Congenital nasolacrimal duct mucocele – a case report

Andrzej Brodkiewicz[1], Anna Zakowska[1], Jarosław Peregud-Pogorzelski[1], Maria Gizewska[2], Mirosław Burak[3], Magdalena Pastuszak-Gabinowska[1]

[1]Division of Pediatrics, Hematology and Pediatric Oncology, and [2]Division of Pediatrics, Endocrinology, Diabetology, Metabolic Diseases and Cardiology, and [3]Department of Diagnostic Imaging and Interventional Radiology, Pomeranian Medical University, Szczecin, Poland

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Congenital nasolacrimal duct mucocele (CNDM) is a very rare condition in newborns. Prolapse or expansion of the mucocele into the nose may lead to respiratory distress and difficulty in feeding. The triad of cystic medial canthal mass, dilatation of the nasolacrimal duct and a contiguous sub-mucosal nasal mass on computed tomography (CT) is indicative in the diagnosis of CNDM. The case of a five-week-old girl with infected CNDM is described. The authors aim to emphasize the very rare incidence of CNDM in Polish newborns, delayed diagnosis in the case described and the paramount importance of CT of the head for the correct diagnosis and treatment.

Key words: tumor, nasolacrimal duct mucocele, child, computed tomography.

Congenital nasolacrimal duct mucocele (CNDM) is a very rare condition in newborns. Prolapse or expansion of the mucocele into the nose may lead to respiratory distress and difficulty in feeding, as newborns are preferential nose breathers[1]. A lacrimal sac mucocele (LSM) occurs when the normal flow of tears through the nasolacrimal duct is obstructed, resulting in medial canthus mass. CNDM occurs when a LSM is directly contiguous with a cystic intranasal mass via a dilated nasolacrimal duct[2]. A mucocele is usually lined by ciliated columnar epithelium with goblet cells and secondarily filled with mucus and epithelial debris. Should this closed system develop a secondary infection, it is then called a mucopyocele[1]. Embryologically, the nasolacrimal passageway arises from a thickening of the ectoderm in the naso-optic fissure[1,3]. As the ectoderm thickens, it sinks into the mesenchyme, detaching itself from the surface ectoderm, and forms an epithelial cord. Caudally, the epithelial cord becomes the nasolacrimal duct[3]. At birth, the lower nasolacrimal duct is not usually open to connect with the nasal cavity. Most nasolacrimal ducts spontaneously open within the first year of life. Differential diagnosis of upper airway respiratory distress secondary to nasal pathology includes a variety of congenital malformations (e.g. choanal atresia, meningoencephalocele, hemangioma, dermoid cyst, Thornwald cyst, pyriform aperture stenosis), neoplastic tumors and hamartomas, trauma, and local infections[2,4-6]. Computed tomography (CT), magnetic resonance imaging (MRI), ultrasonography (USG), and nasal endoscopy play key roles in the diagnosis of CNDM and differential diagnosis of other lesions of this region (especially including neoplasms)[3,5]. Several treatment modalities have been proposed for the management of CNDM[5,7,8]. Spontaneous resolution may also occur.

Case Report

The five-week-old girl was admitted to the Department of Pediatrics, Hematology and Pediatric Oncology due to suspicion of a nasopharyngeal tumor. Pregnancy and delivery were uneventful. The parents were healthy and unrelated.

When the child was born she presented narrow lid slit and gradually increasing thickening in the right internal canthus. Additionally, the girl presented a mild degree of respiratory

distress when she was fed. Due to purulent conjunctivitis, the patient had been treated with antibiotic eye drops on several occasions (4 times) since the fourth day of life. Such therapy proved to be only temporarily beneficial. Lack of permanent improvement prompted ambulatory nasal fiberscopic examination leading to the diagnosis of a tumor-like lesion adjacent to the right-sided aspect of the nasal septum. On the day of admission, physical examination revealed only narrowed right lid slit with visible thickening in the right internal canthus. Only a mild degree of respiratory distress was noted when feeding. The infant was sleeping comfortably with oxygen saturations above 98% and with normal temperature. The inflammation parameters were negative. Blood morphology was within the normal range. Contrast-enhanced CT examination performed on the first day of hospitalization revealed intranasal, avascular mass in enlarged nasolacrimal canal and lacrimal sac, which confirmed the NDM diagnosis. Dislocation of inferior nasal concha due to the pathological mass was also present. There was small narrowing of the right posterior nasal cavity (Figs. 1-3). CT examination was performed on multi-detector (MD)CT scanner (MDCT Somatom Sensation Cardiac 64, Siemens) using a low-dose regimen (110-125mA, 120kV). For contrast-enhanced phase, nonionic iodinated contrast material (Visipaque 320 mgl/ml; 10 ml) was injected intravenously (IV) manually. Radiation dose was reduced with CARE Dose4D techniques – with automatic tube current modulation during each tube rotation according to the patient's angular attenuation profile; estimated CT radiation exposure: Total DLP = 649, CTDIvol = 17.52 and 19.22. 0.6 mm-thick images with a 0.6 mm-interval were acquired craniocaudally before and after contrast injection. Image postprocessing was undertaken on a workstation (Syngo, Leonardo Workstation, Siemens); standard and oblique multiplanar reconstruction was used.

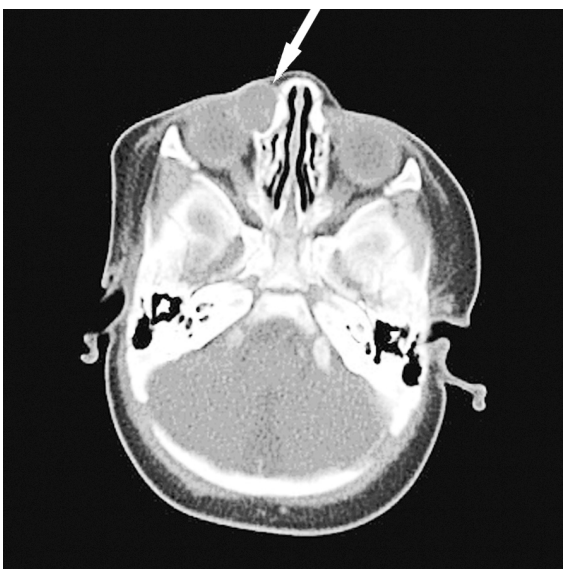

Fig. 1. Transverse CT scan shows the hypo-attenuating mass in the medial part of the right orbit (white arrow).

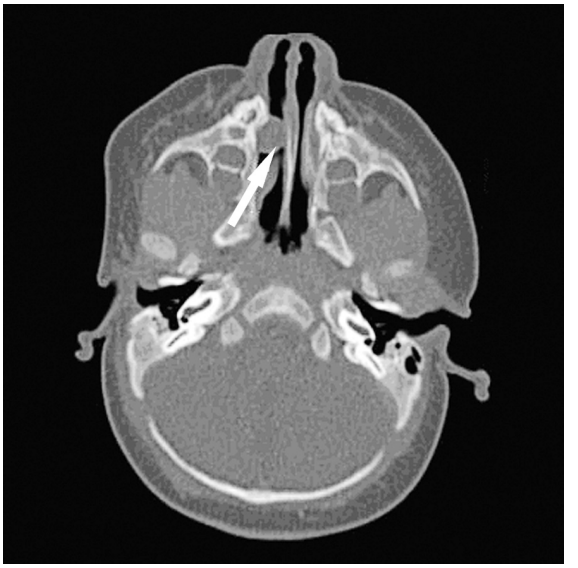

Fig. 2. Transverse bone window CT scan shows pathological mass in the right inferior nasal meatus (white arrow).

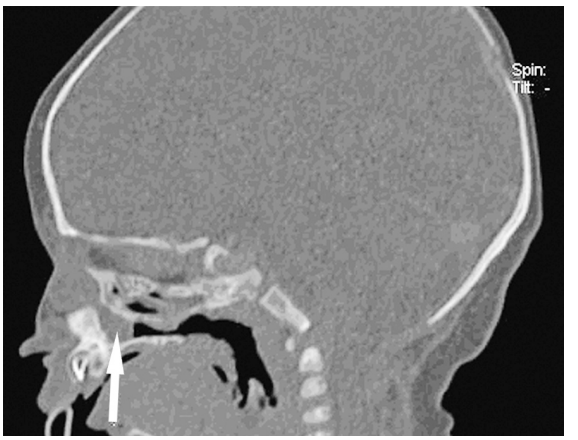

Fig. 3. Sagittal reconstruction bone window CT scan shows communication between the hypo-attenuating mass in the medial part of the right orbit with the dilated nasolacrimal duct (white arrow).

Right nasolacrimal duct probing was performed (under local anesthesia in the operating room) by the ophthalmologist, and 3 ml of mucopurulent discharge was evacuated. The cultured discharge revealed the presence of *Staphylococcus aureus*. Additionally, the treatment included massage and topical (tobramycin 0.3% for 5 days) and intravenous antibiotic (ceftriaxone 2 x 250 mg IV for 10 days), resulting in prompt clinical improvement. The child did not require surgical treatment.

Follow-up CT scheduled by an ophthalmologist performed four weeks later was normal. The patient did not show any pathological clinical symptoms. Currently, the child in under follow-up in the Outpatient Laryngology Department.

Discussion

Congenital nasolacrimal duct mucocele was believed to be rare before 1995, when 21 cases were reported[6,9]. Currently, it is believed that CNDM prevalence is higher than previously thought. Between 1998 and 2002 in the Hospital for Children (Falls Church, Virginia), Brachlow et al.[9] diagnosed symptomatic CNDM in 10 neonates. Within the past few years, several pediatric centers have reported series of children ranging from 5 to 22 cases with CNDM[6,9-11].

These data are not similar to our observations. Between 1990 and 2009, only one case, the currently described CNDM patient, has been hospitalized in the 1st Department of Pediatrics/ Department of Pediatrics, Hematology and Pediatric Oncology. One additional patient was hospitalized in the same period in the Department of Neonatology (verbal information). The extremely low number of CNDM patients hospitalized in our departments may be the result of hospitalization of these children in other hospitals (which seems rather unlikely), low prevalence of CNDM and/or lack of proper diagnosis of CNDM (also rather unlikely).

The presenting symptoms range from none or only benign nasal stuffiness to frank cyanotic spells[9]. It should be emphasized that neonatal and infant nasal ducts are significantly narrower when compared to elder children. Additionally, infants are obligate nasal breathers for the first 2-6 months of life (except during crying)[4,12]. Thus, even mild viral infection at that age may cause impaired nasal patency and significant breathing difficulty[1,4,9].

According to Levin et al.[7], Brachlow et al.[9] and Duval et al.[12], CNDM shows higher rates in girls than in boys. This finding is concordant with our observation, since the child described was a girl. Predisposition to CNDM development with regard to ethnic group is questionable[7]; according to Brachlow et al.[9], the prevalence of the disease is higher in patients of Hispanic ethnicity. The only child described is of Polish origin.

Various techniques are applied in diagnostics of CNDM. According to Schlenck et al.[10], ultrasound is a simple, straightforward and gentle method to reliably distinguish CNDM from other pathologies. However, a majority of authors favor CT scan for many reasons (mainly due to excellent soft tissue and bony definition), and thus it plays a crucial role both in the diagnosis of this entity and in its differentiation from a variety of other causes of impaired nasal patency. This very uncommon finding presents usually with a diagnostic triad on CT: an intranasal cystic mass, and a dilated nasolacrimal duct and lacrimal sac[3,5]. This is also consistent with our findings. The girl described was referred to our Department following initial diagnosis of nasopharyngeal tumor (suggested rhabdomyosarcoma [RMS]) that was made after nasal endoscopy in the 5th week of life. Due to the child's age and suspicion of malignancy on admission to the Department of Pediatrics, Hematology and Pediatric Oncology, the CT head scan was performed, which finally and indubitably confirmed the diagnosis of CNDM.

It should be emphasized that discrete symptoms suggestive for CNDM developed immediately after birth. Parents and physicians observed narrow lid slit, gradually increasing thickening in the right internal canthus and permanent purulent conjunctivitis. Additionally, the girl presented with a mild degree of respiratory distress when feeding. Initial diagnostic difficulty may be justified by the mild symptoms presented by the patient that might have misled neonatologists, pediatricians and ophthalmologists in numerous exams. In addition, poor knowledge of that entity caused by the rare prevalence of CNDM in the Polish

population might have contributed to the delay in diagnosis.

Several treatment modalities have been proposed for the management of CNDM, including massage, warm compresses, topical or parenteral antibiotics and, when still symptomatic, nasolacrimal probing, silastic stenting and intranasal endoscopic cyst marsupialization with e.g. mucosal wall resection[7,8,11]. In the described case, the nasolacrimal duct was mechanically opened by the ophthalmologist and mucopurulent discharge was evacuated. The conservative treatment (including massage and topical/intravenous antibiotic) was started, resulting in rapid clinical improvement.

Follow-up CT scan (MDCT Somatom Sensation Cardiac 64, Siemens) was performed at the request of the ophthalmologist four weeks after admission to the Pediatric Department. The scan revealed no pathological changes. Prompt clinical improvement following the treatment should have excluded a consecutive CT. As a result, the child acquired additional radiation dose and faced unnecessary stress.

Neonatologists, pediatricians, general practitioners, ophthalmologists, and laryngologists should be familiar with CNDMs. According to Brachlow et al.[9], anterior rhinoscopic examination should be a part of routine newborn examination, especially when a newborn or neonate presents with symptoms of nasal obstruction. It is particularly important to rule out malignant tumors requiring immediate treatment. The authors would like to underline the very rare incidence of CNDMs in Polish newborns and young neonates, the delayed diagnosis in the described case and the paramount importance of CT head scan for the correct diagnosis and treatment.

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