Congenital inclusion dermoid cyst of the Posterior fontanel - A case report

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Abstract

Background: Dermoid cysts are rare and benign tumors originating from failure of the normal embryologic development. Congenital inclusion dermoid cysts are usually located over the anterior fontanel. Surgical treatment is curative and provides good aesthetic results. The authors present a rare case of congenital inclusion dermoid cyst over the posterior fontanel and discuss the location, differential diagnosis, treatment and prognosis of this unusual pathological entity. Case description: A two-year old boy presented with a lesion on the posterior aspect of the head which had dated since birth. Imaging studies revealed an extracranial lesion over the posterior fontanel without intracranial extension. Surgical removal was performed and the histopathological examination confirmed the diagnosis of a dermoid cyst. The patient was discharged free of any medical complications. Conclusion: Although congenital inclusion dermoid cysts are more common over the anterior fontanel, they can occur over the posterior fontanel as well. Surgical treatment is curative in both locations with favorable aesthetic results.

Keywords: Dermoid cyst, congenital inclusion cyst, posterior fontanel.

Introduction

Congenital inclusion dermoid cysts (CIDC) are rare and benign lesions originating from a failure during normal embryologic development of the neural tube. Usually, they are found over the anterior fontanel during childhood. The diagnosis of CIDC is suspected on the basis of clinical and radiologic features of the lesion. However, only after surgical excision and histopathological analysis can it be fully confirmed. The goal of treatment is the complete surgical removal of the tumor, which provides very good outcome for the patients with low recurrence rates and satisfactory aesthetic results.

The authors report for the first time a case of a CIDC over the posterior fontanel. No other similar cases could be retrieved from the world literature.

Case report

A two-year-old boy presented with a history of a bulging lesion on the posterior aspect of the head which had dated since birth. The physical examination revealed a painless, not pulsatile lesion, adherent to the posterior surface of the cranium and not associated with any kind of draining secretions (figures 1 and 2). The head computed tomography (CT) illustrated an extracranial tumor over the posterior fontanel without intracranial involvement (fig. 3). The lesion was completely resected and the patient was discharged free of any complications. The histopathological analysis was consistent with a dermoid cyst.

Discussion

Dermoid cysts are rare lesions representing 0.1 to 0.2% of the cranial tumors. Pathologically, they can be classified in three types: the congenital teratoma type, the acquired implantation type, and the congenital inclusion type. The last one, unlike the teratoma, is not neoplastic and develops due to in-
clusion of ectodermic and mesodermic cells during the third to fifth week of gestation.\textsuperscript{8, 10}

The CIDC is usually diagnosed in childhood,\textsuperscript{5, 6, 11} being rare in the adult population.\textsuperscript{5, 9, 14} Its main location is over the anterior fontanel.\textsuperscript{1, 2, 5, 7, 11, 13, 14} The authors describe herein a case occurring over the posterior fontanel which, to their knowledge, is the only case of CIDC of the posterior fontanel reported in the literature.

During clinical examination, the CIDC is characterized as a soft and painless not pulsatile lesion, lacking inflammatory signs.\textsuperscript{2} Recently, it has been reported that it can be associated with spinal dysraphism,\textsuperscript{14} but normally there are no neurologic or systemic complications associated with CIDC.\textsuperscript{2} Histological features include sebaceous and sweat glands, hair follicles and a thick stratified squamous epithelium that can undergo dystrophic calcifications.\textsuperscript{3} The cystic secretion can be rich in lipids, due to the sebaceous glands, and have a high concentration of sodium, chloride, potassium, and glucose.\textsuperscript{2, 3, 12}

The presented case is in agreement with the aforementioned characteristics but it is unique in the sense that it is located on the posterior fontanel. In addition to the anterior fontanel,\textsuperscript{2, 3, 5, 7, 8, 15} there have been reports in the literature of dermoid cysts occurring in the neck,\textsuperscript{16} sacrococcygeal region,\textsuperscript{17} and spleen.\textsuperscript{18}

The differential diagnosis related to CIDC includes lipoma, hemangioma, epidermoid cyst, sebaceous cyst, en-
References


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