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Case presentation

“Cyrano nose” associated with hepatic hemangiomas successfully treated with propranolol

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Abstract

Hemangioma of the nasal tip is commonly described as “Cyrano nose” and his treatment is extremely difficult because of its location and possible severe aesthetic complications like definitive nasal deformation. We describe a patient who presented at two months of age with a “Cyrano nose” associated with multiple hepatic and cutaneous hemangiomas, which completely resolved after therapy with propranolol. Treatment was well tolerated and aesthetic result was excellent.

Keywords: Cyrano, propanolol, hemangioma

Introduction

The “Pinocchio” or “Cyrano” nose is a rare condition in which deformity of the nasal tip is produced by a soft tissue hemangioma that infiltrates the alar cartilages. Unlike the usual spontaneous involution observed in infantile hemangiomas, nasal tip hemangiomas tend to regress slowly. If untreated they give rise to permanent scarring and disfigurement, resulting in a Cyrano appearance with a profound psychological impact [1]. Herein, we report a female infant with Cyrano nose associated with multiple hepatic hemangiomas, both of which completely resolved after treatment with propranolol.

Case synopsis

Our patient was born at term after a normal pregnancy. The physical examination at birth revealed three millimeter cutaneous hemangiomas and a nasal tip hemangioma, the so-called Cyrano nose (CN). The patient came to our attention at two months of age for an episode of bleeding of the nasal hemangioma. Abdominal ultrasound and MRI revealed the presence of multiple hepatic hemangiomas, the largest of which was 2.3 cm in diameter. Laboratory investigations showed normal thyroid function tests. After one month both nasal hemangioma as well as hepatic hemangiomas rapidly increased in size. Taking into account the severe cosmetic complications of CN hemangioma and considering the rapid evolution of the lesions, at three months of age we decided to start a treatment with propranolol at an initial dosage of 1 mg/kg/day that was gradually increased to 2 mg/kg/day. The therapy was well tolerated, except for a mild loss of appetite and insomnia during the first month of treatment. After only one week, the small cutaneous hemangiomas disappeared and the nasal hemangioma changed from intense red to purple. Also the hepatic hemangiomas completely disappeared after five months of therapy. In the following months the nasal tip hemangioma continued to gradually reduce in size until a near complete disappearance at 15 months of age, when the propranolol dose was reduced and discontinued one month later (Fig. 1A,B). At the last follow-up obtained at 4 years of age,
33 months after cessation of treatment, abdominal ultrasound was normal and no recurrence of the nasal hemangioma was noted.

![Figure 1. (A) Patient at 2 months of age with a nasal hemangioma leading to a disfigurement of the nasal tip. (B) Patient at 15 months of age, after 12 months of propranolol therapy; the hemangioma of the nasal tip is completely resolved.](image)

**Discussion**

Despite the high risk of severe cosmetic complications associated with Cyrano nose, the modality and the timing of treatment is still controversial and include corticosteroids, conventional surgery, cryotherapy, and laser therapy [2, 3]. These treatments are associated with a considerable risk of iatrogenic induced deformity and scarring. Propranolol was recently introduced as a treatment of life and function-threatening infantile cutaneous hemangiomas and our patient also demonstrates the effectiveness also in hepatic hemangiomas [4]. Eivazi B et al [5] reported a retrospective study of 23 children with CN, two of which were successfully treated with only propranolol. The good response of nasal tip hemangioma to propranolol has recently been described in other case series, as reported in Table 1 [6, 7, 8]. Some cases of regrowth of hemangioma after propranolol withdrawal is reported, especially when treatment is stopped before 1 year of age [7, 8, 9]. Too-early withdrawal of the treatment and a late proliferative phase seem to be responsible for recurrence, but it is difficult to determine which hemangiomas will require longer propranolol therapy. Therefore, the duration must be individualized according to the intrinsic features of the hemangioma and its clinical evolution, which are largely unpredictable. Our case, together with the previously reported cases, confirm the efficacy of early and prolonged treatment with propranolol in Cyrano nose hemangioma, and suggests that it could be proposed as the first treatment option for proliferating CN. Moreover our case emphasizes the importance of performing abdominal ultrasound in patients with Cyrano nose in order to exclude the presence of hepatic hemangiomas, particularly if associated with other cutaneous hemangiomas.

**Table 1. Case-series of nasal tip hemangioma treated with propranolol**

<table>
<thead>
<tr>
<th>Author</th>
<th>Patients</th>
<th>Results</th>
<th>Age at propranolol, months (range)</th>
<th>Treatment duration, months (range)</th>
<th>Relapses</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ben-Amitai et al, 2012</td>
<td>10</td>
<td>8 good 2 partial</td>
<td>6 (2-22)</td>
<td>9.7 (5-13)</td>
<td>0</td>
</tr>
<tr>
<td>Bagazgoita et al, 2011</td>
<td>6</td>
<td>6 good</td>
<td>5.8 (1-45)</td>
<td>7</td>
<td>1</td>
</tr>
<tr>
<td>Fuchsman et al, 2011</td>
<td>13</td>
<td>13 good</td>
<td>6.1 (1-15)</td>
<td>9</td>
<td>3</td>
</tr>
<tr>
<td>Our patient</td>
<td>1</td>
<td>1 good</td>
<td>3</td>
<td>12</td>
<td>0</td>
</tr>
</tbody>
</table>

**References**


