RESEARCH PAPER | Radiology | Volume : 6 | Issue : 3 | March 2016 | ISSN - 2249-555X | IF : 3.919 | IC Value : 74.50

Dandy Walker Malformation with Parietal Encephalocele- Case Report.

KEYWORDS: Dandy-Walker malformation, encephalocele, posterior cranial fossa, magnetic resonance

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ABSTRACT *Dandy-Walker malformation is congenital malformation characterized by dysgenesis of the foramina of Magendie & Lushka in the upper 4th ventricle, hypoplasia of the cerebellar vermis and agenesis of the corpus callosum. Cephaloceles are congenital malformations with herniation of intracranial structures through a defect in the cranium. It is one of the rarest neural tube defect. A 3 year old male child came with 2x2 cm encephalocele on physical examination. On magnetic resonance imaging, the posterior fossa was enlarged with cysts and vermian hypoplasia. A connection was seen between the ventricle and the cysts in the posterior fossa. These findings were significant from the aspect of Dandy-Walker malformation. Bony defect in the right parietal region with extension reaching 7 mm was seen in accordance with encephalocele. It is rare for Dandy-Walker syndrome to occur together with parietal encephalocele.*

CASE REPORT:
A 3 Year old male child was brought by his mother with complaint of swelling over parietal region since birth. The swelling was gradually increasing in size and at time of presentation measured approximately 2x2 cm. (FIGURE 1)

There was no history of trauma or developmental delay. Birth history was insignificant.

USG of the swelling revealed possible brain parenchymal tissue with vessels within the lesion. (FIGURE 2) Subsequently MRI Brain was done which showed herniation of CSF and isointense tissue of brain parenchyma into the extracalvarial subcutaneous plane through skull defect measuring approx 7 mm (seen on relevant CT sections)(FIGURE 3) in right parietal region in the paramedian location near confluence of sinuses, suggestive of encephalocoele. The swelling measured approx. 1.5x1.7x2.2 cm (APxTRxCC). Hypoplastic inferior vermis with prominent 4th ventricle and cisterna magna was noted suggestive of Dandy Walker malformation. There was agenesis of the distal body and splenium of corpus callosum. (FIGURE 4)

Figure 1: Clinical photograph of the swelling.

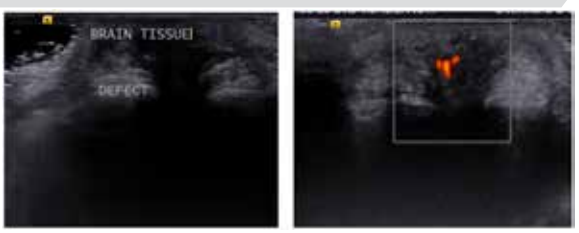

Figure 2a) Ultrasonographic image showing skull defect and brain tissue herniation through it. 2b) Colour Doppler image showing vascularity with vessels within the herniated tissue.

Figure 3: CT angiography reformated sagittal image of brain showing skull defect in right parietal region with herniated brain tissue.

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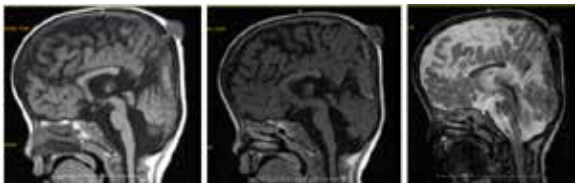

Figure 4a) MRI T1 WI sagittal section of brain showing encephalocole, hypoplastic inferior vermis with prominent 4th ventricle and cisterna magna. Also there is agenesis of the distal body and splenium of corpus callosum.4b) MRI post contrast T1 WI sagittal section of brain showing same findings as in Fig.4a, with enhancing vessels in encephalocoele.4c) T2 WI sagittal section showing CSF within the lesion.

DISCUSSION:

Agenesis of corpus callosum is the most common CNS anomaly associated with Dandy-Walker syndrome. [1] Only few cases of Dandy-Walker syndrome with meningocele or encephalocele have been reported. Curnes JT, Oakes WJ reviewed 3 cases of parietal cephalocele, finding all 3 cases had associated hindbrain deformities; two with Dandy-Walker malformation, the third with a Chiari II malformation. [2]

theories have been proposed for the pathogenesis of Dandy-Walker syndrome. The original theory was based on atresia of the foramina of Luschka and Magendie. [3] Carmel et al[4] postulated that Dandy-Walker syndrome is a primary developmental anomaly that affects the closure of the neural tube at the level of the cerebellum before the 8th week of embryonic life. In contrast, Padget [5] explained the syndrome as the result of reopening of the neural tube rather than failure to close.

In the present case, however, the coexistence of the encephalocele suggests that the origin is much later than neural tube closure. Furthermore, histological and immunohistochemical analyses of human neonate autopsy and surgical specimens demonstrated that, in contrast to anencephaly which is associated with interruption of the neuronal developmental process, cranium bifidum cysticum presents with completed cortical formation of the telencephalon with an intact central neural canal, implying that cranium bifidum cysticum originates later than anencephaly. Based on this evidence, the associated encephalocele in our case suggests that Dandy Walker syndrome may not be an early embryonal maldevelopment, but an event occurring much later than the closure of the neural tube.

Dandy-Walker malformation is best diagnosed with the help of ultrasonography (US) and magnetic resonance imaging (MRI). US may be the initial examination performed because it can be done portably and without sedation. [7, 8] US, however, is limited because it is heavily operator-dependent.

CT is used to diagnose Dandy-Walker malformation and to follow ventricular shunt function. Nonenhanced CT examination successfully delineates multiple components of Dandy-Walker malformation and encephalocoele, and is mainly used to delineate the skull defect.

Malformations of the CNS are best delineated using MRI. MRI is usually required for better anatomic resolution prior to surgical intervention.[8] Sagittal MRI can help evaluate an abnormally high position of torcular herophili, high tentorial insertion (lambdoid-torcular inversion), hypoplastic and compressed brainstem, and obstructive hydrocephalus secondary to cystic dilatation of the fourth ventricle in patients with Dandy-Walker malformation. MRI also is the procedure of choice for displaying communication of the cephalocele with intracranial structures as well as associated venous vascular anomalies. [2]

Classic abnormal findings described on cranial CT and MRI scans can also be demonstrated on cranial sonography. Transducers of 5-7.5 MHz are used for newborns, and transducers of 3-5 MHz are used for older infants.

surgical methods used to treat Dandy-Walker syndrome include shunt of the lateral ventricle and in refractory cases shunt of the cyst. This is considered to have a poor prognosis, more so if associated with other CNS anomalies.

CONCLUSION:

There is herniation of CSF and brain parenchyma through skull in right parietal region in paramedian location near confluence of sinuses, suggestive of encephalocoele.

Hypoplastic inferior vermis with prominent 4th ventricle and cisterna magna is noted, suggestive of Dandy Walker malformation.

This is also associated with agenesis of the distal body and splenium of corpus callosum.

ACKNOWLEDGEMENTS-

I would like to thank **Dr Santosh Konde** for providing me the necessary insight to evaluate the case presented in our institution and encourage me to publish this case. I would also like to thank **Dr Punit Agrawal and Dr Swapnil Raut** for always being there as a constant support and a gem of a colleague to work with. My gratitude also goes out to my guide **Brig. Dr Hariqbal Singh** for always being a constant source of support for the entire faculty.

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