theory could not be proven in that particular case. Unfortunately we were not able to study the genetics of our patient’s tumors or his constitutional genotype.

The present case raised a significant diagnostic dilemma. Due to the history of a previously resected meningioma and the recognized tendency of recurrence, and because of its radiographic appearance, the right temporal tumor was originally suspected to be a meningioma. However, unexpectedly, it was associated with intense concentration of MBG. In view of the rarity of cranial pheochromocytoma metastases we speculated on the possibility of meningioma-associated catecholamine secretion, but this was rejected by the histopathologic findings that were compatible with metastatic pheochromocytoma, as well as by the lack of secretory characteristics in the meningioma tissue.

Two previous reports on coexistent meningiomas and pheochromocytomas referred to benign pheochromocytomas [1,2]. To the best of our knowledge, the present case is the only available report of a patient with meningioma associated with malignant (metastatic) pheochromocytoma.

Following removal of the right frontotemporal metastasis of pheochromocytoma, a marked decrease in urinary catecholamine excretion was observed; however, urinary catecholamines excretion subsequently increased beyond their preoperative level [Figure B]. Administration of MBG, used as a pharmacologic vehicle for conveying radioactive iodine to the pheochromocytoma metastases, produced an initial apparently encouraging response, based on an observed decrease in urinary catecholamines and metanephrine excretion after each administration. However, evidence for remission was short-term [Figure B], as post-MBG scans failed to show regression of the metastases and catecholamine excretion subsequently increased. The discrepancy between the observed fall in catecholamine secretion on the one hand and the consistency of MBG scans on the other underlines that caution should be applied to the interpretation of urinary catecholamines and metanephrine excretion as markers for monitoring the therapeutic response in patients with malignant pheochromocytoma.

References

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Bilateral Congenital Nasolacrimal Duct Cyst: An unusual Cause of Respiratory Distress in the Neonate

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Neonates are obligate nasal breathers. Nasal obstruction causes acute respiratory distress immediately following birth and is especially evident during feeding and sleeping. During crying there is improved airway, so that the clinical finding is “cyclic cyanosis.” If both nasal airways are completely obstructed, the neonate has a potentially life-threatening disease. When a child is born with respiratory distress, catheters are passed through the nostrils to rule out choanal atresia. However, other pathologies can cause nasal obstruction in the neonate including: pyriform aperture stenosis, nasopharyngeal teratoma, meningoecephalocoele and others. Congenital obstruction of the nasolacrimal drainage system occurs commonly [1] but rarely causes nasal obstruction. We describe a rare case of a newborn with congenital bilateral nasolacrimal duct cyst located in the nasal cavity who presented with respiratory distress and required prompt surgical treatment.

Patient Description
A full-term male infant was born by vertex vaginal delivery with Apgar scores of 9/9 at 1 and 5 minutes. Soon after birth he was noted to have respiratory difficulties. Phy-
Physical examination was normal except for the respiratory findings and purulent discharge in the right medial canthal area. A 5F catheter could be passed on both sides. Nasal endoscopy revealed a bilateral smooth and soft cystic mass located in the inferior meatus obstructing the nasal airway (Figure A). A computerized tomography scan confirmed the diagnosis of nasolacrimal drainage system cysts (Figure B). On day 5, endoscopic marsupialization of the cysts under general anesthesia was performed, yielding 2 ml purulent discharge from the right cyst while it was being marsupialized. Postoperatively the infant did well and had no recurrence of symptoms. Follow-up endoscopy was normal without evidence of the obstructing cysts. Of note was the resolution of the infection in the right eye.

Comment

Embryologically, the nasolacrimal apparatus begins developing in the third fetal month from a core of surface epithelium found between the maxillary and frontal nasal recesses. Canalization of this cord occurs uniformly throughout the entire length, and final communication with the inferior nasal meatus usually occurs by the sixth fetal month. If complete canalization fails to occur, it most commonly leaves a membranous barrier between the duct and the nasal cavity at the level of the valve of Hasner and may last up to or beyond the time of birth [2]. Such distal obstruction results in expansion of the duct and sac and may cause epiphora and mucoid discharge [3]. A congenital NLD occurs when there is a concomitant imperforate nasolacrimal duct distally and a valve-like obstruction at the junction of the lacrimal canal and sac proximally. Another 32 cases of congenital NLD have been published. Congenital NLD cyst presents clinically at birth or at a few weeks of age when tear production increases. NLD cysts occur more commonly in females than in males, and may be unilateral or bilateral. The differential diagnosis of intranasal mass includes nasal encephalocele, dermoid, hemangioma and nasal glioma. Other types of tumors have also been described; such as fibroma, lipoma, teratoma or, more rarely, carcinoma and sarcoma. Radiologic imaging is important for delineating the extent of the mass and is essential for ruling out any intracranial communication prior to biopsy or surgery. The recommended treatment of a NLD cyst is marsupialization [4]. This can be performed with the operating microscope or under endoscopic guidance.

It should be noted that nasopharyngolaryngoscopy (using a flexible endoscope) is recommended for diagnosis when symptoms of upper airway obstruction, stridor, dysphonia, or feeding difficulties appear in a neonate.

In summary, congenital NLD cysts with nasal obstruction are rare but must be included in the differential diagnosis when a neonate shows respiratory distress symptoms. Nasal endoscopy and CT are necessary to make the diagnosis. Surgical intervention is indicated when obstruction is symptomatic and results in immediate resolution of the symptoms.

References


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![A] Nasal endoscopy showing nasolacrimal duct cyst. IT = inferior turbinate, NS = nasal septum, C = cyst
[B] Axial CT demonstrating bilateral nasolacrimal duct cyst