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Congenital pediatric nasal dermoid on the lateral part of the nasal root – a series of two cases

Wrodzona torbiel dermatoidalna nosa u dzieci na bocznej części nasady nosa – seria dwóch przypadków

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Keywords

nasal dermoid cyst, dermatoid cysts, intracranial extension

Summary

Nasal dermoid cysts are rare congenital defect resulting from an abnormality in embryonic development. They are usually located along the fusion line of the germ layers. When present superficially, nasal dermoid cysts are easy to detect and appear as firm, non-pulsating, slow-growing masses. The treatment of choice is complete surgical removal, regardless of their size. In any case, computed tomography (CT) should be performed before surgical treatment to exclude a possible intracranial connection.
We present two cases of patients admitted to the hospital for the removal of nasal dermoid cyst. The performed CT examinations excluded the presence of communication with the cranial cavity. No postsurgical complications were observed. Postoperative histopathological examinations confirmed the diagnosis of dermatoid cysts.

Słowa kluczowe

torbiel dermoidalna nosa, torbiel dermatoidalna, naciekanie wewnątrzczaszkowe

Streszczenie

Torbiele grzbietu nosa są rzadką wadą wrodzoną wynikającą z nieprawidłowości w rozwoju embrionalnym. Przeważnie usytuowane są wzdłuż linii fuzji listków zarodkowych. Gdy występują powierzchownie, są łatwe do wykrycia i objawiają się jako spoiste, niepulsujące, wolno rosnące masy. Leczeniem z wyboru jest całkowite chirurgiczne usunięcie niezależnie od ich wielkości. W każdym przypadku, przed leczeniem chirurgicznym należy wykonać diagnostykę obrazową w postaci tomografii komputerowej, aby wykluczyć możliwe połączenie wewnątrzczaszkowe.
Prezentujemy przypadki dwóch pacjentek, które zgłosiły się do Kliniki Otolaryngologii Dziecięcej celem usunięcia torbieli dermatoidalnych zlokalizowanych u podstawy nosa. Wykonane badania TK wykluczyły obecność komunikacji z jamą czaszki. Wykonane zabiegi usunięcia torbieli przebiegły bez powikłań. Pooperacyjne badania histopatologiczne potwierdziły rozpoznanie torbieli dermatoidalnych.

Introduction

Nasal dermoid cyst is a rare congenital abnormality in children, caused by an incomplete obliteration of neuroectoderm during the development of front-nasal process (1). Typically localized along lines of embryonal skin fusion dermatoid cysts are noncompressible, non-pulsatile, slow growing subcutaneous firm masses. When superficial, they are easily detectable (2). Even though most nasal dermoid cysts are isolated lesions, some of them might have an intracranial extension through a persistent nasal dermal sinus tract (3). Complete surgical removal is the gold standard of treatment. Due to the risk of intracranial communication or recurrence, surgical treatment should be carefully planned to avoid complications. Computed tomography (CT) is examination of choice in preoperative evaluation (4).

Hereby we report a series of 2 cases illustrating various presentations of nasal dermoid in pediatric patients.

Case 1

A 16-year-old girl with a slowly growing subcutaneous nodule of the size of a chickpea grain localized on the upper right side of the nasal root with no previous history of

headache, trauma, surgical treatment, or medical complications was admitted to the hospital due to cosmetic complaints. Moreover, no inflammation or purulent discharge from the lesion were observed. The lesion was noticed for the first time at the age of 3 months with its progressive growth observed over the years. The girl remained under the supervision of a neurologist and ophthalmologist.

Physical examination revealed a cystic, well-defined, non-tender, non-compressible and non-pulsatile nodule of the diameter of 10 mm with unchanged surrounding tissue/skin or underlying bones. CT showed a well-defined low-density lesion measuring 13 x 10 x 9 mm on the lateral surface of the nasal bridge, in the right corner of the nose. The lesion was not enhanced after administration of the contrast medium. There was no evidence of bone destruction of the orbit or pressure on the structures of the right orbit (fig. 1). Contrast enhanced magnetic resonance imaging of the head revealed a nasal dorsal lesion (12 x 11 x 12 mm) with a potential intracranial extension of the sinus tract without a definite intracranial component. Additionally, a periradicular cyst at the spinal level Th 10-Th11 was described. The lesion measuring 15 mm in diameter was removed entirely by extracranial excision without a disruption in the continuity of its walls as well as with no intraoperative signs of the intracranial extension (fig. 2a, b). Histopathological examination of the lesion confirmed the diagnosis of a nasal dermoid cyst. No recurrence was observed during the postoperative follow-up period of 8 months (fig. 2c).

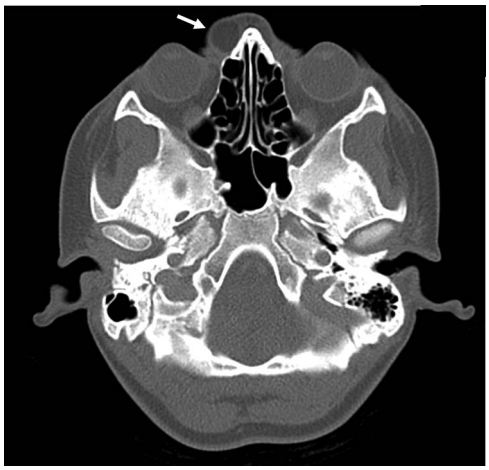

Fig. 1. Contrast enhanced computed tomography (CT) of the head, cross-sectional view, showing a hypodense lesion (arrow) on the surface of the nasal bridge with no evidence of bone destruction of the orbit

Case 2

A 2-year-old girl with a subcutaneous nodule recently noted by her parents around the nasal root and the left eye socket was admitted to the hospital (fig. 3a). The patient was referred to the Clinic by a pediatrician who had seen the child two months earlier, when the previously invisible change became prominent. A periorbital dermoid cyst was suspected. Non-contrast enhanced CT scan showed a 1-cm-sized encapsulated, fat-containing lesion with foci of calcification in the superior aspect of the medial wall of the left orbit (fig. 4a, b). Whereas a discreet depression of the outer lamina of the frontal bone was present, neither disruption in the continuity of the periostium nor signs of bone destruction were observed. Ultrasound examination revealed a well-circumscribed, isoechogenic, round lesion of 8 mm in diameter in the area of the medial canthus of the left eye, on the border with the root of the nose. Surrounding tissue remained unchanged. While no abnormal flow within the lesion was observed, small vessels were visible superficially. Even though the continuity of the cortex of the adjacent bone was preserved, a slight modelling by the lesion could not be excluded. An elective surgery was performed 4 months later. In the meantime, neither increase nor inflammation of the lesion were observed. At admission a smooth, firm, non-tender, subcutaneous nodule localized on the nasal process of left maxilla was noticed during the physical examination. Slight bruising of the skin below the lesion was also present. Laboratory tests showed no abnormalities. A diagnosis of dermatoid cyst of the nasal root was made. The dermatoid was successfully excised and histologic examination confirmed the initial diagnosis. No complications or recurrence of the dermoid were observed during a 15-month follow-up (fig. 3b).

Fig. 2a-c. Nasal dermoid, intraoperative view during resection (a-b) and 8 months after the surgery (c)

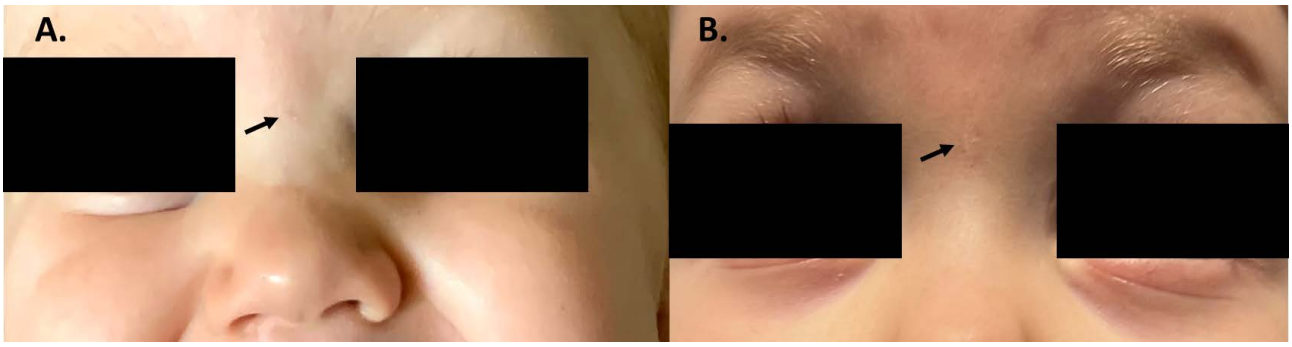

Fig. 3a, b. A 2-year-old girl with a subcutaneous lesion localized around the nasal root and the left eye socket: a) preoperative appearance of the lesion (arrow), b) postoperative appearance of the wound (arrow) 15 months after the removal

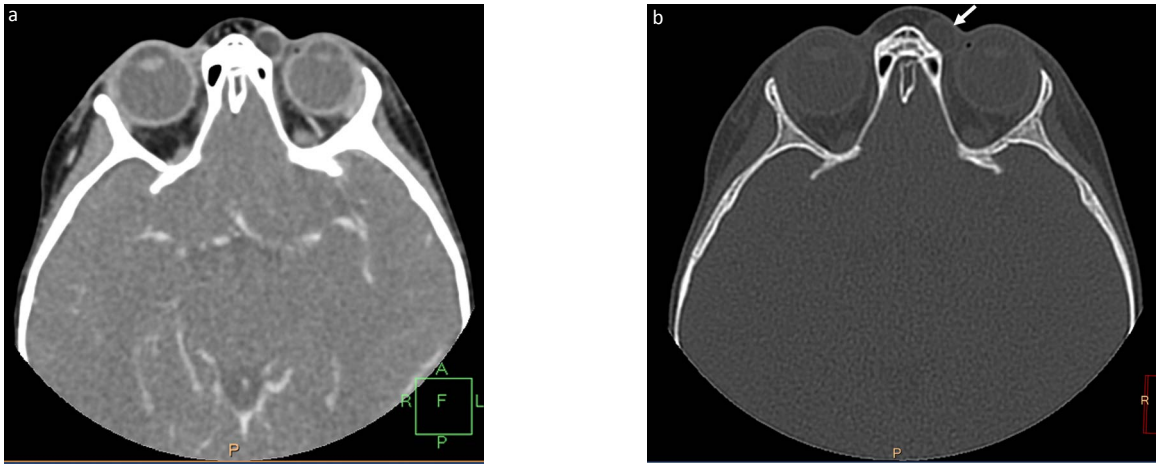

Fig. 4a, b. Non-contrast enhanced CT scan of the head, cross-sectional view, demonstrating an encapsulated, fat-containing lesion (arrow) with foci of calcification localized in the superior aspect of the medial wall of the left orbit (a, soft tissue window) without bone destruction (b, bone window)

Discussion

Medline nasal masses occur in one out of every 20,000--40,000 births of which the most frequently encountered are dermoid cysts (1). Dermatoid cysts are uncommon, isolated developmental anomalies which result from improper embryologic development. Typically localized along lines of embryonal skin fusion dermatoid cysts are noncompressible, non-pulsatile, slow growing subcutaneous firm masses. When superficial, they are easily detectable. Most often dermatoid cysts arise in the periorbital (zygomaticofrontal suture), nasal (frontonasal suture and rhinion), intraoral (floor of mouth), scalp (anterior fontanelle and cranial sutures), and postauricular area (5). Dermoids might cause deformation of the outline of the nose, with its secondary widening, similar to hypertelorism. Moreover, dermoid cysts can undergo recurrent infections leading to induration and, seldom, erosion of skin. In pediatric population they are frequently misdiagnosed as hemangiomas, epidermal cysts or sites of recurrent cutaneous infection (6). Noteworthy, dermoid cysts do not enlarge when the patient cries (6). The incidence of an intracranial extension is estimated to be between 4 and 57% and its presence cannot be determined by clinical examination. There was no relationship between the location of the lesion and the intracranial involvement (7).

In all cases, the diagnosis of a nasal dermatoid cyst requires preoperative radiographic imaging. It suggests or excludes the diagnosis of intracranial extension, which significantly facilitates the planning of the surgical procedure and determines the possible need for neurosurgical consultation (8). CT is considered to be the examination of choice in preoperative evaluation of the patient with nasal dermoid (4). While assessing the risk of intracranial extension, attention should be paid to the anterior cranial soft tissue masses, width or disruption of the nasal septum, the integrity of the skull base, presence of bifidity of crista galli or enlarged foramen cecum (9, 10). Whenever intracranial involvement is suspected, neurosurgical consultation is essential (10). Nasal dermatoid cyst with intracranial connections may require a multidisciplinary approach in the form of cooperation between neurosurgeons, plastic surgeons and otolaryngologists.

Surgical exploration along with histopathological examination are necessary to definitively exclude the presence of an intracranial connection (6). Due to the possibility of an intracranial connection, a biopsy of the lesion should not be performed (6, 13).

If the presence of ectodermal or neurogenic tissues is found, it is necessary to perform another procedure to remove intracranial elements. After their removal, the bone and meningeal defects are obliterated, and the brain is separated from the nasal cavity to avoid neuroinfection (6).

The standard management of nasal dermatoid cysts is complete surgical removal, regardless of its size (11). It is recommended to perform the procedure in the early childhood to avoid bony atrophy that may occur from expansion of the mass. Early removal reduces the risk of local infection and intracranial complications (12). However, choice of the exact moment is the result of a trade-off between the risk of damage to the developing structures and the possibility of later complications. Some consider age of two as the ideal age (12). There is an opinion that the procedure should be carried out immediately after the diagnosis is made (12, 14). Others, emphasizing the possibility of the lesion remain unchanged for a long time or can even become smaller, believe that many nasal dermatoid cysts do not require immediate treatment (12). Recurrence of the lesion may occur if it is incompletely removed (6, 15).

Complications associated with the removal of nasal dermatoid cysts can be severe, especially when there is an intracranial connection. Surgical procedures can cause cerebral fluid leakage and pose a risk of ascending infections. As a result, meningitis and even frontal lobe abscess may occur (6).

The presented cases are consistent with those described in the literature, both in terms of the characteristics of lesions, their histological structure and growth dynamics. Diagnosis and management of patients with nasal dermatoid cysts are not controversial, however, as it is a rare disease associated with potentially dangerous complications and therefore requires reporting of subsequent cases.

Conflict of interest
Konflikt interesów

None
Brak konfliktu interesów

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