Case report

Seromucinous hamartoma of ethmoid sinus in pediatric patient (case report)

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ARTICLE INFO

Article history:
Received 2 April 2021
Received in revised form 19 April 2021
Accepted 20 April 2021
Available online 27 April 2021

Keywords:
Seromucinous hamartoma
Respiratory epithelial lesion
Respiratory epithelial adenomatoid hamartoma

ABSTRACT

Introduction and importance: Seromucinous hamartoma is a rare benign glandular proliferation arising from the respiratory epithelium, which was originally described by Baillie and Batsakis in 1974. Since this time, case reports started to be published on SH, as a middle aged and elderly disease, here we report a case of a pediatric patient who found to have SH.

Presentation of the case: 2-year-old girl, brought by her parent with a complain of a mass at the right medial canthal area for one year.

Clinical discussion: As this pediatric patient presented with long standing history of right medial canthal area, we made out differential diagnosis list, with keeping congenital midline nasal masses such as nasal glioma, dermoid, and encephalocele at the top of our differentials, followed by inflammatory disease and lacrimal system disease. After bedside clinical assessment and imaging, patient underwent endoscopic sinus surgery for surgical excision, histopathology analysis came as Seromucinous hamartoma. Postoperative course was unremarkable, patient is disease-free for 18 months, till her most recent follow up. With no additional treatment or recurrence.

Conclusion: This case report indicates that seromucinous hamartoma should always be considered in the differential diagnosis of pediatric sinonasal disease. According to the literature review we did; this is the first case reported in such an age group.

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1. Introduction

Seromucinous hamartoma (SH) also known as epithelial hamartoma, glandular hamartoma, or microglandular adenosis. It is a rare benign glandular proliferation arising from the respiratory epithelium of the sinonasal tract and nasopharynx, the majority originate from the posterior nasal cavity, it was originally described by Baillie and Batsakis in 1974 as a single case report of polypoid lesion attached to the posterior vomer to the nasopharynx, since its discovered, few numbers of cases have been reported in the literature. Most patients are middle or advanced age, male to female ratio approximately 1:1.5. Patients usually present with long-standing history of nasal obstruction and/or epistaxis [1]. Simple surgical excision found to be curative for such a lesion. Recurrence is rare. Here we report a pediatric patient who found to have SH at her right ethmoid sinus, which is never reported at her age group and not common to appear at the ethmoid sinus.

2. Case report

A 2-year-old girl was referred from a primary health care at her town, brought to our tertiary hospital by her parents who noticed a mass at the right medial canthal area for one year, increasing in size with upper respiratory tract infections. But no changing with crying or feeding. Associated with on/off right eye purulent discharge. She was treated couple of times with oral antibiotics and topical eye drops anti-biotics as bacterial conjunctivitis.

She is full term, product of normal vaginal delivery, with no instrumentments used during delivery. She is not known to have chronic illness, and she had no previous surgeries.

Her family history is unremarkable and no similar presentation in the family, and negative history for relevant genetic disease, she is up to her age. Patient is the first and only child for her parents, living with them, in a good socioeconomic level. She Not known to have any psychosocial illness, not taking any medications regularly, no history of allergy and her vaccines are up to date. Review of all other systems are unremarkable.

On examination: child looks well, all vital parameters are within normal, baby fall in the 50th percentile when plotted in the growth
According to clinical assessment list of diagnosis created, and since congenital midline nasal masses was the first differential diagnosis, we decided to go for imaging before any surgical intervention. CT Fig. 1. MRI of the paranasal sinuses Fig. 2. Revealed an expansile non-complicated right agger Nazi air cell mass. With no intracranial connection.

Depending on the presentation, clinical bedside assessment and imaging, and after considering the patient's age and differential diagnosis, treatment plans was to take the patient for surgical excision by endoscopic approach. Surgery was offered to the family as the curative treatment, and they agreed. so written informed consent, including the plan, risks and benefits, possible outcome and complications, was signed. Patient underwent functional endoscopic sinus surgery under general anesthesia. Surgery performed at our tertiary healthcare center by a Rhinology and skull base surgery consultant, with the assistant of senior trainee ‘5 years of surgical specialty training’.

Under general anesthesia and endotracheal intubation done by anesthesiologist consultant, patient was in supine position with head 30 degree up, prepared in sterile technique, after decongesting the nasal cavity, using 0- and 30-degree telescope, endoscopic sinus surgery was done in the right side, including maxilllectomy, ant total ethmoidectomy. Nasolacrimal duct appeared normal and was preserved. During ethmoidectomy, using microdebrider a mucoid scanty discharge came from the ethmoid cells. Discharge amount was increasing with applying external right eye pressure, discharge was drained and sent for microbial culture and for histopathology study. The specimen was sent for pathology analysis as fresh labeled right ethmoid mass. Pathology results came back to reveal that it was seromucinous hamartoma. After surgery, procedure was explained to the Patient's parents, with the diagnosis and expected outcome and they were happy about it.

Postoperatively, patient stayed for 24 h as an inpatient for observation. She was stable, surgery was tolerated, with no complication developed, she was discharge home the next day on oral antibiotic, saline nasal drops, and analgesia for pain, and home discharge instructions was given to the parents. One-week later patient had her first postoperative visit, doing fine, with absence of any signs or symptoms related to her disease, and good tolerance to the procedure performed. Parents was adherent to the given instructions, and happy about the surgery outcome, as their child's symptoms disappeared and their lifestyle including the patient improved, endoscopic examination was normal, except for some post endoscopic anatomy changes. She continued to follow up every 3 months and was disease-free at the most recent follow-up, 18 months later.

3. Discussion

Seromucinous Hamartomas are considered rare lesions of the sinonasal tract, that should be kept on the differential diagnosis of sinonasal lesion, regardless of patient's age group. Numerous cases of SH have been previously reported, and they typically arise from the posterior nasal septum or the nasopharynx measuring 0.6 to 6 cm, as reported by Weinreb et al. in 2009, who included series of 7 cases of SH. In our reported case, SH raised in the ethmoid sinus which is unusual location as reported by the literature. Moreover, the most common symptom is nasal obstruction and epistaxis in most reported cases, although most patients are asymptomatic, and lesions found incidentally.

According to multitudinous studies, the prevalence of SH is usually middle aged to advanced age patients ranging from 14 to 85 years [3]. However, our findings were the first to be found in a 2-year-old. In regard to differential diagnosis for a pediatric patient who is presenting with a medial canthal area mass, Congenial midline nasal masses should be considered, including Nasal glioma, it is a midline craniofacial malformation consist of heterotopic neuroglial tissue, present at the frontonasal region, encephaloceles as they are the most common congenital midline nasal mass, dermoids, epidermoids, gliomas, teratomas, hemangioma also are differential diagnosis for medial canthal mass. The absence of Furstenberg sign in our patient made masses with intracranial connection to be less likely, all mentioned lesions was ruled out by CT scan that showed no skull base erosion or bone remodeling, and MRI that showed no intracranial connection. Inflammatory disease such as mucocele also considered as differential diagnosis, which often arise in the ethmoid sinuses in pediatrics and can present with a medial canthal mass, it was out after confirming the diagnosis by histopathology. Because the mass was at the medial canthal area, it can
involve the lacrimal system, it was important to perform preoperative CT scan which showed normal lacremial apparatus. malignancy as rhabdomyosarcoma of the orbit if happen will behave more aggressively. Although seromucinous hamartomas are rare, it is important that pathologists be aware of it and keep it in mind as a differential diagnosis of non-neoplastic sinonasal mass. Histologically, the mass is covered by respiratory epithelium, and is comprised of clustered or haphazard proliferations of small to large glands and ducts which are lined by a single layer of ciliated cuboidal or flattened epithelial cells; that is the distinguishable point to make the diagnosis. Nuclei appear monomorphic with no cytologic atypia. They may contain eosinophilic cytoplasmic granules, consistent with zymogen granules, along with clear cell changes and periglandular hyalinization [2] respiratory epithelial adenomatoid hamartoma (REAH), though it is rare, it is the most common hamartoma of the sinonasal tract. It was first described by Wenig and Heffner as a benign overgrowth of submucosal ciliated respiratory lined glands forming a polypoid mass [4]. However, the morphologic features of SH can overlap with other Sinonasal tract epithelial lesions, mainly respiratory epithelial adenomatoid hamartoma (REAH) and low-grade non-intestinal adenocarcinoma (LGNA), its is important distinguishing between the benign hamartoma and Adenocarcinoma that can alter the treatment, but this was not a concern at our case since we are reporting a pediatric case, which make neoplasms least on the differential diagnosis. The treatment of choice for SH is complete surgical excision by transnasal endoscopic approach. With adequate endoscopic tumor excision, recurrence is rare.

4. Conclusion

seromucinous hamartomas are very rare benign tumors of respiratory epithelial origin. Rhinologist and pathologist should keep it in mind and include it in the differential diagnosis of any sinonasal lesion regardless of patient’s age. SH Can be successfully treated via transnasal endoscopic simple excision, and the recurrence rate is low. The work has been reported in line with the SCARE 2020 criteria [5].

Source of funding

All authors declare that this research did not receive any source of funding.

Ethical approval

As an ethical approval, Written informed consent was obtained from the patient’s parents, to publish this case with the accompanying images.