Anomaly of Venous System in Congenital Occipital Dermal Sinus

R. Tanaka¹, Y. Miyasaka¹, K. Yada¹, S. Kan², and F. Ichikawa¹

Departments of ¹Neurosurgery and ²Radiology, Kitasato University School of Medicine, Sagamihara, Kanagawa, Japan

Summary

We report two cases of congenital occipital dermal sinus in which elongation of the vein of Galen, elevation of the straight sinus, division of the superior sagittal sinus, elevation of the confluence of sinuses, elevation of transverse sinus and narrowing of the torcular angle were observed in the venous phase of cerebral angiography. Enhanced computed tomography (CT) revealed enlargement of the supracerebellar cistern, elevation of the straight sinus and of the confluence of sinuses, but no evidence of intracranial lesions. In order to study the relationship between anomalies in the dural venous sinuses and congenital occipital dermal sinus, we examined both cases from an embryological viewpoint.

Keywords: Congenital occipital dermal sinus; dural sinuses; development; embryology.

Introduction

Although there have been detailed reports by Matson¹ and Wright² concerning congenital occipital dermal sinuses, most studies of this anomaly emphasize only the complication of congenital cell rest tumour and its importance as one cause of diseases such as meningitis. There have recently been a few reports examining the characteristics of congenital occipital dermal sinus as seen with magnetic resonance imaging (MRI)³. However, few reports have studied the relationship between congenital occipital dermal sinus and venous system anomalies⁴.

In our two cases of congenital occipital dermal sinus, dural sinus anomalies were observed, and their relationship with congenital occipital dermal sinus was studied from a developmental viewpoint.

Cases

Case 1

A 5-year-old girl had been healthy until the day of admission. She was born after a full-term pregnancy during which the mother had shown no abnormal symptoms, but since birth, a median subcutaneous mass had been present at the occiput. Since no symptoms developed, the mass was left untouched. The patient showed no fever that might suggest meningitis and both physical and mental development was normal. The remainder of the patient’s medical history was unremarkable.

On examination, a median subcutaneous mass 2.5 × 2.5 cm in size was found at the midline of the occiput. It showed no mobility, Fig. 1. Anteroposterior view of skull showing a circular defect of the occipital bone in the midline.
and no pulsation and was evident on palpation. The hair at the site was sparse, although a dimple with an abnormal quantity of hair was present.

There were no findings suggestive of inflammation, nor was any ectasia or enlargement of the subcutaneous mass detected on the application of pressure on the jugular veins.

Neurological examination showed no abnormalities. 

Neurological findings: Plain cranial radiography showed a circular defect approximately 10 mm in diameter in the bone at a site corresponding to that of the subcutaneous mass (Fig. 1). A CT scan showed enlargement of the supracerebellar cistern, elevation of the straight sinus and an abnormally high confluence of the sinuses, but no intracranial lesions (Fig. 2). The depth of the dermal sinus was unable to be determined.

On cerebral angiography, no anomalies were observed in the arteries and cortical veins. However, the following anomalies were recognized: elongation of the vein of Galen, elevation of the straight sinus, splitting of the posterior portion of the superior sagittal sinus at a site corresponding with that of the defect in the bone, elevation of the transverse sinus, abnormally high confluence of the sinuses and narrowing of the torcular angle (Fig. 3).

Surgical findings: When the subcutaneous mass in the midline of the occiput was removed from the skin, a fibrous structure was seen passing through the defect in the bone. When this structure was removed carefully, there was no evidence of penetration of the dura mater, and so complete extirpation was achieved with ease.

Histopathological findings: The walls of the dermal sinus were covered with stratified squamous epithelial cells.

Postoperative course: Twelve years have passed since the operation. No neurological abnormalities have appeared, and the patient is at present leading the normal life of a school girl. Furthermore, on MRI performed nine years after the operation, only the venous system anomalies and the enlargement of the supracerebellar cistern, that had been observed on cerebral angiography, were recognized. There were no findings suggestive of a congenital cell rest tumour (Fig. 4).

Fig. 2. Contrast CT shows the upward course of the straight sinus together with superior extension of the supracerebellar cistern and splitting of the superior sagittal sinus.