Subdural collections of infancy and setting sun sign may be asymptomatic and benign or symptomatic and aggressive – Report of two cases

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Abstract

Objective. Subdural collections of infancy may be asymptomatic and benign or symptomatic and aggressive. We present two cases illustrating both clinical scenarios. The presence of setting-sun sign is an unusual feature in both cases.

Clinical Presentation. The diagnosis of benign subdural collections of infancy depends on several distinct clinical and laboratory findings, which are reflected in these two cases: patient age < 1 yr, macrocrania (Case 1 was 79 cm), normal psychomotor development, no history of CNS infection or head trauma; subdural fluid clear yellow; erythrocyte count < 1 million. Both cases presented with setting sun sign. CT scans showed craniocerebral disproportion, disjunction of the sphenoid, low measures of attenuation coefficient values, relatively normal ventricular size, and no cerebral atrophy. MRI also showed agenesis of the corpus callosum, in case 1.

Intervention. Based on megacrania associated with poor visualization of the cerebral sulci, Case 1 was treated with a subdural collection-peritoneal shunt; Case 2 was treated with clinical observation due to enlarged subarachnoid spaces.

Conclusion. We present two cases with different clinical courses – one more aggressive than the other. We believe these two cases illustrate a disease process that is not always benign, particularly in view of setting-sun sign, and thus warrants a more accurate terminology. Regardless of terminology, however, even symptomatic cases can have good outcome with conservative treatment. Good outcome relies on close observation associated with complete workup that evaluates signs and symptoms of discompensation.

Keywords

Benign subdural collections, hydrocephalus, setting-sun sign, macrocrania, peritoneal shunt.

Sinopse

Coleção subdural na infância e sinal do sol poente com curso assintomático e benigno ou sintomático e agressivo – Relato de dois casos

Coleção subdural da infância pode cursar clinicamente de modo benigno e assintomático ou, ainda, de modo agressivo e sintomático. Os autores apresentam dois casos ilustrativos de ambos os cenários. A presença de sinal do sol poente é um sinal presente em ambos os casos. O diagnóstico das coleções subdurais da infância depende de critérios clínicos e achados laboratoriais, os quais refletem em ambos os casos: macrocrania (Caso 1 – apresentava perímetro craniano de 79 cm), desenvolvimento neuropsicomotor normal, ausência de infecções do sistema nervoso central ou traumatismo craniano, fluido subdural xantocromático, contagem de eritrócitos < 1 mil/mm²; TCC revelando desproporção craniofacial,

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disjunção das suturas, valores rebaixados do coeficiente de atenuação, diâmetros ventriculares relativamente normais e ausência de atrofia cerebral. RN de crânio demonstrou agenesia do corpo caloso no Caso 1. Com base na macrocrania associada a baixa visualização dos sulcos cerebrais, o Caso 1 foi tratado com shunt coleção subdural-peritoneal; o Caso 2 foi observado clinicamente devido ao alargamento dos espaços subaracnoideos apenas.

Palavras-chave
Colecção subdural benigna, hidrocefalia, sinuto do sôlo poente, macrocrania, derivação peritoneal.

Introduction

The diagnosis of benign subdural collections of infancy depends on several distinct clinical and laboratory findings. The patient presents with less than 1 year of age, with macrocrania, normal psychomotor development, and no history of central nervous system (CNS) infection or head trauma. Subdural fluid is usually clear yellow to xanthochromic; the erythrocyte count is less than 1 million/mm³. Neuroimaging reveals normal or slightly enlarged ventricles associated with a normal brain size with or without corpus callosum agenesis.

Little is certain regarding etiology, treatment, or even the terminology of this disorder. It has been variously described as 'benign extra-axial collection of infancy', 'benign enlargement of the subarachnoid spaces, benign communicating hydrocephalus, extra-ventricular obstructive hydrocephalus, and external hydrocephalus.'

Therapeutic options are variable. Treatment for these subdural collections includes observation, percutaneous needle drainage, drainage of the fluid into a closed external system, peritoneal shunting, and craniectomy with removal of subdural membranes.

We present two cases with different clinical courses—a more aggressive than the other. We believe these cases illustrate a disease process that is not always benign and thus warrants a more accurate terminology.

Case report

Case 1

A 6-months old male was referred for evaluation of "a big head" (Figure 1A). Pregnancy and birth history were unremarkable. There was no history of CNS infection or head trauma, no vomiting or alteration of consciousness, and he was feeding normally. The patient was alert and sociable, smiling to his parents. His motor development was normal except for the setting-sun sign (Figure 1A). Moro and bilateral extension plantar were present. Funduscopy was normal. He had a palpable global disjunction of the cranial sutures associated with a bulging and tense cranial vault. Scalp venous engorgement was also seen. Head circumferenc was 79 cm. CT showed an immense subdural collection with no sign of cerebral atrophy. MRI revealed agenesis of the corpus callosum (Figure 1B-D). Hygroma clear yellow fluid was obtained which were clear yellow and contained 3500/mm³ erythrocytes. The patient underwent a subdural-peritoneal shunt with an uneventful postoperative course. He was discharged 30 days post-surgery with head circumference measuring 75 cm. The patient died 6-month thereafter due to complications of respiratory tract infection.

Case 2

A 10-months-old male was evaluated for abnormal head growth. Pregnancy and birth history were unremarkable and there was no history of CNS infection or head trauma. Neurological examination was normal, except for setting-sun sign. Head circumference was at 98th percentile with a normally flat anterior fontanelle associated with a disjunction of the metopic and coronal sutures. Funduscopic examination was normal. CT scan showed a mild ventricular enlargement, wide cerebral sulci, enlarged Sylvian cistern, and interhemispheric fissure (Figure 2). Subdural tap produced 6 ml of clear yellow fluid which contained 1700/mm³ erythrocytes. Monthly neurological observation was the therapeutic option. The patient maintained normal mental and motor development. Later follow-up at 3 months showed improvement in upward gaze palsy and head circumference in 95th percentile.

Discussion

It has been postulated that subdural collections of infancy may result from a delayed maturation of the arachnoid granulations or other CSF resorption sites. Most published reports describe subdural collections of infancy as a benign entity because of a favorable clinical evaluation and good prognosis.

Six months-old baby with macrocrania and setting-sun sign.
Macrocrania and irritability associated with normal psychomotor development are the most common clinical presentations. Although unusual, this so-called benign conditions also present with large effusions, gross abnormalities of CSF flow, or signs and symptoms of intracranial hypertension. Litofsky et al. describe this condition as benign only in cases of asymptomatic fluid collections; they prefer the term chronic extra-axial fluid collection in symptomatic cases.

What dictates a benign or aggressive clinical course is unclear. The two cases we present reflect quite different clinical scenarios, the first being much more intense than the second, despite similar presentations, e.g., normal psychomotor development, macrocrania, impairment of upward gaze and similar erythrocyte count in the subdural collection associated with an unremarkable history for head trauma and CNS infection.

The lack of abnormalities in the psychomotor development and absence of signs of acute intracranial hypertension in both cases is a result of skull mobility seen in infants. Goosknens et al. found fewer neurological symptoms with higher intelligence and/or more advanced development in a group of macrocephalic-megaloecephalic children (including those with subdural collections of infancy) compared with a group of macrocephalic-hydrocephalic children.

Both of our cases came to neurological evaluation for macrocrania associated with a setting-sun sign. Macrocrania is an important sign for several disorders affecting infants, including hydrocephalus, chronic subdural hematomas, subdural hygromas, and subdural collections. The differential diagnosis requires anamnesis, neuroradiological investigations, and at times, CSF examination. Alvarez et al. reported a familial macrocrania in up to 88% of their cases of external hydrocephalus - a relationship unsupported by other authors. No familial macrocrania existed in either of our cases.

The presence of the setting sun sign in both of our cases is an infrequent association with subdural collections of infancy. The absence of ventricular dilatation in both infants without any direct involvement of the prectum or a portion of the posterior commissure areas lead to an anatomical explanation for the upward gaze impairment (i.e., an indirect mechanical compression produced by subdural fluid to the outside mesencephalon and pretectal regions).

Analysis of subdural fluid and erythrocyte count can both be used to distinguish subdural collections/hygromas from subdural hematomas. Red-brown collections are indicative of subdural hematoma and yellow-xanthochromic fluid.
fluids of a subdural collection/hygroma. \textsuperscript{4,12,20,30} Erythrocyte counts < 1 ml/mm\textsuperscript{3} can be used to distinguish a subdural collection/hygroma from a subdural hematoma. In both of our cases, clear yellow fluid with an erythrocyte count < 4000/mm\textsuperscript{3} ruled out the presence of a subdural hematoma.

CT scans in both cases revealed similar patterns, including craniocerebral disproportion, disjunction of the sutures, low measures of the attenuation coefficient values, relatively normal ventricular sizes and absence of cerebral atrophy. MRI further revealed agenesis of the corpus callosum in the first case (an association originally described by Kapila et al.\textsuperscript{14}).

Megalacrania associated with poor visualization of the cerebral sulci were the main criteria used to perform a unilateral subdural collection-peritoneal shunt in the first case. According to some reports\textsuperscript{2,16,18}, no difference in efficacy of shunts was seen in patients treated with bilateral versus unilateral shunting. The unilateral subdural collection-peritoneal shunt reduced the head circumference by 4 cm over the 25 days of hospitalization. Enlarged subarachnoid spaces permitted clinical observation as the treatment option in the second case.

Symptomatic subdural collections of infancy are not benign conditions and should be differentiated as such. Rapidly enlarging head size, severe disjunction associated with normal neuropsychomotor development, however, do not necessarily point to intervention. Even the upward gaze impairment-setting sun sign in the absence of acute signs of intracranial hypertension is not a direction toward surgical intervention, as our second case illustrates. Litosky et al.\textsuperscript{16} divided their cases of subdural collections of infancy into two groups: patients with asymptomatic macrocrania and those with rapidly enlarging head size, full fontanelle, vomiting, and seizures. Considering their data and our two cases, we agree with other findings\textsuperscript{1,14,23} that benign is an inappropriate term because it is too general. Regardless of the terminology, however, even symptomatic cases, such as our second one, can have a good outcome with conservative treatment. What appears to dictate a good or poor outcome is close observation associated with full laboratory workup that evaluates any signs or symptoms of discompensation.

References


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