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Ectopic Preauricular Sinus in a Facial Cleft and Microtia Patient

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Preauricular sinus is a congenital malformation that is very commonly encountered among the general population and it has especially high prevalence among Asians when compared to other ethnic groups. It can often go unnoticed or easily overlooked by the patient or even by doctors because most of them are asymptomatic and is most of the time only a tiny pit that can be trivial in terms of aesthetics. We report a very rare and unique case that has no precedence what so ever; hence no reported case in the literature: an ectopic preauricular sinus in a facial cleft and microsomia patient.

Keywords: Craniofacial abnormalities / Congenital microtia / Branchial region

Case Report

INTRODUCTION

Preauricular sinus was first introduced in the literature in 1864 by Heusinger [1-6]. Similar terminologies that was used interchangeably in the literature are: preauricular pits, fistulas, tracts, and cysts [3,5]. Although asymptomatic disease can be left untreated, once infected however, surgical intervention is the mainstay treatment of choice [4,7]. Its incidence and prevalence are far higher among Africans followed by Asians and least common among Caucasians (African up to 10%, Koreans 5%, Taiwan 1.6%, and less than 1% in the United States and England) [1,2,4-6,8-11]. A retrospective study provides an imaginary tragal vertically extended line that demarcates and categorizes sinus into classical type (preauricular sinus) and variant type (postauricular type) [1,2]. The variant type is further classified into type 1 (middle of crus), type 2 (superior to crus) and type 3 (at the cymba concha) according to the location of the opening pit [1,2]. Although the disease occurs during the process of embryology the exact etiopathogenesis is yet to be elucidated. Currently three theories have been raised: incomplete fusion of the first branchial arch hillocks, isolation of ectodermal folds during auricle formation, and defective closure of the most dorsal part of the first branchial cleft [1]. Although the etiopathogenesis is not clear, we would like to report preauricular sinus that is difficult to explain embryologically.

CASE REPORT

A 61-year-old female patient was admitted to the Department of Plastic and Reconstructive Surgery, National Medical Center, Seoul, Korea complaining of repeated malodorous discharge from a small opening at her left cheek. She had been previously diagnosed of lateral facial cleft, right (Tessier classification #7) and microtia, left in a General Hospital (Junju, Korea) when she was 6–7 years old. During a course of three consecutive surgeries to repair the lateral facial cleft, no surgical intervention however was undertaken to reconstruct her left microtia. Past medical history revealed recurrent bouts of yellow sebum or paste-like discharge that had extremely foul odor throughout her entire life. Her mother had been taking opium-like substance (as part of a folk medicine) when she was pregnant in order to induce abortion.

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Subject was delivered through normal spontaneous vaginal delivery at 40th gestational week. She denied of any hypertension, diabetes mellitus, pulmonary tuberculosis, nor hepatitis. The patient suffered from hyperhidrosis and was diagnosed of hypothyroidism after undergoing blood test in the year 2003. Since then she had been followed-up for monitoring and was medicated in the author's hospital department of endocrinology. After suffering from postpartum stress urinary incontinence, she had undergone trans-obturator tape procedure in the author's hospital department of urology. She neither drinks alcohol nor smokes cigarette. No family history of genetic or congenital disease was noted. On physical examination, there was a 1.5 mm sized orifice. The distance between orifice and left lateral canthus was 34 mm. On the other hand, the distance between root of helix, right ear and right lateral canthus was 75 mm (Fig. 1).

Facial three-dimensional computed tomography scans are done. We can note the severe asymmetry of the facial skeleton including the periorbital area, the midface, and the mandible. Orbital dystopia with the left periorbital area posteriorly and inferiorly located, severe hypoplasia is noted of the left hemiface. Compared to the right side, there is incomplete formation of the left side zygomatic bone, zygomatic arch, maxillary bone, and condyle of the mandibular bone (Fig. 2).

Under local anesthesia, the sinus tract was totally excised. Us-

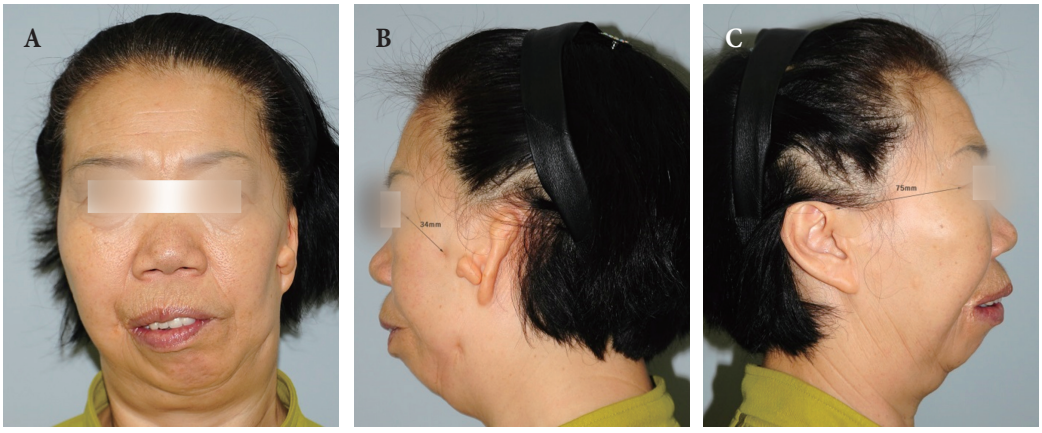

Fig. 1. (A–C) Preoperative photographs. Note the distance between the left lateral canthus and the orifice of the preauricular sinus: comparing distance between the right lateral canthus and the root of helix of the right ear.

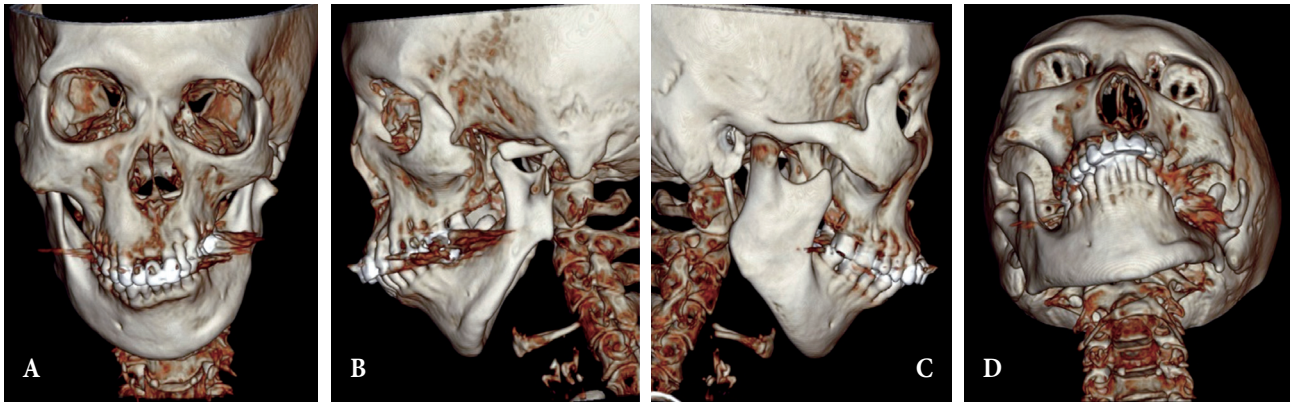

Fig. 2. (A–D) Facial three-dimensional computed tomography scan image. Note the severe asymmetry of the facial skeleton and hypoplasia of the left hemiface.

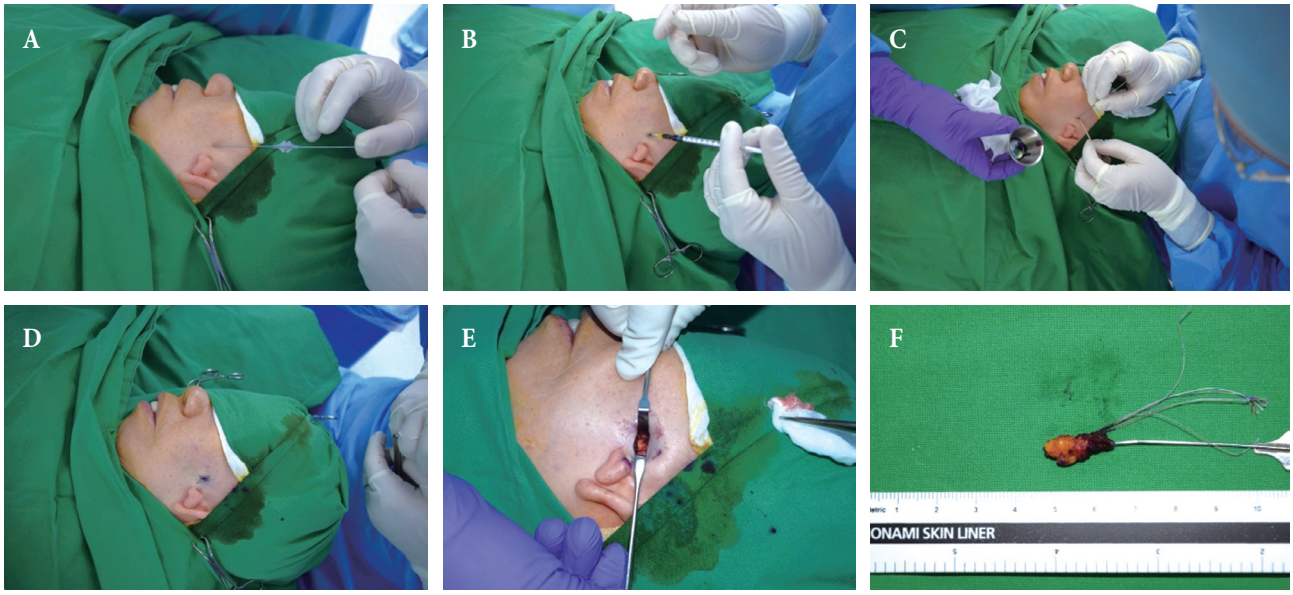

Fig. 3. (A–F) Under local anesthesia, the sinus tract was totally excised avoiding sinus tract rupture with intraluminal staining technique.

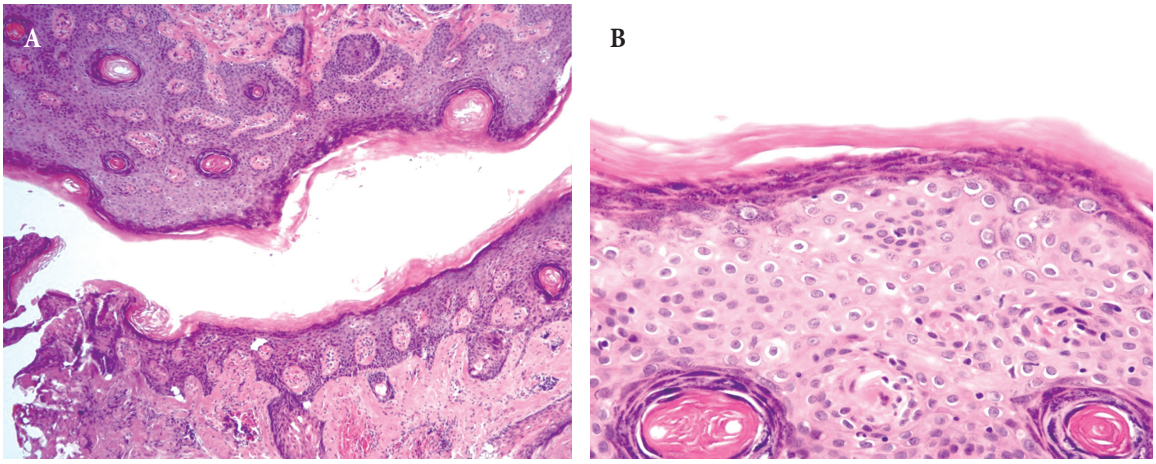

Fig. 4. Pathology findings. (A) H&E, ×100. (B) H&E, ×400.

ing the intraluminal staining with gentian violet solution, we were able to confirm that the sinus tract was not ruptured (Fig. 3).

Pathologic findings were as follows: on gross examination, acquired specimen soft tissue measured 1.5 cm×0.7 cm (Fig. 3F), while tissue microscopy demonstrated pseudoepitheliomatous hyperplasia with interstitial lymphocytic infiltration (Fig. 4).

Judicious administration of antibiotics was done after surgery. The patient was followed-up for more than one year without any complications or recurrences (Fig. 5).

DISCUSSION

The preauricular sinus can be either sporadic or hereditary. Most of them are unilateral (over 50%) affecting the right side more often. The less commonly encountered bilateral cases can be attributed to incomplete autosomal dominant mode of inheritance

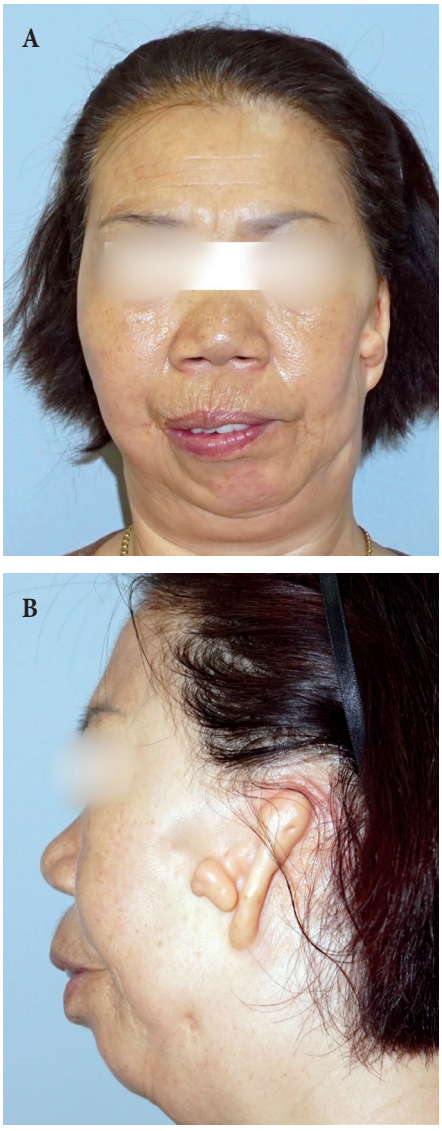

Fig. 5. (A, B) Photograph at more than one year after surgery. No signs of recurrence or complication was noted.

(with penetrance of 85%). Chromosome locus 8q11.1–q13.3 has been reported as possible culprit [5,8,10,11]. Bearing in mind that the external ear originates from fusion of 6 hillocks of His: 3 caudal borders of the first brachial arch (gives rise to tragus and the anterior crus of helix) and 3 caudal border of the second branchial arch (gives rise to rest of the auricle) [1,3,5,6,8-11], it is no surprise the disease manifests in various types of syndromes and these include Branchio-Oto-Renal Syndrome, Branchio-Oto-Ureteral Syndrome, Branchi-Otic Syndrome, Branchio-Oto Costal Syndrome, Tetralogy of Fallot, and clinodactyly, Steatocytoma multiplex, rare syndrome of bilateral defects, deafness, preauricular sinus, external ear anomaly and comissural lip pit syndrome, Cat Eye syndrome, Wartenburg's Syndrome, Floating-Harbour Syndrome of unusual phenotype, Trisomy 22 mosaicisim, and Full Trisomy 22 [5,8]. Concomitant congenital anomalies related to hearing loss accounts for 1.7% of preauricular sinus, while renal problems are associated with 2.6% of the disease.

The rare case report we have provided may shed some lights to the relationships between embryology of three disparate but possibly connected very rare disease entities: facial cleft, microtia, and preauricular sinus. There have been reported syndromes associated with preauricular cysts but two other diagnoses our case report patient additionally had were never recorded. Although the probability of three rare diseases occurring at one individual may simply seem multiplying the three probabilities, clinical manifestations of congenital malformations can be construed as a result of intricate yet profound interplays of actions in the molecular levels of genetics which in turn leads to failure of embryology that manifests clinically as what we identify as congenital abnormalities. Unfortunately, only a handful of diseases conform strictly to the typical Mendelian genetics. Often times multifactorial factors play role that interfere with many pathways which govern metabolic pathways during embryology and these cascade of events along with environmental factors (maternal drug abuse during pregnancy as in our patient's case) may eventually lead to a triggering domino-like effect that brought about three identifiable congenital abnormalities. The exact etiology of diseases labeled as syndrome or sequence are not fully understood whether genetically, embryologically or associations with other diseases. As more and more data accumulate about a syndrome or a sequence, the rest of the missing puzzle can be found and currently these diseases may well be characterized as entities on the same spectrum that spans a broad range of clinical manifestations. The ectopic nature may be closely related to microtia and facial cleft the patient has. Therefore when preauricular sinus with syndromic facial abnomalities is encountered, a thorough investigation of diagnostic workup including physical examination of the whole face and the entire body should be instituted. In addition, it is worthwhile mentioning the impor-

tance of patient warning during consultation that albeit its tiny opening the extent of sinus tract excision may be far greater than what it appears.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

PATIENT CONSENT

The patient provided written informed consent for the publication and the use of their images.

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