CASE REPORT

Cranial fasciitis of childhood (CFC): an unusual clinical case of a rare disease.

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SUMMARY

Cranial fasciitis of childhood (CFC) is a very uncommon tumour of the scalp, which is almost exclusively observed in the first years of life. It is a benign proliferation of fibroblasts, but its rapid growth rate may resemble a malignant disease. This disease may be suspected from clinical and radiological features, but a definitive diagnosis may be achieved only by pathological examination. We report a case whose onset was in late childhood and whose clinical and radiological characteristics were atypical.

BACKGROUND

Cranial fasciitis of childhood (CFC) is a benign disease of the scalp. It is an extremely rare lesion and it was described almost exclusively in young children. CFC was first described in 1980 as a variant of nodular fasciitis with a propensity for the cranium of young children; actually, in young adults, it is located at the forearm and at the trunk, more frequently. Histologically, it is a non-neoplastic fibroblastic proliferation arising from deep fascia or periosteum of the cranium. Causal factors

and pathogenesis of CFC are unknown. It is thought to be a reactive process, although the evidence of local trauma is usually lacking. Clinically, it usually occurs as a single palpable, painless scalp mass, and neurological symptoms are seldom present.^{1–3} Here, we describe an 11-year-old boy developing CFC, which has been almost always described in younger age groups.

CASE PRESENTATION

An 11-year-old child of Caucasian ethnicity came to the Paediatric Emergency Department with two palpable, soft and painless scalp masses, whose onset dated 5–7 days in advance. No history of evident traumatism was reported and the patient presented no symptoms such as seizures, fever, vomiting or skin rash. The scalp masses consisted of two soft and well-circumscribed lesions, 4×3 cm in size, located at both left and right frontoparietal regions of the skull, without any local skin abnormality. The clinical history and physical examination were basically silent, with the exception of the following findings: premature birth (at 30 weeks' gestation); a minimal heart murmur, due to a previously diagnosed coronary fistula in the pulmonary artery, devoid of any haemodynamic significance and a history of recurrent mild nasal bleeding.

INVESTIGATIONS

The skull X-ray did not show any radiological abnormality and the laboratory data were in the normal range (white blood cell count: $6.71 \times 10^9/L$; platelets: $229 \times 10^9/L$, haemoglobin 12.9 g/dL, C reactive protein <0.8 ng/mL, erythrocyte sedimentation rate <20 mm/hour). A CT scan was performed, which confirmed the absence of skull fractures and showed two subcutaneous oval-shaped masses on the frontal regions, characterised with a low-density signal without evidence of inner calcifications and clearly circumscribed by a thin high-density capsule; the administration of contrast medium did not produce any enhancement (figure 1). These findings led to the diagnostic hypothesis of subgaleal haematomas and the surgeon requested no further imaging before the surgical procedure. Therefore, no cranial MRI was performed.

Figure 1 Head CT scan: axial CT section shows bilateral lesions on frontotemporal regions of the skull without radiological signs of bone involvement.

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Figure 2 Histopathology of CFC: low power appearance of the storiform pattern of fibroblastic proliferation and Spindle-shaped fibroblasts.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of such a skull lesion includes a number of other conditions, in addition to CFC: eosinophilic granuloma, juvenile fibromatosis, juvenile fibrosarcoma, primary bone neoplasms, haemangiomas, epidermoid cysts, leptomeningeal cysts, sarcoidosis, tuberculosis, syphilis and others.

TREATMENT

After the exclusion of coagulation and/or platelets disorders, the patient could undergo a surgical procedure: the haematoma was drained after an incision and it resulted to

be included in a fibrous capsule that was completely resected by the surgeon. Fragments of the surrounding bone were collected, too; all these specimens were sent for the evaluation by a pathologist, who reported several features consistent with a primary diagnosis of cranial fasciitis (figure 2) that was complicated by an intralesion bleeding. The 1 year follow-up after neurosurgery confirmed the benign clinical course and, consequently, no further CT scan was performed in the interest of the patient.

DISCUSSION

CFC is a benign fibroblastic proliferative lesion arising from the deep fascia of the skull. It is an extremely rare condition: fewer than 50 cases have been reported in the medical literature so far. It is more predominant in males (male to female ratio=2:1 approximately) and, almost exclusively, the average age of onset is less than 3 years, ranging from newborn to children under 6 years; however, average onset age in females is approximately twice that observed in males.^{4 5}

Here, we report a case of CFC in an 11-year-old boy. To our knowledge, he is the eldest male patient reported in the literature, excluding the three cases described in adults. The clinical presentation as a painless scalp mass without neurological impairment is typical; conversely, the simultaneous onset of bilateral lesions is rather exceptional, having been previously reported only once, in a 20-month-old boy with two soft tissue masses overlying parietal and temporal regions.⁶

CFC is a disease arising from fibrous tissue of deep fascia. Histologically, it is usual described as a proliferation of spindle-shaped normal fibroblasts or myofibroblasts (some with immature aspect, but no atypical mitosis), showing a storiform and/or loose disposition pattern.^{7 8} The involvement of underlying bone, usually consisting of some degree of erosion at the outer table and only occasionally extending to the deep table or to the meningeal sacs, is frequently described.² CT scan and MRI are the elective methods to characterise such a cranial mass and to study the relationship with adjacent structures, like bones and cerebral parenchyma.^{9 10} In our case, CT scan did not document any bone erosion or fracture, while it suggested the presence of a significant haemorrhagic component within a well-circumscribed lesion. The microscopic examination of the excised tissues showed a pattern of circumscribed spindle-shaped cells arranged in a loose stroma, which was consistent with CFC. Despite the absence of any radiological sign of bone involvement, a careful histological examination allowed to demonstrate some aspects of fibroblastic proliferation in the context of osseous fragments, likely indicating that CFC in this patient

Learning points

- ▶ Cranial fasciitis of childhood (CFC) must be considered in well-being children with rapidly growing scalp masses.
- ▶ Radiological investigations are required to ascertain the diagnosis of CFC and to assess the bone involvement.
- ▶ Children diagnosed with CFC must be evaluated for potential platelet and coagulation disorders, if a consistent haemorrhagic component is present inside the lesion.

was recognised and treated early, before the development of radiological skull lesions, as it occurs in most cases.

Contributors DP managed the clinical case, conceived and drafted the paper. NZ helped in writing the paper and in searching the medical literature.

Competing interests None declared.

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