SAS Journal of Surgery (SASJS) **ISSN 2454-5104**
Abbreviated Key Title: SAS J. Surg.

A Unit of Scholars Academic and Scientific Society, India

Median Congenital Fistulas of the Nose of Children (About 3 Cases)

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Original Research Article

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Article History
Received: 20.10.2018
Accepted: 27.10.2018
Published: 30.10.2018

DOI:
10.21276/sasjs.2018.4.10.14

Abstract: Congenital Fistulas of the face, specifically of the nose are embryological malformations infrequent and poorly understood. These medial malformations may be of branchial origin or not. Their formation can be explained by the persistence of an anatomical disposition, normally transient during the embryonic life (2nd month of life in utero). Their circumstances of discovery are variable: in the presence of an external fistula or in episodes of repeated superinfections. These median congenital fistulas must be known earlier for early surgical management because of the significant risk of superinfection. The main differential diagnosis is meningoencephalocele.
Keywords: Median fistula, embryology, child, medial dysraphia.

INTRODUCTION

Cysts and fistulas of congenital origin of the face and neck are infrequent malformations. Dermatologists, pediatricians and ENTs must recognize these lesions early to allow appropriate management. Their formations result from a lack of coalescence of a slot or a gill pocket resulting in the persistence of a residue of the branchial apparatus. Surgical excision is performed before the age of one year because of the risk of superinfection.

MATERIALS & METHODS

We report 3 cases treated in the department of Surgery "C" - Pediatric Plastic Surgery Unit and burns at Rabat Children's Hospital.

Case n°1

This is an 8-year-old girl, who consults after several episodes of cutaneous superinfection interesting the skin of the nose treated medically without results.

She had a biopsy that had objectified leishmaniasis. Specific treatment was instituted without giving results. Then she was referred to our department where the clinical examination showed an external orifice of a cutaneous fistula situated on the median line of the nose at the junction 1/3 average 1/3 inferior with a superinfected lesion opposite the internal canthus left (figure 1).

A CT scan with fistulography was performed confirming the diagnosis of median fistula of the nose with communication with the left internal canthal lesion (Figure 2). After preoperative assessment, the fistula was resected in its entirety under general anesthesia and after infiltration of the fistulous pathway with methylene blue. Dissection removed a cutaneous cord communicating between the two holes and passing under the bones of the nose. The suites were simple and there were no recurrences (Figure 3 and 4).

Case n°2

This is a 6-year-old boy admitted for two medial fistulas and right canthal disinfected (Figure 5) diagnosis was confirmed by MRI.

The fistula was resected in its entirety under general anesthesia and after infiltration of the fistulous pathway with methylene blue. Dissection removed a cutaneous cord communicating between the two holes and passing under the bones of the nose. The suites were simple and there were no recurrences. (Figure 6 and 7)

Case n°3

This is a 5-year-old boy admitted also for median fistula of the nasal root (Figure 8) with a treatment-medically unsuccessful episode of superinfection who had an MRI confirming the diagnosis. Surgical resection was scheduled and removed the entire fistulous path that came into contact with the frontal bone (Figure 9 and 10). In all 3 cases, there was a delayed diagnosis, a specialized consultation was performed after several episodes of superinfection. With an average follow-up of two years, our three patients had no complications (infectious or recurrent).

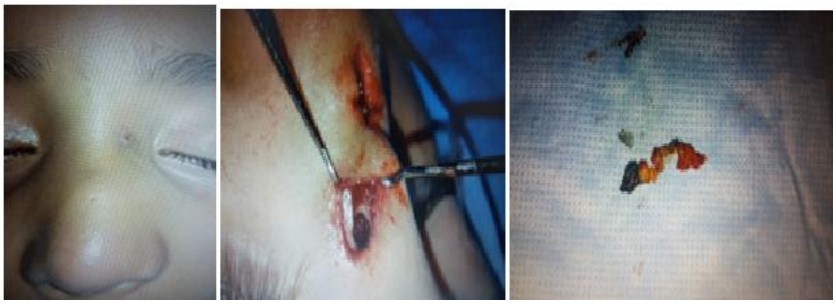

Fig-1: Clinical and intra-operative aspect showing a median congenital fistula and exit point corresponding to fistulization

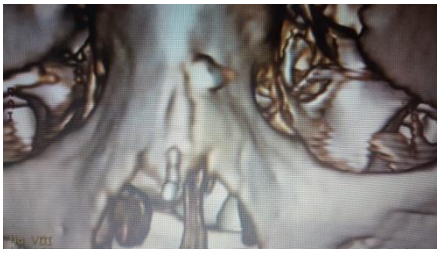

Figure 2: CT scan of a median congenital fistula with fistulogram showing its path

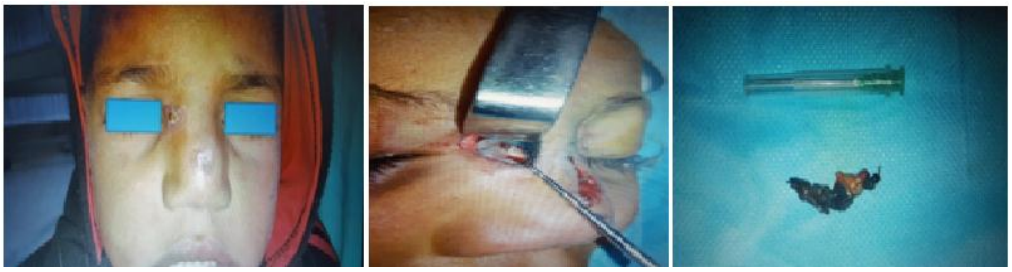

Fig-3: Clinical and intra-operative aspect of a superinfected and resected fistula of the 2nd case

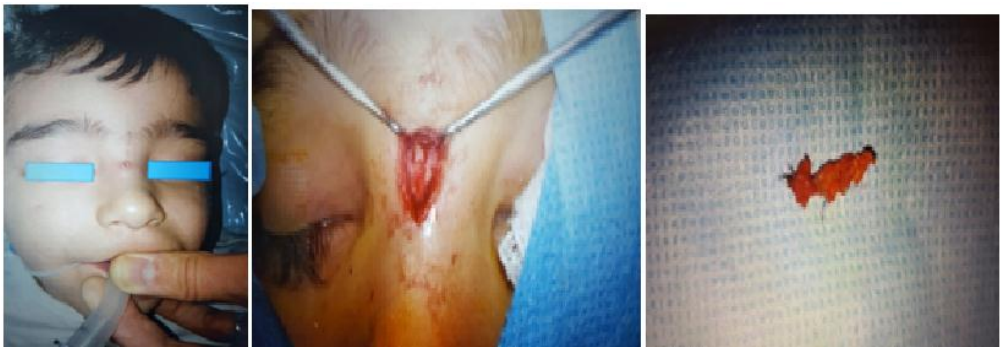

Fig-4: Clinical and intra-operative aspect of fistula in the 3rd case

DISCUSSION

Cysts and fistulas of the back of the nose are rare. These are malformations that remain strictly limited to the median line, extending from the columella to the glabella [1]. Sometimes there is one or more hairs centered on the fistula [2,3]. The frequency of all nasal cysts and fistulas is 1 in 40,000 births. Etiopathologically, it is an ectodermal inclusion during early embryonic development, due to the lack of closure of one of the two openings that are: the small fontanelle and the meridian hard canal [4]. There is no particular terrain, most often sporadic cases, rarely family [5]. There are also associated cranio-facial malformations in 23% of cases, which should be systematically sought, deep extension is possible with presence of contact with lepto-meningeal spaces [6]. CT or MRI imaging is therefore useful preoperatively to clarify the deep extension and the reports of malformation with the screened plate and meninges [7]. The differential diagnoses to be evoked in front of a medial frontal defect or the back of the nose are gliomas, dermoid cysts and meningo-encephaloceles or deep hemangioma [1]. The main risk lies in the meningeal or ependymal communication to be systematically searched for by CT

and / or MRI [8]. The CT remains the examination sufficient to make the diagnosis in case of superficial lesions; MRI is indicated in case of deep lesion with bone extension to clarify the relationship with the meninges as well as the tissue nature [9].

Surgical excision should be performed early because of the major risk of infection [1]. Some authors advise waiting at least 2 years to offer surgery when the majority prefer to operate as soon as the diagnosis is made before the infection occurs [10]. The curative gesture thus consists of an excision of the entire fistulous path by a way that can be discussed.

The medial vertical incision, long practiced, is ideal because it allows, from a small collar around the fistulous orifice, to follow easily along its length. The transcolumellar route is a good choice for short fistulas. In other cases, a horizontal path should be preferred, raising a glabellar flap with an upper pedicle. When intracranial extension exists, a doublé approach is required, associating the glabellar flap with a bitragial scalp with frontal flap [11].

In light of these data it is concluded that a radiological assessment should be carried out in front of any congenital tumefaction or fistula of the nose to see the intracranial extension before any surgical procedure as the case in our patients who have all benefited from a radiological assessment (CT or MRI).

It should also be diagnosed very early in infants before superinfection episodes as the case in our 3 patients who had a delayed diagnosis which caused several episodes of superinfection treated without precise diagnosis. After resection of the fistula at its base we note that there is no more superinfection and thus a complete cure.

CONCLUSION

In front of a swelling or congenital fistula of the nose, a radiological assessment is needed to determine the nature of the mass and to seek communication with the endocranium. MRI is the exam of choice in infants less than two years old. The risk of superinfection, very common, is to know because it is often the mode of revelation. Their management, mainly surgical, should not be delayed.

AUTHOR'S CONTRIBUTION

All the authors have contributed to the redaction of this manuscript.

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