Cervicofacial angioma and the Kasabach-Merritt syndrome

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Abstract We present a neonate with a cervicofacial haemangioma complicated by the Kasabach-Merritt syndrome, respiratory distress due to airway compression and high-output heart failure. This haemangioma and intravascular disseminated coagulation, treated initially by aspirin, ticlopidine and corticosteroids, required more invasive treatment with superselective embolisation and interferon alpha-2a. The clinical outcome was good.

Key words Haemangioma facial  ·  Embolisation  ·  Kasabach-Merritt syndrome  ·  Interferon

Introduction

Haemangioma is the most common benign neoplasm in the neonatal period [1]. The lesion is rarely fully growth at birth. Most are small, harmless birth marks which involute spontaneously without sequelae but up to 20% may reach to a massive size [1, 2]. Kasabach and Merritt [3] described a case of capillary haemangioma with extensive purpura. When associated with thrombocytopenia and consumptive coagulopathy (the Kasabach-Merritt syndrome) or if it interferes with vital structures (airway obstruction, cardiac failure, etc.), the haemangioma may be life-threatening and require unusual therapy [4–7]. We report a large cervicofacial haemangioma in a newborn, which in addition to the recommended treatment, needed superselective embolisation and interferon alpha-2a therapy.

Case report

A girl was born after an uneventful 38-week gestation, weighing 3 kg and measuring 46 cms. On day 3, tumefaction of the left cheek was noticed for the first time. The mass grew rapidly and on day 11 a hard, large left cervicofacial mass was seen, extending from the ear lobe to the lateral cervical area down to the level of the mandible, and under the orbit, with bluish discolouration of the skin. Ultrasonography (US) showed an echogenic, infiltrating mass. CT revealed a homogeneous mass with marked contrast enhancement (Fig. 1). The diagnosis of a left parapharyngeal cervicofacial haemangioma was made and treatment with high dose corticosteroids, ticlopidine and aspirin was instituted.

On day 14, because of respiratory distress due to airway compression and a consumptive coagulopathy (IVDC), the child was transferred to the paediatric intensive care unit (Fig. 2). Treatment with assisted ventilation, platelet and cryoprecipitate transfusions was introduced. The next day, left carotid angiography showed a large hypervascular blush supplied by branches of the external carotid artery, without apparent early venous drainage (Fig. 3a). Following superselective catheterisation, successive embolisation
Fig. 1  Axial contrast-enhanced CT through mandible shows a large, dense, homogeneous left cervical mass with retropharyngeal extension

Fig. 2  The patient on day 14 on admission to the paediatric intensive care unit

Fig. 3  a The hypervascular tumour is fed by multiple distal branches of the left external carotid artery. b Postembolisation left common carotid angiogram shows devascularisation of the tumour, except for a small residuum near the origin of the external carotid artery.

of the distal branches of the left internal maxillary (sphenopalatine and descending palatine), lingual (inferior maxillary), facial, auricular, occipital and ascending pharyngeal arteries was performed with polyvinyl alcohol particles (150–250 μ). Major devascularisation was achieved, except for small residuum fed by small arteries arising from the main trunk of the left external carotid artery (Fig. 3b). These were not embolised owing to the quantity of contrast medium injected and their proximity to the origin of the left internal carotid artery. There were no complications. The next day, marked decrease in size of the cervical mass was clinically evident. On day 16, after extubation, the child was transferred to the neonatology unit with IVDC controlled. On day 21, US confirmed a marked decrease in size of the mass, with a residual pretracheal component. The child was discharged with cardiorespiratory monitoring on day 23.

She was readmitted on day 32, following respiratory distress and a recurrence of the IVDC. Angiography showed persistent occlusion of the arteries previously embolised and a residual blush. New collateral channels had developed from the inferolateral trunk of the left internal carotid artery and occipital and muscular branches of the left vertebral artery (Fig. 4). Embolisation of the left internal maxillary and lingual arteries and the occipital branch of the left vertebral artery was performed without complications. Medical treatment was pursued and the clinical status of the child improved.

On day 41, she presented with cardiac failure. Additional embolisation was considered too risky and treatment with interferon alpha-2a was instituted. The child showed progressive clinical improvement with control of the IVDC. On day 57, she deteriorated again, with thrombocytopenia and consumptive coagulopathy. MRI showed a large residual left parapharyngeal cervicofacial mass with mixed signal and intense contrast enhancement extending into the retropharyngeal space, with adjacent airway compression (Fig. 5). The clinical state remained stable. From day 81, a decrease in the size of the tumour was observed. The IVDC onwards was controlled; interferon alpha-2a was stopped on day 96 and assisted ventilation on day 100. The child was discharged from the hospital at the age of 5 months.

At 1 year, neurological, clinical and laboratory tests were normal. MRI showed marked regression of the mass with a small focus of residual left parapharyngeal angiomatous tissue (Fig. 6). The skin of the cervicofacial region on the left was normal and there was no visible residual angioma (Fig. 7).

Discussion

Immature haemangiomas of the newborn result from proliferation of capillary endothelium of non-neoplastic embryologic type [8]. Haemangioma is the most frequent tumour being present in more than 10% of normal newborns, with a strong female predominance. Its natural course is rapid postnatal growth for 8–18 months (proliferative phase) followed by slow regression over the next 5–8 years (involutional phase) [9]. In 95% of cases, the tumour will regress spontaneously, without sequelae [10]; some leave a residual fibrofatty scar or