Lymphangioma in nasal tip and vestibulum: Case report

Linfangioma en punta y vestíbulo nasales: reporte de caso

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SUMMARY

A ‘Pinocchio’ or ‘Cyrano’ nose is a condition with a deformity at the tip of the nose caused by an underlying soft tissue tumor. The deformity of the nose with secondary involvement in the form of lymphangioma is a sporadic case, and few studies have reported it. Lymphangioma itself is a benign tumor that originates from the lymph vessels. The diagnosis of lymphangiomas can be made based on the clinical picture and computed tomography (CT) or magnetic resonance imaging (MRI) scans. Treatment options for nasal tip lymphangioma and nasal vestibule are radiotherapy, cryotherapy, and sclerosant injection. However, excision is still the treatment of choice. We report a 4-year-old (Asian) boy complaining of a lump on the tip of his nose that has been getting bigger for the last year. The results of a non-contrast CT scan showed a mass in the subcutis tip of the nose. The patient underwent tumor excision using a gull-wing incision in the transcolumella. The results of the anatomic pathology examination of the tumor tissue identified lymphangioma. Lymphangiomas at the tip of the nose and the vestibule are accompanied by their presence. Very satisfactory results were obtained at the time of control four months postoperatively. The patient had no symptoms, and from physical examination, there was no palpable mass at the tip of the nose and the nasal vestibule.

Keywords: Lymphangioma, nasal tip, nasal vestibulum, case report.

RESUMEN

Una nariz de 'Pinocho' o 'Cyrano' es una condición con una deformidad en la punta de la nariz causada por un tumor de tejido blando subyacente. La deformidad de la nariz con afectación secundaria en forma de linfangioma es un caso esporádico y pocos estudios lo han informado. El linfangioma es un tumor benigno que se origina en los vasos linfáticos. El diagnóstico de los linfangiomas se puede hacer con base en el cuadro clínico y la tomografía computarizada (TC) o la resonancia magnética nuclear (RMN). Las opciones de tratamiento para el linfangioma de la punta nasal y el vestíbulo nasal son la radioterapia, la crioterapia y la inyección de esclerosante. Sin embargo, la excisión sigue siendo el tratamiento de elección. Se reporta sobre un niño (asiático) de 4 años que se queja de un bulto en la punta de la nariz que se ha ido agrandando durante el último año. Los resultados de una tomografía computarizada sin contraste mostraron una masa en la punta subcutánea.
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de la nariz. El paciente fue sometido a extirpación del tumor mediante una incisión en ala de gaviota en la transcolumela. Los resultados del examen anatomopatológico del tejido tumoral identificaron linfangioma. Los linfangiomas en la punta de la nariz y el vestibulo nasal se acompañan de su presencia. Se obtuvieron resultados muy satisfactorios al momento del control a los 4 meses del posoperatorio. El paciente no presentaba síntomas y al examen físico no se palpaba masa en punta de la nariz y vestibulo nasal.

Palabras clave: Linfangioma, punta nasal, vestibulo nasal, reporte de caso.

INTRODUCTION

Lymphangiomas are uncommon, benign malformations of the lymphatic system that can occur anywhere on the skin and mucous membranes. Lymphangiomas can be categorized as deep or superficial based on the depth and size of the abnormal lymphatic vessels or as congenital or acquired. Approximately 60% are present from birth or arise in childhood, and most are found in the head and neck area (1). The ‘Pinocchio’ or ‘Cyrano’ nose is a rare condition in which an underlying soft tissue tumor produces nasal tip deformity (2). The deformity of the nose due to lymphangioma is a very rare case; few studies have reported it (2,3).

Diagnosis in cases of lymphangioma can be made based on the clinical picture. Still, computed tomography (CT) or magnetic resonance imaging (MRI) scans will determine the size, location, and relationship to essential structures more accurately and help plan surgery (4). Managing lymphangiomas is challenging, and many treatment modalities have been described over the years. Treatment strategies vary depending on the anatomical location, size, and involvement of surrounding structures (3,4).

Treatment options include radiotherapy, cryotherapy, and sclerosant injections. But permanent excision is the first choice of treatment (3,5). Uysal et al. (3) performed tumor excision using a gullwing incision by combining the two-incision techniques previously performed by Wynn and Koopman (6,7). Here, we report a case of lymphangioma at the nasal tip and vestibule, which was successfully treated by tumor excision using a gullwing incision in the transcolumela.

CASE REPORT

A 4-year-old boy (Asia) with the main complaint of a lump on the tip of his nose, which he felt was getting more extensive in the last one year (Figure 1). Initially, the lump was bluish, then purplish red spots appeared the previous month. From the physical examination, a painless lump was found at the tip of the nose, and the left nasal vestibule felt supple, mobile, and bluish with a solitary spot, tiny and consisting of purplish-red vesicles. No other disturbances were found on anterior rhinoscopy, otoscopy, and pharyngoscopy examination.

Figure 1. Clinical photo of the patient. A mass is seen at the tip of the nasi and the nasal vestibule (red arrows).
The results of a non-contrast head CT scan showed a well-defined isodense lesion originating from the subcutis tip nasi without any damage to the surrounding structures (Figure 2). Laboratory examination results and chest X-ray were within normal limits. It was planned a tumor excision at the tip of the left nasal vestibule in this patient.

![Non-contrast head CT scan. Coronal (A), sagittal (B), and axial (C) sections show a mass at the tip of the nose (yellow arrow).](image1)

Under general anesthesia, the patient underwent an open approach technique, a gull-wing incision was made in the transcolumella, and the operating area was expanded so that the entire mass boundary was clearly visible. A complete tumor excision was performed (Figure 3). The results of the anatomical, pathological examination of the tumor tissue showed lymphangioma results (Figures 4 and 5).

![Tumor excision surgery. Transcolumella gull wing incision (A). The operating field was extended to reach the tumor (BC). Excision of the tumor at the tip of the nose and the nasal vestibule (D). Suturing the incision (E). Tumor excised completely (F).](image2)

Follow-up was carried out on patients in polyclinic care within one month and four months after surgery and found no symptoms complained of by patients. The examination results showed no visible mass at the tip of the nose and vestibule nasi, with minimal surgical scars in the columella.
Table 1. Lymphangioma classification (1)

<table>
<thead>
<tr>
<th>Classification</th>
<th>Description</th>
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<tbody>
<tr>
<td>Capillary lymphangioma</td>
<td>Consists of lymphatic vessels the size of thin-walled capillaries, usually asymptomatic and presenting as small vesicles or small warts on the skin or mucosa the same color as the surrounding normal skin, filled with lymph.</td>
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<tr>
<td>Cavernous lymphangioma</td>
<td>40% of all lymphangiomas, consisting of dilated lymphatic spaces, often with a fibrous adventitia, usually occurring on the tongue, cheeks, and lips, and presenting as a diffuse, painless swelling with a soft consistency, such as a sponge with a normal color like the surrounding tissue.</td>
</tr>
<tr>
<td>Cystic lymphangioma</td>
<td>In the form of cysts and sinuses trying in diameter from a few millimeters to several centimeters, they usually appear as a cystic mass containing acellular lymph fluid subcutaneously or in deep places. They are often found in the neck (hygroma colli), axilla (hygroma axillare), and so on.</td>
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Figure 4. Histopathological results of tumor tissue on the nasal tip with magnification (A) 10x and (B) 40x show that lymph vessels coated with endothelium are not atypical and do not contain erythrocytes.

Figure 5. Histopathological results of tumor tissue in the nasal vestibule with magnification (A) 10x and (B) 40x show tissue covered with epidermis. There are lymph vessels covered with endothelium that is not atypical and does not contain erythrocytes.
DISCUSSION

Lymphangiomas are generally present at birth but may sometimes manifest for the first time in young adults. This disease can appear anywhere on the head and neck (5). On palpation, it is soft, cystic, and transilluminated. It may persist or regress but may not regress spontaneously and, in some cases, enlarge, especially after internal bleeding or infection (8). Hobby et al. (2) first reported a case of a 3-year-old girl who had nasal tip deformity caused by cavernous lymphangioma (2). Uysal et al. (3) also reported a similar case in a 17-year-old boy with ‘Pinocchio’ or ‘Cyrano’ nose deformity due to circumscrip lymphangioma (3).

Several types of lymphangiomas exist, including capillary, cavernous, and cystic (Table 1) (1). The lymphatic system arises from five primitive sacs (two jugular sacs, two posterior sciatic sacs, and one retroperitoneal sac), which develop from the venous system (9). Two main theories explain the origin of lymphangiomas: sequestration of lymphatic tissue and proliferation of endothelial fibrillation membranes.

There are two reported techniques for surgical treatment of nasal deformities: Pinocchio, Wynn with an elliptical incision technique on the nasal dome, and the use of soft intestinal clamps because they cause a smaller level of scarring (6). Koopman has modified and utilized an open incision rhinoplasty, where the scar is more aesthetically pleasing (7). Uysal et al. reported that the two techniques were combined due to skin involvement using a gullwing incision and excision. Patient satisfaction appears good, producing acceptable aesthetic results (3).

CONCLUSION

Lymphangiomas at the nose tip and vestibule are accompanied by their presence. Pinocchio’s nose deformity is a rare and benign lymph vessel malformation. No previous literature study report reported cases of lymphangioma in the nasal tip and nasal vestibule. The main and best treatment option is surgery for this benign lesion, which has proven very effective. The recurrence rate is low if complete removal of the cystic epithelium has been achieved.

REFERENCE