CASE REPORT

An extensive bilateral cervicofacial hemangioma managed successfully with propranolol, a case report and literature review

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Received 21 January 2013; revised 14 April 2013; accepted 16 April 2013
Available online 24 May 2013

KEYWORDS
Infantile hemangiomas; Propranolol

Abstract  Infantile hemangiomas are the most common benign childhood tumors, they are more common in girls and premature babies, and all races are affected with a slight preponderance in whites (Metry). These lesions most commonly occur on the head and neck area, but they can occur anywhere on the body (Paller, 2011). Here, we describe a case of bilateral facial mandibular segment hemangioma with deep and superficial components involving both parotid glands which was managed successfully with propranolol.

1. Case report

Here we present the case of a full term baby boy born at 37 weeks of gestation by an elective cesarean section after an uneventful pregnancy. One to two weeks after birth, the parents noticed small telangiectasias and strawberry colored macules on the skin overlying both parotid areas, lower lip, tip of the tongue and the anterior aspect of the chin and neck. The lesions started a growth phase at the age of 3–4 weeks, and the patient was seen in our clinic at the age of three and a half months, at that time he had extensive bilateral parotid hemangiomas with both superficial and deep components, and had superficial hemangiomas on the lower lip, chin, preauricular area, neck and anterior chest (Figs. 1 and 2). The lesion on the lower lip was partially ulcerated and was associated with bleeding upon feeding. The patient was noticed to have a stridor which was related to the extensive size of the hemangioma partially obstructing the airway.

The infant was admitted to the pediatric ward for full investigation and evaluation for internal organ involvement as well as other associated congenital anomalies such as PHACES syndrome. (Posterior fossa malformations, Hemangiomas, Arterial anomalies, Cardiac anomalies and aortic coarctation, Eye abnormalities and Sternal clefting and supraumbilical raphe) (Paller, 2011). The evaluation for systemic involvement and radiological studies was carried out by the pediatric cardiologist.

No evidence of associated congenital anomalies or internal organ hemangiomas were found upon appropriate radiological studies of the head (Fig. 3), neck, chest, and abdomen. An echocardiography was also done, with no signs of cardiac anomalies.

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Peer review under responsibility of King Saud University.
The blood work showed a microcytic hypochromic anemia (Hemoglobin of 8.4 gm/dl), iron level was 13 μg/dl (low), and this was dealt with iron supplementation. Liver, renal and thyroid function tests were within normal. Cultures taken from ulcerated areas were negative for any bacterial growth.

The decision to intervene was made due to the extensive size of the hemangioma, difficulty of feeding due to recurrent bleeding from the ulcerated areas on the lower lip, and the stridor the patient was noticed to have. A potential complication that was considered and emphasized the need for intervention was the risk of delayed language development due to partial obstruction of the auditory canals which was noticed upon examination.

After full evaluation by the pediatric cardiologist, the infant was started on propranolol 0.78 mg/kg/day in EDD (equally divided doses), with gradual increasing of the dose as tolerated, while monitoring vital signs and blood glucose levels periodically. The final dose reached was 1.8 mg/kg/day given in EDD, on which the patient was discharged, and followed up in the outpatient clinic on a monthly basis, both in the pediatric cardiology clinic and the dermatology clinic. The hemangioma showed a significant improvement in size and color after 2 weeks from starting therapy and continued to shrink in size with each follow up visit (Figs. 4–6). Finally, at the age of 12 months propranolol was stopped after gradual tapering started in the last month of treatment under the recommendation and supervision of the pediatric cardiologist, the decision to taper treatment was based on the assumption that the active growth phase of the hemangioma usually ends by the first year. The patient had no regrowth on 6 months, 12 months, and 18 months of follow up after the discontinuation of treatment, but a fibrofatty residue remained in the lower lip area and the anterior chest area.

2. Discussion

Infantile hemangiomas are the most common soft tissue tumors occurring during infancy. They are noticed more in baby girls and in premature babies, and they are more common in whites (Metry and Hebert, 2000). Other risk factors may include advanced maternal age, multiple pregnancies, and pregnancies complicated with placenta previa and pre-eclampsia (Metry and Hebert, 2000; Paller, 2011).

Infantile hemangiomas may be superficial, deep or may be composed of both a superficial and a deep component as in our case.
It is well known that infantile hemangiomas pass through three phases, a growth phase known as the proliferative phase which usually lasts up to the age of 12 months and occasionally longer. A plateau phase as the growth seizes which usually occurs at 12–18 months of age, and a third phase of spontaneous involution which occurs between the ages of 3 and 10 years in most cases (Metry and Hebert, 2000; Paller, 2011).

Hemangiomas with a deep component particularly ones involving the parotid glands as our case usually take a longer time to involute and some may not undergo complete involution (Metry and Hebert, 2000). Some superficial hemangiomas located on the lips as our patient had, may also fail to involute completely (Metry and Hebert, 2000) and may be associated with recurrent bleeding due to ulceration and therefore lead to difficulty of feeding as was experienced by our patient.

The decision to intervene is based on several factors including the size and location of the infantile hemangioma, as hemangiomas located in critical areas such as the eyes or the preauricular areas may lead to a compromise or loss of vision and delayed language development due to conductive hearing loss respectively (Greene et al., 2004). In general, infantile hemangiomas that interfere with function or lead to destruction of vital structures or obstruct airways or cause significant disfigurement or are associated with life threatening complications such as congestive heart failure need intervention (Greene et al., 2004; Menezes, 2011). Our patient had massive bilateral cervicofacial hemangiomas involving the parotid glands which were partially obstructing both auditory canals with the possible risk of delayed language development. Along with the stridor due to partial airway obstruction caused by the hemangioma involving the anterior neck area, and in addition to the difficulty of feeding that the infant experienced, as well as the disfigurement and parental anxiety that it caused, we decided that intervention was necessary.

Management options for infantile hemangiomas that do not interfere with vital functions or cause any disfigurement or complications are mainly active nonintervention (Vibhu and Masarat, 2010). But those that require intervention, management options include, medical therapy, LASER therapy usually PDL (pulse dye LASER) and Nd:YAG LASER (Neodymium–Yttrium aluminum garnet), cryotherapy, compressive therapy, and surgical resection (Sinno et al., 2010; Vibhu and Masarat, 2010). Medical therapy includes topical, intralesional or systemic corticosteroids, topical imiquimod, Interferon alfa 2a or alpha 2b, vincristine, cyclophosphamide, and intralesional bleomycin (Paller, 2011; Bagazgoitia et al., 2011; Greene et al., 2004; Vibhu and Masarat, 2010). Recently, successful treatment with captopril was also documented by S.T Tan et al. (Tan et al., 2012). All of these treatments have very well known side effects (Metry and Hebert, 2000). Systemic glucocorticosteroids are reported to cause fussiness, irritability, gastriac irritation, hypertension and HPA suppression (George et al., 2004). Decreased rate of growth and weight gain have also been documented (Vibhu and Masarat, 2010). Intralesional steroids may cause skin atrophy, depigmentation, necrosis, and if used for periorbital hemangiomas may cause embolism to the retinal artery (Vibhu and Masarat, 2010). Interferon alfa 2a or 2b use is usually reserved for life threatening hemangiomas and can be associated with spastic diplegia in 20% of patients (Paller, 2011), more commonly encountered side effects with interferon alpha include fever, irritability, and malaise. Neutropenia, anemia and elevated liver transaminases have also been reported (Paller, 2011). Side effects related to captopril use included hypotension with subsequent renal impairment, skin rashes and gastrointestinal upset (Tan et al., 2012). Cryotherapy may be associated with scarring and surgery is also risky as it may lead to scarring, facial asymmetry and temporary or permanent facial nerve paralysis, along with other complaints and possibly even death (Sinno et al., 2010).

Less commonly used modalities of treatment include Beacplermin (recombinant platelet derived growth factor) which is mainly used for ulcerated infantile hemangiomas especially in the diaper area (Vibhu and Masarat, 2010).

In 2008 Léauté-Labrèze et al. reported the use of propranolol a non selective beta blocker in the management of infantile hemangiomas that require intervention (Léauté-Labrèze et al.,...
A dose of 2 mg/kg/day in EDD has been suggested by Léauté-Labrèze et al., 2008. We used a dose of 1.8 mg/kg/day in EDD in our patient as suggested by the pediatric cardiologist. The speculated mechanism by which propranolol works is through vasoconstriction, blocking of the proangiogenic signals such as vascular endothelial growth factor (VEGF), basic fibroblast growth factor (bFGF), and matrix metalloproteinases 2 and 9 (MMP), and on the long term, propranolol may induce apoptosis in the proliferating endothelial cells. (Morais et al., 2011; Sans et al., 2009). Propranolol therapy requires careful monitoring of the infant for bradycardia, hypotension, hypoglycemia and hypothermia (Morais et al., 2011; Bonifazi et al., 2008; Bagazgoitia et al., 2011; Menezes, 2011; Sans et al., 2009). Other side effects of propranolol include bronchospasm especially in patients with reactive airway disease, skin rashes, fatigue, sleep disturbance and gastrointestinal discomfort (Morais et al., 2011; Sans et al., 2009) and hyperkalemia was reported in one case as a potential life threatening complication (Pavlakovic et al., 2010). Therefore, we opted to admit the patient for the first 2 weeks of therapy for evaluation and monitoring purposes, and our patient seemed to have tolerated the treatment very well without any adverse events.

3. Conclusion

Our patient responded wonderfully and promptly to propranolol therapy, and has tolerated the therapy with no serious side effects. Parental anxiety was reduced and prolonged disfigurement was avoided. No regrowth was experienced on 6 months, 12 months, and 18 months of follow up after discontinuation of propranolol.

Conflict of interest

None.

Acknowledgment

We would like to thank Dr. Alaeddin Dajani, the pediatric cardiologist who helped us in the management and monitoring of the patient while on propranolol treatment.

References


