CHAPTER 10

Benign Pericerebral Collections in Children

SYROS SGOUROΣ AND CHRISTOS TOLIAS

Introduction

Excess extracerebral accumulation of CSF over the brain and under the arachnoid mater in infants was first described by Dandy, who introduced the term "external hydrocephalus", indicating that CSF is collecting in a site external to the brain itself [14]. The plethora of terminology used in the past for extracerebral collections has been confusing and reflected the poor knowledge of the condition. Terms such as "benign subarachnoid collections of childhood", "benign enlargement of the subarachnoid spaces", "external hydrocephalus", "widened subarachnoid space", "subdural haematoma" (collection of blood), "subdural effusion" (collection of proteinaceous fluid) and "subdural hygroma" (collection of CSF) have all been used to describe extracerebral collections. Most of these terms predate the discovery of computed tomography (CT) and derive their origin from the clinical observations of the fluid recovered during subdural taps. Several early clinical series included patients with subarachnoid and subdural collections, contributing to the confusion over management and outcome issues [8]. A clear distinction should be made between benign pericerebral collections of CSF, previously called "external hydrocephalus", and subdural collections. The wider use of CT in the 1980s and magnetic resonance imaging (MRI) in the 1990s has contributed significantly to differentiating between the two conditions. In the former, excess CSF is accumulating underneath the arachnoid mater, whereas in the latter excess fluid - usually altered blood mixed with CSF - is accumulating between the arachnoid and the dura mater. The clinical presentation of the two conditions can be similar with progressive enlargement of the head, but the natural history is often different and consequently the management differs. For subarachnoid collections of CSF the term "benign pericerebral collections" probably reflects the anatomical basis of CSF distribution and the natural history of the condition better, and is therefore to be preferred.

Incidence of Benign Pericerebral Collections

The incidence of benign pericerebral collections among infants is unknown. It appears to be more common in boys. Apart from the absence of any population-based studies, an additional problem associated with the diagnosis of the condition is the lack of clear definition of what constitutes a pericerebral collection in an infant. It is established that in normal neonates and infants the subarachnoid spaces are wider than in older children, and this may persist for up to the 1st year of life [21, 42]. It is not clear, though, when the normal prominent subarachnoid spaces become benign pericerebral collections of CSF. In most published series the diagnosis is made on the basis of disproportionate enlargement of the frontoparietal subarachnoid spaces on CT or MRI, without any attempt to quantify that enlargement. Differentiation from cerebral atrophy is equally unclear and has usually been based on the presence on non-increasing head circumference, associated with a uniform enlargement of subarachnoid spaces.

Pathophysiology of Benign Pericerebral Collections

The pathophysiology of benign pericerebral collections is not well defined. Many authors regard this condition as a variation of the normal growth pat-
Clinical Features of Benign Pericerebral Collections

Clinical presentation can be with progressive enlargement of the head in the first few months of life, disproportionate to the remaining body growth. The head circumference usually crosses the centiles and often climbs well over the 95th parallel [2, 12, 26, 30, 33, 34, 48], although pericerebral collections have been observed in children with normal head circumference who underwent a CT scan for some other reason. Indeed, in a series of 67 children with widened subarachnoid spaces only 23% had macrocrania [56]. In another series of 63 patients, none had increased head circumference [35]. A significant proportion of children (20%) with benign pericerebral collections have been born prematurely [1]. In a large proportion of patients, up to 88%, a history of macrocrania among parents has been described [1, 48]. A history of abnormal delivery is reported on some occasions [35, 48]. Normal development is usually reported by the parents, although some developmental delay has been reported in up to 25% of children [1, 35]. Features of intracranial hypertension are usually absent both in the history and on examination. In the majority of patients no predisposing cause can be identified for the CSP collections. In a small proportion of patients other associated conditions coexist, such as epilepsy or a variety of genetic syndromes, such as achondroplasia, craniosynostosis, Beckwith syndrome, Soto syndrome, Goldenhar syndrome, Weaver syndrome and others, most of which can be associated with various forms of developmental delay and should be considered when advising parents on long-term prognosis.

Neuroradiological Findings

Before the development of modern digital imaging, a typical investigative approach to children with enlarging head circumference would be the performance of diagnostic subdural taps, followed by subdural drainage, radionuclide scan, air ventriculography or encephalography as appropriate [18, 39]. Technological progress in the field of radiology has made invasive techniques obsolete, which in turn lead to a lower threshold of investigation, resulting in more children undergoing imaging and in consequence a clearer knowledge of the condition. Diagnosis can be made with ultrasound imaging [13, 15, 44], but the methods of choice are CT and MRI. Ultrasound is available more readily to community paediatricians and can be used as a first quick measure in the investigation of a child with an enlarging head, excluding large space-occupying lesions. It can demonstrate clearly the deeper position of the cortical surface, and the widened subarachnoid spaces and interhemispheric fissure [44]. It can differentiate between subarachnoid and subdural collections. In a study comparing ultrasound and MR findings in seven children – four with benign pericerebral CSP collections and three with subdural collections – it was noted that in all cases ultrasound could differentiate between the two pathologies correctly [15]. The use of a 10-MHz probe was particularly helpful in identifying different components of the collections. The combined use of colour-Doppler imaging has also been reported as useful in discriminating between the two conditions [13]. The positive cortical vein sign has been described: colour-coded veins are seen crossing subarachnoid fluid collections at the cerebral convexities [13]. This sign was regarded as pathognomonic in a comparative study of 18 patients: concordance of diagnosis was seen in all patients when ultrasound and MR imaging were compared [13]. While the non-invasive nature of ultrasonography is appealing, the end-result is operator-dependent, which can be a problem when appropriate expertise is not available.
A more reliable and objective modality is the CT scan, which shows a considerable amount of CSF over the convexity of the brain parenchyma, usually more prominent in the frontal and parietal regions, and an enlarged interhemispheric fissure anteriorly (Fig. 1a). In the majority of patients (80%) the collections are bilateral [25, 36]. Mild or even moderate ventriculomegaly has been observed in up to 70% of the patients, but ventricles are normal in up to 30% of the patients [12]. Contrast-enhanced CT can help differentiate subarachnoid from subdural collections by the position of enhancing cortical blood vessels: in subarachnoid collections the vessels are seen traversing the collection, whereas in subdural collections the vessels are seen compressed on the cortical surface of the brain. In addition, subdural collections often are of greater density than CSF [22, 25, 28, 32, 33-36, 48]. The collections can be classified as to extent according to whether they occupy a frontal disposition only, or whether they extend and separate the anterior interhemispheric fissure, or whether in addition they dilate the cortical sulci as well (grade I, II and III or convexity-faix-sulcus type, respectively) [32, 36].

CT scan is not always reliable [30], and imaging in the brow-down position has been employed to enhance the detection rate. In cases where CT cannot resolve the difference between a subarachnoid collection and an isodense subdural one, MRI scan can significantly help to differentiate between the two conditions, in particular when a combination of T1-, T2-weighted and proton density sequences are employed [4, 50]. In subarachnoid collections the fluid has an intensity similar to that of CSF, the vessels that cross the subarachnoid space are clearly seen as flow voids within the extracerebral collection, and the collected fluid follows the brain parenchyma in the sulci, often spaying them open (Fig. 1b, c). In subdural collections the fluid has an intensity greater than that of CSF, no flow voids attributable to blood vessels are seen within the collections, the cerebral sulci are compressed and the arachnoid membrane is often clearly seen as a separate structure [4, 15, 22, 48] (Fig. 2). It is not unusual to have coexistence of subarachnoid and subdural collections, which can vary in appearance between the two sides of the head [32, 34].

An important issue is differentiation from cerebral atrophy. This is more important in children who have an apparent degree of developmental delay, and some other potential cause of cerebral atrophy. Differentiation between the two conditions has a significant bearing on prognosis. Radiological criteria for differentiation between the two conditions are not very rigid. Cerebral atrophy is usually diagnosed in children with normal or low head circumference for age, whose scans show a uniform enlargement of subarachnoid spaces, with wide dilatation of cortical sulci in all regions of the brain, and an element of ex vacuo ventriculomegaly. Sometimes, though, differentiation between pericerebral CSF collections and cerebral atrophy is not easy.

![CT scan images showing pericerebral collections.](image)

**Fig. 1a-c**: Benign pericerebral collections. **a** CT scan of a 4-month-old boy who presented with macrocephaly and seizures. Enlarged subarachnoid spaces are seen in the frontal regions. The anterior interhemispheric and right sylvian fissures are splayed open. Mild ventriculomegaly is present. **b** T1-weighted MRI scan, showing signal intensity in the pericerebral collections similar to ventricular CSF. The fluid of the collections "enters" the sulci, splaying them open. **c** T2-weighted MRI scan, showing clear signal voids from cortical veins, traversing the collections from the dura to the surface of the brain.
Natural History of Benign Pericerebral Collections

Differentiation between benign subarachnoid CSP collection and subdural collection is important from the management point of view; the former requires no surgical treatment, the latter some form of drainage. The outcome of benign subarachnoid collections is usually good without any surgical treatment: most children develop well, the head enlargement tends to arrest by 15–18 months and in most cases the collections disappear by 2 years of age. In a small proportion of children subtle motor developmental dysfunction can be identified with detailed testing [41]. In a study of 74 megalen-