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Case Report

Presentation of a case of a dermoid cyst with a rare location

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ABSTRACT

Dermoid cysts are choristoma that accumulate normal embryonic tissue in abnormal locations, making up 3% to 5% of congenital orbit conditions. They are found in various parts of the body, including the head and neck, frontal, periorbital, scalp, cervical, and nasoethmoidal regions. Nasoethmoidal dermoid cysts can compromise deep structures and may extend intracranially, with a frequency of 1% to 45%. They are usually diagnosed in the first 3 years of life, but some may go undetected until adulthood. Two theories explain the origin of nasal dermoids: the cranial origin theory and the superficial inclusion theory. Nasal dermoids can be pale, flesh-colored, pearly, or erythematous and may be accompanied by congenital anomalies. Different diagnoses include epidermal cysts, gliomas, meningoencephaloceles, tumors of vascular origin, fibromas, neurofibromas, osteomas, and lipomas. Imaging evaluations are necessary for diagnosis, determining the degree of involvement of neighboring structures, rule out associated malformations, and establish potential intracranial extension. The only curative treatment for nasoethmoidal dermoid cysts is surgical removal, as untreated lesions can cause nasal deformities, recurrent infections, and intracranial extension.

Keywords: Dermoid cyst, Choristoma, Benign tumor, Congenital tumor, Midline dermoid cyst

INTRODUCTION

The dermoid cyst is a choristoma (accumulation of normal embryonic tissue in an abnormal location). It represents 3 to 5% of congenital conditions of the orbit, where it is the most frequent benign tumor in childhood. They are usually well-demarcated lesions consisting of a fibrous capsule with a squamous epithelium lining and containing a yellowish pasty material, with skin attachments (hairs, sebaceous glands and sweat glands) also characteristically found.[1] Most cases are sporadic, however, in some cases there is a familial predisposition.

Dermoid cysts can be found anywhere in the body, especially in relation to sutures and embryonic clefts. They are more frequent in the head and neck, especially in the tail of the eyebrow. Other sites of presentation are the frontal, periorbital, scalp, cervical and nasoethmoidal regions.[2] The latter is an infrequent location and was first described in the literature by Bramann in 1890.[3] Nasoethmoidal dermoid cysts differ from those of other locations, since they have the potential to compromise deep structures and may extend intracranially, which according to different series fluctuates between 1 and 45% of the cases.[4] Other possibilities of extension include the frontal bone and the palate. It is important to always consider this possibility, and each case should be carefully evaluated in order to plan the surgical approach and treatment that allows complete removal of the lesion.

Nasoethmoidal dermoid cysts are usually diagnosed in the first 3 years of life, although some may go undetected until adulthood.[5] Their incidence is 1 per 20 000 live newborns, without differences by sex, they constitute 1 to 3% of all dermoid cysts and 11 to 12% of those of the head and neck. Among children, they represent 61% of nasal midline lesions.[6,7]

Two theories attempt to explain the origin of nasal dermoids:[8]The cranial origin theory: nasal dermoids result from defective recession of the dura mater from the prenasal space during normal embryogenesis. The superficial inclusion theory: nasal dermoids are formed by submucosal entrapment of the ectoderm during fusion of the lateral nasal processes. Nasal dermoids are classically described as noncompressible masses on the nasal dorsum, with a midline pit or midpoint located anywhere along the dorsal surface of the nose.[9]

Lesions have been variably described as pale, flesh-colored, pearly or erythematous. An unusual, but important, presentation is that of an ill-defined yellowish plaque of the midline nasal dorsum.[10]

The ability to express sebaceous material or the protrusion of a hair from the pit or punctum suggests the presence of a dermoid.[8] Pits may not be noticeable unless they are draining or infected; they may end near the skin surface, but often extend deep into the cribriform plate.

Associated congenital anomalies such as cleft defects, auditory atresia/ hydrocephalus have been reported in up to 41% cases. Presence of associated anomalies increases the frequency of intracranial extension from 31-65%.[11]

Differential diagnosis of nasal midline lesions includes epidermal cysts, gliomas, meningoencephaloceles, tumors of vascular origin, fibromas, neurofibromas, osteomas and lipomas.[7] Every patient with suspected dermoid dermoid requires an adequate imaging evaluation, especially in cysts of long evolution due to the greater probability of deep bone involvement and meningo-encephalic extension and in midline lesions as in the case of nasoethmoidal dermoids, which is performed with high resolution CT and/or MRI. In addition to confirming the diagnosis, these allow us to determine the degree of involvement of neighboring structures, rule out presence of associated malformations, present in up to 19% of cases, and establish a potential intracranial extension.[9]

Only curative treatment for nasoethmoidal dermoid cysts is surgical removal. Untreated lesions grow progressively and may cause nasal deformities, recurrent local infections and nasal airway obstruction. Intracranial extension may cause meningitis and brain abscess. Dermoid must be completely excised, even small amount of residual epithelium can be source of recurrence.[12]

Surgical approach to nasoethmoidal dermoid cysts can be through a direct nasal route over the lesion as in our case. Another alternative surgical approach in selected cases is open rhinoplasty, which avoids incisions in the nasal dorsum, but only allows access to low nasal lesions, which is why it was not considered in our patient.[5,7] In cases in which preoperative evaluation shows intracranial extension, coronal approach with frontal craniotomy and transcranial excision of the lesion is proposed, in addition to a direct approach to the nasal dorsum, being therefore indispensable the participation of a neurosurgeon.

CASE REPORT

The case of a 2-year-old female patient is presented. The mother reports that since birth she noticed a bulge at the level of the inner canthus of the right eye that has grown more rapidly in recent months not associated with epiphora/other relevant orbital or nasosinusal symptoms, except for cosmetic alteration caused by lesion described.

Ophthalmological examination showed a visual acuity of 0.4 logMAR with Teller chart in both eyes, noting on inspection discrete facial asymmetry due to an overelevation of the skin of the right inner canthus just below the insertion of the medial canthal ligament caused by nodule-shaped lesion of ±15 mm in diameter, painless, soft, smooth, well delimited and adhered to deep planes with no outflow of any material to acupressure (Figure 1).

Figure 1: Nodule-shaped lesion of ±15 mm in diameter, painless, soft, smooth, well delimited and adhered to deep planes with no outflow of any material to acupressure.

Bilateral biomicroscopy showed absence of lower lacrimal puncta in the right eye and permeable lacrimal puncta in the left eye. The rest of the ophthalmologic examination was within normal parameters.

A tomographic study was requested in order to better characterize the lesion, which showed the presence of a homogeneous tumor with well-defined borders of approximately 15 mm located in the preseptal space of the left inner canthus, with similar density to the encephalic mass and muscle bellies. No expansive or lytic bone lesions associated with the tumor were observed. Compression of the lacrimal duct was observed obstructing the duct lumen by the lesion (Figure 2).

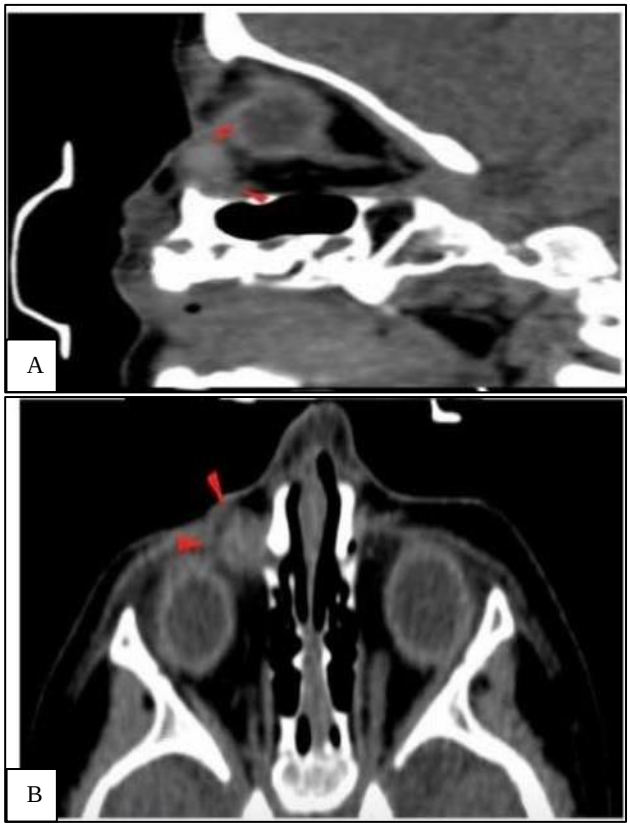

Figure 2 (A and B): Compression of the lacrimal duct was observed obstructing the duct lumen by the lesion.

After anesthetic evaluation with paraclinical examinations in normal parameters, under general anesthesia, an approach is performed through a direct nasal route over the lesion. A spindle-shaped incision is made with a scalpel blade no. 15 and dissection by planes is started with metzenbaum forceps until the base of the cystic mass is located, it is completely removed respecting adjacent anatomical structures. The skin is sutured with 6-0 nylon, hemostasis is checked and the procedure is finished.

The specimen obtained was sent for histopathological study. After surgery, the patient's evolution was satisfactory. The histopathological study showed a cystic formation consisting of stratified squamous epithelium suggestive of true dermoid cyst. The patient evolved satisfactorily, she is being followed up asymptomatically, with no evidence of recurrence after two years of follow-up (Figure 3).

Figure 3: No evidence of recurrence after two years of follow-up.

DISCUSSION

Dermoid cysts, although congenital, often manifest after birth and may present at various sites in the body, particularly in areas related to embryonic sutures and clefts. Consistent with previous studies, nasoethmoidal dermoid cysts represent a rare but important subset of head and neck dermoids, diagnosed predominantly within the first three years of life. Their incidence of approximately 1 in 20,000 live births aligns with reports by Harrison et al where no gender predilection was observed, and they comprised 11-12% of head and neck dermoid cysts, similar to our findings.[1,5,7]

Clinical presentation was the typical presentation of nasoethmoidal dermoid cysts includes a mass on the nasal dorsum or inner canthus, which is consistent with our patient's lesion at the right inner canthus. Our case supports observations by Pratt et al which emphasize the importance of early diagnosis and intervention due to the potential risk of intracranial extension in 19% of cases, as noted in MRI and CT imaging. This parallels earlier studies by Gorlin et al which also underscored the importance of imaging in excluding central nervous system involvement.[2,4]

Diagnostic evaluation imaging played a crucial role in our case, confirming the absence of bony involvement and central nervous system extension, consistent with Lafferty et al recommendations for utilizing high-resolution CT and MRI for diagnostic confirmation. The cyst's location, midline nasal, required differentiation from other nasal midline lesions, such as gliomas or meningoencephaloceles, echoing findings by Feijoo et al where similar differential diagnoses were proposed.[8,9]

Surgical management in terms of treatment, complete surgical resection is the definitive approach for nasoethmoidal dermoid cysts, as supported by several studies. In our case, a direct nasal approach was used, ensuring complete removal of the lesion. This mirrors the approach suggested by Rosbe et al who emphasize that incomplete excision increases the risk of recurrence. Our patient's two-year follow-up showed no recurrence, consistent with literature indicating that thorough excision results in low recurrence rates.[11,12]

Comparison with previous studies the characteristics of the cyst in our case-a soft, well-demarcated, and painless nodule with no signs of infection-are consistent with descriptions in the literature, including findings by Narang et al where similar clinical features were noted. Additionally, our patient had no associated anomalies, such as cleft defects or hydrocephalus, which have been reported in up to 41% of cases in studies like those by El-Hakim et al further supporting the variability in presentations.[6,8]

CONCLUSION

According to the bibliographic references consulted, the incidence of dermoid cysts of the dorsum of the nose, internal canthus or midline is low, since those related to embryonic sutures and clefts, especially eyebrow tail cysts, are more frequent and that no recurrences are observed after surgical treatment if it is completely removed.

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