Intradiploic Ciliated Epithelial Inclusion Cyst of the Skull
—Case Report—

Hidetoshi NAKAMOTO,1 Takemasa KAWAMOTO,1 Sakiko SUZUKI,1
Kenzo HIROSHIMA,2 Masayuki NAKANO,2 and Takakazu KAWAMATA1

Departments of 1Neurosurgery and 2Pathology,
Tokyo Women’s Medical University Yachiyo Medical Center, Yachiyo, Chiba

Abstract
A 56-year-old woman presented with a cystic skull lesion in the right temporal bone detected after resection of breast cancer. She underwent resection of the skull tumor for pathological diagnosis and treatment. The tumor was covered with ciliated epithelium and there were no malignant findings. The pathological diagnosis was ciliated epithelial inclusion cyst. Intradiploic inclusion cysts of the skull presenting as a calvarial defect include epidermoid cysts and dermoid cysts, which are clinically difficult to differentiate. Ciliated epithelium lining an intradiploic inclusion cyst is very rare. Surgical resection is essential for a definitive diagnosis and differentiation from a neoplasm.

Key words: cilia, dermoid cyst, epidermoid cyst, inclusion cyst, metastasis

Introduction
Intradiploic epithelial inclusion cysts, which appear as lytic defects of the skull, account for a very small percentage of primary intracranial tumors.1,4) Lytic defects in the skull bone are also associated with epidermoid cysts, dermoid cysts, hemangiomas, eosinophilic granulomas, and others.1,4,6) The differential diagnosis of circumscribed lytic defects of the skull includes benign cysts as well as neoplasms. A definitive diagnosis can only be made by performing an open biopsy.

We encountered a case of intradiploic epithelial inclusion cyst lined by ciliated epithelium in the right temporal bone, which appeared as a hot spot on bone scintigraphy. The patient had a history of breast cancer, and differentiation from a bone metastatic lesion was difficult preoperatively. Histological examination of the lining cells demonstrated ciliated epithelium, which is very rare.

Case Report
A 56-year-old woman was referred to our department with suspicion of temporal bone metastasis of breast cancer. She was diagnosed with right breast cancer and referred to the breast surgery department of our institute in December 2008. The breast cancer was T1N0M0 (stage I). She underwent partial removal of the right breast with sentinel lymph node biopsies (SLNBs) in January 2009. The tumor was 10 mm in size, and the SLNBs were all negative. Axillary lymph nodes were not removed. The postoperative course was good, and she was discharged on postoperative day 3. Bone scintigraphy conducted before discharge showed a hot spot in the right temporal bone. Since bone metastasis was suspected, she was referred to our department.

A slightly elevated bulge was palpated under the skin in the right temporal area without tenderness. The overlying skin showed no inflammatory signs. No abnormal neurological findings were noted. Computed tomography revealed a solitary low density lesion without contrast enhancement in the caudal part of the right temporal bone, measuring 3 cm × 3 cm × 1.5 cm (Fig. 1). The outer table was slightly bulged, and both the outer and inner tables

Fig. 1 Bone window computed tomography scan demonstrating osteolytic changes with bulging of the inner and outer tables (arrow).
were thinned. The lesion was circumscribed within the mastoid air cells and the petrous bone, and partial lytic destruction of the underlying bones was suspected. Magnetic resonance imaging showed a high intensity lesion on T2-weighted and fast fluid attenuated inversion recovery images, which appeared slightly hyperintense on the T1-weighted image. Slight peripheral enhancement was observed with gadolinium. There was no obvious extension to the dura or to the brain parenchyma.

The possibility of bone metastasis should be investigated because of the history of breast cancer. Lesion removal was performed via a right temporal craniotomy. The bone surface over the lesion was slightly reddish with mild exudation. There was no obvious erosion. A craniotomy with sufficient margin was made (Fig. 2). The bone flap was easily dissected from the dura without adhesion, and there was no obvious invasion to the dura. A dark yellow liquid was observed in the cystic lesion. The cyst invaded into the mastoid air cells, and sufficient resection of the bone was additionally performed. The opened air cells were covered with a pedicle galeal flap and a deep layer of the temporal muscle. Cranioplasty was performed with a titanium mesh plate. The postoperative course was uneventful.

Examination of the 6 cm × 4.5 cm skull specimen showed a partial bone defect enclosing a cystic lesion with fibrous structure (Fig. 2). The cyst was covered with pseudostratified epithelium consisting of columnar ciliated, cuboidal, or flat epithelial cells (Fig. 3). Goblet-like cells with mucin that reacted with periodic acid-Schiff staining with diastase digestion were also present in the epithelium. The cilia were approximately 10 μm in length. The epithelium was atrophic, with no malignant changes or cellular atypia. Small cysts covered with ciliated or non-ciliated cells were observed in the fibrous structure, and had the same composition as the large cyst. Immunohistochemical studies were conducted. Epithelial membrane antigen (EMA) immunoreactivity was observed clearly on the cilia of ciliated cells and on the cytoplasmic membrane of non-ciliated cells (Fig. 4). CAM5.2 and cytokeratin 7 (CK7) immunoreactivities were positive in the cytoplasm of the epithelial cells (Fig. 4), but no immunoreactivities to CK20, estrogen receptor (ER), and progesterone receptor (Pgr) were detected. The final diagnosis was intradiploic ciliated epithelial inclusion cyst of the skull.

Discussion

The differential diagnosis of lytic defects in the skull bone includes dermoid cysts, epidermoid cysts, hemangiomas, and eosinophilic granulomas.1,4,6) Dermoid cysts are most frequently diagnosed in childhood and epidermoid cysts in adulthood. Intradiploic epithelial inclusion cysts are usually diagnosed at infancy to late adulthood without sex
preference. However, the present case of intradiploic epithelial inclusion cyst lined with ciliated epithelium is extremely rare, and the histology was completely different from the squamous cell lining of epidermoids. In a case of ciliated epithelium cyst of the skull, the lining was histologically compatible with the epithelium of upper respiratory tract, which was the only previously reported case of intradiploic ciliated epithelium inclusion cyst.

In the present case, the epithelium lining the cyst consisted of ciliated cells and non-ciliated cells. Cilia can be differentiated from microvilli by size: cilia are 5 to 10 μm in length and approximately 0.2 μm in diameter, whereas microvilli are 0.5 to 1 μm in length and 80 to 90 nm in diameter. The size of the projections observed in this case was approximately 10 μm, compatible with the size of cilia. Goblet-like cells were also observed in the epithelium. Immunohistochemical studies of epithelial markers showed membrane staining for EMA, clearly demonstrating the cilia structure, and cytoplasmic staining for CAM5.2 and CK7 in the epithelial cells. The absence of immunoreactivities to ER and PgR ruled out the possibility of Mullerian heterotopia in this case. The histological and immunohistochemical findings thus led to a diagnosis of intradiploic ciliated epithelial inclusion cyst, and supported the idea that the cyst in the present case originated from the upper respiratory epithelium. The cyst of the present case was located in the vicinity of the mastoid, and we speculate that the epithelium of the paranasal sinuses might have migrated accidentally to the skull during fetal development. Intradiploic epithelial inclusion cysts are considered to be mainly primary and congenital in origin, theoretically derived from misplaced cell rests during weeks 3 to 5 of embryogenesis. The inclusion of ectodermal cells causes epidermoid or dermoid tumor, whereas the involvement of dysplastic neuroectodermal, mesodermal, and/or endodermal cells causes teratoma. The ectopic stem cells may also be transported accidentally later in fetal development, by tagging along during invagination of the optic vesicles or be pinched off during development of the sutures, meninges, or diploe. Accordingly, lesions of this type commonly arise off-midline in the orbits and temporal bones in relation to the suture lines.

Although a tentative diagnosis can be made based on clinical and imaging features, a definitive diagnosis of intradiploic inclusion cyst requires excisional biopsy. In the present case, breast cancer was previously diagnosed, and subsequently bone scintigraphy showed a hot spot consistent with the radiological findings of an intradiploic cyst. Such a hot spot in the bone scintigraphy scan is not necessarily diagnostic of a malignant tumor, because benign tumor, fracture, or inflammation may also show up as a hot spot, so the preoperative diagnostic findings could not exclude a metastatic lesion to the skull. Considering the possibility of skull metastasis and poor prognosis because of the history of breast cancer, tumor removal with biopsy was necessary for diagnosis and treatment.

The present case of intradiploic ciliated epithelial inclusion cyst of the skull is very rare. Differentiation from metastatic bone lesion is required. Ciliated epithelial inclusion cyst of the skull has unique histological features. Histological evaluation is essential for a final definitive diagnosis and differentiation from a neoplasm.

Conflicts of Interest Disclosure

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices in the article. All authors who are members of The Japan Neurosurgical Society (JNS) have registered online Self-reported COI Disclosure Statement Forms through the website for JNS members.

References


Address reprint requests to: Takakazu Kawamata, MD, PhD, Department of Neurosurgery, Tokyo Women’s Medical University, 8–1 Kawada-cho, Shinjuku-ku, Tokyo 162–8666, Japan.

E-mail: tkawamata@nij.twmu.ac.jp

Neurol Med Chir (Tokyo) 53, April, 2013