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REVIEW ARTICLE

The preauricular sinus: A review of its aetiology, clinical presentation and management

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Summary The preauricular sinus is a not uncommon finding in the paediatric population. Recent reports have added to our knowledge of this benign malformation. We review the current literature with respect to the aetiology of the condition, its clinical features, and associations with other congenital malformations. In those patients in whom a preauricular sinus is identified, we recommend associated congenital anomalies be sought. In selected cases, a renal ultrasound scan may be appropriate. Where no associated abnormalities are identified, and where the preauricular sinus is asymptomatic, there is consensus opinion that no further action is indicated. In the acute phase of infection, treatment comprises administration of appropriate antibiotics, and incision and drainage of an abscess if present. In the symptomatic preauricular sinus exhibiting recurrent or persistent infection, opinion regarding optimal management varies. Latest evidence suggests definitive surgical treatment offering the most favourable outcome is by wide local excision of the sinus, as opposed to the previously preferred technique of simple sinectomy. Magnification employed during surgery, and opening, and following from the inside as well as outside, branching tracts of the sinus may further minimise the risk of recurrence.

Contents

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1. Introduction

First described by Van Heusinger in 1864 [1], the preauricular sinus is a benign congenital malformation of the preauricular soft tissues. It is variably also termed a preauricular pit, preauricular fistula, preauricular tract and preauricular cyst. It has an estimated incidence of 0.1—0.9% in the United States, 0.47% in Hungary [3], 0.9% in England [4], 2.5% in Taiwan [5] and 4—10% in some areas of Africa [6]. In most instances, it is noted during routine ear, nose and throat examination, though can present

Table 1 Syndromes associated with preauricular sinus

Syndrome	Clinical features	Comments
Branchio-Oto-Renal (BOR) Syndrome [17,18]	Structural defects of the outer, middle, or inner ear and associated conductive, sensorineural or mixed hearing loss, preauricular sinus, renal anomalies, lateral cervical fistulas, cysts, or sinuses; and/or nasolacrimal duct stenosis or fistulas	
Branchio-Oto-Ureteral Syndrome [19]	Sensorineural hearing loss, preauricular sinus and duplication of ureters or bifid renal pelvices	Two families reported
Branchio-Otic Syndrome [20]	Branchial anomalies, preauricular sinus, and hearing loss with no renal dysplasia	A variant of BOR syndrome
Branchio-Oto-Costal Syndrome [21]	Conductive deafness, bilateral commissural lip and preauricular sinuses, unilateral branchial fistula and rib anomalies	Three cases in a single family reported
Tetralogy of Fallot and clinodactyly [22]	Tetralogy of Fallot, characteristic appearance, preauricular sinus and fifth finger clinodactyly	Six cases reported
Steatocytoma multiplex [23,24]	Facial steatocytoma multiplex associated with pilar cyst and bilateral preauricular sinuses	Case series affecting four generations
Rare syndrome of bilateral defects [25]	Bilateral cervical branchial sinuses, bilateral preauricular sinuses, bilateral malformed auricles and bilateral hearing impairment	A family of male to male transmission through three generations
The deafness, preauricular sinus, external ear anomaly and commissural lip pit syndrome [26]	Commissural lip pits, pinna dysplasia, preauricular sinus and mixed or conductive hearing loss	Large family described
Cat Eye Syndrome [27]	Coloboma of the iris, preauricular sinus, imperforate anus and down slanting palpebral fissures	Two cases reported
Waardenburg's Syndrome [28]	Typical features of Waardenburg's syndrome with exception of the white forelock (poliosis), but with additional anomalies including syndactyly, absence of the fourth left toe, bilateral preauricular sinus and dacrocystitis	Case report
Floating-Harbour Syndrome of unusual phenotype [29]	Floating-Harbour syndrome, trigonocephaly due to metopic suture synostosis, preauricular sinus, hypoplastic thumb, subluxated radial head and Sprengel deformity	Case report
Trisomy 22 mosaicism (46,XX/47,XX,+22) restricted to skin fibroblast [30]	Growth failure, microcephaly, hypertelorism, epicanthal fold, preauricular sinus, congenital heart defect, hypotonia and delayed development	Case report
Full Trisomy 22 [31]	Primitive and low set ears, bilateral preauricular sinuses, broad nasal ridge, antimongoloid palpebral fissures, macroglossia, enlarged sublingual glands, cleft palate, micrognathia, clinodactyly of the fifth finger, hypoplastic finger nails, hypoplastic genitalia, short lower limbs, bilateral sandal gap and deep plantar furrows	Case report

as an infected and discharging sinus. This article provides an up to date review of the condition, outlining recent developments in the understanding of its aetiology, and describing other congenital anomalies with which it has been associated. Management of the symptomatic preauricular sinus varies greatly. Recommendations for optimal treatment are made, based on critical review of the limited literature available.

2. Embryology and development

The formation of a preauricular sinus occurs during embryogenesis and is closely associated with the development of the auricle during the sixth week of gestation [7]. The auricle develops from six mesenchymal proliferations, known as the hillocks of His; three from the caudal border of the first branchial arch and three from the cephalic border of the second branchial arch [8]. These hillocks fuse to form the definitive auricle [9]. The most frequently cited and generally accepted theory attributes the development of a preauricular sinus to incomplete or defective fusion of the six auditory hillocks [1,10]. The other, less well-known and published theory, is that the sinus develops as a result of isolated ectodermal folding during auricular development [11,12]. The preauricular sinus is often confused with a branchial fistula. Whereas branchial cleft anomalies can be intimately related to and involve the external auditory meatus, tympanic membrane or angle of the mandible) [1,8,13], a pre-auricular sinus is not. Similarly, a preauricular sinus does not typically involve branches of the facial nerve, though treatment can place the facial nerve at risk [14].

3. Genetics and associated syndromes

A preauricular sinus occurs either sporadically or is inherited. Over 50% of cases overall are unilateral [8], and most often sporadic. It occurs more commonly on the right side [2]. Bilateral cases are more likely to be inherited [6]. When inherited, the pattern is of incomplete autosomal dominance with reduced (around 85%) penetrance [8,15]. Research recently undertaken in China has mapped a possible locus for congenital preauricular fistula to chromosome 8q11.1-q13.3. The work used linkage analysis of a family comprising affected and non-affected members [16].

The preauricular sinus has been described as part of a number of syndromes (Table 1).

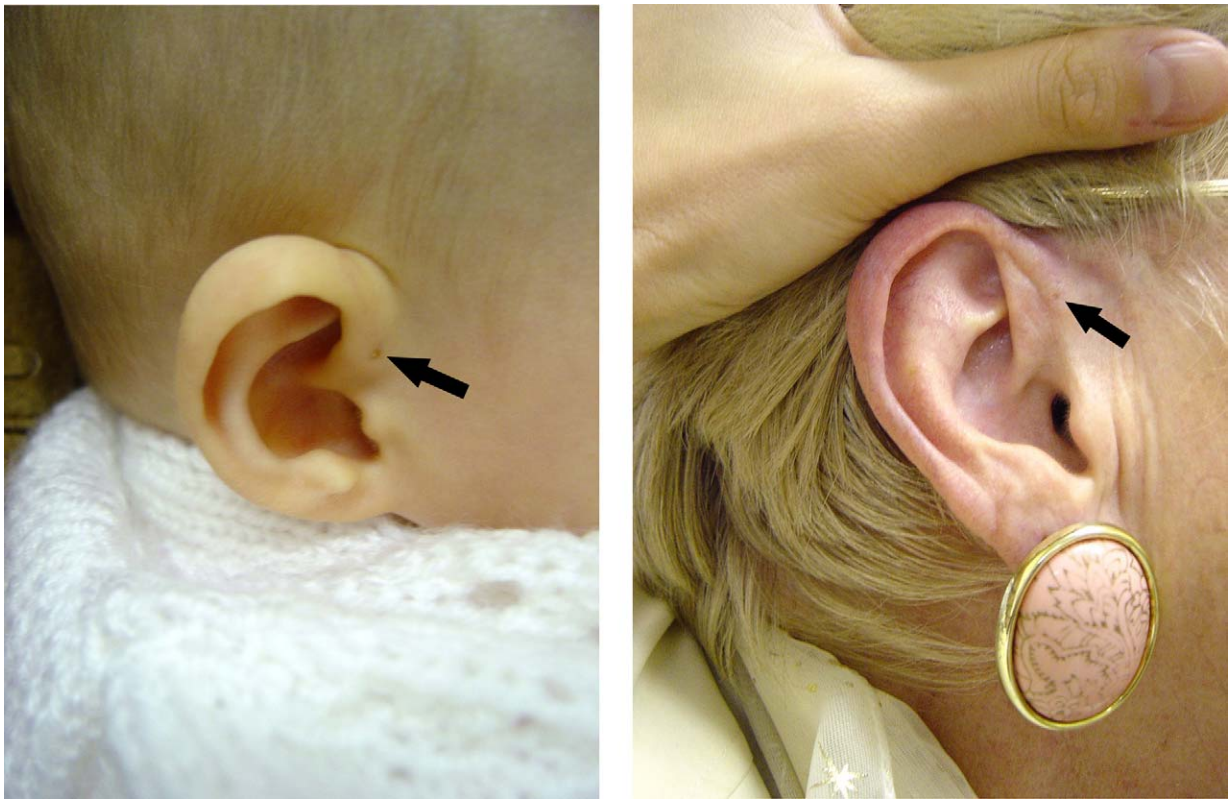

Fig. 1 (a) Infant with preauricular sinus located on the anterior margin of the ascending limb of the helix. (b) Infant's grandmother with preauricular sinus.

4. Clinical presentation

A small pit is often noted adjacent to the external ear, usually located at the anterior margin of the ascending limb of the helix [17] (Fig. 1a and b). The opening of the preauricular sinus has also been reported along the posterosuperior margin of the helix, the tragus or the lobule [1]. The visible pit may represent the full extent of the deformity, or mark a sinus tract that can vary in length, branch and follow a tortuous course. Preauricular sinus may lead to the formation of a subcutaneous cyst that is intimately related to the tragal cartilage and the anterior crus of the helix. In all cases, part of the tract blends with the perichondrium of the auricular cartilage [32]. The sinus tract is lateral and superior to the facial nerve and parotid gland, in contrast to the tract of an anomaly of the first branchial cleft, which tends to be intimately related to these structures.

Not infrequently, patients present with discharge from the sinus either as a result of desquamating epithelial debris or infection. Erythema, swelling, pain and discharge are familiar signs and symptoms of infection. The most common pathogens causing infection are Staphylococcal species and, less commonly, Proteus, Streptococcus and Peptococcus species [6].

5. Management

The majority of patients with preauricular sinus are asymptomatic [8,33]. A sinus pit in a typical site is highly suggestive of the diagnosis. A thorough history and head and neck examination is mandatory in all cases, seeking evidence of associated anomalies. Where it is an isolated, asymptomatic finding no treatment is required.

There is evidence, arising from a prospective study of over 32,000 live births, to suggest there is a slightly increased risk (odds ratio 1.3) of renal anomalies in children with ear anomalies [34]. After patients with syndromic diagnoses are excluded from the analysis, there continues to be a strong association between preauricular pits and renal defects. Leung and Robson [17] in Calgary, Canada, carried out a prospective study to investigate the incidence of renal anomalies associated specifically with preauricular sinuses. They found that, on renal ultrasonography, 3 of 69 children with a preauricular sinus also had a renal anomaly (however one of these children had BOR syndrome). They concluded such anomalies were significantly more common in patients with a preauricular sinus than the 1% incidence of renal anomalies reported in the general population. On the basis of their results, and that the clinical implications of detecting a renal anomaly are important, they suggested that renal ultrasonography should be performed on all patients with a preauricular sinus. Their view is not shared widely, though. Evidence to support the cost-effectiveness of large population screening for this condition, as they recommend, is lacking.

Wang et al. in California, U.S.A, refined Leung's indications for renal ultrasound [34]. They performed a retrospective review of 42 children with external ear anomalies who had undergone renal ultrasound in two genetics medical centres between 1981 and 2000. They suggested that renal ultrasound should only be performed on patients with a preauricular sinus *and* one or more of the following:

1. Another malformation or dysmorphic feature.
2. A family history of deafness.
3. An auricular and/or renal malformation.
4. A maternal history of gestational diabetes.

Unfortunately, their sample was biased by including only clinic patients whose ears (or other organ systems) were anomalous enough to have been referred for a clinical genetics evaluation. Also, patients who had not undergone ultrasonography were excluded, which will have affected the calculated incidence of renal malformations. Despite these limitations, these guidelines, proposed for considering renal ultrasound, appear both reasonable and appropriate for application in clinical practice.

Audiometry is another investigation proposed as an investigation proper for patients with an isolated preauricular pit/tag [15,34]. A prospective study was undertaken in Israel in 1997 [15]. Its authors reported that the incidence of conductive and/or sensorineural hearing impairment in 4-month-old infants with an isolated preauricular pit/tag, identified on brainstem evoked response audiometry, was significantly higher (4 out of 23 children, or 17%) than the incidence reported in preschool children (0.4%). As in much of the literature regarding the preauricular sinus, the numbers studied were, again, small. Moreover, however, a comparison between 4-month-old infants and pre-school children is inappropriate (there is a well recognised decline in the prevalence of otitis media with effusion, which often causes a conductive hearing loss, with increasing age). There has been no evidence presented to support their conclusion that hearing assessment should be carried out in the routine evaluation of the newborn with isolated preauricular pits, or at least no more so in these children than in those without preauricular pits.

In the acute phase of infection of a preauricular sinus, intervention involves administration of appro-

priate antibiotics active against the causative pathogen, and, where an abscess is present, it is common practice to undertake incision and drainage. Coatesworth et al. have described a technique for drainage of a preauricular abscess using a lacrimal probe [35]. They claim this negates the need for an incision, causing little or no disturbance to the underlying sinus and making any subsequent surgery more straightforward. Their described technique involves anaesthetising the overlying skin with a topical anaesthetic and inserting a blunt ended lacrimal probe into the sinus, to allow drainage of the abscess. The procedure can be repeated if necessary. They report to have used this technique successfully in six out of the seven patients, the seventh patient having required incision and drainage. It is, possibly, an alternative management option to be considered, though the authors fail to mention that lacrimal probe trauma to the tract may be disadvantageous in causing deeper scarring and subsequent difficulty in excision [13].

Recurrent or persistent preauricular sinus infection requires surgical excision of the sinus and its tract during a period of quiescence [36,37]. Various surgical techniques, aimed at ensuring complete dissection, have been described in the literature. Incomplete excision is believed to be the cause of recurrence of a preauricular sinus; recurrence rates have been reported between nil [13] and 42% [38]. The standard technique is to excise an ellipse of skin surrounding the preauricular sinus opening and to dissect out the individual tract: the simple sinectomy. A comparison of this technique with a more radical supra-auricular approach (wide local excision) was made by Prasad et al. in 1990 [38] and by Lam et al. in 2001 [39]. Based on these reports, the supra-auricular approach has a lower recurrence rate 5% (of 21) versus 42% (of 12) [38] and 3.7% (of 27) versus 32% (of 25) [39]. The supra-auricular approach involves a postauricular extension of the elliptical incision around the orifice of the sinus [38]. Dissection is carried out to identify the temporalis fascia which is the medial limit of the dissection, and continues over the cartilage of the anterior helix, which is regarded as the posterior margin of dissection. Tissue superficial to the temporalis fascia is removed together with the preauricular sinus. A portion of the cartilage or perichondrium of the helix at the base of the sinus should be excised to ensure complete removal of the epithelial lining [36]. Dead space should be closed by means of a layered closure, with or without a drain or compression bandage [13].

More recently, Baatenburg de Jong has described a modification of the wide local excision technique [40], which appears to be of at least equal, if not superior effectiveness in minimising the risk of recurrence. He describes the new procedure as the ''inside-out'' technique, first introduced by Jesma in Rotterdam, but at that time unpublished. The method involves mandatory use of magnifying glasses or a microscope. An elliptical incision is made in the skin around the sinus pit and the sinus is opened. The sinus is viewed and followed from both outside (as in the classic procedures) and inside. Each subsequent branching tract is opened and followed until every dead end is identified and excised. Baatenburg de Jong describes a recurrence rate of 0% in 23 patients in whom the inside-out technique was employed. This is not at present a widely employed method but certainly deserves interest, and further trial.

Currie et al. carried out a retrospective review over a period of 8 years in Hong Kong looking at a range of factors that influenced the outcome following surgical excision of a preauricular sinus [13]. A total of 159 operations were performed in 117 patients. They found that previous excision, the use of a probe to delineate the sinus, post-operative wound sepsis and operating under local anaesthetic all increased the chance of recurrence (though surgeon and patient factors were not taken into account, and no statistical analysis was made). They observed a number of factors that appeared to reduce the chance of recurrence. These included meticulous dissection of the sinus by an experienced head and neck surgeon under general anaesthetic, the use of the supra-auricular approach with clearance down to the temporalis fascia, avoidance of sinus rupture and closure of wound dead space.

To aid complete resection of the sinus and its tracts, by whichever chosen surgical method, several adjunctive techniques have been suggested. Pre-operative sonographic imaging, pre-operative sinograms, intra-operative methylene blue injection and the use of a lacrimal probe have all been described [14,33], but have been found to be of variable benefit. Individual preference currently dictates which of these methods is used, since it is still not known which adjuncts are most useful. Other authors have recommended the destruction of the sinus tract with either sclerosant solution or electrodiathermy as an alternative to dissection, though again outcome is variable, with no clear advantage proven [33].

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