Acquired Occipital Dermal Sinus: Tip of the Iceberg

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Introduction

The most frequently occurring pediatric skull lesions are dermoid and epidermoid cysts, which occur due to failure of dysjunction during development, leading to intraosseous retention of epithelial and dermal elements. Rarely a sinus tract may be associated which communicates with intradiploic space, intracranial space and more unusually intadural space. These lesions have propensity of becoming infected, which can thereby lead to infection of subgaleal space and, theoretically the intracranial space in communicating lesions. Here we describe the unusual case of a 2 year old female with an acquired dermal sinus of the occipital region that presumably resulted from the disruption of an intradiploic dermoid cyst.

Case report

A 2 year old female presented to Neurosurgery OPD with chief complaint of occasional pus discharge from occipital region since 5 months. Patient had past history of incision and drainage of scalp abscess over occipital region before 6 months at some peripheral hospital following which patient became asymptomatic. 1 month after incision and drainage, patient developed complaint of pus discharging sinus from occipital region about 2 cm below the previous incision. On examination patient was conscious and vitally stable. Midline occipital sinus with scanty pus discharge was seen. (Figure 1, lower arrow). Scar of previous incision and drainage was seen. (Figure 1, upper arrow). Computed tomography scan (CT scan; Figure 3) showed evidence of 10*5*6 mm sized soft tissue density lesion seen in diploic space of occipital bone in midline with 4 mm sized bony defect in outer table with minimal adjacent soft tissue thickening of scalp. No evidence of intracranial extension was seen. Patient underwent sinus tract exploration under general anesthesia. Sinus tract was seen extending from external opening up to previous scar. Sinus tract was excised and outer table was opened which revealed intradiploic mass (Figure 2) of 1 cm * 1 cm which was excised and sent for histopathological examination. Post operatively patient was stable and asymptomatic during discharge and follow up at 1 week and 1 month. Histopathology report (Figure 4) revealed Ruptured Dermoid cyst with active chronic inflammation.

Figure 1
Dermoid cysts are well-circumscribed lesions lined by stratified squamous epithelium. They contain thick, viscous, and yellowish material composed of secretion of sebaceous glands, calcifications, desquamated epithelium, and whorls of hair. Intradiploic dermoid cyst was first described by Cushing in 1922. Intradiploic dermoid cysts represent 0.04-0.7% of cranial tumors. The occurrence of dermoid cysts of the cranium associated with midline dermal sinus tract is well documented. The reason for reporting this case is the significance that should be given to dermal sinus tracts due to the risk of meningitis following rupture of the intracranial part of the track and the poor results of treatment following meningitis. A dermal sinus consists of a tract lined by epidermis that can connect the skin and the deeper tissues of the central nervous system. It is often clinically occult, but in infants; the presence of a midline cutaneous pit or mass should raise awareness, particularly in the occipital or lumbosacral regions. Dermoid cyst is a congenital benign neoplasm resulting from inclusion of ectodermal elements within the neural tube during its closure between the third and the fifth weeks of embryonic development and, thus, occurs in the midline. It accounts for 0.1%-0.7% of all intracranial tumors. The association of dermal sinus with dermoid cyst, which is well-known, should be suspected and investigated in depth, especially when a cutaneous lesion, such as a lump or pit, lies on the midline. This association can predispose to recurrent meningitis and intracranial suppuration by allowing cutaneous micro-organisms to penetrate into the cyst by means of the sinus tract. Treatment consists of complete surgical removal of the tract and the cyst.

Conclusion

Intradiploic dermoid cysts are relatively rare especially with discharging sinus. In case of patient presenting with dermal sinus over scalp, meticulous exploration of sinus tract should be done to search for intradiploic or intracranial dermoid cyst. Complete resection is recommended due to the potential adverse effects that may occur if these cysts are left untreated.

References

1) Handbook of Pediatric Neurosurgery, page 85


