CASE REPORT

Dural sinus filling defect: intrasigmoid encephalocele

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SUMMARY

Filling defects of dural venous sinuses are considered to be a challenging problem especially in case of symptomatic patients. Many lesions have to be ruled out such as sinus thrombosis, arachnoid granulations and tumours. Encephalocele into dural sinus is also a rare cause of these filling defects of dural sinuses. Here, we report an extremely rare case with spontaneous occult invagination of temporal brain tissue into the left sigmoid sinus and accompanying cerebellar ectopia.

BACKGROUND

Filling defects of dural venous sinuses have long been considered to be a challenging problem. There have been many studies about these filling defects caused by arachnoid granulations (AGs) or sinus thrombosis. Although these filling defects are frequently detected incidentally, radiologists feel troublesome about notification of this finding in their final reports. This situation becomes more problematic if the patient is symptomatic, because many lesions have to be ruled out, especially sinus thrombosis. There are several causes of dural sinus filling defects including AGs, sinus thrombosis, tumours and encephalocele into dural sinus. Here, we report an extremely rare case with spontaneous occult invagination of temporal brain tissue into the left sigmoid sinus and accompanying cerebellar ectopia.

CASE PRESENTATION

A 31-year-old woman was admitted to our neurology department because of recent onset of headache (especially in standing). The physical examination and laboratory revealed normal findings. There was no history of previous trauma or surgical management and any kind of seizure in her medical records.

INVESTIGATIONS

Cranial MRI with a 1.5 T scanner revealed loss of signal void in the left sigmoid sinus (figure 1) as well as herniation of the cerebellar tonsils through foramen magnum (figure 2). Thin slice, multiplanar, three dimensional (3D)—fast spoiled gradient echo T1-weighted (brain volume imaging (BRAVO)) and 3D—fast imaging employing steady-state acquisition (FIESTA) images showed filling defect of left sigmoid sinus which gave the impression of left temporal brain tissue extension (figure 3A,B). On contrast-enhanced coronal and sagittal cranial BRAVO sequence, a pedunculated soft tissue filling defect measuring 18×15×7 mm within the left sigmoid sinus was clearly demonstrated to be a cerebral herniation into left sigmoid sinus (figure 4A,B). A final diagnosis of spontaneous occult invagination

Figure 1 T2-weighted axial image showing loss of signal void in the left sigmoid sinus (arrow).

(encephalocele) of temporal brain tissue into the left sigmoid sinus and accompanying cerebellar ectopia was made. Additional MRI images obtained to evaluate the whole spinal canal showed no evidence of spinal osseous deformity or cavitation in spinal cord.

TREATMENT

The patient was referred to the neurosurgery department. She was recommended to have an operation and also a detailed information was given

Figure 2 On T2-weighted sagittal image, cerebellar ectopia is clearly visible.

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To cite: Karatag O, Cosar M, Kizildag B, *et al.* *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-201616

Figure 3 Precontrast coronal BRAVO (A) and FIESTA (B) images demonstrating filling defect of left sigmoid sinus which gives the impression of left temporal brain tissue extension (arrows). On coronal FIESTA image, right sigmoid sinus has normal signal (asterisk).

to her about the operation type, the benefits and possible complications that she might face. However, she did not accept the surgical management and was on 1 week bed rest and analgesic medication at the hospital. She has been undertaken for clinical and radiological follow-up.

DISCUSSION

The inner layer of duramater compose wall of dural venous sinus which shows different specialties from classical venous vessels.¹ The drainage of cerebrospinal fluid (CSF) through arachnoid villi leak into dural venous sinus, deep and superficial venous structures of the brain.²⁻³ So, dural venous sinuses carry all removable extracts of the brain via CSF and venous blood. Brain tissue invagination, AGs, tumours and thrombosis may occlude the dural venous sinuses thus leading to dramatical pathological changes.²⁻⁴⁻⁶

Encephalocele into dural venous sinus is an extremely rare entity which can appear as filling defects of dural venous sinuses on radiological images. There have been a few studies talking about this condition in which other filling defects of dural venous sinuses such as AGs and calvarial lesions are mostly discussed. There is no detailed information about the mechanisms, symptoms and treatments of encephalocele into dural venous sinus in the literature.

AGs are normal variants which appear as focal filling defects within dural venous sinuses during neuroradiological imaging.²⁻⁷ They are thought to be enlarged arachnoid villi.²⁻⁴⁻⁵⁻⁷ Both AGs and arachnoid villi are responsible for resorption of CSF from subarachnoid space into venous

circulation.²⁻³ AGs rarely exceed 1 cm in diameter.⁴⁻⁷ If they become excessively large, they can cause impression and erosion of the skull.²⁻⁵ So, a differential diagnosis of AGs with pathological processes becomes essential.

AGs are described as isointense structures relative to CSF in all MRI sequences.⁴⁻⁶⁻⁸ Controversely, the fluid content of AGs, does not always have the same MRI features with CSF.⁴ Round/ovoid shape and the absence of solid contrast enhancement may be helpful in the differentiation of AGs from other lesions.⁴

Vascular structures identified as linear flow voids or contrast enhancement within or adjacent to AGs have been featured in many studies.⁴⁻⁷ Non-vascular soft tissue content compatible with stromal collagenous tissue, hypertrophic arachnoid mesangial cell proliferation, or encephalocele have also been reported in giant AGs.²⁻⁴⁻⁷⁻⁸

Several lesions can be misdiagnosed as AGs on radiological images, including sinus thrombosis, calvarial lesions, meningiomas, metastases, arachnoid cysts, dermoids, epidermoids, extra-axial haemangiomas, intrasinus tumours and encephalocele.²⁻⁴⁻⁶ Thrombus typically appears as an elongated filling defect extending along the sinus.⁴ Dermoids include fat signal⁴ and epidermoids show restricted diffusion with MRI.⁴⁻⁶

The filling defect in our case within left sigmoid sinus was round-shaped and sharply margined similar to AGs. However, the dimensions of our lesion were larger than AGs previously reported.⁴⁻⁷⁻⁸ Besides, we did not detect any linear signal void or contrast enhancement indicating venous structure entering the pouch. Thin slice multiplanar 3D-BRAVO and 3D-FIESTA MRI sequences clearly demonstrated the continuity of the filling

Figure 4 Postcontrast coronal (A) and sagittal (B) BRAVO images clearly show that the filling defect within the left sigmoid sinus has continuity with the left temporal brain tissue consistent with cerebral herniation (arrows).

defect with brain tissue. Thus we diagnosed the filling defect as encephalocele into left sigmoid sinus.

We really do not know whether headache in our patient is related to encephalocele or not. There is not enough information concerning the nature of the related headache in the literature, and long-term follow-up studies are strongly needed to understand if symptoms are in relation with radiological findings or not.⁶ Besides, cerebellar ectopia can also be responsible for the headache in our patient. Although tussive occipital headaches have been reported to be the most frequent type of headache in cerebellar ectopia, there is no objective way to differentiate headaches associated with cerebellar ectopia from headaches which are not.⁹

In addition, it is well known that the tension of the duramater and vessels is one of the most frequent mechanisms of headache.¹⁰ The pressure of the encephalocele may change with the position of the patient under the effect of gravity. This may reflect to intrasinus blood pressure and may cause tension of duramater. Increase in severity of the headache especially in standing position and decrease during bed rest in our case may be explained this way.

The commonest of the associated syndromes with encephalocele has been reported to be the Meckel Gruber syndrome.¹¹ Other cerebral malformations often associated with encephalocele are deformities of the tentorium, complete or partial agenesis of the corpus callosum and myelomeningocele.^{11–12} On the other hand, occipital encephalocele mostly containing much of the cerebellum has been well known to be the component of Chiari III malformation.^{13–14} In a case report by Udayakumaran *et al*,¹⁵ the association of the temporal lobe herniation and Chiari II malformation has also been reported. The most common accompanying findings in Chiari I malformation has been listed as spinal cavitations (syringomyelia, hydromyelia and syringohydromyelia), atlas assimilation often associated with Klippel-Feil anomaly, and hydrocephalus.^{16–20} However, there has been no literature finding supporting the relationship between encephalocele and Chiari I malformation. Thus we think that the findings in our case may be coincidental.

Surgical management is required for temporal encephaloceles if the patient has temporal lobe epilepsy. There have been approximately 15 cases in the literature dealing with temporal encephalocele which were excised in order to control seizures resistant to medical treatment.²¹ The temporal encephaloceles causing epilepsy discussed in the literature were seen either congenital or acquired where the acquired form had occurred due to trauma, infection, tumour or previous surgery.^{22–25} It has been postulated that the pathogenesis of developmental temporal encephaloceles are different from those associated with neural tube defects.²¹ Main mechanisms mostly discussed in the literature are either non-union of ossification centres in temporal bone²⁶ or variations of temporal bone thickness or occurrence of some tiny pores through which brain tissue herniates by the pressure of the brain tissue or CSF.²⁷ The improvement in resolution of MR scanners as well as detailed thin section coronal images has provided a better visualisation of these pathologies before surgery.²¹ We did not come across any case report regarding temporal encephalocele without epilepsy that had undergone surgical intervention. In addition, according to our knowledge this is a unique case report of a temporal encephalocele herniating into the sigmoid sinus.

As most of the filling defects within dural sinuses have been reported to be AGs, with the possibility of many lesions mimicking this condition, the diagnosis of encephalocele into dural venous sinus should always be kept in mind in the differential

diagnosis. Cross-sectional imaging methods, especially MRI plays a critical role in recognising these filling defects within dural sinus.

Learning points

- ▶ Encephalocele into dural venous sinus is an extremely rare entity which can appear as filling defects of dural venous sinuses on radiological images.
- ▶ The mechanism of a developmental temporal encephalocele is either non-union of ossification centres in temporal bone or variations of temporal bone thickness or occurrence of some tiny pores in which brain tissue herniates due to pressure phenomena. The mechanism of developmental temporal encephaloceles is different from those associated with neural tube defects.
- ▶ There are several causes of dural venous sinus filling defects including arachnoid granulations, sinus thrombosis, tumours and encephalocele.
- ▶ The differential diagnosis of these filling defects has to be made carefully in order to avoid unnecessary and wrong treatment procedures.
- ▶ MRI should be the preferred method for the best visualisation of these filling defects.

Contributors OK and BK were involved in the collection of the data, and wrote the manuscript. MC also wrote the manuscript and HMS was involved in the collection of the data.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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