Cranio-Facial Cirsoid Aneurysm in Childhood:
A Report of Two Cases

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Summary

Otu AA. Cranio-Facial Cirsoid Aneurysm in Childhood: A Report of Two Cases. Nigerian Journal of Paediatrics. 1984; 11:87. Two Nigerian male children with cirsoid aneurysm of the head and face are described. This lesion, which is also known as arterial racemose angioma, although well documented, is rare in clinical practice. The aneurysm was observed at birth in one child and at the age of three years in the other, suggesting a congenital aetiology. The lesion was successfully excised in one child; the second child was, however, lost to follow-up. A plea is made that whenever possible, surgery should be offered to affected children to obviate the unpleasant cosmetic appearance which is always seen in the untreated case.

Introduction

IRVING, Thakur and Walker recently described a single 12-year old male who presented with cirsoid aneurysm of the scalp. Following ligation of right superficial temporal artery, the patient returned 7 years later with recurrence and an ulcerated bleeding area. Although the ulcer healed, the aneurysm gradually increased over the next 14 years to cover most of the scalp and the forehead. Further surgery including ligation of the right external carotid artery, transection of vessels over the bridge of the nose and raising an extensive scalp flap and dissection of the enlarged vessels from the undersurface of the scalp was required to effect cure. This patient highlights some of the problems in the management of this rare lesion. The present communication concerns two children with this condition who were seen at the Department of Surgery, University of Calabar Teaching Hospital (UCTH). To the knowledge of this author, cirsoid aneurysm has not been reported before in Nigeria.

Case Reports

Case 1

A six-month old male infant presented with an extensive pulsatile mass in the scalp measuring 3 by 4 cm (Fig 1) and occupying the left parietal region of the head. The lesion was associated with an ulcer which had been present for one month. The infant was the first child of a recent marriage; both parents were young, being in their early twenties. The pregnancy had been supervised in hospital and the birth of the baby was by spontaneous vaginal delivery. The mother had noticed
soon after birth, that the baby had two or three
wine-coloured marks in the left parietal region;
these increased in size rapidly and eventually
coalesced.

On examination, a large pulsatile lesion, 3 by
4 cm across, in the left parietal region of the scalp
was found. The condition gave the child a gro-
tesque appearance (Fig 1). A bruit was heard
over the tumour and the growth of hair in the
area was relatively sparse.

The child was examined for associated con-
genital anomalies; none was found. The child's
development was apparently normal. Pseudomo-
nas pyocyanea was isolated from the ulcer; it was
sensitive to gentamicin. Skull radiographs showed
no bone erosion. Carotid angiography was con-
sidered but could not be carried out for lack of
facilities.

Initial treatment consisted of antibiotic therapy
for the scalp sepsis. This led to a healing of the
ulcer within three months leaving a puckered scar
at the centre of the lesion. (Fig 1). A biopsy of a
portion of the tumour was then taken. However,
while arrangements were being made for angio
graphic studies elsewhere, the child was lost to
follow-up.

Case 2

A four-year old boy was seen with a one-year
history of a swelling in the lower lip. The swelling
was small at first but increased slowly in size. The
child was the fifth of seven children in the family
and both parents were middle-aged farmers. The
development of the child had been normal in
every other respect.

On examination, a pulsatile mass, 3 by 2 cm
was found occupying most of the lower lip, much
more to the right than the left (Fig 2). A bruit
was heard over the lesion. Two other much smaller
but similarly pulsatile lesions, were seen in the
right side of the face (Fig 3). No associated con-
genital anomalies were found.
Fig 3 Two small aneurysmal swellings on the right side of the face of the same child as in Fig 2.

The tumour in the lip was successfully excised. A year after surgery, the cosmetic effect was considered satisfactory.

Histological examination of sections from both the biopsy material and the excised surgical specimen stained with haematoxylin and eosin showed an overgrowth of small arteries. These intertwined in racemose fashion forming a highly vascular tumour which is sometimes likened to a pulsating mass of earthworms.

Discussion

Cirsoid aneurysm is a rare condition. It is known by a variety of other names including arterial angioma, and arterial racemose angioma, both names indicating the histological appearance of the lesion. Clinically, it is most often seen to occupy the forehead or the side of the scalp; it may, however, be seen in other parts of the body including the face. These features are demonstrated by the two patients in this report.

In general, cirsoid aneurysm is regarded as a congenital arteriovenous malformation. Like other angiomata, it is therefore, a hamartoma. The congenital nature of this condition is exemplified by the patients described in this communication. In one child (case 1), the lesion was observed by the mother shortly after birth; in the other child (case 2), it became evident at the age of three years. This observation is in agreement with previous reports. However, there is sometimes, a history of trauma such as a direct blow on the site. Although trauma may serve to draw attention to an already existing lesion in some instances, it is possible that in others, the aneurysm is of traumatic origin and, therefore, not a true neoplasm. It is noteworthy that there was no history of trauma in the two children herein described.

Cirsoid aneurysm of the scalp can give rise to skin ischaemia resulting in ulcer formation; this was clearly the case in the first child. Furthermore, skull erosion and intravascular connections may result in cerebral impairment. The present patient showed no evidence of such impairment. However, the unpleasant cosmetic appearance which always accompanies this condition can be readily seen.

These aneurysms have been classified by Oldfield and Addison as small, intermediate and large. Case 1 falls into the latter class and case 2 into the intermediate group. Although spontaneous thrombosis with subsequent arrest of growth has been observed, most cirsoid aneurysms are progressive. The two children demonstrate this feature.

Wherever possible, surgery should be undertaken to prevent the complications: grotesque appearance and cerebral impairment. The surgical procedure adopted in a given case varies considerably from ligation of the vascular supply to the tumour, excision of the aneurysm area, wide resection with rotation flaps, to excision of dilated vascular channels. The aneurysm was excised successfully in the second case. In the first case, however, lack of facilities for angiographic
studies prevented early operation and the child
was lost to follow-up. In this child, angiography
was considered necessary as large cirsoid aneur-
ysms are known to possess large feeder arteries
which enter them from all sides making surgery a
formidable problem.

The operation of choice which was planned for
this child was extirpation of the arteriovenous
channels and dilated veins; in the hands of other
operators this procedure has resulted in cure in
some patients. Selective angiography with arterial
embolisation using thrombotic materials might
be attempted but the extent of arterial collaterals
would make this less satisfactory.

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