IJN
Iranian Journal of Neonatology
http://ijn.mums.ac.ir

Open Access
Case Report

Coffee Ground Vomiting as the First Presentation of Salivary Gland Anlage Tumor: A Case Report and Review of the Literature

Seyed Hossein Saadat[1], Ali Atash Ab Parvar[2*], Fatemeh Kheiry[3]

1. Clinical Research Development Center of Children Hospital, Hormozgan University of Medical Sciences, Bandar Abbas, Iran
2. Endocrinology and Metabolism Research Center, Hormozgan University of Medical Sciences, Bandar Abbas, Iran
3. Student Research Committee, School of Nursing and Midwifery, Shiraz University of Medical Sciences, Shiraz, Iran

ABSTRACT

Background: Salivary gland anlage tumor (SGAT) is a rare but benign tumor of the nasopharynx that most commonly presents with symptoms of obstruction, including severe respiratory distress. There are multiple reports of SGAT in the literature proposing it as an important differential diagnosis of nasal obstruction in the early weeks of life. However, primary manifestations other than respiratory distress have not been reported with SGAT.

Case report: An18-hour-old neonate presented with feeding intolerance and coffee ground vomiting since the 6thhour of birth and was admitted to the neonatal intensive care unit. An orogastric tube was inserted for irrigation, and secretions were persistently bloody after 24 hours. At the 36th hour of birth, mild subcostal retraction developed with a slight increase in the respiratory rate as well as bloody secretions from the nose. At the 48th hour, subcostal retraction became severe accompanied by tachypnea. All symptoms resolved completely within 2 hours of spontaneous vomiting containing a polypoid mass.

Conclusion: The SGAT should be considered a differential diagnosis of nasopharyngeal obstruction in the early weeks of life.

Keywords*:* Case report, Nasal obstruction, Oropharynx, Respiratory distress, Salivary gland anlage tumor

Introduction

Salivary gland anlage tumor (SGAT) is a benign but rare tumor of the nasopharynx, which usually presents with nasal obstruction, followed by respiratory distress in the first year of life (1). The SGAT was first described in a case series by Jones et al. in 1994. They reported the clinical findings of a group of infants with respiratory distress due to midline polypoid lesions. Additional pathologic examinations led to the introduction of the SGAT (2). To the best of our knowledge, 43 cases of SGAT have been reported so far in the literature (3). In this report, we described a neonate with SGAT that presented with vomiting in the early hours of birth.

Case report

An 18-hour-old male neonate was admitted to the neonatal intensive care unit of our hospital for feeding intolerance and coffee ground vomiting since the 6th hour of birth. His prenatal history was unremarkable. He was the fifth child of the family born through normal vaginal delivery at 39 weeks of gestation with 1- and 5-minute Apgar scores of 9 and 10, respectively. His birth weight was 3250 g, and anthropometric indices were within normal limits. Upon physical examination, vital signs were normal with a respiratory rate (RR) of 52/min, and except for bilateral hand polydactyly, other examinations were normal. The patient was made NPO, and an orogastric tube (OGT) was inserted showing bloody secretions. Laboratory tests, including electrolytes, arterial blood gas, serum glucose level, sepsis workup, and coagulation tests were requested. Test results

* *Corresponding author*: Ali Atash Ab Parvar, Endocrinology and Metabolism Research Center, Hormozgan University of Medical Sciences, Bandar Abbas, Iran. Tel: +980763333328; Email: Dr_atashparvar@yahoo.com

Please cite this paper as:
Saadat SH, Atash Ab Parvar A, Kheiry F. Coffee Ground Vomiting as the First Presentation of Salivary Gland Anlage Tumor: A Case Report and Review of the Literature. Iranian Journal of Neonatology. 2022 Jan: 13(1). DOI: 10.22038/IJN.2022.54503.2013

showed prolonged prothrombin time (PT) and considering the presence of blood in gastric secretions, coagulopathies were suspected as the initial diagnosis; therefore, intravenous vitamin K (1mg/kg) was administered. Other laboratory test results were as following: white blood cells, 11800/µl (60% neutrophils and 40% lymphocytes); hemoglobin, 14.6 g/dl; platelet count, 245000/µl; C-reactive protein, 5 mg/dl; blood glucose, 60 mg/dl; serum calcium, 10.2 mg/dl; blood urea nitrogen: 11.5 mg/dl; creatinine, 0.68 mg/dl; sodium, 137 mEq/l; potassium, 3.7 mEq/l; thyroid-stimulating hormone, 1.9 mIU/l; T4, 11.4 µg/dl; total bilirubin, 7.8 mg/dl; and direct bilirubin, 0.4 mg/dl. Moreover, blood culture and Coombs' test were negative, and a peripheral blood smear was normal. Arterial blood gas analysis showed a pH of 7.45, PCO2 of 24 mmHg, PO2 of 100 mmHg, and HCO3 of 17.8 mEq/l. Abdominopelvic ultrasound was performed which was normal, and echocardiography demonstrated a mild-to-moderate tricuspid regurgitation and patent foramen ovale.

The OGT bloody secretions persisted despite the normalization of PT at the 24th hour of birth, and yet there were no signs of respiratory distress. At the 36th hour of birth, mild subcostal retraction developed with a slight increase in the respiratory rate from 52 to 58/min (which was still within normal limits) as well as bloody secretions from the nose. At the 48th hour, subcostal retraction became severe accompanied by tachypnea with an RR of 68/min. All symptoms resolved completely within 2 hours of spontaneous vomiting containing a polypoid mass.

At the 60th hour, the OGT secretions contained fresh blood. Spontaneous vomiting occurred containing a firm red polyp-shaped mass measuring 1×2 cm which was sent for pathologic examination (Figure 1). Subsequently, within 2 hours, all respiratory symptoms resolved completely. At the 72nd hour, the patients had no respiratory distress or gastric bleeding. On the 4th day of birth, he resumed feeding, and stayed in the hospital for another three days and was monitored for any complications. Consequently, he was discharged from the hospital on the 7th day with the satisfactory general condition. Histopathologic and immunohistochemical studies revealed a polypoid mass covered by squamous epithelium with proliferated blood vessels beneath the surface epithelium composed of two components in favor of SGAT (Figure 2). Upon

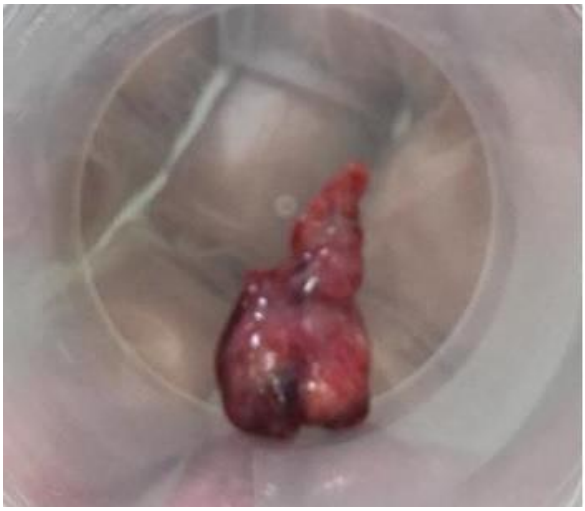

Figure1. A red firm polyp-shaped mass measuring

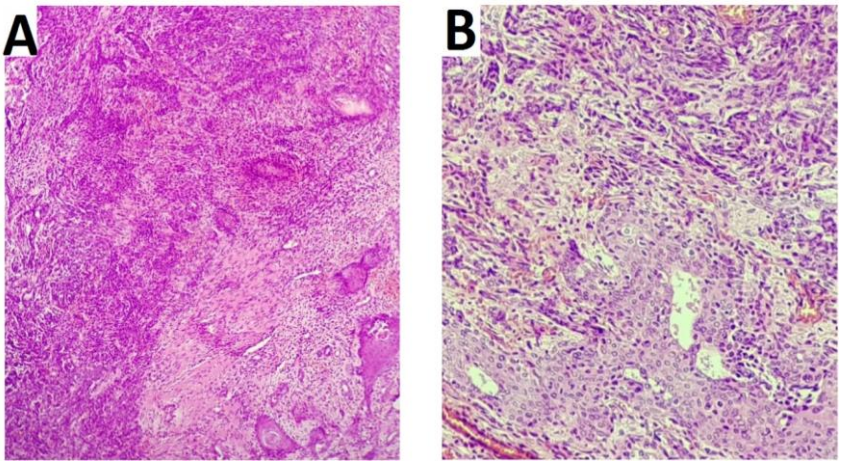

Figure 2. A: Lobulated lesion covered by non-keratinized stratified squamous epithelium, included epithelia-ductal unit and spindle cells, depicted foci of hypocellular area (H&E staining, 200× original magnification). B: Biphasic growth of bland-looking epithelial-ductal component with nests of squamous cells intermingle with monomorphic fascicles of spindle cells accompanied by proliferative blood vessels (H&E staining, 400× original magnification)

receiving the pathology results, parents of the patient were contacted and advised to return for further imaging or endoscopic examination to determine the presence of any remnants of the tumor in the nasopharynx; however, despite several phone calls, they failed to come back for follow-up most probably due to the long distance of their residence from the hospital and the good general health of the patient.

Discussion

The SGAT is an extremely rare, benign, congenital, and biphasic tumor with mixed epithelial and myoepithelial elements located near the midline in the nasopharynx or posterior nasal septum (4). It usually presents within the first six weeks of life with airway-related symptoms and/or nasal obstruction manifesting as respiratory distress and feeding difficulties (5).

In our case, the patient presented with coffee ground vomiting 6 hours after birth without any respiratory distress typical of SGAT. In other words, although the patient later developed progressive respiratory distress, the primary manifestation and the prolonged PT were suggestive of coagulation disorders leading to gastrointestinal bleeding. Nevertheless, with the persistence of bloody secretions even after the administration of vitamin K, coagulopathies were ruled out. As a matter of fact, the diagnosis of SGAT in the index case was made retrospectively when histopathological examination of the polypoid mass was performed. The later development of respiratory symptoms was only indicative of the presence of some kind of nasopharyngeal obstruction, and as there is a long list of possible causes of nasal obstruction, including nasal mucosal edema, infectious causes, structural abnormalities, cystic lesions, and midline lesions (e.g., glioma and meningo-encephalocele) (5), it was difficult to suspect a rare entity, such as SGAT.

Diagnosis of the SGAT is challenging and is usually based on clinical suspicion, magnetic resonance imaging (MRI), and computed tomography (CT) scans showing a lobular, midline soft tissue mass in the naso-oropharynx without infiltration into the surrounding structures (6). In one case report, MRI showed T1 isointense and T2 iso-hyperintense homogeneously enhancing mass measuring 1 to 2 cm (7). Imaging was scheduled for our case after the definitive diagnosis based on pathologic examination which was unfortunately canceled due to the patient's loss to follow-up.

For conclusive diagnosis of SGAT, the pathological examination should be performed on the resected mass revealing a lobulated nasopharyngeal proliferation with mixed epithelial and myoepithelial components (8). The tumor shows tubules and ducts with variable keratinization attached to the surface squamous epithelium accompanying multiple cellular spindle cell nodules (8, 9). Furthermore, the focal chondromyxoid matrix is noted, and the epithelial component shows diffuse and strong immune reactivity for cytokeratin and CK7 (10). The spindle cells show variable to strong immunoreactivity with CK7, p63, SMMS, and S100 (6). In our case, the immunohistochemical study demonstrated immunoreactivity with S100, CD31, CD 34, Ki67, CK, SMA, and P63. The most common differential diagnosis of SGAT includes dermoid cyst and respiratory epithelioid adenomatoid hamartoma which could be distinguished from SGAT using histopathological studies (11).

Consistent with the report by Swayampakula et al. (12), no interventions were required for our case to remove the lesion since it came out spontaneously with vomiting; however, SGAT can be treated with surgical resection. One of the limitations of our study was that we were unaware of the exact diagnostic or therapeutic measures taken for the patient's condition prior to referral to our hospital. The nasal mass may have previously been manipulated causing bleeding leading to coffee ground vomiting.

Conclusion

Our report demonstrates that in case of nasopharyngeal obstruction in neonates, especially in the first weeks of life, SGAT can be considered one of the differential diagnoses, and histopathological studies are necessary for definitive diagnosis.

Acknowledgments

None.

Conflicts of interest

The authors have no conflict of interest.

References

1. Başak K, Günhan Ö, Akbulut S, Aydin S. Salivary gland anlage tumour of the nasopharynx: A case report and review for histopathological characteristics. Malays J Pathol. 2019; 41(3):345-50.
2. Dehner LP, Valbuena L, Perez-Atayde A, Reddick RL, Askin FB, Rosai J. Salivary gland anlage tumor (" congenital pleomorphic adenoma"). A clinicopathologic, immunohistochemical and ultrastructural study of nine cases. Am J Surg

Pathol. 1994; 18(1):25-36.
3. Peters SM, Turk AT. Salivary gland anlage tumor: molecular profiling sheds light on a morphologic question. Oral Surg Oral Med Oral Pathol Oral Radiol. 2019; 127(5):e108-13.
4. Cohen EG, Yoder M, Thomas RM, Salerno D, Isaacson G. Congenital salivary gland anlage tumor of the nasopharynx. Pediatrics. 2003; 112(1):e66-9.
5. Daniller T, Goedhals J, Seedat RY. Salivary gland anlage tumour-a rare cause of congenital nasal obstruction. South Afr J Child Health. 2014; 8(1):31-3.
6. Khurana A, Singaravel S, Mukherjee U. Salivary gland anlage tumor. Indian J Pathol Microbiol. 2016; 59(3):431-3.
7. Mogensen MA, Lin AC, Chang KW, Berry GJ, Barnes PD, Fischbein NJ. Salivary gland anlage tumor in a neonate presenting with respiratory distress: radiographic and pathologic correlation. AJNR Am J Neuroradiol. 2009; 30(5):1022-3.
8. Hellquist H, Paiva-Correia A, Vander Poorten V, Quer M, Hernandez-Prera JC, Andreasen S, et al. Analysis of the clinical relevance of histological classification of benign epithelial salivary gland tumours. Adv Ther. 2019; 36(8):1950-74.
9. Michal M, Sokol L, Mukenšnabl P. Salivary gland anlage tumor. A case with widespread necrosis and large cyst formation. Pathology. 1996; 28(2):128-30.
10. Gauchotte G, Coffinet L, Schmitt E, Bressenot A, Hennequin V, Champigneulle J, et al. Salivary gland anlage tumor: a clinicopathological study of two cases. Fetal Pediatr Pathol. 2011; 30(2): 116-23.
11. Stelow EB, Wenig BM. Update from the 4th edition of the World Health Organization classification of head and neck tumours: nasopharynx. Head Neck Pathol. 2017; 11(1):16-22.
12. Swayampakula AK, Ischander M, Zuppan CW, Krishnan M, Tong K, Qureshi S. Newborn with congenital salivary gland anlage tumor presenting with respiratory distress. Int J Pediatr Otorhinolaryngol Extra. 2015; 10(3):42-4.