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An unusual tumour causing neonatal respiratory distress

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Abstract. *An unusual tumour causing neonatal respiratory distress*. *Problem*: We present the case of a term neonate referred shortly after birth because of breathing and feeding difficulties.
Methodology: Fiber-endoscopic examination of the nasal cavity showed a pendulating mass in the nasopharynx.
Results: A complete surgical resection was performed and the baby recovered completely. Microscopic examination of the mass showed an overlying non-keratinized squamous cell lining with an atypical cell population in some fragments. Histological features were compatible with a high-grade epithelial tumour like a midline carcinoma, but a final diagnosis of a salivary gland anlage tumour was established.
Conclusion: Flexible fiber endoscopy is the method of choice for examining the nasal passages and oropharynx in neonates with respiratory distress. Congenital salivary gland anlage tumour is a rare cause of neonatal nasal obstruction; it is benign and complete excision results in a cure. Histologically, it may mimic a malignant tumour owing to the high mitotic index.

Introduction

Since neonates are obligate nasal breathers, any degree of nasal obstruction can lead to respiratory distress and/or feeding difficulties. A wide range of congenital, inflammatory, and neoplastic entities can cause nasal obstruction. Neoplasms are a rare cause of nasal obstruction during the neonatal period. The majority of these neoplasms are benign; however, malignancies such as rhabdomyosarcoma, neuroblastoma, and lymphoma do occur.

Salivary gland anlage tumour (SGAT) is a rare benign neoplasm of the nasopharynx. To date, there are approximately 28 cases reported in the literature. The term was first used by Dehner *et al*.[1] although other authors previously described what is probably a similar tumour.[2,3] This paper describes the case of a neonate with a SGAT and discusses the clinical evaluation and diagnostic challenges.

Case report

We present the case of a baby boy who was referred to the neonatal intensive care unit (NICU) on the day of birth because of breathing and feeding difficulties. The pregnancy had been uneventful, and he was delivered at term using vacuum extraction. He weighed 3.290 kg, and APGAR scores were 7 after 1 minute, and 9 after 5 minutes. At clinical examination, he exhibited respiratory distress with subcostal retractions, noisy nasal breathing (stertor), and occasional grunting worsening when he was supine. Chest auscultation was normal. There were no other clinical abnormalities or dysmorphic features.

Blood gases showed signs of respiratory acidosis. Insertion of a Mayo canula resulted in only a slight improvement in his clinical condition. Flexible fiber optic endoscopy of the nasal cavity showed a pendulating mass in the nasopharynx. To further identify the lesion, computed tomography (CT) was executed. This examination provided little additional information regarding the type of lesion, but a skull-base defect could be excluded.

At day 10, a complete surgical resection through a combined endonasal endoscopic and transoral approach was performed under general anesthesia. Macroscopically, the specimen resembled a papilloma or adenoid tissue. The baby recovered completely without signs of

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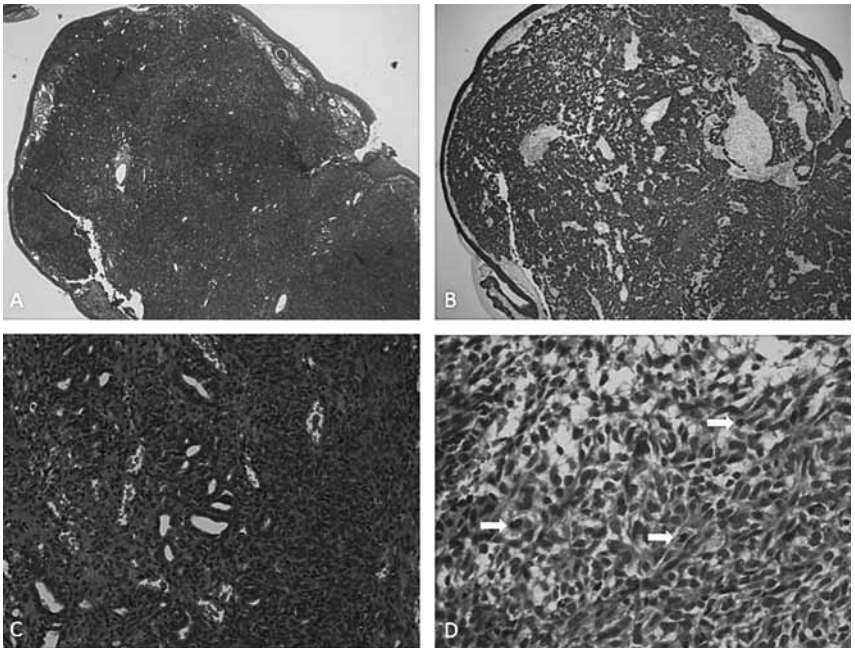

Figure 1
A: Overview of a polypoid tumour mass at the surface lined with non-keratinizing squamous mucosa. The tumour consist of solid nests, tubular to cord-like epithelial cells with a limited fibrovascular stroma in between them (hematoxylin-eosin, 4×).
B: Immunohistochemistry shows that both the mucosa and the tumour mass are pan-cytokeratin positive (pan-CK, 4×).
C: On the right side, a more solid growth pattern is seen, whereas more glandular structures are present on the left side (hematoxylin-eosin, 20×).
D: The tumour consists of more or less bland looking cells with round to ovoid nuclei, which are surrounded by a limited amount of ill-defined cytoplasm. The tumour cells show little cytonuclear atypia. Although mitoses are rare, they can be present. In this case, three mitoses (arrows) were seen in one high power field (hematoxylin-eosin, 40×).

respiratory distress or feeding difficulties.

No signs of recurrence of the lesion have been observed with follow-up to date (up to 10 months), and the patient continues to be in excellent clinical condition.

Microscopic examination of the mass showed an overlying non-keratinized squamous-cell lining with an atypical cell population in some fragments (Figure 1). Histological features were compatible with a high-grade epithelial tumour like a midline carcinoma. However, only few mitoses were present. International review analysis (Dr Fletcher, Harvard Medical School) revealed a superficially-benign glandular and squamous proliferation with numerous smaller glandular structures admixed in other areas merging with cytologically uniform spindled stromal cells. These findings are compatible with a SGAT. There was no sign of malignancy.

Discussion

Nasal obstruction during the neonatal period can be caused by a wide variety of entities, such as neoplastic, congenital, and inflammatory processes. Symptoms can also vary widely depending on the age of presentation and the severity of the obstruction. Symptoms can include potentially life threatening respiratory difficulties, but also more subtle signs such as nasal infections, feeding difficulties, nasal discharge, and cyanotic spells.

A thorough evaluation of the upper airway is mandatory in unexplained respiratory distress and obstructive breathing.[4] Flexible fiber endoscopy is the method of choice to examine the nasal passages and choanal opening, and to exclude obstructive lesions at the level of the nose, rhinopharynx, and oropharynx.[5] Identifying nasopharyngeal masses with endoscopy should be followed by radiographic imaging to evaluate the relationship with the surrounding tissues, and to exclude intracranial extension. In our case, CT did not provide any further information; this was possibly because the examination was performed with the Mayo canula in situ, which may have compressed the lesion. No intracranial extension was seen in our case. SGAT is characterized by a homogeneous appearance and lack of bone destruction on CT, although magnetic resonance imaging is the method of choice for soft tissue evaluation.

As shown in our case, pathologic examination is not always straightforward in infants because benign tumours at this age can mimic malignancies, usually owing to cells with a high mitotic index. Because SGAT is uncommon, international expertise may be required in the definitive diagnosis. Recognition of SGAT is, however, very important because simple excision of the lesion results in a complete cure.

Congenital SGAT is a rare cause of neonatal nasal obstruction. It is benign, and recurrence

has not been reported after complete surgical excision.[6,7] Dehner *et al*.[1] initially reported 9 cases of SGAT presenting at birth or within the first few days or weeks of life. The histological and architectural features of SGAT were similar to those of the developing salivary gland. It was proposed that SGAT is probably a hamartoma of minor salivary gland origin, and not a true neoplasm. Other authors previously described what is probably a similar tumour.[2,3] There is a male predilection. In 2005, there were a total of 23 cases described[8]; since then, another 5 cases have been reported.[6,8-10] Of these 28 cases, 19 occurred in males.

Cohen *et al*.[11] described a case were accidental dislodgement of the mass occurred following probing with a nasal catheter. The mass acted as a foreign body in the airway and there was profuse bleeding. SGAT are usually attached by a thin vascular pedicle to the posterior septum or posterior nasopharyngeal wall.[8]

Fiber optic endoscopy is the first diagnostic step, followed by radiologic imaging.

Microscopic features are usually consistent from one case to another,[8] with epithelial and mesenchymal components, and tubular structures that are found in the nodular and perinodular areas where they blend into stromal-mesenchymal nodules. In our case, there was also glandular and squamous proliferation and numerous smaller glandular structures merging with cytologically-uniform spindled stromal cells.

Surgical excision is the treatment of choice, followed by regular follow-up.

Conclusion

A nasopharyngeal mass should be suspected in neonates with unexplained respiratory distress and obstructive breathing. Flexible fiber endoscopy is the method of choice for visualizing the nasal passage to exclude obstructive lesions at the level of the nose, rhinopharynx, and oropharynx. If a mass is identified, radiologic imaging should be obtained to exclude intracranial extension. Congenital salivary gland anlage tumour is a rare cause of neonatal nasal obstruction. It is benign, and does not tend to recur if completely excised. Pathologic examination is not always straightforward in infants, because benign tumours at this age can mimic malignancies (usually due to the presence of cells with a high mitotic index).

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