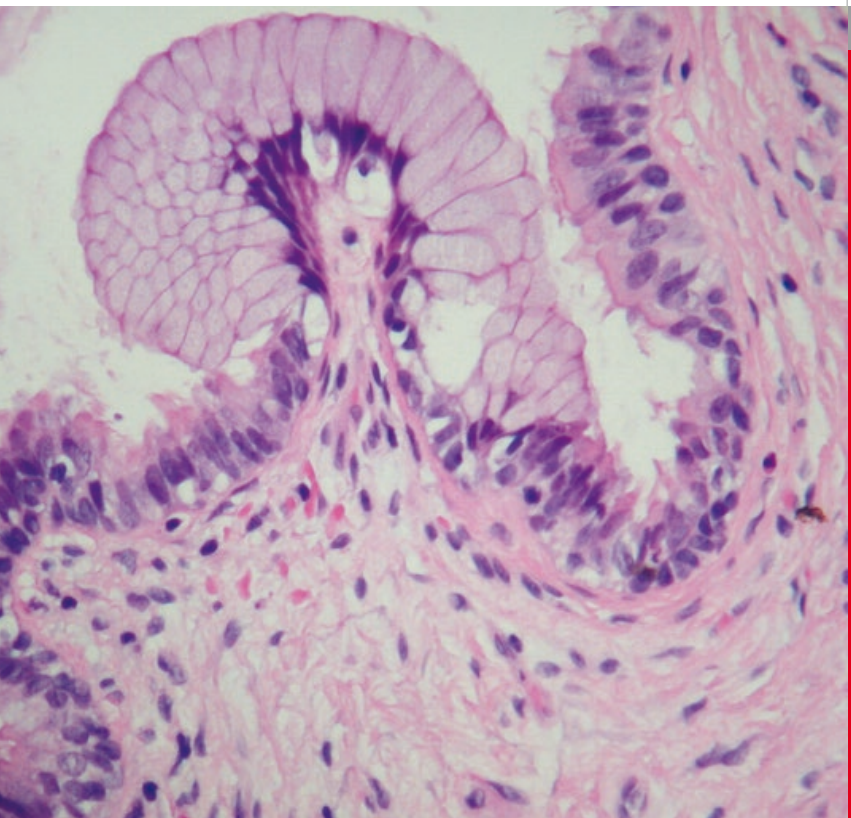
SWISS SOCIETY OF NEONATOLOGY

Congenital sublingual cyst



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INTRODUCTION

Oral and cervical tumors are very rare in the perinatal period, yet they may have severe effects after birth. Although most of these lesions are benign, their localization may have significant consequences. Large masses that hinder respiration at birth are usually announced by the presence of a polyhydramnios on prenatal ultrasound. However, masses may still continue to grow after birth and subsequently lead to severe respiratory obstruction, particularly in association with upper airway infection. Smaller tumors may not produce airway obstruction but can still interfere with feeding. Spontaneous regression of antenatal oro-pharyngeal masses has been reported, similar to post-natal oro-pharyngeal tumors.

CASE REPORT

We present the case of a male infant born by vaginal delivery at 39 weeks gestation. At 20 weeks gestation lingual cysts were discovered at the routine morphological ultrasound. Two cysts were identified (Fig. 1, 2), which increased in size during the last trimester of pregnancy, growing up to 9 mm by 36 weeks of gestation. The mother's medical and obstetrical history was unremarkable. There was no family history of any congenital anomalies.

At birth, adaptation was excellent, Apgar scores were 9,10,10 at 1, 5 and 10 minutes and arterial cord pH was 7.26. On clinical examination the infant had a protruding tongue with two bluish cysts bulging on

the inferior aspect of the tongue (Fig. 3). The boy did not appear to have breathing difficulties and was able to bottle feed. However, feeding on the breast could not be achieved as the child was unable to latch sufficiently around the nipple. He was admitted to the special care nursery for observation.

Ultrasound imaging confirmed two cysts measuring 10 mm in diameter probably communicating and containing mucoid fluid. There was no sign of any vascular malformations. On day three, the infant underwent surgical resection of the cysts (Fig. 4). After excision in toto (Fig. 5), dexamethasone was administered for 48 hours, in order to prevent local swelling. The postoperative period was uneventful, and the child was discharged home five days after surgery, fully breastfed. Four weeks later, the child was healthy, showed a normal sized tongue and had no feeding difficulties. The histological analysis was compatible with a sublingual cyst (Fig. 6, 7) and confirmed communicating cysts of 20 x 10 x 6 mm, covered by a mixed pattern of epithelium. Some sections were covered by a multistratified flat epithelium, whereas others had a pseudo-stratified ciliated epithelium or a cylindrical epithelium. Two islets of accessory salivary glands were found in the cyst wall.

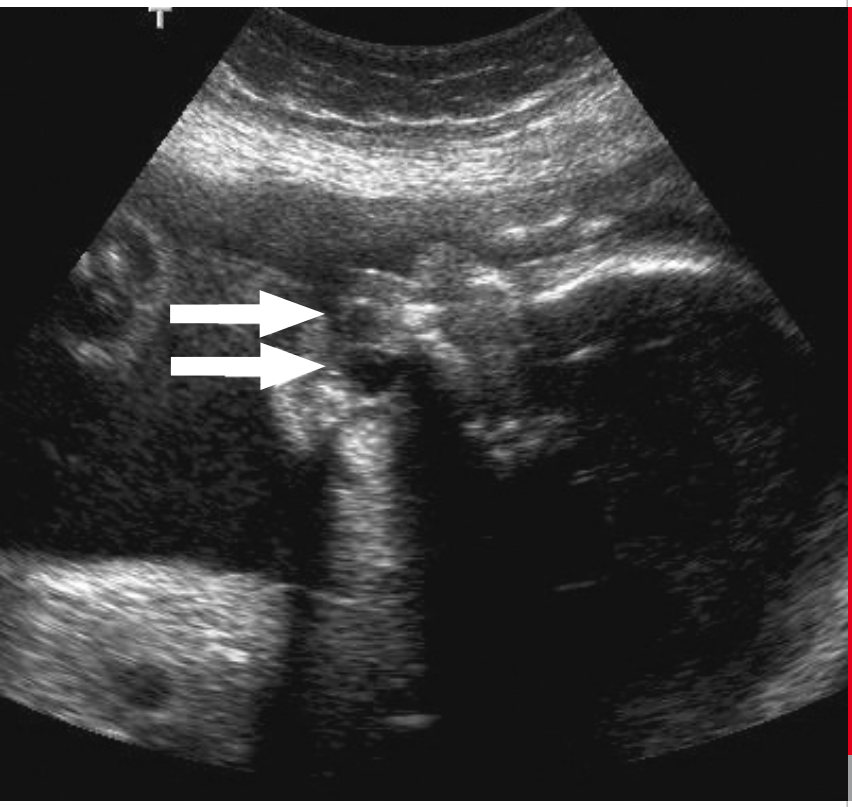

Fig. 1

Antenatal ultrasound scan at 22 weeks of gestation (arrow points to cyst).

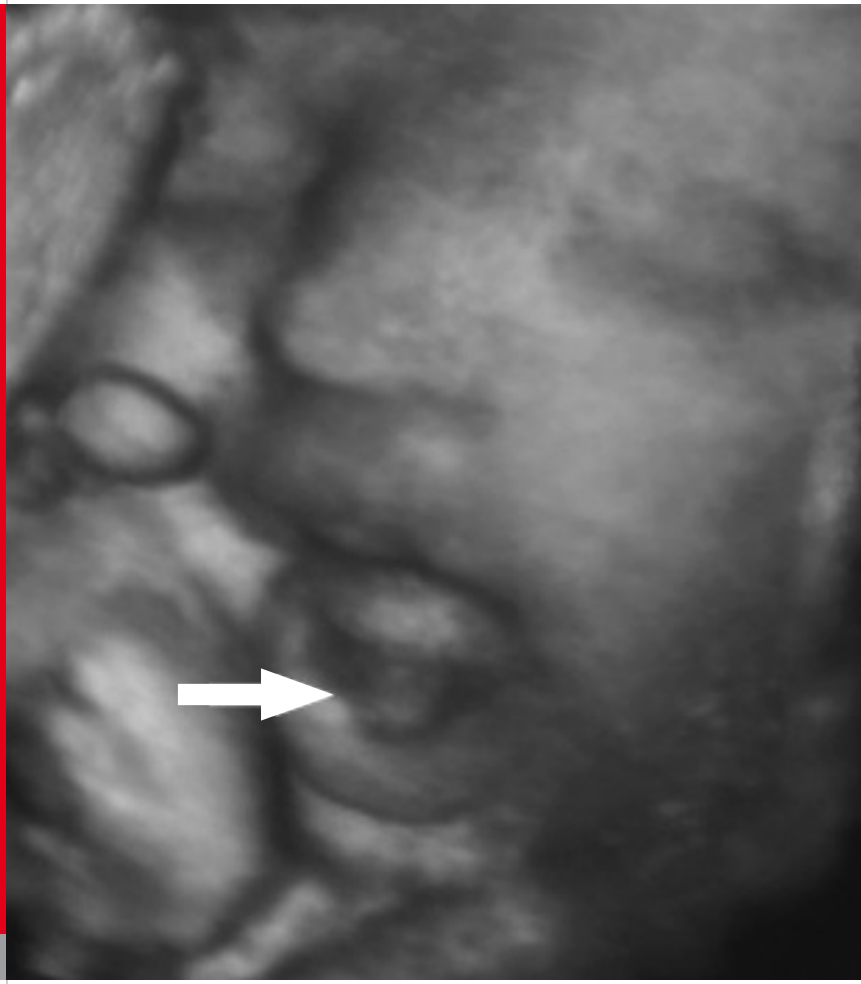

Fig. 2

3D ultrasound scan a few days before birth (arrow points to the protruding cyst).

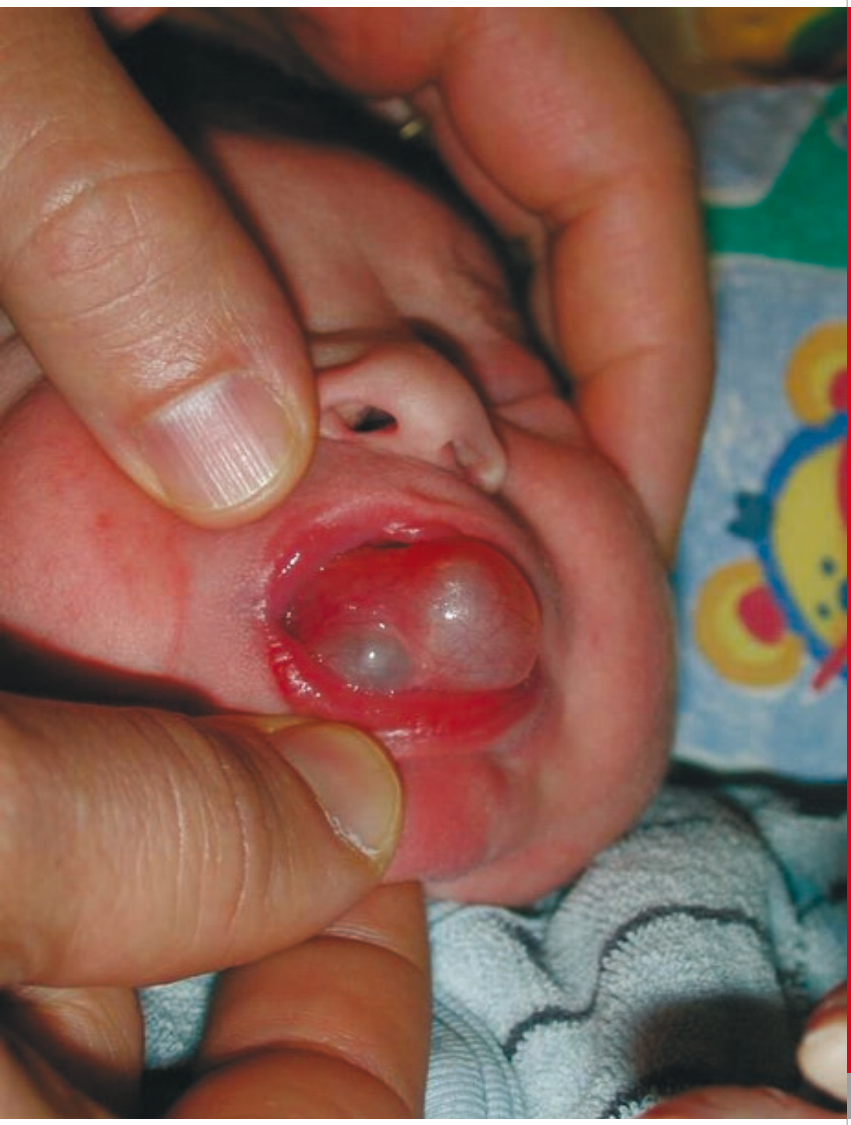

Fig. 3

Initial clinical appearance of the ranula, protruding out of the mouth.

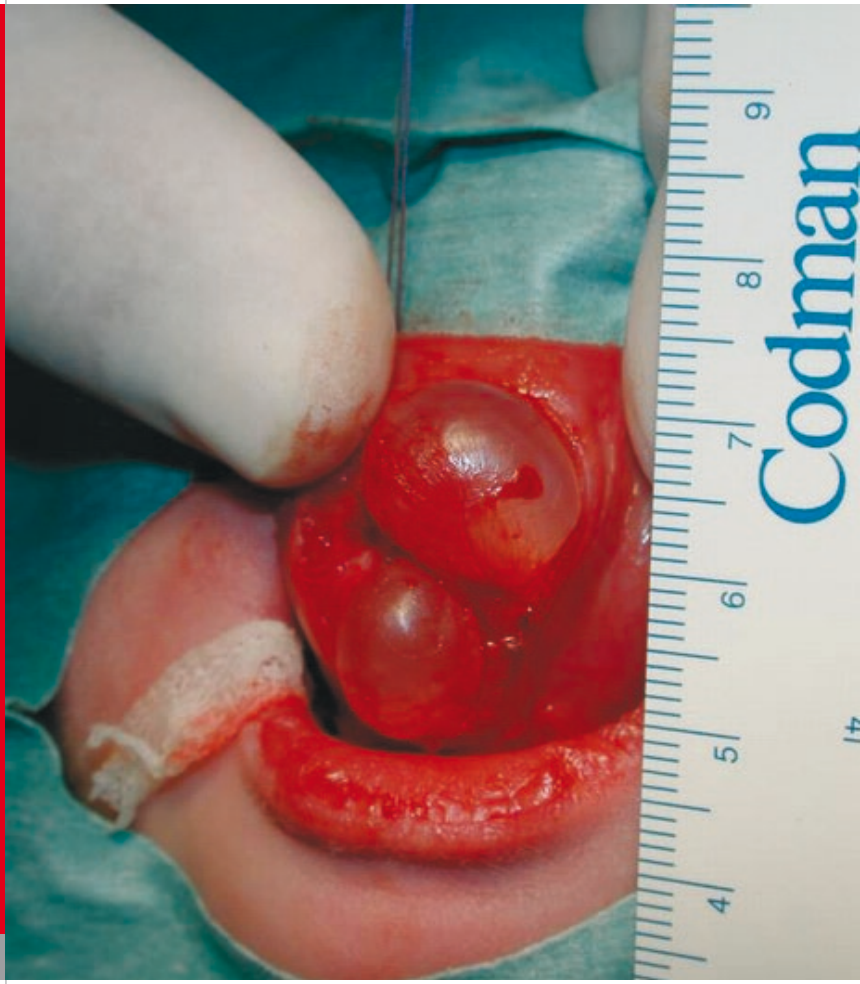

Fig. 4

Appearance of the ranula at operation, on the inferior aspect of the tongue.

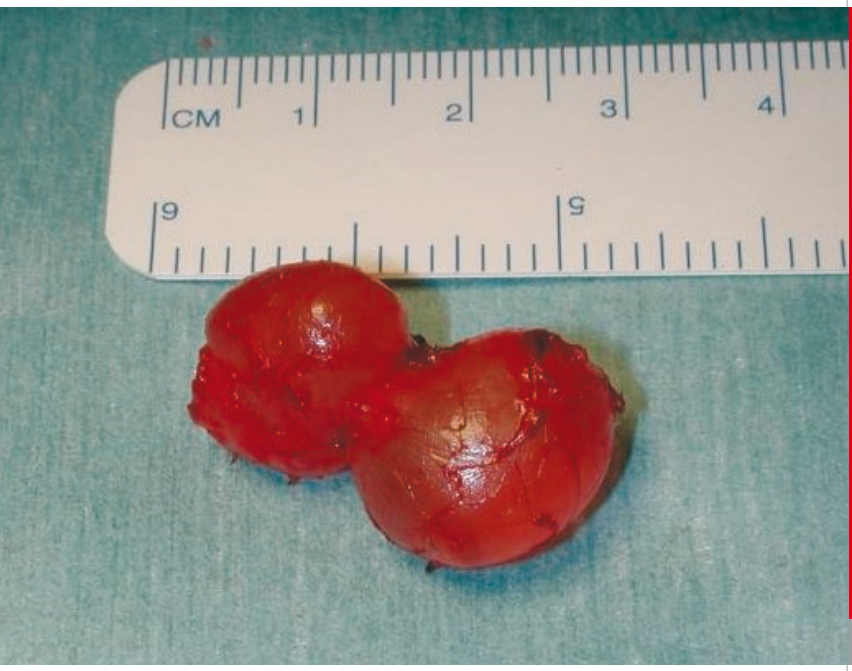

Fig.5

Macroscopic view of the cyst, showing two communicating chambers.

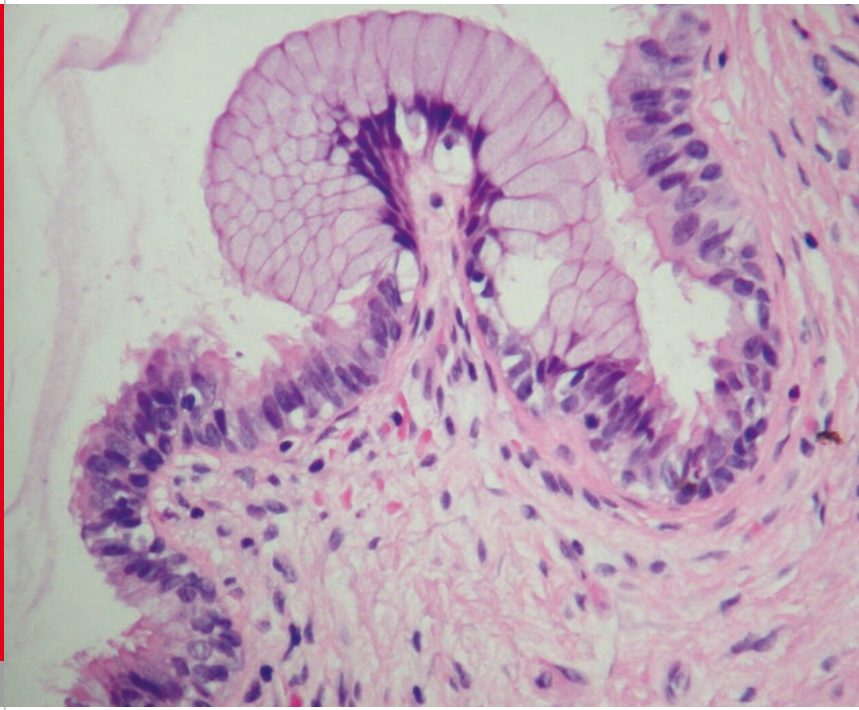

Fig. 6

Microscopic view: transition between a colon-like cylindrical epithelium and a respiratory-like pseudostratified ciliated epithelium.

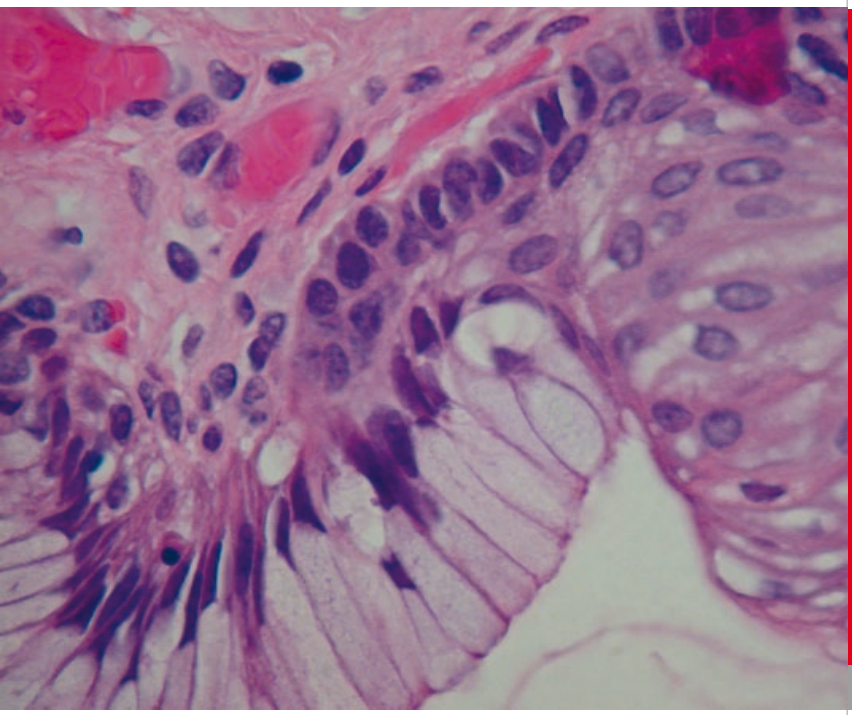

Fig. 7

Microscopic view: high power view of the transition zone described in Fig. 6.

DISCUSSION

Congenital sublingual cysts are a rare anomaly seen in newborns. Their prevalence is still unknown and only small series have been published so far. A congenital sublingual cyst, or ranula, is a dilatation of a sublingual gland, localized anteriorly on the floor of the mouth, between the mylohyoid and hyoglossus muscles (1, 2). Clinically, a fluid-filled cyst elevates the tongue. The cyst may plunge under the mylohyoid muscle with cervical extension. Congenital sublingual cysts usually go unnoticed at birth, but may grow enough to represent a real threat to the airway, causing stridor and dyspnea (3). When small at birth, they are often diagnosed in late childhood, at occasions when they swell (2, 5). Ranulas may also be acquired and occur following tongue trauma.The differential diagnosis (1) of ranulas (Table) includes lymphatic malformations (solid-cystic masses with indistinct margins on ultrasound), foetal cervical tumors such as teratomas (solid-cystic tumors with mixed areas of hypo- and hyper-echogenicity, often associated with polyhydramnios), thymomas and thyroid tumors, thyroglossal duct cyst (quite unusual in the floor of the oral cavity), heterotopic gastric cysts and enterocystomas. An antenatal ultrasound is helpful in differentiating between solid and liquid tumors. After birth, a radiologic workup may help establishing the diagnosis. For the precise location and determination of cyst content, computed tomography and particularly magnetic resonance imaging may be of help. In our case, the postnatal ultrasound deter-

mined the exact anatomy of the cyst and confirmed that it was polycystic.

In our case, the cyst was lined by an epithelium. Usually, congenital sublingual cysts are epithelium-free. They develop from a mucus leak following disruption of the sublingual gland elements and are considered pseudo-cysts. However, 7.5% of cysts are mucosal retention cysts with epithelium (7). The glands involved may then frequently display metaplasia of the epithelium lining. As the histological pattern of our patient's cyst demonstrated three different types of epithelium, the histopathology diagnosis was a sublingual mucosal retention cyst.

Different techniques are described to treat sublingual cysts. As mentioned, spontaneous resolution has been observed (5), but usually, a surgical approach is recommended (4-6). It can involve marsupialization or unroofing of the cyst, simple excision of the cyst or excision combined with excision of the ipsilateral sublingual gland. The recurrence rate in epithelium-free cyst is lowest after excision including the ipsilateral sublingual gland.

Table

Differential diagnosis of lingual/sublingual cystic lesions in the perinatal period
- mucosal retention cyst
- lymphangioma
- vascular malformation
- hemangioma
- ranula (salivary cyst)
- ectopic thyroglossal duct cyst
- foregut duplication cyst

CONCLUSION

Congenital sublingual cysts are rare congenital tumors of the oro-pharynx that may exceptionally be diagnosed in utero. The diagnosis should be suspected when the tongue appears to protrude or appears elevated by a fluid-filled cyst in the lingual mucosa. The lesion can enlarge enough to compromise the airways and lead to life-threatening events or may go unnoticed until late childhood. Surgery is only needed or recommended when the cyst hinders feeding or breathing, otherwise, potential spontaneous resolution can be awaited. After an observation of a few months however, complete excision of the cyst, usually with the ipsilateral gland is the treatment of choice with the least recurrences.

ACKNOWLEDGEMENTS

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