



Case report

Gigantic vascular anomaly in a PHACE syndrome patient managed with photodynamic therapy

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Vascular anomalies are congenital vascular tissue aberrations, which are commonly identified in the head and neck and known to affect approximately 1 in 22 children. The most recent classification of vascular anomalies includes two main categories: vascular tumours (haemangiomas) and vascular malformations [1].

Several problems can arise from vascular tumours, including functional (through mass effect), aesthetic impairment, pain, bleeding (platelet sequestration) and disability [2]. They are known to be associated with a number of syndromes involving the head and neck, including PHACE (posterior fossa malformations – haemangiomas – arterial anomalies – cardiac defects – eye abnormalities) [3].

PHACE syndrome is a rare association between cervicofacial haemangiomas, structural brain abnormalities (mainly of the posterior fossa), arterial and cerebrovascular anomalies, aortic and cardiac abnormalities as well as eye defects. It represents a fairly common syndrome, identified in 1996 and more common in females. The haemangioma component in PHACE is very small or

not even visible at birth but it is known to grow very fast over the face and head and neck region. The genetic component is still undergoing investigations and no familial cases have been identified. Multi-disciplinary team is required for managing this unique group of patients [3].

Conventional management of cervicofacial haemangiomas can include systemic or intra-lesional corticosteroids, beta-blockers, chemotherapeutic agents, surgery, laser or a combination of these therapies. Where lesions are large, surgery can have marked adverse effect on form and function and due to difficulty in delineating these lesions, recurrence can be high. The side effects of chemotherapy are well known and radiotherapy carries the risk of inducing new tumours [1,5].

Photodynamic therapy with interstitial illumination (iPDT) is a technique developed for the treatment of deep-seated malformations and falls within the minimal surgical approach. Previously PDT for vascular anomalies has been used primarily for the management of age-related macular degeneration. It has been described as effective in the treatment of port-wine stain birthmarks, alone and in combination with pulsed dye laser therapy. There are relatively few studies in the literature describing the use of PDT for deep-seated vascular anomalies [4].

We report the use of ultrasound guided iPDT (US-iPDT) in the management of a gigantic haemangioma of an 11-year-old PHACE

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Fig. 1. US-iPDT: patient draped, prepped and eye shield applied; Left: examination under anaesthesia (EUA). Top right: ultrasonic examination of the haemangioma. Bottom right: needle insertion under ultrasound guidance.

syndrome Asian male with an on-going breathing, swallowing and speaking problems as well as severe facial deformity.

An 11-year-old Asian male was referred to the University College London Hospital Head and Neck Centre, London for management of a gigantic haemangioma involving various parts of his head and neck as part of PHACE syndrome. The child syndromic features included posterior fossa abnormalities, extensive haemangioma of the nasopharynx, oropharynx and supraglottis with extensions to many tissues and structures in the cervicofacial region, arterial cerebrovascular anomalies, cardiac defect (aortic coarctation) and eye anomalies.

The patient has received a number of conventional interventions over the years to manage the problems arising from the extensive size of the lesion, including breathing, swallowing and speaking problems. At time of presentation, he had difficulty controlling the food and saliva in the mouth and it was difficult to initiate swallowing and coughing. He was unable to swallow solid food, and had difficulty managing fluids and semi-solid food. His breathing was laboured and difficult at rest most of the time; and there was difficulty in speaking with gurgling sound. The patient suffered regularly from lower respiratory tract infections and sometimes required to be on antimicrobials for several months. Local bleeding and infections of the haemangioma caused continuous problems and required multiple hospital admissions in the past. At the time of referral the patient had exhausted all conventional interventions that were applicable to his age and no other options were being considered.

Physical examination of the lesion was a challenge. The haemangioma involved the cervicofacial tissues, especially the mid and lower face, upper one third of the neck, oral cavity, nasopharynx and oropharynx. Severe oedema, ecchymosis petechiae and purpura were noted over the facial skin. Posterior fossa abnormalities, arte-

rial cerebrovascular anomalies and eye anomalies were assessed by a number of investigations and found to be stable. Cardiac defects were assessed by an echocardiogram and were found to be stable. Due to the worsening breathing pattern, an urgent surgical tracheostomy had to be undertaken to maintain the airway. Flexible fiberoptic laryngoscopy revealed lesional extension to the tongue base and supraglottis. Fine needle aspiration cytology (FNAC) was performed and malignant change was ruled out. The FNAC report highlighted the presence of haemangioma with fatty metaplasia.

A pre-intervention scan revealed a soft tissue mass extending from the face down to the larynx. A large number of feeding vessels were identified, deeming the lesion unsuitable for embolization or surgical excision. Bilateral jugular stents were noted extending almost to the skull base. There were a number of phleboliths (small local calcifications within a vein) noted throughout the mass suggestive of a venous component. The pathological process extended into the superior mediastinum. There was almost complete circumferential involvement of the subglottis causing 50% narrowing of the airway. Due to its large size, surgical resection carried a significant risk to the vessels and neurological structures close to the lesion.

Following discussion at a multi-disciplinary team meeting, it was decided that the most suitable option was to treat the lesion with ultrasound-guided interstitial photodynamic therapy (US-iPDT) under general anaesthesia. The patient's family signed an informed consent for the procedure to be carried out. Meta-tetrahydroxyphenyl chlorine (mTHPC) was administered at a dose of (0.15 mg/kg) intravenously into the mid-cubital vein 96 h prior to pathology illumination.

18 Gauge 70 mm long spinal needles were inserted under US guidance into the right and left cervicofacial portions of the haemangioma. Other needles were inserted to manage other com-



Fig. 2. US-iPDT: Left: light delivery to the oral tongue. Top right: Light delivery to the right facial part of the haemangioma. Bottom right: light delivery to the left facial part of the haemangioma.

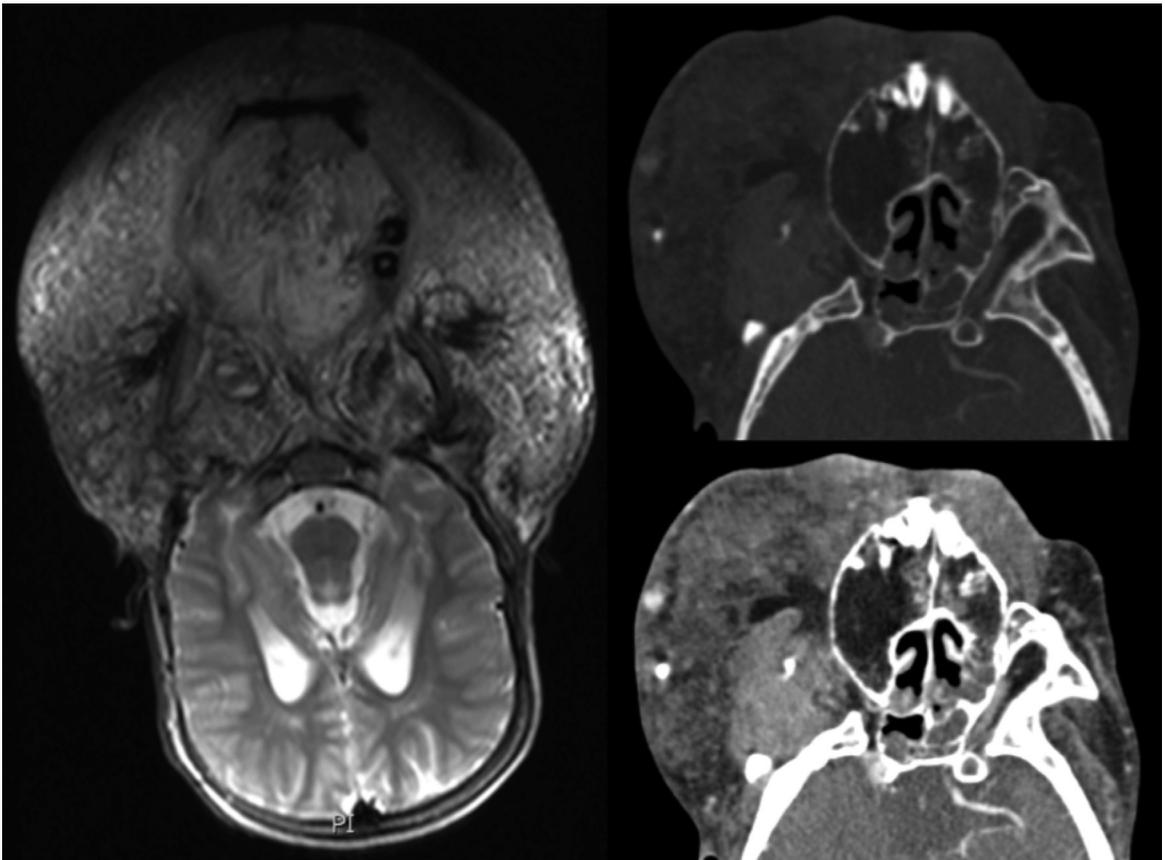


Fig. 3. Left: pre-PDT scan showing the extensive haemangioma. Top right and bottom right: 6 weeks post-PDT scan showing significant shrinkage of the lesion.

ponents including the oral tongue and tongue base. The area of treatment was scanned by ultrasound (EMP 1100 with high resolution) prior to needle insertion, to ensure accurate identification of the target volume (Fig. 1). Diffuser fibres were used instead of bare polished tip fibres. The structure of the vascular tumour allows maximal tissue illumination when using diffuser fibres. The fibres with core diameter of 400 μm were introduced into the spinal needles. A four-channel 652 nm diode laser was used for illumination and 20J/cm² was delivered per station to the target tissue (Fig. 2).

No immediate complications were reported after this intervention. On day 5 post-PDT, the tracheostomy tube was removed without any problems. During the first 4 weeks post-illumination, a gradual improvement of breathing, swallowing and speaking symptoms were noted. Gradual re-exposure to light was carried out at an incremental rate of 100 lx per day. The patient was advised about the need to avoid direct exposure to sunlight for up to 2 weeks after the photosensitiser administration and this was explained to the parents and they were provided with light exposure guidelines.

The clinical assessment revealed an excellent response of the haemangioma after only one round of photodynamic therapy. At the last clinical review, the dysphagia has significantly improved along with the breathing problems, which has become less labored, and only difficult after exercise and no gurgling sound. The patient reported a better control of fluid and saliva in the mouth. The patient started to manage small chunks of solid food and it became easier with fluids and semi-solid food. A 6-week post-PDT scan showed significant reduction in the size of the haemangioma by about 70% (Fig. 3). The patient's pathology continues to be in a stable state 4 years after the intervention. The rate of local bleeding and infections, along with lower respiratory tract infections became less after one treatment round of US-iPDT.

The management of vascular anomalies continues to be extremely challenging. Although several modalities have been developed and the literature reports successful treatment in many, data from long-term studies reports relapse in many and the need for re-treatment or another intervention. US-iPDT is not superior

to other modalities, but it is characterised by being one of the least invasive, being repeatable with no residual toxicity and with minimal bystander effect [1,4–6]. This case report proves US-iPDT to be an appropriate treatment option for this complex vascular anomaly.

Conflict of interests

We declare none.

Authors' contributions

All authors designed and performed the study, carried out the literature search and manuscript preparation. All authors were responsible for critical revision of the scientific content and manuscript review. All authors approved the final version of the manuscript.

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