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Case reports & case series

Conservatively managed extradural haematoma in a child with progeria

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ABSTRACT

Hutchinson-Gilford progeria syndrome (HGPS) is a rare genetic disorder characterised by premature ageing. We report a case of a 13-year-old girl with HGPS who presented with an extradural haematoma following head injury. A conservative approach was successfully employed in this case, avoiding the risks of general anaesthesia and surgery.

1. Introduction

Hutchinson-Gilford progeria syndrome (HGPS) is an extremely rare genetic disorder with a prevalence of one in 4–8 million [3]. It is characterised by premature ageing, severe growth failure, skin atrophy and early onset atherosclerosis [1,2]. It is caused by a gene mutation that results in abnormal Lamin A, a key supportive protein found within the nuclear membrane [3]. Abnormal Lamin A results in an unstable nuclear envelope, progressive nuclear damage and premature cell death. The average age of survival is 13.5 years (range 8–21 years) and death occurs most frequently secondary to cardiovascular complications [2,3]. We report a case of a child with HGPS who presented with an extradural haematoma which we managed conservatively.

2. Case report

A 13-year-old girl with a known diagnosis of HGPS fell down a flight of stairs at home sustaining a head injury and right clavicular fracture. She did not lose consciousness and was asymptomatic. On examination, she had a Glasgow Coma Score of 15 and was neurologically intact with a right parietal scalp swelling. Imaging demonstrated a 5.5 × 2.2 cm right fronto-parietal extradural haematoma and an overlying right parietal bone fracture (Fig. 1).

Given she was asymptomatic and considering the potential anaesthetic and perioperative risks associated with HGPS, a conservative approach was taken with close neurological observation on the paediatric high dependency unit. She remained an inpatient for one week and remained neurologically stable. Interval imaging demonstrated stable appearances. She was discharged home and reviewed in clinic with repeat imaging at one month which revealed complete resolution of the haematoma. She remained clinically well.

3. Discussion

Patients with HGPS are healthy at birth but develop characteristic skin, craniofacial and skeletal abnormalities by the age of 1–2 years [1,3]. General features of the condition are listed in Table 1. As an estimate, over the course of one year, children with HGPS biologically age by 10 years [3]. Rapidly progressive atherosclerosis of the cerebral vasculature and abnormally fragile connective tissue predispose HGPS patients to intracranial haematomas, with relatively minor trauma [1,2].

Extradural haematomas have previously been reported in patients with HGPS; our literature search revealed 2 such reports (Table 2). In both cases, patients developed extradural haematomas following minor head trauma and were treated surgically. Mandera and colleagues reported a case of a 10-year-old boy who presented with reduced conscious level and right hemiparesis. CT of the head revealed two extradural haematomas exerting significant mass effect. Both haematomas were evacuated and at discharge from hospital the boy had made a good neurological recovery with only minimal right hemiparesis [2]. Hansda et al. published a case of a 7-year-old boy who presented with confusion and symptoms of raised intracranial pressure. Imaging revealed a right parietal extradural haematoma which was evacuated. The child made an unremarkable post-operative recovery [1]. In comparison to these previously reported cases, our patient was neurologically intact and had no symptoms of raised intracranial pressure. Therefore, a conservative approach was successfully employed avoiding

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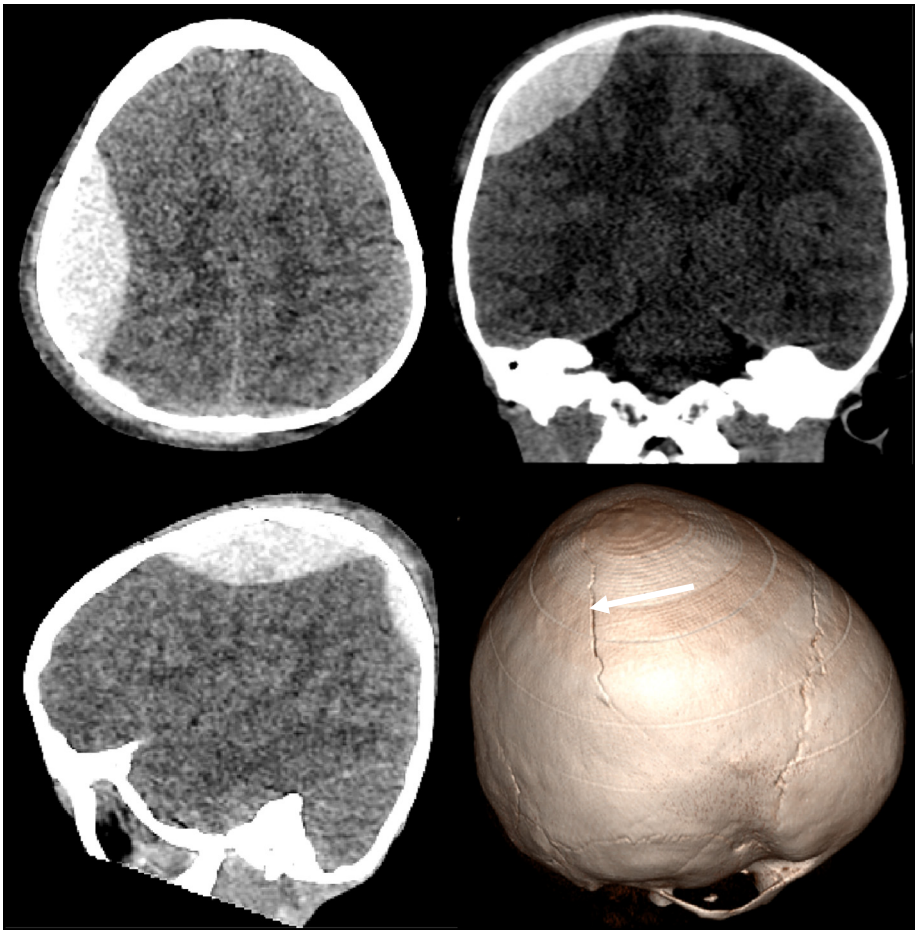

Fig. 1. Unenhanced axial, coronal and saggital CT scan showing a right fronto-parietal extradural haematoma. 3D reconstruction demonstrating a linear right parietal fracture (white arrow).

Table 1
General features of Hutchinson-Gilford progeria syndrome.[1,4,5]

System involved	Abnormalities
Craniofacial	Small face relative to head size (craniofacial disproportion), prominent eyes, sculpted 'beaked' nose, mandibular and maxillary hypoplasia, generalised alopecia, patent anterior fontanelle, high arched palate, narrow glottis, abnormal dentition, thin lips, protruding ears with absent lobes
Skin	Skin thin, wrinkled, "sclerodermatous", Subcutaneous fat almost absent
Skeletal	Short stature, decreased weight for height, joint stiffness, osteoarthritis, osteoporosis, wide-based gait and shuffling
Cardiac and cerebrovascular	Atherosclerosis, coronary artery disease, hypertension, heart failure, carotid and cerebral aneurysms
Multi-systemic involvement	Diabetes mellitus, restrictive lung disease, abnormal lipid profiles, vision and hearing loss

Table 2
Reported cases of extradural haematomas (EDH) in patients with Hutchinson-Gilford progeria syndrome.

Reference	Age (yrs), gender	Neurological symptoms/signs	Location of EDH	Size of EDH	Treatment
Mandera et al., 2003 [2]	10, male	GCS 10, right hemiparesis	2× EDH - Left posterior fossa Left temporal	 2.5 cm thick 8 × 2.5 cm	Both haematomas evacuated surgically
Hansda et al., 2013 [1]	7, male	Headaches, nausea, confusion	Right parietal	N/A	Surgical evacuation
Rajwani et al., 2019	13, female	None	Right fronto-parietal	5.5 × 2.2 cm	Conservative

the risks of general anaesthesia and surgery.

There are a number of anaesthetic concerns that should be considered in the HGPS patient. It is important to remember even though these patients have an advanced physiological age, they are psychologically young with a normal childhood intelligence [5]. They should be treated psychologically as children. Pre-operative evaluation of their airway is extremely important. Multiple craniofacial abnormalities such as micrognathia, small mouth, diminished subcutaneous fat,

a short stiff neck and poor dentition make intubation challenging [1]. In addition, severe osteoporosis may predispose to easy fracturing of the jaw with mask ventilation and laryngoscopy [6]. Sebastian and colleagues (2016) recommended using ultrasonic measurements of the subglottic area to help chose the correct endotracheal tube diameter. They also advised maintaining spontaneous respiration until a difficult laryngoscopy had been ruled out. Liessman et al. [4] secured the airway by performing a laryngoscopic-assisted fiberoptic intubation with use of a guide wire and bougie.

Children with progeria tend to develop severe atherosclerotic heart disease, angina and heart failure [5]. Up to 40% of patients have cardiac murmurs due to degenerative calcification of mitral and aortic valves [7]. Cardiac disease results in an increased risk of perioperative myocardial ischaemia [1]. Pre-operative assessment of cardiovascular function, intra- and post-operative ECG monitoring are essential in these patients [4]. Skin and skeletal abnormalities also make these patients susceptible to pressure injuries during surgery and extra care should be taken during positioning.

4. Conclusion

Children with HGPS are prone to developing intracranial haematomas. We recommend a high index of suspicion for intracranial pathology in the context of minor head trauma to ensure early diagnosis. A conservative approach with close neurological monitoring is advisable in neurologically intact and asymptomatic patients, provided the imaging encourages this. Prompt surgery should be performed in the presence of neurological deterioration.

Declarations of interest

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Conflicts of interest

We declare that we have no conflicts of interest in the authorship or publication of this manuscript.

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