal phalanges.

DISCUSSION

Short stature bird like facies and normal intelligence were distinctive features of progeria in our cases. Normal intelligence excluded pangeria. Typical facial changes ruled out acrogeria. Lack of photosensitivity, absence of disproportionate large extremities and ocular changes ruled out Rothmund Thompson syndrome.

GHP rare genetic disorder phenotypically characterised by many clinical features of clinical ageing. Hutchinson described the first case of progeria in 1886. Gilford was the first to propose the term progeria in 1904. The Greek word geraios meaning old refers to progeria¹. DeBusk renamed this condition as achutchinson Gilford syndrome³.

Generally the rate of ageing in progeria is accelerated to 7 times that of normal. The average life span is 13 years with a range of 7-27 years they are reports of survival till the age of 45 years⁴.

The cause of death in these patients is cardiovascular complications like myocardial infarction or congestive heart failure⁶.

The basic cause of this syndrome is still not cleared, the most probable cause proposed is lamin-A/C gene. The mutation of this gene is responsible for premature ageing,

increase in hyaluronic acid is responsible for sclerodermatous changes and cardiovascular complication and so for failure to thrive⁶.

The effected children are normal at birth and start showing the changes after 1 year ie alopecia with loss of subcutaneous fat with scleroderma changes in the skin giving typical plucked bird appearance. Scalp, hair and eye lashes are progressively lost with prominent scalp veins. These patients are usually short and thin with an average height of 100cm & weight of 12-15 kg⁷.

Patient with progeria exhibit normal bone age and normal mental age¹. The differential diagnosis includes Werner syndrome (WS), Acrogeria, Rothmund-thompson syndrome cockayne syndrome (CS). Werner syndrome is also known as progeria adulatorum, progeria of the adult and part common of the premature ageing disorders. The onset might occur in individuals in their mid-teens or individual is as old as 30 years. Both sexes are affected equally. Death occurs when patients are having atherosclerosis or malignant tumors. Acrogeria is a progeroid syndrome of premature ageing of the skin and internal organs seen in the Hutchinson-Gilford progeria syndrome. It is seen mainly in females and in familial cases are also seen (Gottron type). Acrosteolysis of the distal phalanges, delayed cranial sutural bones, linear lucent defects of the metaphyses and antegoneal notching of the mandible are the predominant features Rothmond Thomson syndrome is a hereditary and familial disease characterised by short stature, cataract, baldness, bone, nail, teeth changes. Cockayne syndrome type 1, 2, 3 of mild to severe forms characterised by growth deficiency, premature aging and pigmentation. To date, no definitive treatment is available and patients are treated conservatively.

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